STUDY GUIDE OF PEDIATRIC SURGERY

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From the Authors

Training of foreign students is a big responsibility for higher educational institutions teachers. It is shaping Institutions' international image and prestige. When teaching is put into practice in a language which is not the teacher's native one, the responsibility raises exponentially. Both teacher's competency and language skills are essential.

We have managed to fill out a powerful group of authors, renowned specialists in a certain field of pediatric surgery, who are also quite proficient in English. The study guide briefly presents modern approaches to the diagnosis and treatment of most common surgical diseases in the pediatric population. We expect this study guide to be equally helpful for foreign students, teachers, and Ukrainian students, who want to improve their level of English

We have tried to adhere to international classifications and terms that would have facilitated communication between doctors of different nationalities. It was taken into account that English is the main language of international medical forums as well as medical periodicals. At the same time the study guide retains traditions of Ukrainian pediatric surgery: all the clinical cases and illustrations represent authors' original studies and clinical practice.

By regret, there are a limited number of academic hours for pediatric surgery studying; therefore we have tried to provide structured presentations by means of tables and drawings. Each chapter is supplied with self-control tasks and essential clinical cases. We expect this manual to be useful not only for academic success in pediatric surgery, but also in further career guiding and clinical practice.

Editors,
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ABBREVIATIONS

ABCs - airway, breathing, and circulation
AIS - Abbreviated Injury Scale
ALT – alaninaminotransferase
ALV - artificial lung ventilation
AP - alkalin phosphatase
ARDS-Acute respiratory distress syndrome
ARM - Anorectal malformations
AST - aspartataminotransferase
ATLS - Advanced Trauma Life Supports
AVM - arterio-venous malformations
BA - Biliary atresia
BCV - blood circulation volume
BJI - Bone and Joint Infection
CBC - Complete blood count
CC - Choledochal cyst
CPAM - Congenital pulmonary airway malformations
CRAMS - Circulation, Respiration, Abdomen, Motor, and Speech
CRP – C-reactive protein
CT - computed tomography
DAA - Double aortic arch
DCO- damage control orthopedics
DCS - damage control surgery
DSRS - distal splenorenal shunt
EA - Esophageal atresia
EPVO - extrahepatic portal vein obstruction
ERCPG - Endoscopic Retrograde Cholangiopancreatography
ESR - erythrocyte sedimentation rate
FAST - focused assessment with sonography in trauma
GAS - group A streptococcus
GCS - Glasgow coma score
GGT - gammaglutamilaminotransferase
GI – gastrointestinal
HDN - Hemorrhagic disease of the newborn
ISS - Injury Severity Score
LM – lymphatic malformations
MISS - modified injuryseverity scale
MODS - multiple organ dysfunction syndrome
MRCP - Magnetic Resonance Cholangiopancreatography
MRI - magnetic resonance imaging
MRSA - methicillin-resistant S. aureus
NG - nasogastric
OM - Osteomyelitis
PALS - Pediatric Advanced Life Support
PDABCDE - P - Protection, D - Decision, A - Airway Maintenance with Cervical Spine Protection, B - Breathing and Ventilation, C - Circulation with Hemorrhage Control, D - Disability (Neurologic Evaluation), E - Exposure / Environmental control
PLP - Pathologic Lead Point
PTFE – polytetrafluorethylene
PTS - Pediatric Trauma Scale
PTS - pediatric trauma score

PVL - Panton-Valentin-leukocidin
RLQ - right lower quadrant
SA - septic arthritis
TB - tuberculosis
TEF - tracheoesophageal fistula
VM - Venous malformations
WBC - white blood cell

CONTENTS

THEME 1

Acute abdomen in children. Appendicitis. Primary peritonitis. Intussusception.

1. **Overview**: Abdominal pain is one of the most common chief complaints of children seen in the emergency department, and appendicitis, peritonitis, and intussusception represent the most common abdominal surgical emergency in the pediatric population. The results of treatment depend directly on from timely diagnosis and time of operational response since the moment of illness. It is especially important to study clinical picture and diagnostic techniques for diagnosis of acute appendicitis in children before 3 years. The topic urgency is stipulated for difficulty of diagnosis, high morbidity, and frequent complications with unfavorable consequences.
2. **Aims of the lesson:**

1. To analyze anatomic-morphological peculiarities of the abdominal cavity in children of different age groups.
2. To learn peculiarities of etiology, diagnosis and clinical picture of acute appendicitis, peritonitis, intussusception in children.
3. To analyze peculiarities of clinical picture of acute appendicitis at atypical localization of the appendix.
4. To master peculiarities of examination of young children with abdominal pain.
5. To learn peculiarities of the techniques of operative intervention in acute appendicitis in children.
6. To know peculiarities of the conservative treatment of intussusception.
7. To know possible complications of acute appendicitis, intussusception.
8. To know recommendations for postoperative dispensary surveillance of children after laparomy, laparoscopy.
9. To discuss peculiarities of diagnostic and treatment of appendicular mass in children.
10. To know principles of treatment of peritonitis in children.
11. To know peculiarities of etiology, diagnostics and clinical development of peritonitis in the newborn.

3. **Basic skills and knowledge, necessary to know: (interdisciplinary integration)**

Subjects	**Skills obtained**
Anatomy, topographic anatomy	Palpation of different parts of intestines, knowledge of peculiarities of abdominal cavity in different age periods.
Physiopathology	Clinical interpretation of laboratory analyses.
Pathology	To compare morphological changes, peculiar of certain kinds of acute appendicitis, peritonitis.
Microbiology	To master technique of collecting material for investigation. To interpret the results.
Pediatric diseases propaedeutics	To collect complaints, medical history, to examine the child, to use additional methods of examination at abdominal cavity diseases.

Topographic anatomy and surgery	To master methodics (schemes) of operative access and methods of operative intervention on abdominal cavity organs in children of different age groups.
General surgery	To display methods of patient preparation for treatment, diagnostics and operation.
Hospital surgery	To perform differential diagnostics, to ground methods of diagnostics and treatment of acute abdominal pain in children.
Clinical pharmacology	To classify medications necessary for treatment of acute surgical diseases

4. Tasks for individual self-preparation for the lesson.

4.1. The list of key terms and characteristics a student must know:

Term	Definition
Acute appendicitis	Inflammation of the vermiform process.
Peritonitis	Inflammation of the peritoneum and abdominal cavity, primary, secondary.
Primary peritonitis	Primary peritonitis (idiopathic or spontaneous peritonitis) has been defined as an infectious process involving the peritoneal cavity without an intra-abdominal source
Appendicular mass	An appendix mass results from appendicitis that is localized by edematous, adherent omentum and loops of small bowel.
Appendiceal abscess	The appendiceal abscess is a localized suppurative process that may occur at any time in the course of appendicitis, or may complicate an appendiceal mass
Plain roentgenogram of the abdominal cavity.	It is performed in vertical position, we appraise the abdominal cavity organs condition, presence of liquid and free gases, signs of intestinal obstruction.
Intussusception	Full-thickness invagination of the proximal bowel into the distal contiguous intestine

”Red currant jelly” stools	The combination of intraluminal fluid, blood, and mucosal tissue fragments results in stools with appearance, color, and consistency of red currantjelly

CONTENT

Acute appendicitis

Appendicitis is the most common childhood surgical emergency. However the diagnosis can be challenging, especially in small children, often leading to either unnecessary surgery in children without appendicitis, or a ruptured appendix and serious complications when the condition is missed.

In the first 3 years of life appendicitis occurs rarely - 3-7%, before 1 year - 0,9-1%. Appendicitis in the newborn occurs in single cases. After three years the disease frequency increases and reaches its highest position at 9-11 years. The frequency of destructive forms of appendicitis in children before 3 years is twice as high as in older children, and it makes 70-84%.

It is especially important to know anatomical peculiarities of the right inguinal area in children of young age to be able to diagnose and differentiate acute appendicitis.

The anatomy of abdominal wall. The skin of the abdomen is tender and elastic. The subcutaneous tissue is well developed. The superficial fascia isn’t expressed. The muscle layers aren’t differentiated, they are intimately related with aponeurosis. The preperitoneal tissue is absent. The peritoneum is very thin and tender.

The caecum in the newborn is situated high, in the right subcostal region. It is explained by the fact that in the newborn the third stage of physiological rotation of the “middle gut” isn’t finished yet. The caecum assumes position in the right inguinal area by three years, as a rule.

The vermiform process comes off the posterior-interior surface of the caecum *in the place of* thenia convergence. It is short and broad in the newborn, of conical shape with wide lumen, no definite borders with the caecum. The valve, covering entrance into the process (Gerlach's valve) isn’t expressed.

The position of the process can be descending (35%), medial (26%), retrocaecal (20%) or lateral (15%).

The anatomy of the vermiform process wall doesn’t differ from the anatomy of the caecum. The process walls are hydrophilic. Blood supply is more intensive than in children of older age. In young children the nerve plexuses are characterized by immaturity, they are undifferentiated. There are no lymphoid follicles in the appendicular wall of the newborn, in three years there are 7-8 of them, in 9-10 years-12-15 (as in the adults).

The greater omentum in children of the first year is short and thin, that is why it doesn’t perform the barrier function. This explains the fact that in young children at destructive appendicitis we can meet local peritonitis forms (abscess, infiltrate) very rarely, most frequently generalized peritonitis occurs.

The omentum in children before three years possesses low plastic characteristics, it is susceptible to expressed exudation.

Appendicitis results from luminal obstruction followed by infection. But the cause of obstruction is not always clear. It may be fecal stones, helminthes, congenital anomalies of the process (kinking, flexion, membranes) and also process’ carcinoid. In this case secretions of the mucosa are accumulated in the lumen of the appendix, distend it and ischemia develops. The intestinal bacteria penetrate into the mucosa and cause inflammation of all layers of the appendix. Another cause of appendicitis may be lymphoid follicles hyperplasia, which also may cause obstruction of the lumen of the appendix. Another cause of appendicitis may be lymphoid follicles hyperplasia, which also may cause obstruction of the lumen of the appendix.

Alimentary factor is also very important. The diet, rich in meat and fats contributes to appendicitis development.

Patients with cystic fibrosis have an iccreased incidence of appendicitis, which presumably results fromalterationsin the mucous-secreting glands.

Clinical and morphological classification (after Shprengel).

Simple (congestive [catarrhal] appendicitis)

The process looks normal or there is little hyperemia and swelling of the process. There is no exudate on the serous layer. In the serous layer there may appear focal haemorrhages. But catarrhal changes in the appendix may be the secondary ones, caused by another abdominal cavity pathology.

Phlegmonous appendicitis. As a rule, we may observe obstruction of the process. It is oedematic, hyperemic, intense, thickened, in some parts it is covered with fibrous layers. Sometimes the process is swollen, with fluctuation in it, there is pus in its lumen (process empyema). We determine serous, serous-purulent or purulent exudate in the abdominal cavity.

Gangrenous appendicitis is characterized by deep destructive changes of all appendiceal wall. The process is thickened, earthy-grey, with purulent and fibrinous layers. Its wall is flaccid, so perforation may easily happen. The parietal peritoneum may often possess such characteristics: it is swollen, infiltrated, covered with fibrin.

Perforating appendicitis. The appendiceal wall is perforated in the necrosis area. The purulent exudate with fecal odor is determined in the abdominal cavity. The perforating orifice is often situated near the apex on the side opposite to the mesentery, where blood supply is poor. Mesenteric edge perforation of the appendix in young children leads to general peritonitis. The parietal peritoneum is swollen, hyperemic, thickened and may be easily broken.

Clinical picture of acute appendicitis in older children.

The classic presentation of appendicitis has been well described, and includes fever, anorexia, nausea, guarding, and migration of pain from the umbilical region to the right lower quadrant.

The main symptom of acute appendicitis is abdominal pain. The pain first appears in the umbilical region, it is continuous and abrupt, with time - more intense. Later the pain is located in the right inguinal area - Koher symptom. The pain may diminish with time due to necrosis of the nerve endings of the vermiform process in gangrenous appendicitis. Later, with peritonitis development, the pains appear again and they are more intense.

The patients refuse food, they have nausea and vomiting (once or twice). Anorexia is a helpful sign, particularly in children. So a hungry child rarely has appendicitis. The body temperature is increased to the subfebrile. Some patients have stool retention or diarrhea. A child with acute appendicitis looks ill: moves slowly, bends the shoulders, spares the right leg at walking, lies down very carefully, lies on the right side with legs pulled to the abdomen, the face expresses suffering.

The tongue and lips are dry, tachycardia is noted. The characteristic symptom is discrepancy of pulse and temperature (normally every degree of temperature's increase provides for pulse rate increasing for 10 beats, in appendicitis - pulse rate greatly exceeds temperature increase.)

Examination of the abdomen: at uncomplicated appendicitis the abdomen is of usual shape, but the patient tries to spare it while breathing. The classical symptoms at palpation are: local tenderness in the right inguinal area - Filatov's sign, passive rigidity of muscles, the Blumberg's sign (abrupt pain intensification in the right inguinal area at quick releasing of the hand pressing on the abdominal wall). Sometimes some other symptoms may be positive: Rouzing's symptom (symptom of visceral tenderness caused by reverse direction of flatus from the sigmoid colon to the caecum), Voskresensky's symptom (the " shirt" symptom - there are local tenderness and vibration of the abdominal wall under the place where gliding motions stop), Sitkovsky's symptom (increased tenderness at changing position from the back to the left side), Razdolsky's symptom (abrupt tenderness at percussion in the right inguinal area), Bartomier - Michelson's symptom (increased tenderness at palpation of the right inguinal area when the patient is lying on the left side), Obraztsov' symptom (when a hand is pressing deeply onto the right inguinal area, the patient

can't raise his straightened leg because of acute pain) and others. At auscultation of the abdomen we determine weak peristaltic murmurs.

The patients with suspected acute appendicitis must have rectal temperature measured, at appendicitis it is increased more than for 0,5-1°C in comparison with the axillary.

Clinical picture of acute appendicitis in young children is characterized by general symptoms prevailing over the local ones; this is caused by generalized reaction of the child's body onto the inflammatory process. These symptoms (restlessness, high temperature, vomiting) are characteristic for many diseases, not only appendicitis. This provides for difficulty of diagnosis of appendicitis in young children.

The diagnosis difficulty is also explained by complicated anamnesis collection and negative reaction of the child towards the examination. In older children complaints about right inguinal pain prevail, while in young children of the first three years there are no direct indications of pain, so it is possible to put the diagnosis just by the indirect signs. The most important sign is child's changed behavior. The child becomes restless, capricious, uncooperative. The condition disturbs sleep, the child doesn't want to play, refuses food. If the mother touches the child's abdomen, agitation increases.

Body temperature is increased, unlike the older children it reaches 38-39°C. Quite often multiple attacks of vomiting appear. The vomiting in the onset of the disease is reflex, it doesn't bring relief. The vomiting contents are that of the stomach. Later vomiting is caused by intoxication.

A loose stool is present in nearly 15-20% of young children; it is encountered in children with deeply expressed destructive changes in the vermiform process.

At objective examination of the child tachycardia and dryness of the lips and tongue are determined. The symptom of pulse and temperature discrepancy is not characteristic for children of the first three years. At abdominal examination it is worth noting if the child spares the abdomen at breathing.

The Blumberg's and other symptoms of peritoneal irritation are quite rare to be determined, but the symptom of dosed percussion (by Shurinok) is quite informative. At auscultation of the abdomen - peristaltic murmurs are slow and weak, at peritonitis they may not be heard at all.

It is necessary to remember that clinical picture of acute appendicitis may vary depending on the process localization.

Retrocaecal localization. In this localization the disease develops with signs of expressed intoxication at mild local symptoms. Local tenderness at palpation and rigidity of muscles of the anterior abdominal wall are less expressed than in typical appendix localization. Later peritoneal symptoms appear.

Retroperitoneal localization. Pain appears in the right lumbar area, irradiating into the genitals or ureter, causing dysuria. The abdomen participates in breathing, at palpation it is soft, not painful, Blumberg's symptom is negative. Kokher's symptom is positive. At palpation in the right lumbar area there is expressed tenderness, muscle rigidity, positive Pasternatskiy's symptom

Pelvic appendicitis. The characteristic for this are pains in lower part of the abdomen, above the bosom, irradiating towards the urethra position, into the testicle (right labia pudendi) , or the rectum. The patients may have diarrhea or frequent painful urination. At palpation of the abdomen there is tenderness and rigidity of muscles above the bosom. General analysis of urine often shows proteinuria, leucocyturia, epithelium, erythrocytes.

Left-side appendicitis. Such localization may be caused by mobile caecum, uncomplete bending of the " middle intestine", reverse localization of the internal organs. It is met in 0,5% of all cases. Clinical manifestations are the same as in right-side localization, but local manifestations are expressed in the left.

Subhepatic appendicitis. The clinical picture is like that in destructive cholecystitis: multiple vomiting, pains in the right subcostal region, local tenderness and muscular rigidity, positive Ortner's symptom. Sometimes there is slight skin icteritiousness. The lower parts of the abdomen are not painful, soft at palpation.

Laboratory studies

Complete blood count (CBC) is the most commonly advised laboratory investigation in children with suspected acute appendicitis. Although the white blood cell (WBC) count is increased in acute appendicitis, still it is non-specific and insensitive. WBC count is also elevated in other disease processes such as gastroenteritis, mesenteric lymphadenitis, pelvic inflammatory disease and certain other infections. Furthermore, the WBC count cannot differentiate between a complicated and an uncomplicated acute appendicitis. Elevated neutrophil count along with the total WBC count further helps in the diagnosis of acute appendicitis. However, in cases of less chances of acute appendicitis, a high WBC count warrants further radiological evaluation and clinical observation.

C-reactive protein (CRP) is a nonspecific inflammatory mediator. However, it is more sensitive than WBC count in diagnosing appendicular perforation and abscess formation, which are more common in children.

Urine analysis is advised to rule out urinary tract infection. However 7–25% of pediatric patients with acute appendicitis have more than 5 WBCs or RBCs per high power field in the urine sample.

The decision to advise WBC count, neutrophil count, and CRP, or urine analysis is usually based on the clinical impression, duration of symptoms, and the preference of the emergency room physician or consultant surgeon.

Imaging studies

Ultrasound

Demonstration of a noncompressible appendix that is 7 mm or larger in anteroposterior diameter is the primary criterion for the diagnosis. A target sign with five concentric layers, high echogenicity surrounding the appendix, pericecal and perivesical free fluid, and thickened bowel loops with deceased peristalsis are the additional signs of appendicitis.

Despite the benefits of US, the diagnostic utility of the modality may be greatly limited by operator experience and lack of availability at many hospitals during nights, weekends and holidays.

The diagnostic criteria on CT scan include swollen appendix (diameter more than 7 mm), fat streaking, focal caecal apical thickening, lymphadenopathy, presence of an appendicolith, abscesses.

Currently, no significant data is available on the sensitivity and specificity of CT scan for the detection of acute appendicitis in young children. Despite this relatively high diagnostic accuracy, concern over ionizing radiation has prompted efforts to minimize use of CT as the initial diagnostic imaging test, and to explore the efficacy of radiation-sparing protocols.

MRI has not been routinely used in diagnosis of acute appendicitis. Disadvantages of MRI investigation are lack of availability at many hospitals, lengthy image acquisition time, and relatively high cost compared to CT and US. Furthermore, MRI is generally not considered appropriate for younger children and those who cannot lie still for the duration of the test.

Differential diagnosis of acute appendicitis in children presents great difficulties, because acute appendicitis has common symptoms with many somatic, infectious, urologic and surgical diseases.

General and infectious diseases: acute nonspecific mesenteric lymphadenitis, acute respiratory viral infection, pneumonia, acute otitis, intestinal infections, pseudotuberculosis, acute hepatitis, children infectious diseases (measles, scarlet fever, chicken-pox, mumps).

Surgical diseases of the abdominal cavity organs: coprostasis; ileus; intussusception; acute cholecystitis; ulcer disease; acute pancreatitis; abdominal tumors; Crohn's disease; Jackson's membrane; Lane's adhesion; uncomplete bending of the " middle gut".

Urologic diseases: urolithiasis; nephroptosis; acute pyelonephritis; acute cystitis.

Gynecological diseases: primary pelvic peritonitis; premenstrual cramps; hematocolpos; hematosalpinx; ovarian apoplexy; tumors of internal genitals.

Systemic diseases: rheumatism; haemorrhagic vasculitis; diabetes mellitus.

The treatment of acute appendicitis is operation. Indications for operation are determined by the surgeon on the background of clinical picture and differential diagnosis. In doubtful cases the dynamic surveillance is sufficient to decide the question concerning operative treatment. But examination and surveillance must be performed in short terms to avoid severe complications. In difficult cases the surgeon must decide whether to perform the operation or delay it. At unnecessary operation there may appear aggravations of some other diseases, at delayed operation some complication that will end terminally may develop. In such case laparoscopy may help. Historically, open appendectomy has been practiced in young children all over the world for acute appendicitis. (Fig. 1.1).

However with the advent of minimally invasive techniques, laparoscopic appendectomy has become increasingly popular among pediatric surgeons. (Fig.1.2).

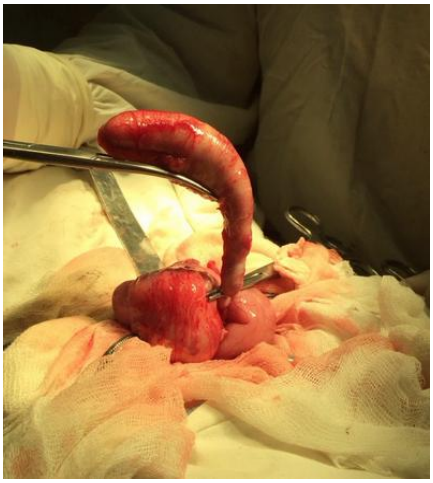
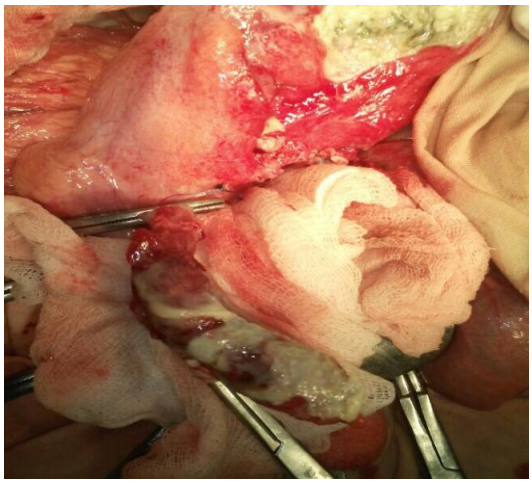
A.  B. 

Fig. 1.1. Open appendectomy. A – Flegmonous appendicitis, B – gangrenous appendicitis

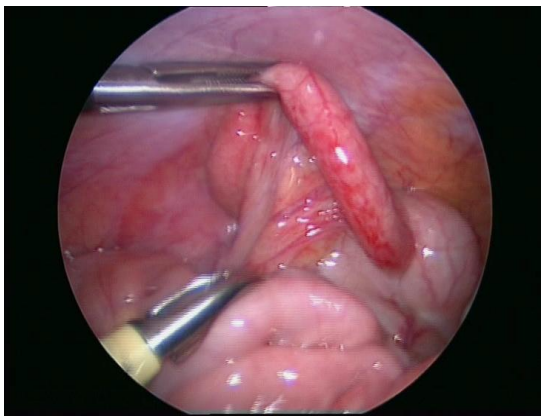


Fig. 1.2. Laparoscopic appendectomy

At uncomplicated appendicitis the patient doesn't require special preoperative management, just no food and cleansing enema before the operation. General anesthesia is administered.

In the open technique, a transverse or oblique (Volkovich-Dyakonov) right lower quadrant incision is made through the McBurney point. In this incision muscles and nerves of the abdominal cavity are traumatized minimally, also the abdominal wound burst and postoperative ventral hernia are rare to appear. If necessary, the incision may be extended. After the abdomen is entered, the cecum and appendix are mobilized and the appendix is brought out through the incision. The mesoappendix is then divided, and the base of the appendix is ligated. The stump is managed by simple ligation, ligation with inversion using a purse-string or Z-stitch suture, or inversion without ligature. For simple appendicitis, irrigating the wound is unnecessary. The wound is closed in layers, and no drains are placed.

As of today, laparoscopic appendectomy is increasingly being used and introduced. Three trocars are usually employed: one at the umbilicus for the scope, one in the suprapubic area, and one in the left lower quadrant, although a single-incision multiport approach may also be employed. The appendix is found by following the cecum, and the mesoappendix is grasped near the tip to lift the appendix toward the abdominal wall. After mobilization of the appendix, the mesoappendix is divided, the appendiceal stump is ligated with endoloops or an endoscopic stapler, and the appendix is delivered through the umbilical trocar site.

At appendicitis complicated with peritonitis treatment starts with detailed preoperative management. It consists of decompression of the alimentary tract, antibiotics, infusive therapy aimed to correct the dehydration, improved rheological blood properties, energetic deficiency reinforcement, correction of water-electrolyte impairment, disintoxication. The preoperative management shouldn't last for more than 5-6 hours.

The operative treatment in general appendicular peritonitis is performed under endotracheal anesthesia.

The complications of acute appendicitis are divided into preoperative, postoperative and complications during the operation.

Complications in the abdominal cavity (preoperative): peritonitis, periappendicular abscess, appendicular mass.

Postoperative: adhesive intestinal obstruction, paralytic ileus, insufficiency of appendicular stump, postoperative peritonitis, intestinal fistulas, postoperative infiltrate and abdominal cavity abscess.

Complications during the operation: haemorrhage, intestinal perforation.

Also, we determine complications in the *abdominal* cavity, wound, other organs and systems.

Complications in the wound: haemorrhage, suppuration, infiltrate, ligature fistula, separation of sutures, intestinal eventration, ventral hernia, keloid cicatrix.

Complications in other organs: pneumonia, sepsis, hepatic insufficiency, renal insufficiency, cerebral edema, toxic encephalopathy.

The prevention of acute appendicitis complications is early diagnosis and timely treatment of appendicitis. To reduce the diagnostic errors, according to the order of Ministry of Public Health of Ukraine, all children before 3 years with abdominal pain must be hospitalized in the pediatric surgical department with their subsequent examination and surveillance.

*Complications of acute appendicitis***.** Peritonitis is an inflammation of the peritoneum and abdominal cavity. According to the generally accepted classification, the following forms of peritonitis are distinguished:

The peritonitis of appendicular genesis is common with the children brought to hospital on late stages of the disease. Peritonitis develops in 5 to 25,6% of all cases of acute appendicitis. The children before 3 years have it 4-5 times more often; this is explained by quick destruction of the appendix and late diagnosis, general symptoms predominating over the local ones and wide administration of antibiotics which makes recognition of peritonitis extremely difficult.

Clinical picture of appendicular peritonitis depends on character and speed of pathological process development. The disease symptoms may develop continuously or very quickly (at process' perforation).

Introduction of a modern treatment protocol a protocol of preoperative antibiotics, intraoperative irrigation of the peritoneal cavity, primary subcuticular skin closure and a short course of postoperative antibiotics showed good results in treatment of appendicular peritonitis.

Appendicular mass (infiltrate)

The appendicular infiltrate (appendix mass) appearance is an evidence of child's body ability to demarcate the inflammatory focus. Most frequently this complication develops in older children (10-14years), which is connected with great omentum localization, becoming quite long (2 cm lower than the umbilicus) only at 3 years. An appendix mass results from appendicitis that is localized by edematous, adherent omentum and loops of small bowel. In contrast, the

appendiceal abscess is a localized suppurative process that may occur at any time in the course of appendicitis, or may complicate an appendiceal mass. Clinically, it is not possible in most cases to distinguish with certainty between the two conditions.

The condition of children with appendix mass is moderate. Abdominal pains are less intense, intoxication is observed. Body temperature is 38-39°C, there may be slight tachycardia. At palpation in the right inguinal area there is determined abruptly painful malformation without definite borders. Sometimes it occupies almost all right part of the abdomen or even over the middle line. According to infiltrate localization we may observe dysuria, tenesmus. Depending on age, body reaction, degree of inflammatory process development and treatment intensity this stage lasts for 12-14 days.

Appendicular infiltrates are treated conservatively with antibiotics. An exception is infiltrate suppuration, in which operation is indicated. When conservative treatments have a positive result at first, there is often a dilemma whether or not to perform an interval appendectomy or maintain nonoperative approach. However, the real disadvantage of the latter is not having the appendix submitted to histological analysis. Initial conservative management of appendiceal masses, followed by interval appendectomy, is most common tactics in pediatric surgery.

If infiltrate is determined accidentally during operation on occasion of acute appendicitis, the surgeon's tactics depends on the circumstances. In cases when the process is surrounded with loose adhesions and is easily demarcated from the surrounding organs and tissues, appendectomy is performed. At presence of solid infiltrate in the abdominal cavity it is necessary to apply drainage. The postoperative management of the child includes general supportive and antibacterial therapy. In two months the child is indicated to undergo appendectomy.

Primary peritonitis

The peritonitis, which doesn't result from inflammation of the abdominal cavity' organs (appendicitis, diverticulitis, intestinal perforation, etc) is named idiopathic, primary, haematogenous, etc.

We can determine 2 groups of patients according to clinical picture, peculiarities of clinical treatment and prognosis: 1) peritonitis without accompanying diseases; 2) peritonitis appearing on the background of ascites.

In the majority of the first group of patients peritonitis develops relatively mildly, its clinical picture is similar with acute appendicitis. But quite often the disease develops quickly and very severely. So, we can determine two forms of primary peritonitis: simple and toxic.

Clinical picture of simple form of primary peritonitis is characterized by subacute onset. The child complains about development of generalized continuous pain in the abdomen, gradually increasing and localized in the right inguinal area. The child refuses food, body temperature reaches 37,5-38,5°C. There appears vomiting. The stool is normal. Gradually general condition aggravates, abdominal pain becomes more intense. Pulse is rapid. The tongue is damp, a little coated. Palpation and percussion of the abdomen are painful in all regions, but especially- in the right inguinal area, here we may observe rigidity of muscles of anterior abdominal wall. In older children Blumberg's symptom is expressed. The CBC shows leucocytosis within $15\text{-}20*10^9$/l, neutrophilosis shifted to the left.

Clinical picture of *toxic form* of primary peritonitis is characterized by rapid onset. It is most frequently to be encountered in preschool children and children of early school age, especially in girls. The main way of infectioning in this case is ascending infection of the peritoneum through the female genitals (cryptogenic peritonitis.). The prepubertal cervix lacks the endocervical glands that may harbor bacteria, so ascending infections in this age group would more likely be associated with some traumatic force that pushes the bacteria up through the vagina, as in sexual abuse cases or by jumping feet first into a swimming pool or lake water. Soon there appears diffuse abdominal pain, multiple vomiting. The body temperature increases to 39-40°C. There may be loose frequent stool. General condition of the patient progressively aggravates, the tongue is dry, coated with white lining. In a few hours since the disease onset the child may have cramps, dizziness. Pulse is rapid, and small. The abdomen has ordinary shape, it doesn't participate

in breathing, and there is expressed rigidity of muscles of anterior abdominal wall, positive symptom of Blumberg. If the patient was brought to hospital late, we may determine swelling of the anterior abdominal wall, CBC shows high leucocutosis, that reaches 35-45g/l with abrupt shift to the left. Girls may have mucopurulent vaginal discharge.

Ultrasound studies and computed tomography (CT) of the abdomen have been used to differentiate primary from secondary peritonitis (caused by such common pediatric processes as appendicitis) when the clinical picture is confusing.

Treatment of primary peritonitis is usually conservative. In all instances of peritonitis without an overt source of infection, failure to improve with intravenous antibiotics is an indication for laparoscopy or laparotomy. An appendectomy can be done safely even in the presence of cloudy exudate, and the bowel surface can be inspected for secondary causes of the infection.

The clinical picture of primary peritonitis developing on the background of ascites at impaired hepatic function, cirrhosis, ascites, lipoid nephrosis or nephrosonephritis, depends on the development of the main disease. Such patients are susceptible to infection, which often affects the patient and influences his condition. The most severe manifestation of infection is peritonitis. Administration of antibiotics has reduced its morbidity, but its development and prognosis still stay quite severe.

Spontaneous bacterial peritonitis has been described in children with indwelling ventriculoperitoneal shunts without evidence of cerebrospinal fluid infection.

In children maintained on chronic peritoneal dialysis, peritonitis is the primary complication compromising survival. Risk factors for the development of peritoneal dialysis-related peritonitis are the duration of dialysis (longer than a year), chronologic age younger than 2 years, and decreased serum IgG levels.

Clinical symptoms include abdominal pain, fever, and an elevated neutrophil count in the cloudy dialysate. Once suspected, the fluid should be cultured and both intravenous and intraperitoneal antibiotics should be instituted. Symptoms should begin to improve within 24 to 36 hours after initiation of therapy.

Intussuseption

Intussusception is among the commonest emergencies in children requiring the attention of a pediatric surgeon. The most common cause for a small bowell obstruction in an infant without an abdominal scar is an ntussusception.

Intussusception means by definition that one portion of the gut is telescoping in another distal part with further motion of the intussusceptions into the intusscepiens by ongoing peristalsis. Intussusception occurs most frequently between the ages of 3 months and 3 years. Peak incidence is 4-12 months. The most common form is ileo-colic in 80-90% of cases, less often ileo-ileal occurs in up 15% and rarely caeco-colic, jejuno-jejunal or even ileo-ileo-colic occur in a double or three-fold manner. Most ileocolic intussusceptions involve the distal ileum, cecum and part of the colon and may rarely prolapsed out the rectum.

Intussusception is related to the mixed or combined type of mechanical obstruction since it is a combination of strangulation (strangulation of the mesentery of the invaginated intestine) and occlusion (closure of the intestinal lumen by the invaginated segment of the intestine). The proximal invaginating intestine is termed the intussusceptum, the distal receiving bowel (outer part) is the intussuscipiens. The cause of intussuseption is most often unknown (idiopatic, nonpathogenic lead point) and is best though to be viral in origin. Several viral gastrointestinal pathogens (rotavirus, adenovirus, etc) may cause hypertrophy of the Peyer's patches of the terminal ileum which may potentiate bowel intussusception. Although only proven to be causative factor in 20% of cases, the intussusceptions often follows a viral illness (upper respiratory, gastrointestinal) which produces an enlargement of the distal ileal lymphoid tissues.

A small percentage of intussusceptions are caused by PLPs (Pathologic Lead Point), the most common of which are a Meckel's diverticulum .Other anatomic leads points include polyps, ectopic pancreatic or gastric rests, lymphoma, lymphosarcoma, enterogenic cyst, hamartomas (i.e., Peutz-Jeghers syndrome), submucosal hematomas (i.e., Henoch- Schonlein purpura),inverted

appendiceal stumps , foreign bodies, and anastomotic suture lines.These anatomic lead points tend to increase in proportion to age, espetially after 2 years of age. PLPs occur in 4 % to 8% of intussusceptions and more commonly found in intussusceptions that are not ileocolic. Ileocolic intussusceptions most often (95%) occur in children from 6 months to 12 months of age.

Ileocolic intussusceptions usually present with complete small bowel obstruction. The mesentery of the invaginated intussusceptum is compressed between the layers of the intussusceptum. There is almost immediate venous compression and stasis. This sets up a vicious cycle of more swelling and venous obstruction, which produce an outpouring of mucus and blood from the engorged intussusceptum causing the classic "currant jelly" stools. The arterial inflow continues until the tissue congestion and pressure finally exceed the arterial pressure producing gangrenous changes leading to necrosis.

Clinical presentation. The classic presentation of intussusception is a toddler with intermittent, crampy abdominal pain associated with "currant jelly" stools and palpable mass on physical examination. The principle signs are:

1 - vomiting (85%),

2- abdominal pain (83%),

3 - passage of blood or bloody mucous per rectum (53%),

4 - a palpable abdominal mass,

5 - lethargy.

The classic triad of pain, vomiting, and bloody mucous stools ("red currant jelly") is present in only one third of infants with intussusception. Suspect the diagnosis in any infant or child with a history of episodic paroxysmal abdominal pain with straining, crying, drawing up of knees and inconsolability. The abdominal pain is frequently acute in onset, severe, and intermittent. The pain is sudden in onset in a child who was previously comfortable. During episodes of pain the infant will often draw his/her knees up to the abdomen, scream inconsolably, and become pale and diaphoretic. Between pain' episodes, which may last only briefly, the child may be quiet, appear well and may fall asleep. With time, the child may become more ill and appear lethargic with increasing abdominal distention, vomiting and progression to shock with cardio vascular collapse.

Examining the child, the right lower quadrant seems empty but a tender mass ("sausage - shaped", "olive") – mostly in the right upper quadrant – can be felt in about 85% of cases. The sign of Dance is the absence of cecum in the right iliac fossa during palpation.

The longer the duration of symptoms and signs before the diagnosis is made, the lower reduction rate.

If the diagnosis is suspected by history and physical examination, several radiographic studies can confirm the diagnosis. Early in the course of illness, abdominal plain x-ray may show a normal or nonspecific bowel gas pattern. Later, abdominal films will show a more obvious pattern of small bowel obstruction with a relative absence of gas in colon. However, plain films have limited value in confirming the diagnosis and cannot be used as the sole diagnostic test.

Ultrasonography of the abdomen is reliable means to identify intussusception. Two ultrasonographic signs of intussusception are:

1.the "doughnut" or "target" sign on transverse views, and

2. the "pseudokidney" sign on longitudinal views.

Absence of color flow in color Doppler suggests compromise of the mesenteric vasculature, indicating a difficult reduction and a lower reduction rate.

Barium or air contrast enema is the "gold-standard" diagnostic study for infants with suspected intussusceptions. It is both diagnostic and therapeutic in identifying and reducing intussusception. In comparison to barium enema, air reduction is reported to be quicker, less messy, decreases time of irradiation and shows a higher reduction rate (75 vs.90%).

Treatment options.

Once a presumptive diagnosis of intussusceptions is made, the child should have

1. An intravenous line placed for rehydration (20 cc/kg).

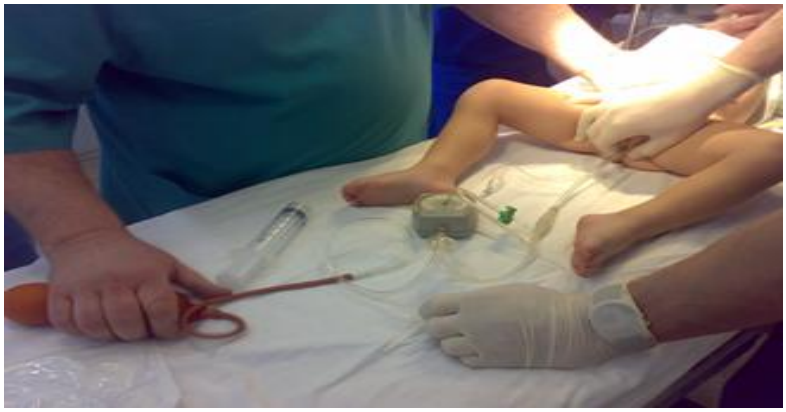
2. A nasogastric tube placed for decompression.

3. Intravenous antibiotics started. A complete blood count (CBC), chemistry panel, and type and screen are obtained.

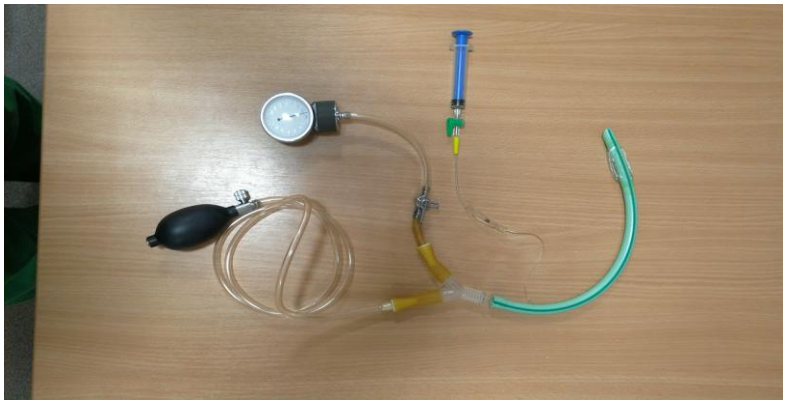
The treatment options for a pediatric patient with an intussusceptions may conservative or surgical: radiologic reduction (hydrostatic - fluid, pneumatic - air), or laparotomy/laparoscory with operative reduction or resection.

Pneumatic (air) enema is quicker and safer than hydrostatic reduction with a barium enema, less messy, easier to perform, delivers less radiation< has an equally high reduction rate, and is more comphortable for the pediatric patient. In air reduction there no risk of barium peritonitis. Non-surgical treatment should not be undertaken in patient with clinical signs of shock, peritonitis, and after 24 hours from beginning of clinical manifestation or 12 hours after rectal bleeding.

The technique of pneumatic reduction (Fig 1.3). The infant lies prone and should be on sedation. A Foley catheter as big as possible is inserted into the ampulla of the rectum. The buttocks are squeezed together and taped to prevent leakage. Under fluoroscopic guidance, pressure within the distal bowel pushes the intussuscipiens proximally. Pressure used for reduction should not exceed 80-110 mm Hg using air (Fig 1.4). Complete reduction is confirmed only when air freely fills more proximal loops of bowel with reflux into the terminal ileum. The clinical criteria of its effectiveness are disappearance of Dance's symptom, phenomenon of "bang", decreasing of pressure till zero, passage of gases into small bowel, belching with air or air coming out of the nasogastric tube. A maximum of three attempts should be made.



(Fig 1.3) Reduction of intussusceptions.



(Fig 1.4) Devise for pneumoirrigography and pneumatic reduction.

If there is a perforation during the air contrast enema, the child may go into extremis due to the high pressure in the peritoneal cavity. If this is the case the abdomen should be rapidly decompressed with a large angiocatheter placed into the RLQ (right lower quadrant). This may need to be done quickly as the rapid and high pressure pneumoperitoneum can cause rapid respiratory embarrassment and abdominal compartment syndrome.

Clinically the infant must show an overall complete improvement and lethargy should disappear in the following routine observation for 24-48 h. Oral diet is resumed on the next morning. If the intussusceptions cannot be completely reduced, operative intervention is indicated.

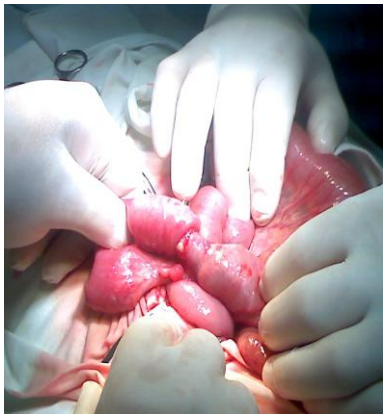
A.  B. 

Fig. 1.5 Laparotomy for intussusceptions (arrow). A-ileoileal, B-ileocolic.

Surgery is indicated in children with: clinical evidence of dead bowel, peritonitis, septicemia, evidence of an anatomic/pathologic lead point, failed enema reduction, hemodynamic instability, perforation, shock, age >3 years, duration of symptoms >24 hours.

Preoperative preparation includes administration of broad-spectrum antibiotics, intravenous fluid resuscitation, monitoring of body temperature and oxygen and placement of a nasogastric tube for decompression.

Surgical exploration for intussusceptions is performed through a right lower quadrant transverse incision. Cloudy or sanguineous fluid raises the suspition that a perforation or necrosis is going to be found. Annual reduction has to be performed very carefully and slowly. Retrograde pressure is applied by sqeezing the intussusceptum within the intussuscipiens in a proximal direction. No “pulling” attempts should be made at the ileal end. Following successful reduction, it is important to assess bowel viability and search for anatomic lead points. Local or segment resection is indicated if:

1. The intussception cannot be reduced ,
2. The segment of bowel appears infracted or nonviable, or
3. A lead point is identified.

The reduced bowel wall is always oedematous with a non-shiny serosa, but may be discoloured or even blue or black. As a test of viability, administering moist and warm wraps may serve in order to check whether there is a regular color coming back after 20 minutes of waiting. Appendectomy is usually performed.

Indications for resection include irreducible intussusceptions, gangrenous bowel or perforatin of the bowel. Primary end-to-end intestinal anastomosis can usually be performed with minimal morbidity. No drainage is used.

Success with air insufflation for reduction is 95%. The recurrence rate of intussception after successful reduction (whether hydrostatic or surgical) is about 5-7%. Recurrence may be slightly lower with reduction using air insufflations. The mortality rate of intussusceptions is less than 1%. Mortality increases with delay in diagnosis, inadequate fluid resuscitation, perforation, and surgical complications.

Laparoscopic approach.

Initially, laparoscopy was only used to confirm either a successful radiologic reduction or the correct diagnosis of intussusception before reduction per laparotomy. Recent studies have reported successful laparoscopic reduction of intussusception in more than 60% of patients.

Most described laparoscopic techniques use three ports (one on the umbilicus and two on the left side of the abdomen). Using atraumatic graspers, gentle pressure is applied distal to the intussusceptum (Fig. 1.6).

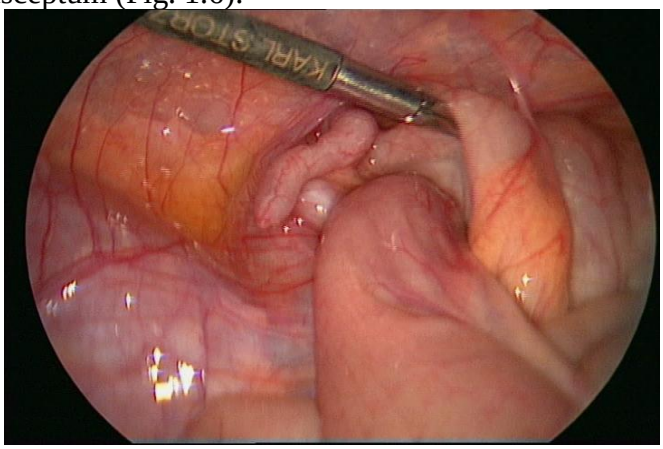


Fig. 1.6. Laparoscopic approach: intussusception.

In difference to the open method, traction must often be applied proximal to the intussuscipiens to succeed with reduction. After reduction, the bowel must be carefully inspected for injury, necrosis, or perforation. Particular attention must be paid to search for a pathologic lead point because most tactile cues are lost.

Recurrent intusssusception has been described in 2% to 20% of cases (average about 5%), with about one third occurring within 24 hours and the majority within 6 months of the initial episode. Recurrences usually have no defined lead point, and they are less likely to occur after surgical reduction or resection. Multiple recurrences can occur in the same patient. Success rates with enema reduction after one recurrence are comparable to those with the first episode and are better if the child did not previously require operative reduction. An overriding concern in recurrent intussusceptions is occult malignancy, although multiple recurrences are not a contraindication to attempted radiologic reduction. Unfortunately the clinical findings or pattern of recurrence do not predict the presence of a pathologic lead point. A careful imaging search is mandatory, and ultrasonography has been recommended as the imaging study of choice.

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Tests for initial level of knowledge

1. What is the most common position of the appendix?
   A. Retrocecal
   B. McBurney point
   C. Pelvic/Descending
   D. Subcecal
   E. Preileal
2. Which of the following is the least common position of appendix?
   A. Retroileal
   B. Retroceacal
   C. Postileal
   D. Pelvic
3. Most common malignancy of appendix is?
   A. Carcinoid tumor
   B. Adenocarcinoma
   C. Squmaous cell carcinoma
   D. Mixed cellularity
4. All are features of acute appendicitis on ultrasound examination except?
   A. A compressible blind ending tube
   B. Diameter of more than 7 mm
   C. Loss of submucosal echogenicity
   D. All are true
5. A child of 11 months of age, was reffered to the surgical department for the third time with the diagnosis of acute intestinal intussusceptions. In the previous cases conservative disinvagination was performed. Which of the causes is the most probable to lead to the recurrent intestinal intussusception?
   A. Irregular introduction of natural feeding.
   B. Disbalanced age nutrition diet.
   C. Gastroenterocolitis.
   D. Disbalansed nutritional regimen.
   E. The anatomic lead point.
6. A child of 6 months of age, in 12 hours after the disease onset there was clinically and roentgenologically diagnosed intussusception. What is the most probable treatment tactics?
   A. Conservative treatment of intussusception.
   B. Planned operation.
   C. Urgent operation.
   D. Pneumoirrigography and pneumatic reduction.
   E. Syphon enema, surveillance.

7. A child of 7 months of age was reffered to a surgical unit 16 hours after disease onset which was sudden. The child became restless, drawed up his knees, refused feeding. An attack of agitation was short, then the boy calmed and fell asleep. He woke up in 20-25 minutes, there appeared vomiting and repeated expressed agitation. The child is pale, adynamic. The diapers are covered with dark-red discharge. What is the tentative diagnosis?

A. Helminth intestinal obstruction.
B. Enterocolitis.
C. Meckel's diverticulum.
D. Intussusception.
E. Tumor of the abdominal cavity.

8. A child of 6 months of age was reffered to a hospital in 6 hours after the disease onset, pale, has attacks of agitation with repeated vomiting. The last defecation occurred 4 hours ago. The child was first in his life fed with "Semolina". At examination: the child is pale, agitated, tachycardia; the forehead is covered with cold sweat. The abdomen isn't swollen, soft, it isn't possible to find the caecum in its usual position, at rectal examination: "raspberry-jelly" stools. What is the diagnosis?

A. Intussception.
B. Dysentery.
C. Gastric ulcer.
D. Anal fissure.
E. Rectal bleeding polyp.

9. A child of 5 months of age first had attacks of abdominal pains, multiple vomiting, flatulence and delayed defecation. At examination: the abdomen is soft; there is a little tender and movable malformation in the right subcostal area. At rectal examination: "red currant jelly" stools on doctor's finger. What diagnosis is the most probable?

A. Intussception.
B. Abdominal tumor.
C. Polyposis.
D. Peritonitis.
E. Intestinal tumors.

10. The factors which contribute for intussusceptions development aren't:

A. Meckel's diverticulum;
B. Disordered nutritional regimen;
C. Insuffitiency of ileocaecal valve;
D. Intestinal polyp;
E. Disordered electrolyte metabolism.

Keys for tests:

1. B. McBurney's point, the junction of the lateral and middle thirds of the line that joins the right anterior superior iliac spine to the umbilicus, is used as a surface marking for the base of the appendix.
2. B
3. B. It was previously believed that carcinoid tumors are the most common tumors of the appendix (Sabiston 17th) but now data from SEER (Surveillane, epidimology and End results) show that mucinous adenocarcinomas are more common
4. D
5. E
6. D
7. D
8. A
9. A
10. E

Tests for final level of knowledge

1. You see a 7 years old girl with her mother during a GP surgery. She has had abdominal pain for one day. The pain is generalised and relieved by rest. The girl walks into the surgery but is clearly in some discomfort. Her temperature is 38°C and her abdomen is tender in the right iliac fossa, with rebound tenderness. You perform a urine dip test that is normal except for the presence of blood. What should you do next?

A. Arrange to admit her to hospital urgently
B. Reassure her and her mother that it is most likely a viral gastroenteritis and review her again if she deteriorates
C. Give a course of broad spectrum antibiotics and ask them to return in three days for further assessment
D. Refer her for assessment of haematuria and give antibiotics for pyelonephritis in the interim period

2. You are called to the emergency department to see a 9 years old girl who has developed abdominal pain and vomiting. She is lying flat and looks pale. On examination there is tenderness in the right iliac fossa, and you think she may have appendicitis. Which of the following signs is NOT suggestive of appendicitis?

A. A palpable mass in the right iliac fossa
B. Rebound tenderness
C. Guarding
D. Microscopic haematuria
E. An olive sized mass in the right hypochondrium

3. You are caring for patients on a ward who have developed vomiting and abdominal distension. You would like to confirm if their symptoms are due to perforation of stomach. Which early simple investigation has the highest sensitivity for detecting the perforation of follow organ?

A. Ultrasound
B. X-ray
C. KT
D. MRI

4. You see a 15 years old girl who has a week history of fatigue, localized abdominal pain in right iliac fossa and subfebrile temperature. She tells you she has also noticed pain shifting from periumbilical to right iliac region. You suspect that she may have appendicular abscess. Which investigation should you order to confirm your suspicion?

A. Ultrasound
B. X-ray
C. Urinalysis
D. EKG

5. Reginald Fitz first described acute and chronic appendicitis in 1886, and it has been recognized as one of the most common causes of some symptom worldwide. What is the symptom?

A. Abdominal pain
B. Vomiting
C. Fever
D. Seizures
E. Hematuria

6. Most appendicitis patients with acute appenditis recover easily with surgical treatment, but complications can occur if treatment is delayed:

A. Gastroenteritis
B. Perforation and peritonitis
C. Colitis
D. Nephritis

7. When the appendiceal inflammation increases, the pain can be localized clearly to one small area, which is:

A. Between the front of the right hip bone and the navel
B. The central portion of the belly
C. The small intestine
D. The left iliac fossa
E. The navel

8. A 8 years old girl hospitalized to surgical department 3 hours after the disease onset with complaints about abdominal pain, temperature 38°C, vomiting. At examination – pain at palpation over all abdominal surface, rigidity of muscles of anterior abdominal cavity, slight mucosal vaginal discharge, blood analysis shows high leucocytosis. What is the diagnosis?

A. Vulvovaginitis.
B. Mesadenitis.
C. Diverticulitis
D. Acute appendicitis.
E. Primary peritonitis.

9.A child of 6 months of age, was brought to the surgical department in 16 hours since the disease onset. The disease developed suddenly. The child became restless, started pulling the legs, refused feeding. An attack of restlessness was short. The boy calmed and fell asleep. He woke up in 20-25 minutes, there appeared vomiting and repeated expressed agitation. The child is pale and adynamic. The diapers are covered with dark-red discharge. What is the tentative diagnosis?

A.Intestinal intussusception.
B.Enterocolitis.
C.Meckel's diverticulum.
D.Helminthic intestinal obstruction.
E.Abdominal tumor.

10.A child of 5 months of age, was brought to a hospital in 6 hours since the disease onset, pale, attacks of agitation, repeated vomiting . Last defecation was 4 hours ago. The child first in his life ate "Semolina". At examination: the child is pale, agitated, tachycardia, the forehead is covered with cold sweat. The abdomen isn't swollen, soft, it isn't possible to find the caecum in right hypogastrium, at rectal examination revealed " raspberry jelly " blood on the surgeon's glove.What is the diagnosis?

A.Intussusception.
B. Dysentery.
C. Gastric ulcer.
D.Anal fissure.
E.Rectal bleeding polyp.

Keys for tests:

1. A;
2. A, E
3. B
4. A
5. A
6. B
7. A
8. E
9. A
10. A

Tasks for final level of knowledge

1. You are called to the emergency department to see a 12 years old boy. He presented with abdominal pain and a two days history of vomiting and anorexia. Based on clinical examination

and laboratory studies (demonstrating a WBC count increasing), your working diagnosis is acute appendicitis.

1. What is your management of this patient?
2. What is the best time of operation in child with acute appendicitis?

Key Answer

1. Appendectomy, preferably laparocopical
2. Timing to surgery has not been shown to greatly affect outcome within the first 24 hours; however, because a risk of rupture theoretically increases over time, an appendectomy should not be delayed if possible.

2. 9 yo girl presents with sever right-sided abdominal pain and vomiting. Pain came on last night and has steadily worsened. Emesis began in morning. ABD exam displays guarding, rebounding and positive Rovsing's signs. WBC 12000, HCT 41, PLT 366000.

1. What is the most likely diagnosis?
2. What should you do next?

Key Answer

1. Acute appendicitis
2. Hospitalization in surgical department

3. You see a 3 years old girl in the emergency department. Her parents have noticed that she has become weakened last three days. Her temperature is 37°C. On examination his abdomen is painful but soft and non-tender with no guarding. You perform the blood and urine tests that are normal. What should you do next?

Key Answer Observe her for the next 24 hours to exclude surgical cause of abdominal pain.

Materials for self-study of the students

Main tasks	Notes (instruction)
Repeat: Anatomy of intestine, appendix vermiformis Abdominal X-ray Pathogenesis of inflammation	To sketch out the anatomy of peritoneal cavity Top represent the methods of diagnosis of acute appendicitis and peritonitis in children
Study: Pathogenesis of appendicitis, primary and secondary peritonitis Features of appendicitis and peritonitis in children The diagnostic possibilities of ultrasound examination, CT, MRI in children	To make differential diagnosis of acute appendicitis and nonsurgical diseases in children To make the indications to surgical treatment in children with abdominal pain To know modern diagnostically methods To know the advantages and disadvantages of classic and laparoscopic appenectomy

THEME 2

GASTROINTESTINAL BLEEDING

Overview

Hemorrhage is a very severe and life-threatening complication in pediatric surgery, which requires urgent treatment. From 5 to 8% of children with gastro-intestinal pathology have gastro-intestinal hemorrhage, and complications of ulcerative disease make up 55% of these patients. It is of great diagnostic importance to determine hemorrhage, and this requires professional knowledge and skills.

Portal hypertension is a syndrome that appears at disordered hemorrhage and increased venous pressure in the portal vein because of congenital and acquired diseases of the liver. In timely diagnosis and treatment of the disease it is possible to expect positive results in most pediatric patients.

1. Aims of the lesson:

The aim of this chapter is to be able to understand under which circumstances gastrointestinal bleeding may occur in children of different ages, know the basic clinical presentations of gastrointestinal bleeding, to be able to understand the mains diagnostic procedures that prove GI bleeding, and to know which type of treatment to assign (conservative or surgical).

1.1.Students must know:

1. The anatomy and physiology of the gastrointestinal tract and its particularities in different age groups.
2. The main causes of gastrointestinal bleeding in children of all age groups.
3. The clinical presentation of gastrointestinal bleeding according to the causes.
4. Classification of diseases that cause GI bleeding.
5. Laboratory and instrumental studies that should be assigned in case of GI bleeding in accordance with the clinical presentation.
6. Differential diagnoses of different diseases that cause GI bleeding.
7. Complications of GI bleeding.
8. Conservative and surgical treatment of GI bleeding and when should either of them be assigned.
9. Preoperative management in children with GI bleeding.
10. Surgical procedures for different diseases that cause GI bleeding.
11. Postoperative complications after surgical treatment of GI bleeding.
12. Postoperative management in children with GI bleeding.
13. Prognoses after GI bleeding.

1.2.Students must be able to:

1. Define an acute gastrointestinal bleeding in children of different age groups according to the clinical presentation.
2. Be able to verify the diagnosis of GI bleeding by laboratory studies and localize the bleeding by instrumental studies.
3. Choose and adequate type of therapy (conservative or operational) for different types of GI bleeding.
4. Describe the patients' pre- operative care.
5. Carry out the post- operative therapy measures.

Terminology	
Gastrointestinal bleeding	Bleeding occurring from any anatomical part of the gastrointestinal tract.
Post- hemorrhagic anemia	Anemia that occurs due to mild or massive bleeding.
Melena	Black or "tarry" feces associated with gastrointestinal bleeding from the upper GI tract.
"Coffee ground" vomitus bleeding	Vomiting blood coming from the gaster.

Esophageal and gastric varicose veins	Enlarged submucosal varicose veins of the esophagus and the stomach that occurs as a severe complication of portal hypertension.
Portal hypertension	Elevation of the pressure in the system of the portal vein more than 10-12 mm of Hg.
Splenomegaly	The enlargement of the spleen.
Hypersplenism	The hyper- function of the sleep that results in the decrease of the WBC, platelets, and RBC, hemoglobin.
Gastro-esophageal reflux	A functional condition at which the content of the stomach enters into the esophagus.
Esophagitis	The inflammation of mucosa of the esophagus.
Peptic ulcer of the stomach	An erosion in the mucosa of the gaster.
Polyps of the GI tract	Abnormal growth of tissue projecting from a mucosa membrane.
Peutz-Jeghers Syndrome (hereditary intestinal polyposis syndrome)	Autosomal dominant genetic disease characterized by development of benign polyps of the intestine and hyperpigmentation of lips and oral mucosa.
Enterocolitis	Inflammation of the digestive tract involving the small intestine.
Intussisseption	A condition at which one part of the intestine is introduced into another.
Meckel's diverticulum	The remnant of the omphalomesenteric duct, that results in a protrusion of the intestine, extending directly from the antimesenteric border of the ileum.
Colitis	Inflammation of the digestive tract involving the large intestine.
Anal fissure	Torsions of the skin around the anus.

2. Basic knowledge and skills necessary for the topic (interdisciplinary integration)

№	Subjects	Skills
1.	Anatomy	To describe anatomy of the abdominal cavity. To appraise peculiarities of possible kinds of the abdominal cavity organs' anatomy.
2.	Histology	To know histological picture of organs of the abdominal cavity. To define peculiarities of histological picture of different parts of gastro-intestinal tract in children of different age groups.
3.	Biochemistry	To demonstrate laboratory methods of diagnostics of child with gastro-intestinal hemorrhage. To appraise data of clinical and biochemical analyses: glucose in blood and urine; protein in blood and urine serum; microelements, hepatic data.
4.	Physiology	To describe physiology of gastro-intestinal tract. To define peculiarities of digestive system in children of different age groups.

5.	Physiopathology	To describe pathological changes at gastro-intestinal bleeding diseases. To define key moments of etiology and pathogenesis at gastro-intestinal diseases complicated with hemorrhage.
6.	Morbid anatomy	To identify pathological anatomical changes in gastro-intestinal diseases complicated with hemorrhage. To define peculiarities of pathological anatomical changes in gastro-intestinal hemorrhage.
7.	Operative surgery	To make a scheme of operative intervention in children. To define peculiarities of topographic anatomy of the abdominal cavity in children of different age groups, discuss operative incision and intervention depending on pathology and age of the child.
8.	Propaedeutics of pediatric diseases	To master the methodic of children with gastro-intestinal hemorrhage examination. To interpret clinical and laboratory data, key symptoms of inflammatory diseases of abdominal cavity organs with hemorrhage.
9.	Infectious diseases	To compare symptoms of infectious diseases for differential diagnosis with abdominal diseases characterized by hemorrhage. To perform differential diagnosis of inflammatory diseases with surgical pathology of the abdominal cavity at abdominal hemorrhage.
10.	Roentgenology, Ultrasound Diagnosis, Blood Pressure, MRI-scan	To interpret data of roentgenologic examination. To appraise data of radiological diagnostics, main roentgenological symptoms. To appraise data of ultrasound diagnostics, blood pressure, MRI-scan, depending on character of pathology and age of the child.
11.	Pharmacology, clinical pharmacology.	To demonstrate peculiarities of administration of medications in children. To determine dosage of medications depending on pathology, child age, peculiarities of treatment of children with acute surgical pathology and gastro-intestinal hemorrhage.

3.1. The list of key terms and characteristics a student must know:

Term	Definition
1.Ulcerative disease.	Presence of gastric or duodenal ulcer.
2. Gastro-intestinal hemorrhage.	Presence of blood in vomiting, feces.
3. Fibrogastroscopy.	Examination of upper part of the digestive system.
4. Fibrocolonoscopy.	Examination of the colon using endoscopic equipment.
5. Proctoscopy.	Examination of the rectum.
6. Portal hypertension.	Syndrome caused by disordered blood circulation and increased pressure in portal vein.
7. Blakemore tube.	Equipment used to stop hemorrhage from dilated veins of the esophagus.
8. Angiography.	X-ray contrast examination of vessels.
9. Polyps.	Malignant and benign malformations.
10.Hemorrhoids.	Varicose dilation of the veins of anal canal and lower department of the rectum.
11. Anal fissures.	Defects of anorectal canal mucosa.

CONTENT

All of GI bleeding can be divided into bleeding from the upper and lower GI tract. By severity of bleeding, they are divided into small, mild, and massive bleeding.

When bleeding occurs from the upper GI tract (gaster, duodenum), it presents with

vomiting of blood - hematemesis, the color of «ground coffee». The reason for such a color of the vomitus masse is the result of a chemical reaction at which blood mixes with the stomach and duodenal acid and hydrochloric acid hematin is formed. It has the color of ground coffee. When bleeding occurs from the esophagus the color of the blood is regular. Blood vomiting occurs when bleeding is massive or mild, because of reflux stretching of the stomach. In cases when blood vomiting doesn't occur, bleeding from the upper parts of the GI tract presents with melena. Melena is black or "tarry" feces associated with gastrointestinal bleeding.

If bleeding is localized in the small intestine, it will present with bloody feces the color of «ripe cherry» or currant jelly stool. When bleeding occurs from the bowl it usually presents with hematochezia- the passage of blood through the anus in or with stool. It presents with streaks of blood in the stool.

EVALUATION OF GI BLEEDING

There are several ways for diagnosing and differentiating upper and lower GI bleeding. For urgent differentiation placement of a nasogastric (NG) tube is recommended. Gastric lavage can be used to help determine if there is ongoing upper GI bleeding. Return of "coffee ground" color material or bright red blood through the nasogastric tube is a sign that bleeding is proximal to the ligament of Treitz and is coming from the upper GI tract. Return of clear contents does not completely rule out upper source of GI bleeding, it signifies that bleeding is distal. Examination of the infant's or young child's diaper may show classic currant jelly stool seen with intussusception, or melena which can be classified as upper GI bleeding. A rectal examination should also be performed for the presence of blood. The best instrumental diagnostic procedure to determine the source of upper GI bleeding is an upper endoscopy (esophago- gastro-duodenoscopy). In cases when currant jelly stool is present an ultrasound (US) should be performed to detect the presence or absence of intussusception. US is the method of choice for detection of intussusception.

For children who are suspected with lower GI bleeding should also have their stool examined along with examination of the anus (to rule out anal fissure) before proceeding with instrumental diagnosing. The most informative examination is colonoscopy. This procedure detects the source of bleeding and offers endoscopic treatment of pathologic lesions. If colonoscopy is entirely normal, the bleeding source is presumed to be localized in the small intestine. In these cases, patient may be a candidate for push endoscopy. This endoscopic technique allows visualization of the distal duodenum and proximal 3 to 5 feet of jejunum. Lesions that can be identified and sometimes treated by these endoscopic techniques include, polyps, and acute ulcers. Identification of the source of bleeding is extremely important even if bleeding cannot be controlled, because location of the bleeding site helps determine the possible surgical approach.

In pediatric surgery there is a frequency pattern focusing on the location of bleeding and different age group (table 1).

GI BLEEDING					
UPPER GI BLEEDING			LOWER GI BLEEDING		
AGE	COMON CONDITION	RARE CONDITION	AGE	COMON CONDITION	RARE CONDITION
Newborn (< 1 mo)	swallowed maternal blood - HDN	- coagulopathy	Newborn (< 1 mo)	- anal fissure - enterocolitis	- intussusception - Meckel diverticulum
Infant (1 mo- 2 yr)	- gastritis - esophagitis	- ulcers	Infant (1 mo- 2 yr)	- anal fissure - Meckel diverticulum	- enterocolitis

				- intussusception	
Children (2-12 yr)	- esophagitis - esophageal /gastric varices	- ulcers	Children (2-12 yr)	- polyps	- intussusception - enterocolitis
Adolescents (> 12yr)	- esophageal / gastric varices - esophagitis / gastritis - ulcers (stress attained)	- Mallory-Weiss tear - NSAIDs	Adolescents (> 12yr)	- polyps - infectious colitis - inflammatory bowl disease	- intussusception - enterocolitis

SOURCES OF UPPER GI BLEEDING

Hemorrhagic disease of newborns (HDN)

Hemorrhagic disease of the newborn (HDN) is a disorder that results from a lack of vitamin K. Infants that have increased risk for the development of this disease include premature infants, , those with fat malabsorption, and breast-feeding infants. Newborn sepsis, as well as metabolic or ischemic injury to the liver, may also cause a deficiency in clotting factors resulting in GI bleeding. Current recommendations for all infants are the administration of 1 mg intramuscular vitamin K at birth with intravenous administration if necessary. In the differential diagnosis one must consider that the infant may have swallowed maternal blood during delivery or during breast feeding. That is why the mothers nipples should also be examined for fissures.

Portal Hypertension

The management of children with portal hypertension has evolved significantly over the past two decades. Improved survival in these patients has resulted from: (a) progress in the pharmacologic control of acute portal hypertensive hemorrhage; (b) improved efficacy and safety of endoscopic methods to treat acute esophageal variceal hemorrhage, which also reduce the risk of rebleeding; (c) recognition of the role for advanced surgical therapy (portocaval shunts); and (d) improved outcomes following pediatric liver transplantation as a definitive treatment for children with end stage liver disease or life-threatening complications of portal hypertension.

Portal hypertension is defined as an elevation of the portal pressure above 10–12 mmHg. In healthy children, portal pressure rarely exceeds 7 mmHg. Elevation of the portal pressure is most commonly secondary to obstruction of portal venous flow due to pre-hepatic, intra-hepatic, or post-hepatic block, although increased splanchnic blood flow might contribute in some cases. Increased pressure within the portal circulation leads to the formation of collateral circulatory pathways connecting the high-pressure portal vasculature to the low-pressure systemic venous system.

The most common and potentially dangerous communications occur within the esophageal wall, connecting the coronary and short gastric veins to the esophageal venous plexus. Esophageal varices developing within this plexus become the site with the highest risk for massive gastrointestinal hemorrhage. Less threatening collateral communications can develop between the recanalized umbilical vein and abdominal wall systemic veins (caput medusa), the inferior rectal veins (hemorrhoids), and in the retroperitoneum. In addition, any surgically created interface between the portal and systemic venous circulations, such as occurs with intestinal stomas, is a potential site of often problematic variceal development. Spontaneous natural splenorenal shunts sometimes develop in the form of favorable collateral vessels within the tissues surrounding the pancreas, duodenum, and left kidney. The progressive development of porto-systemic collaterals

has the beneficial effect of decreasing portal pressure. However, this benefit is countered by the concurrent development of a hyperdynamic circulatory state. Portal hypertension has been associated with the presence of autonomic nervous system dysfunction, as well as an excess of circulating cytokines leading to tachycardia, decreased systemic and splanchnic vascular resistance secondary to vasodilatation, plasma volume expansion, increased cardiac output, and increased portal inflow.

Each of the causes of elevated portal pressure shares the common mechanism of increased resistance to blood flow from the splanchnic portal circulation to the right atrium. In children, the location of this increased vascular resistance can be: (a) pre-hepatic, usually within the portal vein and its primary feeding branches; (b) intra-hepatic, most commonly related to intrinsic liver disease, but may be secondary to pre-sinusoidal obstruction (congenital hepatic fibrosis or schistosomiasis); or, (c) post-hepatic, secondary to hepatic vein outflow obstruction (Tabl. 1).

Tabl. 1. Pediatric diseases associated with portal hypertension

Forms Portal Hypertension	**Portal Hypertension Causes**
Pre-hepatic form PH	Extrahepatic portal vein thrombosis Cavernous transformation of the portal vein Splenic vein thrombosis Congenital portal vein malformation Extrinsic portal vein compression
Intra-hepatic form PH	Hepatocellular disease Autoimmune hepatitis Hepatitis B, C Wilson's disease Alpha-1-antitrypsin deficiency Glycogen storage disease – type IV Toxins and Drugs Histiocytosis X Gaucher's disease Biliary tract disease Biliary atresia Cystic fibrosis Intrahepatic cholestasis syndromes Sclerosing cholangitis Congenital hepatic fibrosis Schistosomiasis Sinusoidal veno-occlusive disease
Post-hepatic causes PH	Budd-Chiari syndrome Inferior vena cava obstructions Chronic congestive heart failure Veno-occlusive disease Postoperative hepatic vein stenosis Prothrombotic disease

The primary factor influencing the prognosis and treatment is the intrinsic functional status of the liver. Pre-hepatic obstruction does not result in impaired hepatic synthetic function, and, therefore, coagulopathy is absent. Treatment should be directed toward the prevention of hemorrhage through palliative interventional procedures while spontaneous collateral venous channels develop. However, in some children who are at higher risk of re-bleeding, portal decompression via surgical shunting may be necessary. In patients with intrinsic liver disease, routine endoscopic surveillance, selective surgical shunting, transjugular intrahepatic portosystemic shunting, and liver transplantation each play a role, depending on the degree of

cirrhosis and extent of hepatic synthetic dysfunction. Post-hepatic obstruction is characterized by hepatic synthetic compromise, coagulopathy, and progressive hepatic failure. Although interventions to prevent or treat potentially fatal complications might be necessary, definitive correction with liver transplantation is often required.

Pre-Hepatic Portal Hypertension

The most common type of pre-hepatic obstruction is extrahepatic portal vein obstruction (EPVO) in children (75-95%). Risk factors for EPVO include umbilical vein infection in infants, and severe intra-abdominal infections (perforated appendicitis, primary peritonitis, inflammatory bowel disease) in older children. It can also develop secondary to primary biliary tract infection or cholangitis, or in the setting of inherited disorders associated with hypercoagulability, such as factor V Leiden mutation, or protein C, protein S, or antithrombin III deficiency. Polycythemia in infancy or umbilical vein catheterization can all lead to secondary thrombosis, especially when accompanied by neonatal dehydration or systemic infection and phlebitis. Embryological malformations resulting in tortuous, poorly developed portal veins, webs, or diaphragms can also be a primary cause for EPVO or predispose to an increased risk of thrombosis. The development of periportal collateral vessels ("cavernous transformation of the portal vein") may result from either a disordered embryologic process or from long-standing portal vein thrombosis. Children with EPVO frequently appear completely healthy prior to an episode of hematemesis or hematochezia. Because their hepatic synthetic function is normal, they are able to recover from their variceal hemorrhage more readily than children with pre-existing liver disease.

Intra-Hepatic Portal Hypertension

Portal hypertension in children is commonly related to progressive hepatocellular injury and fibrosis in the setting of intrinsic liver disease, broadly characterized as hepatocellular disorders and biliary tract disorders (Table 1). The common final pathway of increased intra-hepatic vascular resistance due to hepatic fibrosis and alterations in hepatic microcirculation is the basis for the development of portal hypertension and its associated complications in cirrhosis. The etiologies of chronic liver disease in children are myriad and include recognized disorders such as extrahepatic biliary atresia, metabolic liver diseases such as alpha-1-antitrypsin deficiency, Wilson's disease, glycogen storage disease type IV, and cystic fibrosis. When hepatic synthetic failure is not present or only slowly progressive, direct treatment of portal hypertension or its complications is indicated. However, because many of these conditions are associated with progressive liver failure, the primary treatment in most cases is liver transplantation.

Post-Hepatic Portal Hypertension

Post-hepatic portal hypertension is caused by obstruction of hepatic venous outflow. Hepatic vein obstruction (Budd- Chiari syndrome) can occur secondary to obstruction of the hepatic veins at any point from the sinusoids to the entry of the hepatic veins into the right atrium/inferior vena cava. Veno-occlusive disease, microvascular non-thrombotic occlusion of hepatic venules, also has emerged as one of the most frequent causes of hepatic vein obstruction in children. Most cases occur after total body irradiation, with or without cytotoxic drug therapy, associated with bone marrow transplantation. This condition also occurs after the ingestion of herbal remedies containing the pyrrolizidine alkaloids, which are sometimes taken as medicinal teas.

Diagnosis

Clinical history and physical examination should concentrate on identifying factors that predispose to the development of cirrhosis, including a family history of inherited metabolic disease and possible exposure to viral or toxic pathogens. Clinical examination findings suggesting underlying liver disease (ascites, liver size, nutritional status), hypersplenism (splenomegaly, bruising), or hepatopulmonary syndrome (spider angiomas, clubbing, cyanosis) contribute to diagnostic evaluation and therapeutic planning.

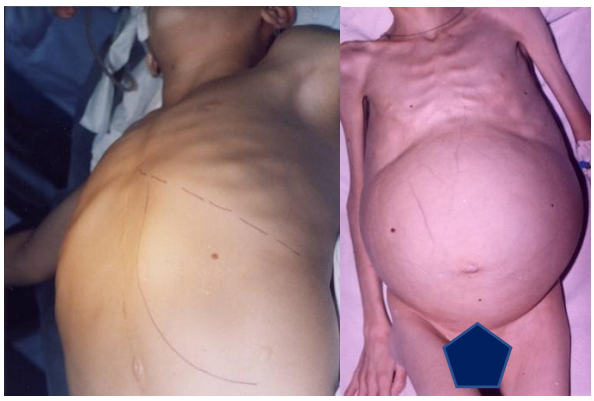


Fig 2.1. Patient with splenomegaly (on the left), patient with ascites (on the right).

Imaging tests are essential to confirm the presence of portal hypertension, define the portal venous anatomy, and formulate options for therapy. Initial screening with ultrasonography can suggest the presence of chronic liver disease and should determine portal venous patency. Doppler examination can demonstrate both the direction of portal venous flow and the degree of hepatopetal flow, which correlates with the risk of variceal hemorrhage. The branches of the portal venous system are examined to exclude splenic vein thrombosis or widespread portal system thrombosis. Magnetic resonance venography or contrast-enhanced computed tomography has replaced mesenteric angiography when further definition of portal anatomy is necessary, such as when liver transplantation or portosystemic shunt procedures are planned. Upper gastrointestinal endoscopy is the most accurate and reliable method for detecting esophageal varices (Fig.2.2) and for detecting the source of acute gastrointestinal hemorrhage. This modality is especially valuable in the presence of acute hemorrhage, where up to one-third of patients with known varices may have bleeding from other sources such as portal hypertensive gastropathy or gastric/duodenal ulcerations. In addition, upper endoscopy can identify features associated with an increased risk for hemorrhage, such as large varices, “cherry-red spots” apparent over varices, representing fragile telangiectasias within the shallow submucosa, and portal hypertensive gastropathy. Endoscopy is also used to intervene therapeutically when acute bleeding varices are identified and when prophylactic endoscopic treatment of varices is warranted.

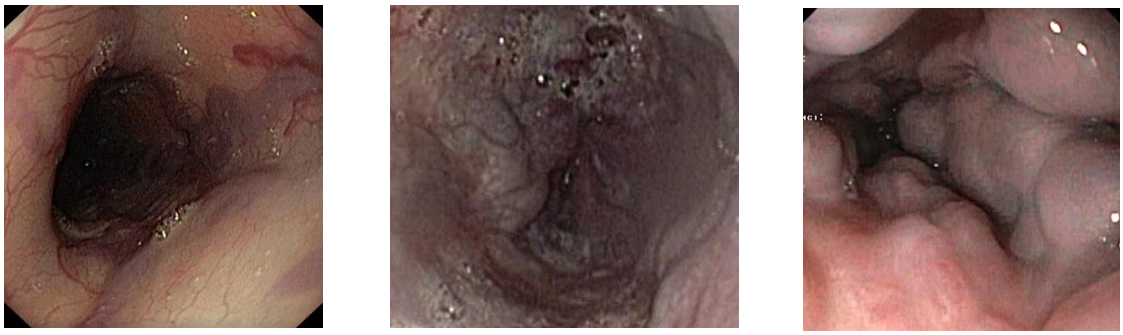


Fig. 2.2. Different stages of esophageal varices

Liver biopsy may be helpful in determining the etiology of intrinsic liver disease and in defining further therapy or the need for transplantation. A percutaneous approach may be utilized, unless significant coagulopathy is present, in which case liver biopsy should be performed using an open/laparoscopic technique under general anesthesia.

Conservative treatment of the portal hypertension in children

The decision to undertake pharmacologic, endoscopic, or surgical treatment for portal hypertension must be based on the natural history of the disease and the possibility of life-threatening complications.

The most common portal hypertensive complication is gastrointestinal bleeding. Regardless of the site and mechanism of bleeding, initial therapy is directed toward fluid resuscitation and, when necessary, blood replacement. A nasogastric tube should be placed to

confirm the upper gastrointestinal source of bleeding and for evacuation of blood from the stomach.

A proton pump inhibitor should be administered to decrease the risk of bleeding from gastric erosions, and antibiotics should be instituted as prophylaxis for bacterial infections or spontaneous bacterial peritonitis. In patients with hepatic synthetic dysfunction and coagulopathy, administration of vitamin K, fresh frozen plasma or cryoprecipitate, and platelets when thrombocytopenia is present also may be necessary.

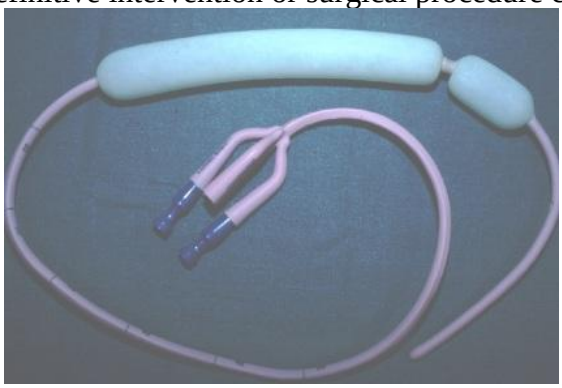
Adequate volume resuscitation is essential; however, volume overload from excessive transfusion or crystalloid administration is counterproductive because this leads to further increase in portal pressure and continued hemorrhage.

Conservative treatment of acute gastrointestinal bleeding.

Somatostatin is a 14-amino acid peptide that reduces splanchnic blood flow by selective mesenteric vascular smooth muscle constriction. Its effects on variceal hemorrhage are similar to those of vasopressin, but it carries a lower risk of adverse systemic side effects. Because the short half-life of somatostatin complicates the management of patients with acute variceal hemorrhage, octreotide, an 8-amino acid synthetic somatostatin analogue, was developed. Octreotide can be administered subcutaneously but is best used as a continuous intravenous infusion (25–50 mg/m2/hour, or 1.0–3.0 mg/kg/hour). In adult studies, both somatostatin and octreotide have achieved excellent results in controlling acute variceal hemorrhage. Recent retrospective studies have demonstrated that octreotide is associated with a high rate of bleeding control in children with portal hypertension without significant adverse events. However, controlled studies in children are necessary to confirm this success. While beta blockers have no role in the treatment of acute variceal hemorrhage, they have been used in an effort to prevent first variceal hemorrhage in high-risk patients, and as an option for secondary prophylaxis. The goal of therapy is a reduction in heart rate by 25%, thereby decreasing cardiac output, portal inflow, and perhaps blocking b-receptor-mediated vasodilatation, allowing unopposed a stimulation within the mesenteric arterioles. Efficacy has been evaluated in two groups: (a) patients with documented varices who underwent beta-blocker treatment in an attempt to prevent the first episode of bleeding (primary prophylaxis); and (b) patients treated following the initial hemorrhage in an attempt to prevent recurrent bleeding (secondary prophylaxis). The efficacy of therapy differs based on the indication.

Mechanical Tamponade

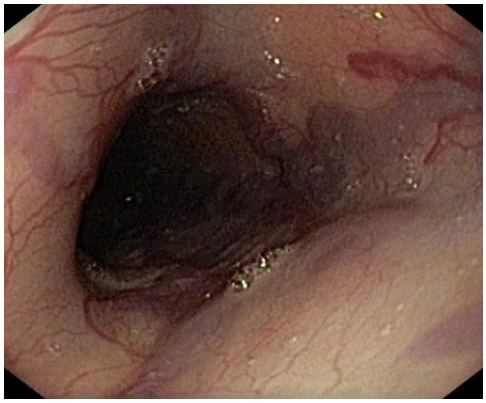
Mechanical tamponade using balloon catheter tubes (Sengstaken-Blakemore or Minnesota tubes) provides mechanical compression of esophageal and gastric fundal varices. Although balloon tamponade is usually successful in stopping refractory hemorrhage, the effect is often transient, and recurrence following removal is common. The significant number of complications and high incidence of rebleeding have limited their use to severe uncontrollable hemorrhage as a temporizing measure until a definitive intervention or surgical procedure can be performed.



Picture 2.3. Sengstaken-Blakemore Tube

Portosystemic Shunt Operations

Numerous surgical procedures have been devised to divert portal blood into the low-pressure systemic venous circulation, thereby decreasing the portal venous pressure. Enthusiasm



for the use of portosystemic shunting in children was limited by early reports suggesting that children less than 8 years of age and those with available vessels less than 8–10 mm would be unsuitable candidates due to the high risk of shunt thrombosis. More recent experience in centers skilled in pediatric vascular reconstruction has established that a high rate of success can be achieved with a minimal complication rate, even in small pediatric patients. In general, portosystemic shunts can be classified into three groups: non-selective shunts, selective shunts, and direct reconstructions of the portal circulation using a vascular graft (Rex shunt).

Non-selective shunts are constructed to communicate with the entire portal venous system and therefore have the potential to divert blood from the normal antegrade perfusion to the liver. H-type mesocaval shunt, which is constructed using a short segment of internal jugular vein to connect the SMV and the IVC Excellent patency (93%) and no significant episodes of encephalopathy support its use in pediatric patients. The limited intra-abdominal dissection required to construct this shunt contributes to its technical ease and if liver transplantation is necessary, the shunt can be easily occluded at that time. Other non-selective shunts have significant disadvantages in children due to the need for splenectomy (proximal splenorenal shunt), or dissection of the main portal vein (end-to-side and side-to-side portocaval shunts), which can compromise subsequent liver transplantation.

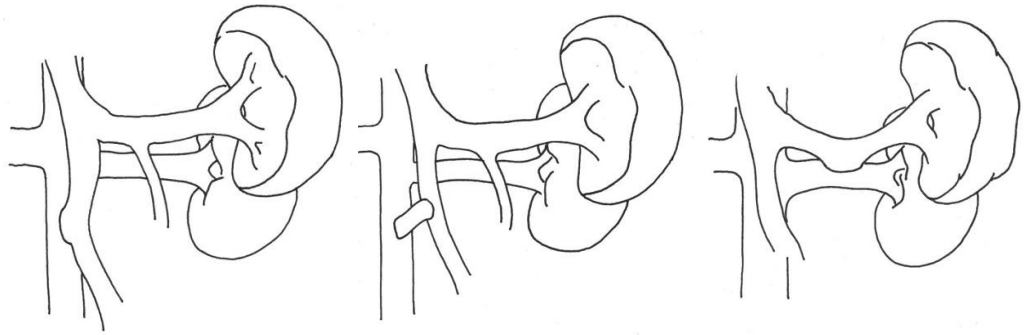
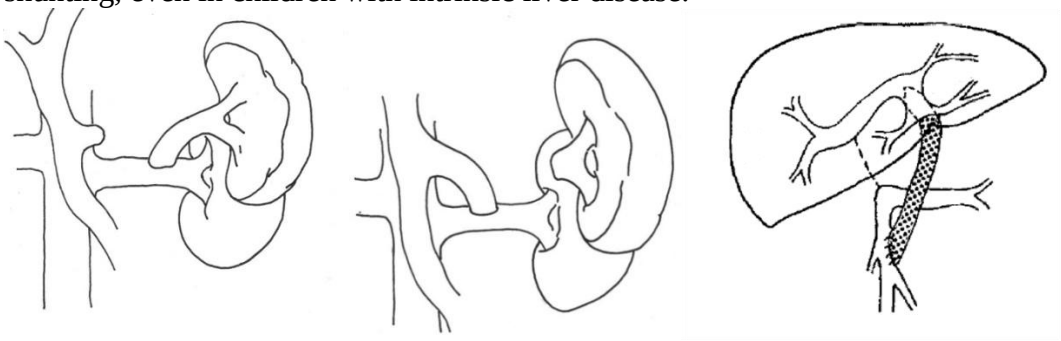


Fig. 2.4. Non-selective shunts (explanation in the text)

Selective shunts are constructed to divert the “gastrosplenic” portion of the portal venous flow into a systemic vein, most frequently the left renal vein or the immediately adjacent IVC. Communication between the “central” mesentericoportal circulation which perfuses the liver and the gastrosplenic portal circulation is severed by dividing the gastroepiploic veins, the coronary vein, and the retroperitoneal pancreatic collateral vessels. The most common and successful selective shunt, the distal splenorenal shunt (DSRS) or Warren shunt, preserves antegrade perfusion to the liver within the mesenteric portion of the portal circulation, while decompressing the esophageal venous plexus through the short gastric veins and splenic vein. We use this shunt as our primary option in children where direct reconstruction of the portal system (Rex shunt) is not possible. When the left adrenal vein is appropriately located and dilated, it serves as an alternative anastomotic site to access the left renal vein. When performed in centers experienced in complex vascular reconstruction of the portal system, as is necessary in pediatric liver

transplantation, shunt patency has ranged from 83 to 100%. When shunt patency is maintained, recurrent variceal bleeding is uncommon, although decompressed varices can sometimes still be identified by upper endoscopy. Encephalopathy is uncommon following successful portosystemic shunting, even in children with intrinsic liver disease.



Picture 2.5. Selective shunts (explanation in the text)

Other causes of upper GI bleeding

Esophagitis – the inflammation of the mucosal layer of the esophagus. It is usually secondary to gastroesophageal reflux. It clinically appears with blood vomiting and pain behind the chest. Medicine treatment (antiacids, thickend feedings, upright positioning and prokinetic agents) is usually effective, and if not a surgical antireflux procedure- fundoplication is performed.

Upper GI bleeding can occur from a variety of sources including gastric, duodenal, and jejunal ulcers. The course of clinical presentation for an ulcer is: recurrent vomiting, refusal of feedings, persistent crying, slow growth, GI hemorrhage («coffee ground» vomitus bleeding). Treatment is usually carried out with endoscopic coagulation and if bleeding control isn't attained then surgery is carried out (closure of the perforation- ulcer).

Rarely the cause of upper GI bleeding may be diffuse mucosal disease, Dieulafoy lesions, and Mallory-Weiss tears.

CAUSES OF LOWER GI BLEEDING

Intussusception

This condition is seen mostly in the age of 3 to 8 months. This may be acquired with the nutritional peculiarities of this age.

Clinical presentation: Intussusception may lead to lower GI bleeding. The presence of currant-jelly stool is seen.

Treatment: Treatment may be conservative or surgical depending on the duration of the disease. If the duration of illness exceeds 24 hours (from the first symptoms) the treatment has to be surgical.

Meckel's Diverticulum

Meckel's diverticulum occurs with a worldwide prevalence of approximately 2-4%. For most patients, this remnant remains clinically silent throughout life. Its presence is not suspected unless complications arise, or it is identified as an incidental finding at autopsy or laparotomy for other abdominal lesions. Males are affected approximately 2-4 times more frequently than females. Meckel's diverticulum is known as the disease of "two's." Two percent of the population is affected, it occurs twice as frequent in males, it is located two feet from the ileocecal valve, it is two inches long and two centimeters wide, and two types of mucosa often exist. Meckel's diverticulum may occur at any age.

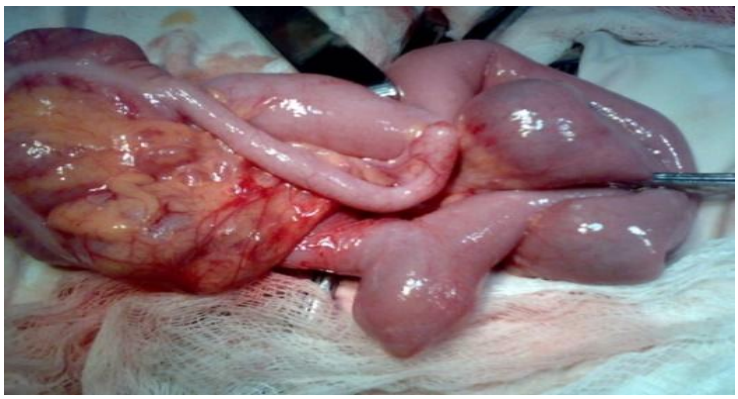


Fig. 2.6. Meckel's Diverticulum

Etiology: During the third week of fetal development, the midgut opens into the yolk sac, remaining temporarily connected by the vitelline duct (or yolk stalk). This duct divides the cranial and caudal portions of the midgut (distal duodenum, jejunum and proximal ileum, distal ileum and cecum, appendix, ascending colon, and proximal two-thirds of the transverse colon, respectively.) As intestinal maturation proceeds, the vitelline duct narrows. By the third month of embryogenesis, the duct disappears as the physiologic intestinal herniation spontaneously reduces. If complete regression fails to occur, an omphalomesenteric duct-related anomaly results. The anomaly that results is solely dependent upon the stage of developmental arrest. A complete diverticulum, the most clinically significant anomaly, extends directly from the antimesenteric border of the ileum. In approximately 75% of patients, the tip remains free and unattached within the abdominal cavity. In the remaining 25%, the Meckel's diverticulum remains attached to the anterior abdominal wall. The connection consists of either a solid fibrous cord (arterial or ductal remnant) or omphalo-ileal fistula extending from the umbilicus to the ileum. Vitelline duct cysts result if a cyst forms in the midportion of the cord. The yolk sac is initially supplied by the paired vitelline arteries. As fetal maturation proceeds, the left vitelline artery disappears, leaving the right vitelline artery to form the superior mesenteric artery. Meckel's diverticulum is commonly supplied from a branch arising from the ileal artery. Infrequently, the vascular supply arises directly from an ileocolic arterial branch or from the mesentery directly. The artery is always an end-artery.

Pathology/Pathophysiology: Meckel's diverticulum is a true diverticulum. It contains all three normal bowel layers: mucosa, submucosa, and muscularis propria. Vitelline duct cells are pluripotential. As a result, 50% of all diverticula contain heterotopic gastric mucosa. If symptomatic, the prevalence is closer to 75%. Heterotopic pancreatic tissue is in the usual antimesenteric position with aberrant tissue in the distal tip responsible for symptoms present in only 5%. Five percent contain both gastric and pancreatic tissue. Heterotopic colonic or enteric mucosa occur infrequently (i.e., 2% each). Many ulcers are nonapparent on inspection and are thus best detected on microscopy. Unlike peptic ulceration of the stomach and duodenum, ileal ulcers from a Meckel's diverticulum are not associated with Helicobacter pylori.

Clinical Presentation: Symptoms are usually absent unless complications occur. The most encountered complications of Meckel's diverticulum are bleeding due to its ulceration caused by hydrochloric acid secreted by heterotopic parietal cells. Bleeding presents with bloody stool of purple color. Bleeding typically occurs in children less than five years of age, with average age of 2 years. Bleeding is characteristically episodic and painless. It recurs frequently and is often severe enough to require multiple blood transfusions for resuscitation. The differential diagnosis includes juvenile polyps, inflammatory bowel disease, peptic ulcer disease of the stomach or duodenum, and blood dyscrasias. Bowel obstruction may occur from intussusception around the diverticulum. The diverticulum may act as the lead point, causing an ileo-ileal intussusception with rapid progression to an ileo-colic presentation. Intussusception is the etiology in nearly 50% of cases. It commonly presents before ten years of age The patient presents with early vomiting, abdominal pain, and often a palpable mass in the abdominal cavity. If untreated necrosis of the gut may occur

just like in cases with general intussusception. Rarely obstruction occurs when a Meckel's diverticulum protrudes through an indirect inguinal hernia (Littre's hernia). Most obstructions due to Meckel's diverticulum are not diagnosed until operation, because it simulates other pathology. Meckel's diverticulum presents similar to acute appendicitis. But it can be differentiated due to other clinical symptoms that appear with appendicitis and aren't present with the diverticulum: elevation of WBC count and the rise of body temperature. If inflammation occurs in the diverticulum it may lead to its' wall perforation causing peritonitis. Infrequently, tumors can develop in Meckel's diverticulum. Carcinoid is the most common tumor, though adenocarcinoma, leiomyoma, lymphoma occur as well.

Diagnosis: In cases when bleeding of the diverticulum occurs an endoscopy may be performed for differential diagnosis. Most of the time it is unable to detect the diverticulum at endoscopy. In such cases an ultrasound of the abdominal cavity is signed. Some times the ultrasound may detect the lesion. In cases when there are signs of obstruction, due to intussusception, an X- ray of the abdominal cavity is performed placing the patient vertical, and an ultrasound is assigned.

Treatment: Treatment of a symptomatic diverticulum requires operative intervention. The method of choice is laparoscopy. In times when laparoscopy can't be performed there may be a conversion. The diverticulum is noted, its' arterial supply is identified and ligated, and the diverticulum is removed. The surgical options include simple diverticulectomy without ileal resection. In cases when obstruction/intussusception are present (and there are signs of necrosis) a wedge resection to include immediately surrounding ileum, or an ileal resection with primary anastomosis. Other conditions require individual judgement.

Outcomes: Upon resection of a Meckel's diverticulum, cure from subsequent bleeding or inflammation is expected. The most common postoperative complication is adhesive small bowel obstruction which occurs in 5-10% of patients.

Polyps of the Gastrointestinal Tract

Intestinal polyps are much less common in children than in adults. The peak age of incidence is between the ages of 3 and 10 years. If they are present, their malignant transformation is less than in adults, and the approach to management is more expectant. Approximately 1% of children have asymptomatic intestinal juvenile polyps, which are benign.

Juvenile polyposis are the most common polyps found in the GI tract. It is an autosomal dominant disorder with polyps in the large and small bowel. Nevertheless these polyps can occur anywhere along the gastrointestinal tract. It is considered a premalignant condition and 6% of these children will eventually develop malignancy. Familial adenomatous polyposis is an autosomal dominant disorder, characterized by hundreds of adenomatous polyps in the rectum and colon causing diarrhea and bleeding. Malignancy may occur in one or more polyps before the age of twenty years.

Although not as common occurs the Peutz-Jeghers Syndrome. This well-known syndrome is associated with the presence of polyps predominantly in the small bowel. They may fill up the whole small bowl, sometimes 2/3 or 1/3 of its length. Its main feature is the pigmented lesions that are observed on the buccal mucosa and lips of these patients. Malignant degeneration can occur, and lifelong surveillance is necessary.

Clinical Presentation: The main two symptoms of polyps of the intestinal tract are rectal bleeding and abdominal pain. Lower intestinal bleeding is the main symptom of most polyps. The blood is usually red, indicating its origin in the lower gastrointestinal tract, and small in quantity, unlike bleeding from duplications or Meckel's diverticulum with peptic ulceration. If bleeding occurs in the result of polyps in the small bowel, the blood will appear darker. In rare cases of duodenal or gastric polyps, rectal bleeding may present with melena. The bleeding is frequently associated with crampy abdominal pain. The pain does not necessarily occur with the bleeding. Intussusception- traction on a polyp may cause intussusception anywhere it occurs. Usually, colocolonic intussusception occurs only when a colonic polyp serves as a lead point. The symptoms include crampy, intermittent pain, bleeding from venous engorgement of the mucosa,

and signs of intestinal obstruction (i.e., vomiting, distention, obstipation). Unlike idiopathic intussusception, intussusception from a polyp occurs in older children.

Diagnosis: The diagnosis of polypoid lesions depends primarily on two modalities: intestinal contrast studies and endoscopy. Endoscopy is advantageous as it can be both diagnostic and therapeutic. Contrast studies for colonic polyps can be very accurate and can be used to follow polyps for changes in number and size. Upper intestinal polyps in the stomach and duodenum are also accurately visualized with contrast studies. Small bowel polyps may be difficult to image even with small bowel enemas. Small bowel polyps are notoriously difficult to diagnose, and thankfully, occur very rarely.

Treatment: The treatment of polyps of the GI tract in children can vary from simple observation of benign lesions to wide excision and chemotherapy for malignant ones. Most are within easy reach of a flexible sigmoidoscope Juvenile polyps should be removed once diagnosed. In cases of Peutz-Jeghers syndrome, when the entire gastrointestinal tract can be affected, excision of all affected areas could easily result in intestinal insufficiency. Endoscopy should be performed to detect polyps which are growing and could be malignant. Large polyps or those causing significant symptoms should be removed. Endoscopic removal should be attempted first. For lesions normally beyond the range of normal endoscopy, a combined endoscopic-surgical approach can be attempted where the surgeon at laparotomy guides the endoscopist through the small bowel for visualization and removal of large lesions. Resection of bowel segments should be performed only for cases where cancer has developed and invaded the submucosa, or in areas where polyps are particularly dense and causing severe symptoms.

Anal fissure

This is a condition that occurs in the result of micro- torsions at the skin to mucose junction of the anus. Clinical it presents with a pain during the act of defecation and a drop of blood on the fecal masses at the end of the defecation act. Such condition is treated conservatively by assigning rectal suppositories and ointments. Rarely this disorder needs surgical treatment.

Other causes

Enterocolitis may be the cause of lower GI bleeding in neonates that are premature or have some form of neonatal distress. Clinically such condition presents with blood noted in the stool. Enterocolitis produces a darker, often maroon or purple stool with varying amounts of mucus. Radiography confirms the diagnosis. 70- 80% of patients recover from conservative treatment, including: antibiotics, bowel rest, total parental nutrition. If there are signs of peritonitis, then the neonates require surgical treatment.

Infectious colitis and inflammatory bowel disease may also be associated with GI bleeding. Clinical features may be abdominal pain and bloody veins in the stool. The use of gastroduodenoscopy and colonoscopy identify most causes of bleeding. However undiagnosed bleeding requires special studies such as small bowel anema arteriography or tagged red cell scanning. Sometimes when conservative diagnostic methods don't clear the diagnoses surgical diagnostic methods can be used, such as a diagnostic laparoscopy.

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Tests for initial level of knowledge

1. What disorder or disease may lead to esophagitis?
   A. Gastro- esophageal reflux.
   B. Portal hypertension.
   C. Gastritis
   D. Polyps of the gastrointestinal tract.
2. What is considered to the normal portal venous pressure?
   A. 15-20 mm Hg
   B. 80-120 mm Hg
   C. 5-10 mm Hg
   D. 12- 15 mm Hg
3. What is the most severe complication of portal hypertension?
   A. bleeding from the esophageal/ gastric varicose veins
   B. Splenomegaly
   C. Hypersplenism
   D. Ascites
4. In the result of what portosystemic shunt procedure blood flow can be restored to the liver.
   A. Splenorenal shunt side-to-side
   B. Distal splenorenal shunt
   C. Mesocaval shunt side-to-side
   D. Mesoportal shunt with graft from the internal jugular vein (Rex- shunt)
5. What hereditary syndrome may causes the presence of polyps covering the whole intestine?
   A. Juvenile Polyposis
   B. Familial Adenomatous Polyposis (FAP)
   C. Peutz-Jeghers Syndrome
   D. Hemangiomatous Polyps
6. In what age does intussusception usually occur?
   A. 1-2 years.
   B. 3-8 months
   C. 5-10 years
   D. 1-2 months
7. What is the clinical presentation of intussusception?
   A. episodic abdominal pain, vomiting, palpable mass in the abdominal cavity
   B. disorders of the stool
   C. vomiting and rise of body temperature
   D. “coffee ground” vomitus bleeding
8. In which group of neonates does enterocolitis usually occur?
   A. neonates that are premature
   B. neonates with physiological jaundice
   C. in male neonates
   D. in those neonates that weigh more that 4 kg at birth

9. What are the complications of a Meckel's diverticulum?
   A. vomiting
   B. abdominal pain
   C. abdominal pain and vomiting
   D. bleeding, obstruction due to intussusception
10. What is the clinical presentation of an anal fissure?
   A. abdominal pain
   B. melena
   C. a drop of blood on the fecal masses
   D. vomiting

Keys for tests

1. A
2. C
3. A
4. D
5. C
6. B
7. A
8. A
9. D
10. C

Tests for final level of knowledge

1. A 4-month-old child is admitted to the emergency department with episodic abdominal pain and vomiting. From the history it is known that the mother just started feeding the child apple mash besides breast milk. Child's mother states that the duration of pain episodes became longer and the intervals between the pain episodes became shorter. At physical examination there is presence of a palpable mass in the abdomen. The primary diagnosis is intussusception. What is the method of choice for instrumental examination that can prove the primary diagnosis?
   A. X- ray of the abdomen
   B. Upper endoscopy
   C. Ultrasound of the abdominal cavity
   D. X- ray contrast study of the digestive tract
2. On your shift a 12-year-old child is admitted to the emergency department with "coffee ground" vomitus bleeding. You are suspecting a gastric ulcer. What diagnostic method will help you detect the lesion?
   A. Ultrasound of the abdomen.
   B. Upper endoscopy
   C. Blood test
   D. X- ray of the abdomen
3. A 5-year-old child is referred to the surgeon with complaints of common constipation during the last month and periodical pain at the anus during the act of defecation and a drop of blood on the fecal masses at the end of defecation. What is that primary diagnosis?
   A. Enterocolitis.
   B. Infectious colitis
   C. Anal fissure
   D. Gastritis
4. On an annual checkup at the pediatrician a 7-year-old child presented with splenomegaly and hypersplenism. For what pathology should the child be examined?
   A. Portal hypertension
   B. Appendicitis
   C. Hepatitis

D. Ascites

5. A 4-year-old child was admitted to the hospital after several episodes of painless bloody stool of purple color. What may be the primary diagnosis?

A. Infectious colitis
B. Intussusception
C. Peptic ulcer
D. Meckels' diverticulum

6. A 9-year-old child was admitted to the hospital with crampy abdominal pain and lower intestinal bleeding, also pigmented lesions that were noticed on the buccal mucosa and lips. What condition may be the reason for bleeding?

A. Peptic ulcer
B. Portal hypertention
C. Intussusception
D. Peutz-Jeghers syndrome

7. A 14-year-old child was admitted to the emergency department with blood vomiting and pain behind the chest. Results of endoscopy showed esophagitis. What caused the presence of esophagitis and such symptoms?

A. Peptic ulcer
B. Gastro- esophageal reflux
C. Portal hepertention
D. Gastritis

8. A premature neonate was noticed to have purple stool on the 5th day after birth. What is the primary diagnosis?

A. Anal fissure
B. Enterocolitis
C. Intussusception
D. Infectious colitis

Keys for tests:

1. C
2. B
3. C
4. A
5. D
6. D
7. B
8. B

Tasks for final level of knowledge

Task 1. A 7-year-old child was admitted to the hospital with intensive vomitus bleeding, splenomegaly, and signs of hypersplenism. Out of the patients history it became evident that since the age of 2 the child was diagnosed with extrahepatic portal hypertension. In the las 6 months this is the second case of bleeding.

1. What list of exams will you assign to the patient and in what term after hospitalization
2. What treatment will you recommend?

Key answers:

1. At the time of hospitalization the child should have a blood test done (WBC and platelet count, level of hemoglobin) and blood pressure should be checked. If the patient is stable then upper endoscopy should be performed right after hospitalization, to detect localization of bleeding. If the patient is unstable endoscopy should be conducted after stabilization. An ultrasound and a contract computed tomography may be performed after the above diagnostic procedures.

2. Taking into consideration that this episode of bleeding isn't the first, the recommended treatment for the patient is portosystemic shunting.

Task 2. A 15-year-old child is admitted to the emergency department with abdominal pain, vomiting. The child has a history of peptic ulcer. On palpation there is abdomen distention, sever pain, tympanic sound on percussion of the stomach.

1. What complication of peptic ulcer occurred?

2. What method of diagnostics should be chosen to detect the possible complication suspected?

3. What treatment should be offered to the patient in case the complication is proven, and in what terms?

Key answers:

1. The complication that occurred is perforation of the peptic ulcer.

2. To detect ulcer perforation an X- ray of the abdomen should be performed positioning the patient vertically, which will help detect the presence of free gas in the abdominal cavity with is seen in the case of ulcer perforation.

3. The patient should be offered surgical treatment in an urgent matter.

Task 3. A 4-month-old child was admitted to the emergency department with episodic abdominal pain and vomiting. From the history it is known that the mother just started feeding the child apple mash besides breast milk. Child's mother states that the duration of pain episodes became longer and the intervals between the pain episodes became shorter. At physical examination there is presence of a palpable mass in the abdomen and signs of peritonitis. The primary diagnosis is intussusception. The time from when the first symptoms started is 32 hours.

1. What treatment should be offered and in what mater?

2. What will be the capacity of the surgical treatment?

Key answers:

1. The treatment should be surgical and in an urgent matter considering signs of peritonitis.

2. Considering signs of peritonitis and the duration of the illness (32 hours), most likely there is necrosis of the gut and resection is needed.

THEME 3

THORACIC TRAUMA. INJURIES TO THE ESOPHAGUS.

Overview

Thoracic trauma is an important cause of morbidity and mortality in children. The spectrum of childhood traumatic chest injuries is wide ranging from simple contusion, multiple rib fractures to rare exsanguinating vascular injuries. Although it accounts for a small minority of pediatric trauma injuries (4%-25%), it is associated with a 20-fold increase in mortality when compared with pediatric trauma patients without thoracic trauma. Thoracic injuries are suggestive of a significant mechanism of injury and herald the likelihood of coexisting injuries to other body regions, particularly the head, abdomen, and spine. Moreover, the mortality rate of head and abdominal trauma in association with thoracic trauma in children increases to 25%. Children with neurosurgical trauma, thoracic trauma, and abdominal trauma may have a mortality rate that approaches 40%. Isolated thoracic trauma in a child is associated with a mortality rate of approximately 5%, which is largely due to penetrating trauma. This mortality and morbidity rate can be improved if the patient is transferred to a pediatric center and managed within "the golden hour"**.**

1.Educational aims: The aim of this part of module is to provide help in identifying those children with thoracic trauma both isolated and associated with other injuries and to provide guidance on understanding of the pathogenesis of thoracic trauma, initial evaluation of the patient, recognition of the variant of injury, diagnosis, and princples of medical aid.

2. A student must know:

1. Anatomy and physiology of thoracic and mediastinal organs.
2. Pathogenesis of thoracic trauma
3. Classification of thoracic trauma.
4. Mechanisms of thoracic trauma and their peculiarities in infants and children.
5. Methods of diagnosis of thoracic trauma and their peculiarities in pediatric patients.
6. Relevant treatment of thoracic trauma in children.
7. Basics of first aid in case of thoracic trauma and its features in pediatric age.
8. The technique of urgent procedures: thoracocentesis, pleural puncture, tracheostomy

3. A student must be able to:

1. Collect complains, determine clinical signs, patient's condition, signs of shock and establish tentative diagnosis.
2. Define the rational set of lab studies and instrumental methods of investigation.
3. Correctly interpret the results of clinical analyses, physical examinations, plain X-ray and CT scans, ultrasonography, tracheobronchoscopy, pleural puncture.
4. Determine the signs of pneumothorax, hemothorax, heart tamponade and know the urgent management of them.
4. Give the first aid in cases of thoracic trauma in children.
5. Define the indications for surgery or other methods of treatment in pediatric patients with thoracic trauma.
6. To administer urgent treatment in thoracic injuries in children, determine the blood group and Rh-factor, measure the arterial pressure, perform pleural puncture.

Terminology

Term	Definition
Pneumothorax	Pneumothorax occurs when the air accumulates in the intrapleural space
Tension pneumothorax	Tension pneumothorax is an immediate life-threatening condition that occurs when the accumulation of air in the pleural space is massive and rapidly increasing thus causing a pressure build up in the affected side, mediastinal shift to the contralateral side, collapse of the ipsilateral lung and compression of the contralateral lung

Hemothorax	Accumulation of blood in the pleural space
Haemo-pneumothorax	Accumulation of blood and air in the pleural space
Traumatic Asphyxiation	Very rare and a potentially immediately life-threatening injury due to severe and sudden compression of the chest and upper abdomen, resulting in decreased venous return from the head, neck, and upper torso and thus extravasation of blood and fluid into the subcutaneous tissues of these areas. These features include subconjunctival and upper body cutaneous petechial hemorrhages, cyanosis, facial edema, and a variable degree of pulmonary and central nervous system abnormalities
Mediastinal emphysema	Accumulation of the air in the anterior or posterior mediastinum due to injury to the trachea or bronchi.
Pericardial tamponade	This involves the accumulation of blood, or any fluid, in the pericardium and can be seen in both blunt and penetrating injuries. The raised pericardial pressure causes reduced cardiac compliance, increased intracardiac pressures, reduced cardiac filling both from systemic and pulmonary circulations, and decreased cardiac output

CONTENT

Thoracic trauma is uncommon in children, comprising only 0.2–7% of all injuries encountered in the pediatric population. Epidemiologic studies have reported a twofold to threefold higher incidence of thoracic trauma in boys as compared with girls. Most injuries (80%-95%) are the result of blunt trauma, typically resulting from a traffic accident in which the child involved as a passenger or pedestrian. Many children will have involvement of other organ systems with a high Injury Severity Score (ISS). When penetrating trauma does occur, older children and adolescents are more likely to be the victims. This is associated with a higher mortality rate. Contusion or laceration of the pulmonary parenchyma is the most common injury and may be associated with rib fractures and pneumothorax or haemothorax. Susceptibility to tension pneumothorax or haemothorax is much greater and these injuries must be treated promptly. This mobility, however, does make it less susceptible to major vascular or airway injuries. Thoracic trauma in children is a marker for the presence of associated injuries, found in more than 50% of these children.

Serious intrathoracic injuries occur in 6% of pediatric blunt trauma victim and require thoracostomy in about 50%. Thoracotomy is seldom needed. Lung injuries (52%), pneumothorax and hemothorax (42%), and rib and sternal fractures (32%) occur most frequently. Injuries to the heart (6%), diaphragm (4%), great vessels (2%), bronchi (<1%), and esophagus (<1%) occur least frequently. Because blunt trauma is nearly 10 times more deadly when associated with major intrathoracic injury, this condition serves as a marker of injury severity, even though it is the proximate cause of death in less than 1% of all pediatric blunt trauma. The child compensates poorly for respiratory derangements associated with serious thoracic injury because of (1) a larger oxygen consumption but smaller functional reserve capacity (which makes the child more susceptible to hypoxia), (2) lesser pulmonary compliance yet greater chest wall compliance (which dictate chiefly a tachypneic response to hypoxia), and (3) horizontally aligned ribs and rudimentary intercostal musculature (which make the small child a diaphragmatic breather). The thorax of the child usually escapes major harm because the pliable nature of the cartilaginous ribs allows the kinetic energy associated with forceful impacts to be absorbed without significant injury, either to the chest wall itself or to underlying structures. Pulmonary contusions are the typical result but seldom are life threatening. Pneumothorax and hemothorax, due to lacerations of the lung

parenchyma and intercostal vessels, occur less commonly but place the child in grave danger of sudden, marked ventilatory and circulatory compromise as the mediastinum shifts.

Mechanism of injury

Blunt trauma accounts for 85–90% of pediatric thoracic injuries. The majority of blunt thoracic injuries result from motor vehicle accidents. Children may be passengers in the motor vehicle or, frequently, may be struck by the motor vehicle. Falls from various heights are the next most common causes of thoracic injuries. Isolated blunt thoracic trauma carries a 5% mortality rate. When associated with blunt traumatic brain injuries, the mortality rate rises to 25%. When associated with brain and abdominal injuries, the mortality rate rises to 40%. Penetrating trauma represents only 10–15% of thoracic trauma but the incidence increases with age, especially for children over 12 years of age. Nearly 14% of children suffering from penetrating thoracic trauma die from thoracic injury. Associated injuries with penetrating thoracic trauma are less common.

There are a number of important anatomical and physiological differences between the pediatric and adult thorax. These differences significantly influence the injury patterns seen in children who have sustained chest trauma. The chest wall of a child is very compliant. As a consequence, the bony skeleton is massively deformable to the degree that the anterior ribs can be depressed until they make contact with the posterior ribs without fracturing. This means that the intrathoracic structures may be significantly compressed without any overlying injury evident. It also suggests that the mechanisms required to cause fractures of these bones must involve substantial force.

Thoracic injuries in children can be categorized by location: chest wall (flail chest, open pneumothorax, rib fracture, traumatic asphyxia); pleural cavity/pulmonary parenchyma (tension pneumothorax, hemothorax, simple pneumothorax, pulmonary contusion/laceration, diaphragmatic injury); and mediastinum (pericardial tamponade, aortic/great vessel injury, tracheobronchial injury, cardiac contusion, esophageal injury).

Clinical variants of thoracic trauma

— Injury to chest wall (contusion, hematoma, rib and sternal fractures, flail chest, open pneumothorax)
— Pulmonary contusion
— Injury to pleura and lungs
— Injury to the trachea and bronchi
— Traumatic asphyxia (Perthes Syndrome)
— Cardiac and great vessels injuries
— Diaphragmatic injury
— Esophageal injury

Incidence of thoracic trauma variants (based on data of Kyiv City Thoracic Trauma Center (1981 – 2003)).

Pulmonary injuries – 11.9 – 19.2%;
Cardiac injuries – 8.65%;
Injury to the trachea and bronchi – 6.4%;
Injury to the diaphragm – 30.76%;
Rib fractures – 8.25%.
Injuries to other organs:
Brain – 16,5%;
Bones – 18,3%

Causes of respiratory and circulation disorders:

- Strong pain causes the decrease of ventilation of the lungs in 15 – 40%;

- Lung collapse, narrowing, kinking and obstruction of airway cause the decrease of lung ventilation as well.

- Arrhythmia, cardiac tamponade, kinking of great vessels, blood loss cause circulation disorders.

Assessment and diagnosis in children with thoracic trauma

Children with suspected thoracic trauma initially require assessment of their airway and breathing. Rarely a thoracic injury will involve trauma to the upper airway resulting in obstruction. In such cases a patent airway must be secured urgently prior to further clinical assessment. The majority of thoracic injuries encountered will impact primarily on the patient's respiration and ventilation. Assessment of patient's condition is made by pediatric trauma severity scale developed by National Center for Pediatric Trauma Registration, USA (1990) (table 1).

Radiological investigations will always involve a chest X-ray, which should be completed in the basic trauma series. If the patient is hemodynamically stable and further information regarding the extent of any injury is required a computed tomography (CT) scan of the chest should be undertaken. CT scan in the initial diagnostic work up of blunt thoracic trauma accurately diagnoses thoracic injuries, influenced the therapeutic management, and was found to be more sensitive than chest CX-ray. However, although CT is a highly sensitive diagnostic modality and influence the thoracic injury management, a chest X-ray still provides valuable information. Given the high dose of radiation associated with pediatric CT scans and the evidence suggesting a fatal malignancy rate of 1 per 1,000 scans this investigation should be used judiciously.

Table 1

Pediatric trauma severity scale

Diagnostic and prognostic component	**Assessment**		
	+2	**+1**	**-1**
Body weight	>20 kg	10 – 20 kg	<10 kg
Airway	Normal	Disorders correctable	Disorders not correctable
CNS (consciousness)	Consciousness safe	Consciousness confused	Coma
Systolic AP	>90 mm Hg	50–90 mm Hg	<50 mm Hg
Open wounds	None	Not much	Severe, penetrating
Fractures	None	Closed, single	Open, multiple

Assessment from +12 points to -6; +12 nonsignificant trauma, -6 – absolutely fatal; +9 and more – no lethal outcomes

Thoracic injuries caused by a blunt mechanism may present diagnostic difficulties when there are no external signs of trauma. This is in contrast to penetrating chest injuries, discussed in the following chapter. In both situations the patient may demonstrate few clinical signs. However, in cases where there has been a penetrating injury there will always be a wound to alert the clinician to the possibility of an intrathoracic injury.

Diagnostic signs of chest wall injury are following wounds, abrasions, hematomas, ribs deformity, asymmetry of chest movements.

Rib Fractures

Rib fractures in children are less common than in adults due to the increased compliance of the chest wall. Therefore, rib fractures are felt to be a marker of increased injury severity. Rib fractures are also commonly associated with child abuse. Nearly two-thirds of cases of rib fractures in children under three years of age result from abuse. The most common cause of rib fractures in older children are motor vehicle crashes. Signs and symptoms include chest pain, dyspnea, and splinting on the side of the fracture which can result in atelectasis and pneumonia. Chest x-ray will confirm the diagnosis and more importantly evaluate for an underlying pulmonary injury, pneumothorax, or hemothorax. Rib fractures typically require no surgical therapy. Pain control and pulmonary toilet significantly reduce the risk of complications. Children with first rib fractures should be evaluated for vascular injury.

Fractures of the sternum occur rarely in children, often in older children, resulting from the direct action of injuring force: the common is trauma obtained from blow into the sternum using the car wheel. Often fractures are localized in upper and middle third, and then the body of the sternum is dislocated backwards. In the area of a fracture we may observe deformation, pain, swelling and pathological mobility of splinters. The main diagnostic method is X-ray of the sternum in lateral projection and computed tomography. In case if splinters are dislocated and diastases develop between them operative treatment (osteosynthesis) – stitching of the sternum or fixation by metals pins.

Flail Chest

Flail chest is caused by multiple rib fractures or disarticulation of the ribs from the sternum resulting in an unstable segment of chest wall. The flail segment demonstrates paradoxical chest wall movement during the respiratory cycle and physical exam is diagnostic. Radiographic findings are only supportive. Treatment involves adequate analgesia, positive pressure ventilation, and possible rib fixation stabilizing the flail segment. Placement of chest tube may be required for associated pneumothorax or hemothorax. As with less complicated rib fractures, pain control and pulmonary toilet are essential.

Chest Wall Laceration

Chest wall lacerations occur from multiple different mechanisms but the treatment remains the same. Wound exploration, foreign body removal, control of bleeding, debridement, and wound closure are the basics of laceration management.

Pulmonary Contusions

Pulmonary contusions are due to alveolar hemorrhage and parenchymal destruction. Pulmonary contusions, the most common injury found in children with thoracic trauma, can result from blunt or penetrating trauma. Only 40% of these patients have associated rib fractures. This low percentage is due to increased pediatric chest wall compliance. Signs and symptoms of pulmonary contusions include chest pain, shortness of breath, hypoxia, dyspnea, coughing, crepitation, distant breath sounds, moister rales, and, less commonly, hemoptysis. Pulmonary contusions may present insidiously during the first 24 h following thoracic trauma with worsening pulmonary symptoms and delayed radiologic findings. Chest X-ray remains the initial diagnostic test and computer tomography is used for complicated cases when further imaging is required. On plain radiograph cloud-like opacity without precise boundaries can be defined. Treatment involves supplemental oxygen, fluid restriction as tolerated, chest physiotherapy to clear secretions and blood, as well as adequate pain control. Intubation and mechanical ventilation are infrequently required. The mortality rate due to isolated pulmonary contusions is low, and there are no long-term respiratory problems.

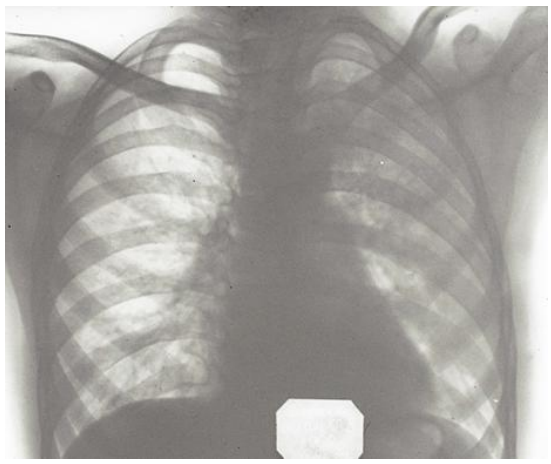


Fig. 3.1. Plain chest X-ray of a child with left lung contusion due to blunt thoracic trauma (motor vehicle accident). Cloud-like opacification of the left lung is sharp boundaries

Injury to pleura and lungs

Pulmonary laceration is usually encountered in penetrating injuries but may occur in blunt trauma as a result of damage from the sharp ends of fractured ribs or secondary to shearing forces such as those experienced in rapid deceleration injuries. Regardless of the causative mechanism the pathology is the same. Pneumothorax (open, closed, simple, tension), hemopneumothorax, hemothorax, hemoptysis and subcutaneous emphysema are reliable symptoms of traumatic injuries to lungs and pleura.

Pneumothorax may occur with penetrating injury to the chest wall or air leak into the pleural space from a pulmonary laceration or disruption of the airway more proximally. It is a relatively common finding in children with blunt and penetrating thoracic trauma.

Simple Pneumothorax

A simple pneumothorax occurs when air gets accumulated in the chest cavity without putting it in any tension. Decreased breath sounds may be observed as well as chest pain and shortness of breath. A simple pneumothorax is often asymptomatic because the lack of increased intrathoracic pressure limits the recognition of symptoms. Chest X-ray is diagnostic and placement of chest tube is therapeutic. In rare cases, a pneumothorax may remain very small due to a transient air leak from the parenchyma that readily stops. In this situation, conservative management may prevail without the placement of chest tube.

Tension Pneumothorax

Tension pneumothorax is a life-threatening condition that requires expeditious decompression of the involved hemithorax. Tension pneumothorax is caused by air entering the potential space between the visceral and parietal pleura resulting in air getting trapped in the hemithorax. This occurs following a tear in the pulmonary parenchymal due to rib fracture and laceration, deceleration injury, crush injury, or due to increased intrathoracic pressure causing pulmonary rupture. Signs and symptoms include agitation that rapidly changes for inhibition, cyanosis or gray skin, chest pain, dyspnea, increased respiratory rate, weak pulse, subcutaneous emphysema, and hyperresonance on the injured side. A pneumothorax can quickly result in cardiovascular demise if tension within the hemithorax develops (Fig. 3.2). Findings of hyperresonance, diminished breath sounds on the affected side, tracheal deviation, and hemodynamic instability indicate a potentially lethal tension pneumothorax requiring immediate intervention. Chest X-ray is not required to make this diagnosis and may waste valuable time. Needle decompression is the most efficient therapy for a tension pneumothorax; it is accomplished by placing a tube in the second intercostal space along the mid-clavicular line just above the third rib. Alternatively, the decompression can be performed in the anterior axillary line just above the sixth rib. This should be followed by placement of chest tube. This results in the expansion of the lung and continued evacuation of air and fluid until the injured lung seals and air leak resolves.

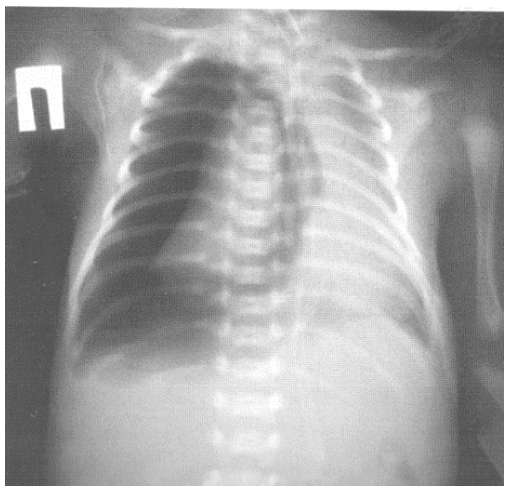


Fig. 3.2 Tension right-sided pneumothorax. Plain chest X-ray. There is no lung tissue picture on the affected side. Right lung is collapsed. Contralateral shift of mediastinum. Intercostal spaces are dilated on the affected side. Right diaphragm cupola is flattened

Open Pneumothorax

An open pneumothorax is caused by a persistent or continuous communication between the environment and the chest cavity, and is sometimes referred to as a sucking chest wound. The negative intrathoracic pressure generated during inhalation results in air from the environment being sucked into the chest cavity and collapse of the lung on the affected side. Signs and symptoms include shortness of breath, chest pain, decreased breath sounds on the affected side, and sound of air being sucked into the chest cavity. This can result in respiratory arrest and cardiovascular collapse if not treated promptly. Treatment involves sealing or closing the hole in the chest wall. Vaseline gauze is commonly used. Placement of a chest tube evacuates residual air from the affected hemithorax and enables lung re-expansion. If there is an associated parenchymal injury, the chest tube is left in place until this air leak resolves.

Hemothorax

A traumatic hemothorax is the accumulation of blood in the hemithorax following blunt or penetrating trauma. Bleeding can result from rib fractures that lacerate the intercostal vessels, pulmonary parenchymal laceration, or, less commonly, great vessel injury. Hemothorax in children can be categorized by the quantity of accumulated blood in pleural cavity as following: small (0.25% of body weight), moderate (0.5%), and severe (1%) (table 2).

Table 2.

Classification of hemothorax in children

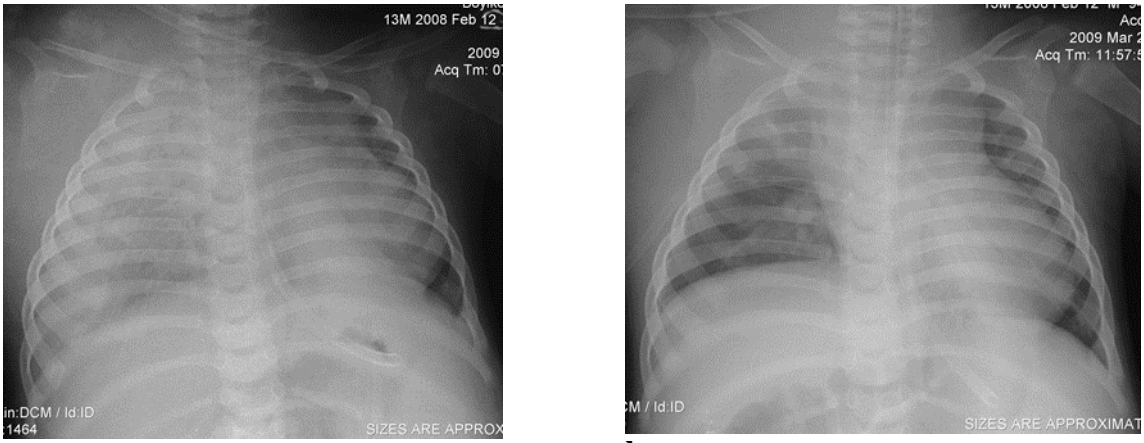
Body weight, kg	**Blood volume, ml**		
	small	**moderate**	**severe**
10	25	50	100
15	37	75	150
20	50	100	200
25	65	125	250
30	75	150	300

A small hemothorax is caused by minor bleeding in the chest cavity due to pulmonary parenchymal injury or chest wall injury. Only the placement of a chest tube is required for drainage if the lung volume is compromised.

A massive (severe) hemothorax results from life-threatening exsanguination into the chest cavity requiring immediate intervention. Signs and symptoms include shock, hypotension, diminished breath sounds and dullness to percussion on the affected side, chest pain, shortness of breath, and oxygen requirement. This is a clinical diagnosis especially in the unstable child. In the stable child, a chest X-ray confirms the diagnosis (fig. 3.3). Standard therapy includes large bore tube thoracostomy for evacuation of fluid and air providing for lung re-expansion. Resuscitation with intravenous fluid and possibly packed red blood cells is required. Thoracotomy is indicated for hemodynamic instability, loss of >25% total blood volume with initial chest tube placement, or persistent bleeding greater than 2 ml/kg/h.

Mild pneumothorax doesn't require special treatment. During 7-10 days the blood is absorbed. Local treatment of hemothorax is combined with general, first of all with hemostatic therapy and eliminating anemia.

Haemothorax is often associated with pneumothorax that worsens its clinical course. This causes infection and empyema. If vessel rupture is accompanied by injury of the bronchus, the sputum may contain admixture of blood.



a **b**

Fig. 3.3. Right-sided hemothorax. Plain chest X-ray before (a) and after (b) pleural punction

Chylothorax

Chylothorax caused by injury to thoracic lymphatic channels is an uncommon complication of thoracic trauma. Chylothorax usually becomes evident 3 to 7 days after injury. The diagnosis is made by obtaining a sample of the pleural fluid and identifying the lymphatic and lipid content. Treatment includes tube thoracostomy and either enteral feedings with medium-chain triglycerides or parenteral nutrition.

Subcutaneous emphysema.

At injured lungs, visceral and parietal pleura, intercostal muscles (that may occur at blunt trauma of the thorax with fractured ribs) air from the pleural cavity may penetrate into the subcutaneous base. It frequently occurs at tension pneumothorax when air penetrates the pleural cavity and tries to find the way out. There are local and general clinical manifestations. Air is accumulated in the subcutaneous base what may be determined by physical examination. At palpation- crepitated riles are determined. At percussion there is tympanic sound, at auscultation- weak breathing. Most frequently air accumulates subcutaneously in injured part of the thorax. Only in severe cases at tension pneumothorax it may spread towards the neck, face and another part of the thorax. The patient resembles inflated rubber toy. General condition of patient deteriorates; patient presents with dyspnea, skin cyanosis and tachycardia.

Mild subcutaneous emphysema doesn't require special treatment. The patient is provided rest, analgesics, anti-cough and cardiac medications. In a few days the air starts to be dissolved. If air in subcutaneous base accumulates quickly and spreads onto the neighbor areas, it is necessary to perform some incisions on skin after finding the cause of subcutaneous emphysema and appropriate operative or conservative treatment.

Mediastinal emphysema.

Mediastinal emphysema occurs resulting from thoracic injury with simultaneous affection of the trachea or bronchi. In such case the air from respiratory ways through the surrounding connective tissue penetrates into the anterior or posterior mediastinum.

Diagnostics of mediastinal emphysema (especially posterior mediastinum) is complicated if subcutaneous emphysema is absent. The air that penetrated into the mediastinum, may compress airway and large blood vessels causing dyspnea, cyanosis, rapid and weak pulse (extrapericardial heart tamponade). Subcutaneous emphysema is progressing quickly at the neck area. The neck and upper jugular veins are overinflated. Patients are agitated.

X-ray imaging is of great importance in diagnostics of emphysema. Roentgenogram shows widening of the mediastinal shade and air accumulation. In severe cases of mediastinal emphysema there may be needed surgery – mediastinotomy or plastic closure of injured areas. In injured trachea or bronchi intubation is performed to relieve the treatment. Forced artificial lung ventilation is contraindicated, because it may cause intensification of emphysema.

Tracheobronchial Injury

Tracheal and bronchial injuries are uncommon in the pediatric population but have been reported following thoracic crush injuries and penetrating injuries. Airway disruption may occur with penetrating injury or with blunt injury such as high-energy acceleration or deceleration. Up to three fourths of these injuries are noted within 2 cm of the carina and almost half occur within the first 2 cm of the right main-stem bronchus. Signs and symptoms include chest pain, dyspnea, hypoxia, stridor, crepitance in the neck and chest wall, mediastinal emphysema, tension pneumothorax, and hemoptysis. Most patients with tracheal injuries have mediastinal air on chest X-ray, although more distal injuries may rupture into the pleural space and present as a tension pneumothorax. Other findings associated with a major airway injury include a persistent large airleak from a chest tube, mediastinal emphysema (fig. 3.4), cervical subcutaneous emphysema without pneumothorax, or florid respiratory compromise. Rarely, complete transection of a distal main-stem bronchus will appear on chest X-ray with total lung collapse and mediastinal displacement. Further diagnostic evaluation is best accomplished via rigid bronchoscopy for direct visualization of the injury and evacuation of blood and secretions. Chest CT with a multiple-array scanner may have a role in visualizing tracheal or bronchial injuries especially if three-dimensional reconstructions of the airway are used (virtual bronchoscopy) Pleural air or fluid collections should be drained until an accurate diagnosis of the airway injury is made.

In general, when a tracheobronchial injury is identified, surgical repair is indicated. Repair of some bronchial injuries can be successful even a year after injury. Occasionally, more distal bronchial injuries may heal with nonoperative management. Distal bronchial injuries, both acute and chronic, are generally well managed by pulmonary resection, whereas more proximal airway trauma is best treated by surgical repair. Flap coverage of the repair with pleura or muscle reinforces and protects the repair. Nonoperative management of a tracheobronchial injury may result in a high incidence of airway stenosis. When the diagnosis of a tracheobronchial injury is substantially delayed, scarring may obliterate the airway lumen and cause chronic collapse of the lung segment or lobe. Complete transections are commonly associated with an obliterated distal bronchus, which may spare the pulmonary parenchyma from infection, thus making repair possible. Incomplete tears, on the other hand, form granulation tissue and scar that result in a patent, but narrowed lumen, predisposing the supplied lung to recurrent infection and retained secretions. This usually necessitates future resection. If the esophagus and the airway are injured near one another, a traumatic tracheoesophageal fistula can occur.

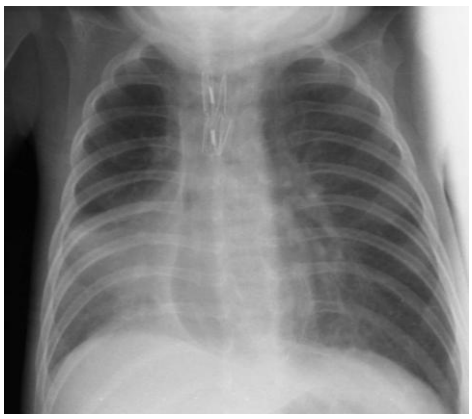


Fig. 3.4. Chest X-ray in 2 months old patient demonstrating peumomediastinum without tension caused by iatrogenic injury to the trachea during tracheal stenting. Resolved with conservative treatment

Traumatic Asphyxia (Perthes Syndrome)

Traumatic asphyxia is the inability to breathe due to compression of the thorax. Removal of the compressive force allows resumption of breathing and recovery. Commonly reported causes are compression from garage doors, motor vehicles, furniture, and other heavy objects. This results in global hypoxia most often rendering the patient unconscious followed by cerebral injury and death if the compression is not promptly relieved. Signs and symptoms include subconjunctival hemorrhage; facial, neck, and chest petechiae; and facial edema. In hospitals, mortality rates are very low if these children survive the actual event.

Heart and Great Vessel Injuries

Injuries to the heart and great vessels of the thorax are rare in young children and comprise 7 to 14% of all thoracic trauma. Blunt cardiac injury such as myocardial contusion, cardiac laceration, or cardiac rupture is rare, occurring in less than 5% of children with blunt thoracic trauma. Penetrating trauma is more often a cause of cardiac and aortic injury in this age group. Mechanisms of cardiac trauma are following: push and impact over the chest (pericordium) (fall, impact by ball, boxing, etc), compression caused by fall of heavy objects, squeeze between the parts of technical devices, crush in blockages or crowded places, hydraulic impact by blood inside the heart chambers, sudden deceleration causing the hit of the heart on chest wall, injuries caused by sharp objects (knives, fall on metal fence, pitchfork, etc), gunshot injuries.

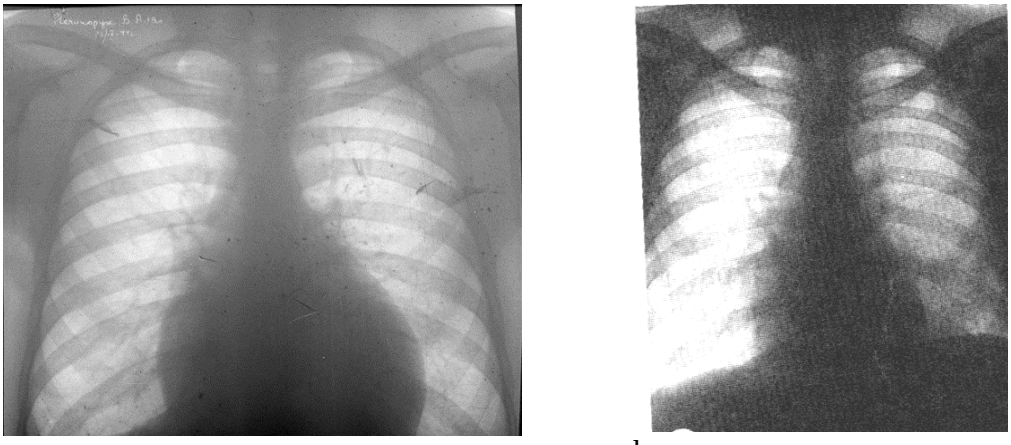
Commotio cordis has become a widely recognized problem in pediatric thoracic trauma. Typically, a young baseball player is struck in the chest with a hit or thrown ball and collapses suddenly. Commotio cordis is characterized by the absence of cardiac contusion, coronary artery abnormalities, structural abnormalities, or conduction system pathology. It is thought that a sudden blow to the chest will result in a disorganized cardiac rhythm followed by rapid cardiovascular collapse. Although chest protective devices seem useful, they do not provide total protection against asystole. Automatic electrical defibrillators may have a place in treating this rare sports-related injury.

Cardiac contusion accounts for 95% of blunt cardiac injuries in the child, followed by valvular dysfunction and ventricular septal defect. Clinical manifestations of cardiac injury after blunt thoracic trauma include arrhythmia, new-onset murmur, and heart failure. These findings, however, may be absent in children, and chest X-ray and electrocardiographic findings are generally nonspecific. The management of a cardiac contusion is supportive. Children with a cardiac contusion who are hemodynamically stable on presentation rarely have deterioration of their cardiac rhythm. Patients should be monitored with continuous electrocardiography and frequent blood pressure determinations. In patients with a suspected cardiac contusion, serum cardiac troponin I levels may be useful to confirm the diagnosis. Inotropic agents are occasionally needed to provide cardiac support in the presence of a cardiac contusion.

Pericardial tamponade involves the accumulation of blood, or any fluid, in the pericardium and can be seen in both blunt and penetrating injuries. The effusion usually consists of blood and

may originate from a cardiac chamber, one of the great vessels, or from the myocardium. The normal volume of pericardial fluid is approximately 25 ml; when this volume is acutely increased by as little as 5–10 ml, it can result in significant hemodynamic compromise. The raised pericardial pressure causes reduced cardiac compliance, increased intracardiac pressures, reduced cardiac filling both from systemic and pulmonary circulations, and decreased cardiac output. This type of injury is poorly tolerated in children, and persistent hypotension despite maximal fluid resuscitation in a child who has sustained a thoracic injury should alert the clinician to the possibility of pericardial tamponade. It should be remembered that the classic symptoms of distended neck veins, a raised central venous pressure, and pulsus paradoxus are not often evident in children with tamponade. Other signs are severe respiratory distress, altered level of consciousness, shock; a wound from a penetrating injury may be present. Heart sounds are soft/muffled. Echocardiogram or simple thoracic ultrasound may be used to confirm the diagnosis. Chest X-ray may show the enlarged spherical heart shadow that significantly decreases after pericardiocentesis (fig. 3.5).

This condition is imminently life threatening and requires urgent intervention. All patients require close monitoring including ECG. Fluid resuscitation is required as well as oxygen supplementation. The main principle in the management of this condition is reduction of the pericardial fluid. Pericardiocentesis in the emergency department is a potentially lifesaving procedure but only provides temporary relief. It should be performed only if the patient is acutely hemodynamically unstable and/or surgical intervention is not immediately available.



a b

Fig. 3.5. Chest X-ray showing hemopericardium resulting in pericardial tamponade. Heart shadow is distended, spherical (a), after pericardiocentesis it looks normal (b)

Children with a blunt cardiac injury should be followed closely and monitored for sequelae such as valvular insufficiency or ventricular septal defect (traumatic heart defects). This is characterized by sudden heart failure and dilation (fig. 3.6) refractory to medical treatment. Very rarely, extracorporeal life support may be needed to manage a child with a severe blunt cardiac injury

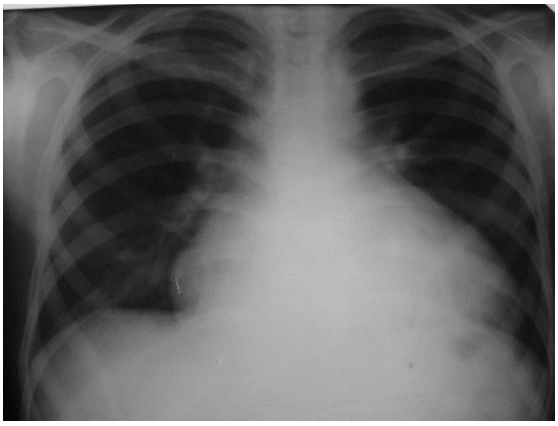


Fig. 3.6. Chest X-ray demonstrating dilation of the heart in patient with traumatic heart defect

The incidence of blunt aortic injury is 0.1% with a mortality rate of over 40%. Eighty percent of children who sustain a thoracic aortic tear will have significant associated injuries to the lung, heart, skeletal system, abdominal organs, or central nervous system. Only one half of these patients will have external evidence of thoracic injury. Most aortic injuries in children are related to falls or motor vehicle collisions, especially when children are unrestrained. The diagnosis of an aortic injury in a child can be difficult. Findings on a chest film may include a left apical cap, pulmonary contusion, mediastinal widening, a shift of the trachea to the right, downward depression of the left main-stem bronchus, and an indistinct aorta. A CT scan may reveal a mediastinal hematoma or an actual aortic injury. Transesophageal echocardiography has also been useful for diagnosing an aortic injury. Whereas thoracic CT and transesophageal echocardiography can diagnose aortic injuries, aortic angiography gives excellent anatomic detail. The most common finding with a traumatic aortic injury is a pseudoaneurysm located at the proximal descending aorta. This is thought to occur secondary to tethering of the aorta by the ligamentum arteriosum at the time of injury, resulting in a tear in the aortic intima and media. There is no large collective study of children with aortic injuries. However, of those who survive until diagnosis, more than 70% will live to discharge. Spinal cord ischemia is a complication with thoracic aortic injury and may be associated with preoperative cardiovascular instability. Although urgent operative repair is thought to be the best treatment in most patients, recent experience has demonstrated the ability to delay operative intervention by using β-adrenergic blockers while other injuries are managed. Several pediatric patients have received aortic endostents to repair an aortic injury. Although this experience is limited, in the future its risk benefit ratio may be better than that of open operative intervention.

Diaphragmatic Injuries

Diaphragmatic injuries are more commonly caused by penetrating trauma. Signs and symptoms include dyspnea, abdominal pain, vomiting, and decreased breath sounds. These injuries can be diagnostic dilemmas. The initial radiologic exam consists of chest x-ray with a nasogastric tube in place. With a left diaphragmatic injury, the stomach may be herniated into the chest and the nasogastric tube confirmatory of the injury. Helical CT is currently the most useful exam with 70–100% accuracy (fig. 3.7). MRI is as accurate but commonly not available in the acute setting and requires much more time. This is costly especially during the golden hour. Up to one-half of diaphragmatic injuries are not found upon initial diagnostic evaluation. Nearly one-third are found intraoperatively, either incidentally or with minimally invasive surgical techniques. Isolated diaphragmatic rupture from blunt trauma is more common in children and occurs more frequently on the left side.

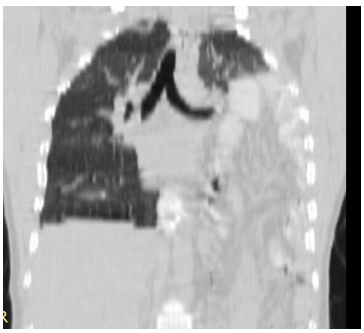


Fig. 3.7. Chest CT coronal ereconstruction in 13 years old patient with traumatic left diaphragmatic hernia. Intestine is identified in the left hemithorax.

Esophageal injuries.

In children esophageal injuries occur relatively rarely in less than 1% of children sustaining either blunt or penetrating thoracic trauma. But severity of development and difficulty of treatment make a problem that should be decided urgently, otherwise serious complications may develop. Determination of esophageal traumas is a responsive and difficult diagnostic problem that may be eased by presence of subcutaneous emphysema and mediastinal process. Subcutaneous emphysema on the neck develops lately, the milder is the trauma, the later it will appear. If in case of perforation of the cervical department it appears in 6-12 hours, then in case of perforation of the thoracic department it may be absolutely absent. The most vivid manifestation of esophageal disruption is increasing emphysema of the neck, anterior chest walls and face during endoscopic examination. It is connected with intense air insufflation into esophageal lumen during the manipulation.

Diagnostics of esophageal injuries is based on anamnesis, patient's complaints, clinical data (symptoms of shock, mediastinitis, disordered breathing), instrumental methods of diagnostics such as esophagography, esophagoscopy. Esophagography with a water-soluble contrast agent and esophagoscopy are typically the studies that will identify an esophageal injury (fig 4.8). A chest X-ray may demonstrate a pneumomediastinum, air or fluid in the pleural spaces (fig. 4.9) and/or subcutaneous emphysema. A CT chest with a water-soluble, oral contrast study will give a definitive diagnosis as well as information regarding the site of the rupture, the compartments with which it communicates, and the size of the leak.

Esophageal injuries may be complicated by concomitant injuries or by a pneumo- or hemothorax. Leakage of saliva and/or gastric contents may result in an empyema or mediastinitis either of which could lead to potentially life-threatening sepsis; therefore, broad spectrum antibiotics are commenced early in the treatment of esophageal injuries.

Careful, long-term follow-up is essential after the repair or reconstruction of an injured esophagus as strictures are an almost inevitable consequence of these injuries. In addition, all patients who have suffered an esophageal injury should be adequately covered for gastroesophageal reflux with proton pump inhibitors.

Operative management of these patients is dependent on the timing of the surgery and the severity of the injury. It is acceptable within 24 hours after injury. Where possible a primary repair of the injured esophagus is preferable; however, in some cases such as when there has been a delayed diagnosis or massive pleural or mediastinal soiling a two-stage repair is advisable. In these situations proximal diversion should be carried out to avoid continued contamination of the mediastinum and to allow effective control of any infection. These patients will also require the formation of a gastrostomy for feeding until the definitive repair can be performed. Regardless of the operative path taken a mediastinal drain should be inserted intraoperatively to assist in controlling the potential infective complications.

Many of these injuries can be managed conservatively with good analgesia and close monitoring in an institution with access to pediatric surgeons and anesthetists. These patients should all have a nasogastric tube inserted under radiological guidance and regular chest physiotherapy. Frequent suctioning of the oropharynx may also be required. In cases where there

is a significant pneumothorax or a pleural effusion is present insertion of an intercostal catheter may be necessary. These patients are at risk of infection and therefore broadspectrum intravenous antibiotics should be commenced. If the esophageal laceration is large or is not responding to conservative management surgical repair of the injury may be required.

Treatment includes the following parts: antibacterial therapy, draining the mediastinum (upper part by Razumovskiy, middle and lower – by intra pleural or extra pleural access after Nasilov); at considerable injuries of the esophagus the measure administered is closure of the last and drainage of the mediastinum, also application of gastrostomy.

Esophageal foreign bodies.

Foreign bodies that lodge in the esophagus are potentially a serious problem that peaks in children between the ages of 6 months and 3 years. Failure to recognize or appropriately manage a foreign body in the esophagus may lead to complications and occasional mortality. Foreign bodies are most likely to lodge where the esophagus is narrow. These areas include the upper esophageal sphincter formed by cricopharyngeaus, the level of the aortic arch (T4) where the esophagus is indented on the left by the aortic arch, and the lower esophageal sphincter at the gastroesophageal junction. The vast majority of esophageal foreign bodies occur in a normal esophagus. However, underlying esophageal pathology should always be considered. The most common cause is an esophageal stricture in conjunction with esophageal dysmotility following repair of esophageal atresia. A foreign body may be the first presentation of peptic stricture of the esophagus. The most common foreign bodies are coins, which account for up to two-thirds of all esophageal foreign bodies in children. Other foreign bodies less commonly encountered include needles, hairpins, disc batteries, and bones (usually fish or chicken). Esophageal foreign bodies can damage the esophagus and lead to strictures. Objects may erode the esophageal mucosa into surrounding tissues and cause esophageal perforation or tracheoesophageal fistulae. This in turn may be complicated by the development of an empyema. If the object erodes into the aorta, rapid exsanguination and death may follow. Disc or alkaline batteries lodged in the esophagus are of special concern; the corrosive effect on the esophagus is caused by the production of sodium hydroxide as current flows from the anode to the cathode.9 Animal studies have shown that full thickness necrosis of the esophagus can occur within 4 h, resulting in esophageal perforation, tracheoesophageal fistula, or aortoesophageal fistula. Following the removal of a disk battery, follow-up is essential to exclude esophageal stricture.

At injuries of the esophagus by a foreign body children often complain about pain at swallowing and dysphagia. The most common symptoms are drooling, gagging, and refusal of oral intake. Respiratory symptoms such as wheezing or tachypnea may be presenting symptoms if the esophageal obstruction causes secondary airway compression.

All foreign bodies are distinguished as X-ray positive which are seen on the film (fig. 3.10) and X-ray negative. Up to 65% of all foreign body bodies are radiopaque. The common exceptions include wooden, plastic, and glass objects, as well as fish and chicken bones, which may not be seen on plain radiographs. If a non-radiopaque foreign body not identified on plain X-ray is still suspected on clinical grounds, then either upper GI contrast or esophagoscopy should be considered.

Most foreign bodies that lodge in the esophagus fail to pass spontaneously once the child presents to hospital and therefore require a surgical opinion. Rigid or flexible endoscopy under general anesthetic and removal of the foreign body would appear to be the most effective means of treatment. Button batteries require urgent removal because of the risk of esophageal damage as discussed previously. Follow-up with contrast studies or endoscopy to exclude a stricture following its removal is recommended in all cases.

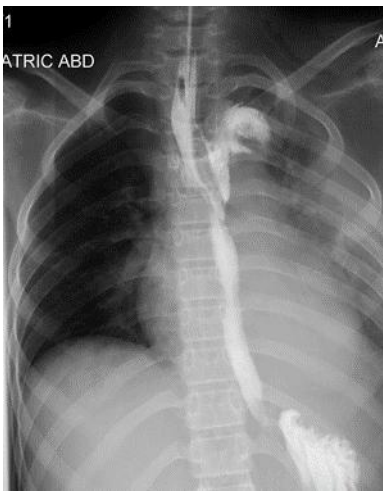


Fig. 3.8. Esophagography demonstrating contrast medium extravasation due to the breach of esophagus

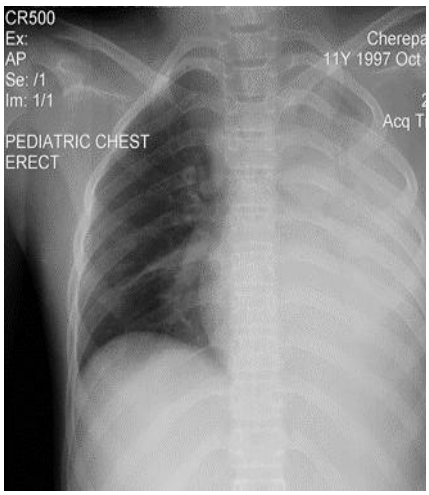


Fig. 3.9. Chest X-ray showing pleural effusion in patient with mediastinitis due to esophageal injury

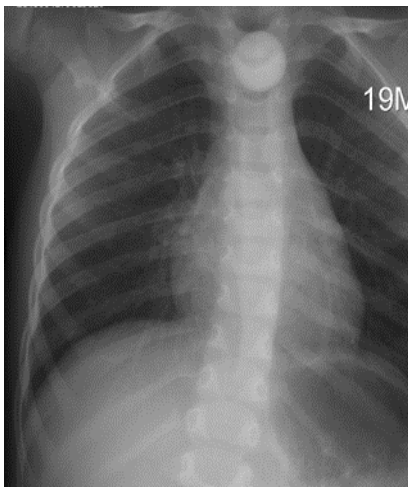


Fig. 3.10. X-ray poisitive foreign body of the esophagus

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Tests for initial level of knowledge

1. A patient in a traumatic shock has AP - 50/0 mm, heart rate -160/min. What is the shock index:
   A. 2,5
   B. 1,5
   C. 3,2
   D. 0,5
   E. 0,3
2. The correct method of treatment of tension pneumothorax is:
   A. Bronchial occlusion
   B. Puncture of the lungs with introduction of antibiotics
   C. Bronchoscopic drainage
   D. Conservative therapy without intervention into the focus
   E. Thoracentesis, pleural drainage by Bulau
3. A patient, 10 years, was referred to the surgical department from the place of road accident with blunt thoracic trauma and fractured ribs on the right side. The patient was indicated puncture and drainage of the pleural cavity. What is the place of the pleural puncture?
   A. In the 3rd -4th intercostum by the medial-inguinal line;
   B. In the 6th intercostum by the posterior-inguinal line;
   C. In the 7th intercostum by the line of the spatula;
   D. In the pleural sinus projection;
   E. In the place of the most expressed dullness determined at percussion

4. A child, 9 years, with destructive pneumonia, the condition abruptly aggravated, there increased dyspnea, the child became agitated, body temperature increased to 38,4 °C. Chest X-ray showed

homogenous opacity up to the third rib on the left, the mediastinum is shifted to the left. What is the most probable diagnosis?

A. Tension pneumothorax
B. Tension pyopneumothorax
C. Diffude pneumonia
D. Diaphragmatic hernia
E. Atelectasis of the lung

5. Cyanosis and dyspnea were revealed in time of casualty exam after the road accident. The condition of the patient is severe, right hemithorax does not take part in breathing, the right intercostal spaces are distended, percussion there reveals tympanic sound, and no breathing at auscultation. What is your diagnosis?

A. Open pneumothorax
B. Pneumoperitoneum
C. Acute purulent pleurisy
D. Tension right pneumothorax
E. Total right haemothorax

6. According to the data of contrast X-ray examination the child has oesophageal perforation. Is endoscopic examination required in this case?

A. Yes
B. No
C. It is contraindicated
D. It is indicated after the operation
E. It is indicated during the operation

7. A 16 years old patient is staying in a hospital with severe traumatic shock, combined thoracic and abdominal trauma. Shallow breathing, BP is 80/60 mm, pulse is 115 beats per minute, breathing is 42 per minute. What are an emergency care of respiratory disorders?

A. Narcotic analgesics.
B. Artificial respiration.
C. Emergency surgery with blood reinfusion.
D. Mechanical ventilation.
E. Central analgesics.

8. The key first aid steps for the patient in traumatic shock are:

A. Effective analgesia and cooling of the body areas;
B. Effective analgesia and glucocorticoids;
C. Effective analgesia and infusion therapy;
D. Effective analgesia and cardiac glycosides;
E. Effective analgesia and transport immobilization

9. What is to be used to wash the stomach if the child swallowed the vinegar essence?

A. Solution of an alkali
B. Acidic solution
C. Neutral liquid (water)
D. Solution of potassium permanganate
E. No correct answers

10. What is the pathognomonic sign of the traumatic injury of the heart?

A. Impaired rhythm
B. Haemopericardium
C. Hypotension
D. Tachycardia
E. Bradycardia

Keys for tests

1 – C

2 – E
3 – C
4 – E
5 – D
6 – B
7 – D
8 – C
9 – C
10 – B

Tests for final level of knowledge, keys for tests

1. The examination of the victim of the road accident showed cyanosis, labored breathing. The patient's condition is severe, right part of the thorax gaps in the breathing, the intercostal areas are distended on the right, box shade at percussion, at auscultation breathing is absent. Your diagnosis:
    A. Open pneumothorax
    B. Pneumoperitoneum
    C. Acute purulent pleuritis
    D. Tension pneumothorax
    E. Total haemothorax on the right
2. What should be used to wash the stomach if the child swallowed alkaline solution?
    A. Alkaline solution
    B. Acid solution
    C. Neutral solution (water)
    D. Potassium permanganate solution
    E. No correct answers
3. The main curative measures to treat the patient in traumatic shock are:
    A. Effective analgesia and cooling the body areas with burns
    B. Effective analgesia and introduction of glucocorticoids
    C. Effective analgesia and infusion therapy
    D. Effective analgesia and introduction of cardiac glycosides
    E. Effective analgesia and transport immobilization
4. A boy, 12 years, who suffered in a catastrophe, has closed fragment fracture of the femur diaphysis, concussion, multiple rib fractures and pneumohemothorax, scalping injury of the lower leg. Which of the injuries is the dominant one?
    A. Closed fragment fracture of the femur diaphysis.
    B. Multiple rib fractures and pneumohaemothorax
    C. Concussion
    D. Scalped injury of the lower leg
    E. All traumas are equal
5. A patient, 15 years, after the road accident complains about dyspnea. Objectively: the skin is pale, cyanotic. Subcutaneous emphysema in the thorax area, abdomen, right side of the neck. At auscultation: no reapiration on the right. Heart contraction rate -130/min.,AP – 80/60 mm, central venous pressure – 140 mm, breathing rate – 30/min., Ht – 0,27, Hb – 90 g/l. The subsequent therapy must include:
    A. Urgent artificial lung ventilation
    B. Massive infusion therapy with crystalloid solutions
    C. Dophamine infusion, 2-5 mkg/kg
    D. Puncture of the pleural cavity on the right.
    E. Oxygenation with 100 % oxygen
6. By what ways does air penetrate to pneumomediastinum?
    A. As a result of disruption of pleura covering the mediastinum organs
    B. By the way of demarcation of the visceral pleura

C. By the connective tissue of the lungs across the vessels and bronchi

D. By the connective tissue of the lungs across the vessels and bronchi with demarcation of the visceral pleura

E. No correct answers

7. A patient was delivered in an hour after the car accident with complaints about pain in the right part of the thorax, hard breathing. At examination: on the right there are superficial scratches, palpation reveals fracture of the IVth and Vth ribs on the right. At auscultation- the breathing isn't heard on the right. At percussion- in the lower departments before the 5th rib there is dullness. AP -100/70 mm, pulse 106 beats per min. What is the diagnosis?

A. Rib fracture, pneumohaemothorax

B. Contusion of the thorax, rib fracture

C. Contusion of the thorax, lung trauma

D. Contusion of the thorax, rib fracture, subcutaneous haematoma

E. Additional examination is required

8. A 12 years old boy after car accident has: diagnosed comminuted fracture of the diaphysis of the femur, concussion, multiple rib fractures and haemopneumothorax, laceration of the shank. Which one of the named injuries is dominant one?

A. Closed comminuted fracture of the diaphysis of the femur;

B. Multiple rib fractures and haemopneumothorax;

C. Brain concussion;

D. Laceration of the shank;

E. All injuries are equal

9. A 15 years old patient after the road accident complains about sudden dyspnea. Objectively: pale and cyanotic skin. Subcutaneous emphysema is at the right hemithorax; right half of abdomen, and neck. Auscultation: no breathing on the right, heart rate is 130 beats per minute, BP 80/60 mm, central venous pressure is 140 mm, respiration is 30 per minute, Ht – 0,27, Hb – 90 g/l. Emergency therapy must include:

A. Artificial ventilation

B. Massive infusion therapy by crystalloid solutions;

C. Infusion of dopamine, 2-5 mcg/kg min;

D.Puncture of the right pleural cavity;

E. Oxygenation by 100 % oxygen.

10. A 4 years old child received blunt chest injury after falling down from the ladder onto the packed soil. Aggravated dyspnea and cyanosis, left hemithorax is out of breathing, no breath on the left chest, heart sounds are shifted to the right. There is subcutaneous crepitation. Plain X-ray shows left hydropneumothorax, pneumomediastinum. The initial diagnosis is blunt chest trauma, hemopneumothorax, pneumomediastinum. What is your first aid management?

A. Administration of analgesics, left thoracentesis, anti-shock therapy;.

B. Analgesics, anti-shock therapy, suprajugular mediastinomy;

C. Left thoracentesis, anti-shock therapy

D. Anti-shock therapy, artificial respiration, antibiotics, suprajugular mediastinomy;

E. Anti-shock therapy, left thoracentesis

Keys for tests

1 – D
2 – C
3 – C
4 – B
5 – D
6 – D
7 – A
8 – B
9 – D

10 – C

Tasks for final level of knowledge

1. A patient was hospitalized with complaints of acute sharp pain in the left hemithorax, dyspnea. A day ago he fell down from 2.5 m of height. Fractures of the 6th, 7th and 8th ribs and horizontal level of fluid at the 4th rib were revealed at plain chest X-ray. Left haemopneumothorax was diagnosed. What treatment should be performed?
   The answer is **Puncture of the left pleural cavity and thoracentesis in the 5th intercostal space by the medium axillary line**
2. Cyanosis and dyspnea were revealed at the time of casualty exam after the road accident. The condition of the patient is severe, right hemithorax does not take part in breathing, the right intercostal spaces are distended, percussion there reveals tympanic sound, and no breathing at auscultation. What is your diagnosis?
   The answer is **Tension pneumothorax**
3. A 15 years old patient after the road accident complains about sudden dyspnea. Objectively: pale and cyanotic skin, subcutaneous emphysema at the right hemithorax, right half of abdomen, and neck. Auscultation: no breathing on the right, heart rate is 130 beats per minute, BP 80/60 mm, central venous pressure is 140 mm H_2O, respiration is 30 per minute, Ht – 0,27, Hb – 90 g/l. Emergency therapy must include:
   The answer is **drainage of the right pleural cavity**
4. A 4 years old child had blunt chest injury after falling down from the ladder onto the packed soil. Aggravated dyspnea and cyanosis, left hemithorax is out of breathing, no breath on the left chest, heart sounds are shifted to the right. There is subcutaneous crepitation. Plain X-ray shows left hydropneumothorax and pneumomediastinum. The initial diagnosis is blunt chest trauma, hydropneumothorax, pneumomediastinum. What is your first aid management?
   The answer is **Left thoracocentesis, anti-shock therapy**
5. A 10 years old patient was hospitalized at surgical department after the road accident, with blunt chest trauma. Ribs' fractures on the right chest and right pneumothorax were diagnosed. The tube thoracostomy was indicated. What is the point of procedure?
   The answer is **the 2nd intercostal space by the medium-clavicular line**
6. A patient was delivered to hospital in one hour after the road accident with complaints about pain in the right hemithorax, breathlessness. Examination revealed bruises on the right chest, and fracture of the 4th and 5th ribs was revealed absence of breath sounds in the right side, percussion under 5th right rib revealed dullness. BP is 100/70 mm, pulse is 106 beats per minute. What is the initial diagnosis?
   The answer is **Rib fracture, right hemothorax;**

Materials for self-study of the students

Main tasks	Notes (instruction)
Repeat: 1. Anatomy of organs of mediastinum, lobar structure of lungs, anatomy of pleural cavity, features of pulmonary circulation 2. Physiology of respiratory system 3. Pathogenesis of respiratory disorders in thoracic trauma	To sketch out the anatomy of lungs To represent the methods of diagnosis of diseases of lungs and pleura
Study: Technic of thoracic drainage Tactics in tension pneumothorax and hemothorax Tactics in thoracoabdominal trauma	To make differential diagnosis of injuries of mediastinal organs

	To make the indications to surgical treatment of variants of thoracic trauma

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THEME 4
POLYTRAUMA IN CHILDREN
Overview

Polytrauma patients are expected to have a higher risk of mortality than that obtained by the summation of expected mortality owing to their individual injuries. This study was designed to investigate the outcome of patients with polytrauma, which was defined using the new Berlin definition (2014), as cases with an Abbreviated Injury Scale (AIS) ≥ 3 for two or more different body regions and one or more additional variables from five physiologic parameters (hypotension [systolic blood pressure ≤ 90 mmHg], unconsciousness [Glasgow Coma Scale score ≤ 8], acidosis [base excess ≤ −6,0], coagulopathy [partial thromboplastin time ≥ 40 s or international normalized ratio ≥ 1,4], and age).

Aims of the lessons:

1. To teach students the main notions of polytrauma.
2. To interpret additional methods of examination (ultrasound diagnostics, roentgenology. Blood pressure).
3. To examine the abdomen, thorax and musculo-skeletal system: examination, palpation, percussion.
4. To offer an algorithm of doctor's actions at polytrauma in a patient.
5. To demonstrate abdominal examination: visual examination, percussion, palpation, auscultation).
6. What are the main principles of urgent therapy for patients with polytrauma?
7. Modern approaches to treating polytrauma, indications for operative intervention.
8. Determination of anatomical structures that are injured at polytrauma.
9. To interpret the data of additional and laboratory diagnostics.
10. To demonstrate pleural puncture and point the areas of puncture.
11. To define general principles of treatment.
12. To use the additional methods of diagnosis and define the indications for operative treatment.
13. To make a plan of examination and interpret the additional methods of examination (roentgenological, ultrasound, computed scanning, etc.), laboratory and biochemical analyses, hemodynamic indicators (Нв, Ht, blood circulating volume, central venous pressure, AP, Ps).

Basic knowledge and skills necessary for the topic (intradisciplinary integration):

Subjects	Skills
1. Anatomy	To describe peculiarities of possible variants of localization of organs of the abdominal cavity, skeletal system and blood circulation depending on the child's age.
2. Pharmacology	To define dosage of antishock blood-substituting, spasmolytic and analgetic medicationsdepending on the child's age, the peculiarities of treatment of children with polytrauma.
3. Physiology	To determine peculiarities of respiratory and digestive system in children of different age.
4. Physiopathology	To determine key moments of etiology andpathogenesis at traumatic injuries of the skeletal system, thoracic and abdominal cavity in children of different age.
5. Propaedeutics of padiatric diseases	To examine the digestive, respiratory and skeletal systems in children.
6. Operative surgery	To show schematically topography of the thorax, abdominal cavity and retroperitoneal space.
7. General surgery	To discuss clinical and laboratory examinations, main symptoms at traumas of the abdominal and thoracic cavities.
8. Roentgenology and ultrasound diagnostics	To make roentgenological examination, estimate the results, define main roentgenological symptoms. To appraise the data of

	ultrasound diagnostics, computed tomogram depending on character of pathology.

4. Tasks for individual self-preparation for the lesson.

4.1. The list of key terms and characteristics a student must know:

Term	Definition
1.Polytrauma	(AIS) ≥ 3 for two or more different body regions and one or more additional variables from five physiologic parameters (hypotension [syst. BP ≤ 90 mmHg], unconsciousness [GCS score ≤ 8], acidosis [BE ≤ −6,0], coagulopathy [PTT ≥ 40 s or INR ≥ 1,4], and age.
2. FAST-protocol	Focused assessment with sonography in trauma
MISS	Modified injuryseverity scale
ISS	Injury Severity Score
PTS	Pediatric trauma score
GCS	Glasgow coma score
"Waddell's triad"	Femur fracture after the bumper strike, a thoraco-abdominal injury after contact with the car hood, and a head injury after striking the ground
Salter–Harris fracture classification	Classification of fracture that involves the epiphyseal plate or growth plate of a bone, specifically the zone of provisional calcification
ARDS	Acute respiratory distress syndrome
4.Oliguria	Decrease of urine volume
5.Pneumohaemothorax	Blood and air in the pleural cavity
6.Mediastenal emphysema	Air in the anterior or posterior mediastinum resulting from thoracic trauma with simultaneous injury of the trachea or bronchi.

CONTENT
PEDIATRIC POLYTRAUMA

Description

Polytrauma is a combination of two or more injuries, one of which or their combination poses an immediate threat to the victim's life and is the direct cause of the development of traumatic illness.

Polytrauma is the leading cause of death and disability in children over a year old. Every year in Ukraine, 2-2.5 thousand traumatic deaths are registered in children aged 1 to 18 years, the number of admissions to medical institutions with injuries of varying severity is about 140 thousand. The intensive growth of transport and industrial injuries, as well as the increased number of natural and man-made disasters, local military conflicts and terrorist acts, significantly changed the structure of mechanical damage in polytrauma. Polytrauma is usually an unforeseen event that happens at home or on the street. Severe multiple and concomitant traumatic injuries have become dominant, in which it is almost impossible to separate the treatment of injuries of the musculoskeletal system and internal organs. There is a need for urgent transportation of a child with significant injuries from the scene or from one hospital to another. In the prehospital phase, the time allocated for emergency care should be effectively used to implement the basic principles of trauma resuscitation.

Initial assessment

The modern emergency trauma care system began to take shape in the middle of the last century. The history of the creation of the first emergency trauma care protocol named after the American podiatrist Jim Steiner, whose private plane crashed in February 1976 in Nebraska (USA). His entire family was taken to a peripheral hospital, where care was far from adequate. D. Steiner realized the inadequacy of helping his family and decided to change something.

Thanks to the work of a group of physicians at the American College of Surgeons, the first ATLS (Advanced Trauma Life Support) protocol for physicians was created in 1978. His ideology was based on the fact that timely and correct assistance to victims can significantly improve the outcome. At present, the ATLS course is compulsory in more than 50 countries around the world. This is a basic course for teaching doctors the basics of providing qualified care for polytrauma. The ATLS course provides the most up-to-date and effective clinical guidelines based on evidence-based medicine principles. Initial care for a child with an injury should follow the basic principles of trauma resuscitation. The recommendations of the latest revision are based on the sequence of therapeutic and diagnostic measures - **PDABCDE**, which must be observed in all patients with traumatic injuries: P - Protection, D - Decision, A - Airway Maintenance with Cervical Spine Protection, B - Breathing and Ventilation, C - Circulation with Hemorrhage Control, D - Disability (Neurologic Evaluation), E - Exposure / Environmental control**.**

When a casualty is admitted to the intensive care room, the first priority is to provide reliable personnel protection (**P-Protection**), including face shields and protective suits, in addition to the usual masks, gloves and gowns. This item is especially relevant given the spread of pandemic respiratory and vector-borne diseases, as well as the threat of bioterrorism.

The next important step is to make a decision to start or stop resuscitation (**D - decision**), taking into account the severity and potential resuscitation of the injured person (for example, decapitation, disruptionof most of the trunk, long duration of extrahospital life support, etc.). This is of particular importance in the field of triage in case of mass admission of victims. Effective assessment and forecasting on the scales – Trauma Score (TS), Revised Trauma Score (RTS), Injury Trauma Score (ISS), Glasgow Coma Scale (GCS). The initial examination is aimed at identifying life-threatening injuries: level of consciousness, frequency and efficiency of breathing, heart rate, pulse, blood pressure (BP), presence of external bleeding, condition of the head, chest (exclude pneumothorax, hemothorax), abdomen, dysfunction of the limbs , body temperature. Laboratory tests (with priority emphasis): group and Rh-belonging of blood, hemoglobin, erythrocytes, blood gases, parameters of the acid-base state, glucose, clotting indicators (platelets, activated partial thromboplastin time (APTT), international normalized ratio (INR), prothrombin index), electrolytes (Na, K, Ca, Cl), WBC, urea, creatinine, protein, transaminase (ALAT, ASAT), alpha-amylase, urinalysis.

A (Airway) - maintaining airway patency is necessary when assessing the Glasgow Coma Scale (GCS) £ 8–10 points, in case of multiple rib fractures, chest flotation, progressive decrease in SaO2 £ 80% with FiO2 ³ 0.3.

There is no need to dwell on the structural features of the child's airways, since every doctor knows that they differ significantly from adults in size and anatomical structure. You should only pay attention to the fact that the tongue in children is relatively larger, so it can often move up and back, which will lead to airway obstruction, especially when trying to ventilate with a bag and mask. The glottis in children is located closer to the oral cavity and anteriorly than in adults, and the larger the size of the soft tissues of the tongue and epiglottis, the more justified it is to use a straight blade for tracheal intubation.

If the child is conscious, then you can assess the patency of the airway by asking his name. A clear and understandable answer or any spoken word indicates that the airway is passable. If the child is unconscious, then it is very difficult to assess. In many children, in the supine position, partial obstruction of the airways is often noted due to hypertrophy of the tonsillar and adenoid tissues, and it is possible to judge the patency of breathing in such children only in the lateral position. Since most injured children are transported on their backs, their airway patency may be slightly impaired for these reasons. It is a mistake to intubate a child if stabilization can be achieved without invasive intervention, but one should always ensure that there is no foreign body. The position of the child's head is important. This problem can be easily solved by placing a pillow under the back of the head, which slightly unbends the neck and makes the airway more passable. Carefully manipulate the cervical spine. If the child is

unconscious and no manipulations restore the patency of the airways, use an oro- or nasopharyngeal airway tube. Its size should not be more than the distance from the corner of the jaw to the chin. A poorly fitted airway tube can displace the tongue posteriorly and cause obstruction, oropharyngeal airways that are too long can stimulate reflex activity, leading to vomiting and possible aspiration of gastric contents.

Endotracheal intubation is required in a small number of traumatized children, usually with brain damage. Patients in a state of clinical death or coma do not require premedication before intubation.

B (Breathing) - providing immediate oxygenation, adequate assisted breathing, or mechanical ventilation based on a protective artificial lung ventilation (ALV) strategy.

The assessment of the ventilation quality of injured children at the scene of the accident is based on clinical indicators. The frequency and depth of breathing efforts, combined with the presence of well-conducted breathing over both halves of the chest, are the best indicators of its adequacy. Factors such as fear and pain can significantly affect the respiratory rate and must be taken into account when assessing breathing.

All affected children receive oxygen therapy. The physician should not be bothered by the fact of oxygen toxicity, since the therapy is short-term and the children have no previous pulmonary disease. If the child has fear of suffocation in the face mask, oxygen can be supplied through nasal cannulas.

In young children, obstruction of the upper airways is accompanied by significant inspiratory efforts with retraction of the sternum and front of the ribs. In such a situation, at first, the patency of the upper airways is restored (correct body position, bringing the jaw forward, oropharyngeal airway tube), then, if the result is not achieved, endotracheal intubation is performed to establish mechanical control over breathing. Auscultation over the surface of the lungs should rule out the presence of hemothorax, pneumothorax, or cardiac tamponade. If a child has any of the listed problems, but his condition is stable and will soon be delivered to a hospital, then it is better not to intervene. If, during transportation, symptoms appear in the form of rapid breathing, an increase in "congestion", hypoxia or cyanosis, in combination with physical data, and the clinic worsens, it is necessary to perform simple and quick interventions, the essence of which is aimed at increasing oxygen delivery and/or providing ventilation.

In the presence of a growing pneumothorax, we suggest that in all children, a catheter should be inserted along the anterior axillary or mid-axillary line at the level of the fifth intercostal space (nipple level). It is necessary to penetrate into the pleural space only above the rib. After evacuating the air, the catheter should be secured and connected to a unidirectional valve (Bulau type).

C (Circulation) - control of blood circulation, stopping external bleeding, ensuring venous access, controlling blood loss, replenishing blood loss, diagnosing internal bleeding, eliminating dangerous injuries to prevent the lethal triad (hypothermia, coagulopathy, acidosis).

Assessment of the state of circulation includes: determination of the peripheral pulse, blood pressure, skin perfusion (“white spot” symptom), rhythm and heart sounds, as well as the child's consciousness. It is rather difficult to determine the degree of intravascular volume deficit at the prehospital stage; it is necessary to focus on all of the above parameters at the same time.

Bleeding is the main problem with trauma. In this regard, its control and replenishment of the BCV (blood circulation volume) is the main therapeutic effect in such a situation, since not only the volume of blood loss must be reimbursed, but also the extravascular and extracellular fluid lost in this case.

If the initial therapy includes the infusion of crystalloid fluids, such as 0.9% NaCl solution or Hartman's solution, the amount needed to replace the loss is roughly three times the estimated blood loss. The accuracy of its assessment in a child is determined only in the hospital. A lot of blood loss is noted with tears in the face and skull, as they are extremely well supplied with blood, therefore all deep tears in this area must be adequately bandaged using pressure bandages at the scene. If in adults intracerebral bleeding does not lead to hypotension, then in

infants the opposite picture is observed. In this regard, blood loss in infants in the cranial cavity must be treated with intensive fluid therapy along with other measures of cerebral resuscitation.

Intrathoracic, intra-abdominal and pelvic bleeding can also lead to significant hypotension. In a child with a hip fracture, blood extravasation is much less pronounced than in adults due to the lower muscle mass. Therefore, if hypotension is noted with an isolated hip fracture, it is necessary to look for another source of bleeding.

Tachycardia and peripheral vasoconstriction are two early symptoms of hypovolemia, in which the change in blood pressure in the child occurs somewhat later. It is an indicator of significant blood loss only if the BCV deficiency is at least 30-40%. A more reliable indicator of a decrease in intravascular volume is the heart rate, which should be carefully monitored, since the change in heart rate is observed long before the decrease in blood pressure. It is necessary to take into account the fact that most children experience fear, which can also increase their heart rate, this feeling is always present in the corresponding situation. In a state of fear, the child has normal blood pressure and tachycardia, but the rate of filling of the capillaries is not increased and the filling of the pulse is not worsened.

Measuring blood pressure in children is often problematic; using the wrong cuff size will not provide accurate results, which entails a large number of errors.

At the scene, only percutaneous intravenous access is used with catheters on a needle with the maximum possible size, for peripheral cannulation - the elbow, the back of the hand, the femoral vein on the ankle, with significant blood loss and subsequent vasoconstriction, attempts to percutaneous catheterization of the vein can be difficult. Preferred starting intravenous solutions in children, as in adults, are 0.9% sodium chloride solution, Ringer's or Hartman's solution without dextrose, they contain physiological amounts of sodium and chloride. Lactate added to Hartmann's solution is readily metabolized by the liver under normal perfusion and does not cause lactic acidosis. Immediately after injury, glucose cannot be administered intravenously, since the normal endocrine response of the child at this moment, like in an adult, leads to the secretion of a large number of hormones, including catecholamines, glucagon, growth hormone and various steroids, each of which mobilizes glucose from the glycogen stores of the liver and muscles. The added exogenous glucose will be not utilized by the body and will cause an increase in its concentration to exorbitant figures. The developed hyperglycemia will lead to osmotic diuresis, a decrease in vascular volume and dehydration, as in diabetic ketoacidosis, and incomplete utilization of the glucose molecule in the damaged brain tissue will cause its damage.

Therefore, the ideal solution for intravenous use in children after traumatic injury is Ringer's lactate or Hartman's solution without dextrose. In case of symptoms of hypovolemia, to restore volume, these solutions are injected bolus (two or three boluses) at a rate of 20 ml / kg, they compensate for both intravascular and extravascular fluid loss by the body. Further need for fluid and blood will be determined by ongoing bleeding and the need for surgery.

D (Disability) - impaired consciousness: exclude or correct hypoxia or hypotension; evaluate pupils (diameter, symmetry, photoreaction); quickly assess the level of neurological deficit on the Glasgow Coma Scale; evaluate blood glucose levels to exclude hypoglycemia. All patients with blunt skull trauma should be evaluated for possible injury to the cervical or thoracic spine.

There are a large number of scales to assess the severity of trauma in children: trauma index, injury severity index, sorting index, trauma scale, hospital trauma index, CRAMS (Circulation, Respiration, Abdomen, Motor, and Speech) scale, Glasgow Coma Scale (GCS), Pediatric Trauma Scale (PTS). GCS is one of the most used scales, but it is only suitable for injuries of the central nervous system; its modification should be used to assess infants and children. The PTS is the most widely used pediatric scale. It uses the relative values of six components of injury in children that are relevant to mortality and disability, which include weight, airway status, blood pressure, level of consciousness, open wounds and fractures. A score of less than 8 points indicates a threat of disability or death, and such children should be

urgently hospitalized in a specialized medical institution. Contusion and trauma to the chest with rib fractures, especially when combined with other injuries, increase the likelihood of disability and death in children. The ribs in children are extremely pliable and deform significantly without breaking. The force of impact, leading to fractures of the ribs in children, is much greater than in adults. Therefore, when rib fractures are present, severe pulmonary injury must be assumed, as well as severe damage to organs in the immediate vicinity. With a combination of rib fractures and severe brain damage, the mortality rate is 71%.

E (Exposure / Environmental control) - examination of the patient. For adequate examination, the patient should be nude, provided that heat loss is minimized. Urgent measures: assessment of the severity of injuries; empirical calculation of the degree of blood loss.

Children cool down very quickly. There are two main consequences of significant heat loss and temperature drop. This is a violation of peripheral perfusion in favor of vital organs and the development of lactic acidosis, as well as a violation of the normal mechanisms of blood coagulation due to an increase in prothrombin time and partial activation of thromboplastin. Therefore, every effort should be made to rewarm the child after injury.

Damage in a child to other areas of the body, where pain is felt more strongly, always distracts the doctor's attention from identifying spinal and vertebral injuries. In this regard, in children, it is necessary to pay great attention to the immobilization of the cervical vertebrae, as well as the thoracic and lumbar spine. In children, various injuries of the cervical spine are most often observed at the level of C_1, C_2, C_3, therefore, maintaining and maintaining the position along the midline will be the best way to immobilize it. In this case, a well-fitted neck collar must be used. Control of limb injuries: stopping massive external bleeding by temporarily pressing or applying a tourniquet, taking into account the permissible compression time, fixing fractures, treating compartment syndrome (fasciotomy, laparotomy, thoracotomy); the fight against crash syndrome, the use of efferent methods (hemofiltration, hemodialysis, plasmapheresis). In case of damage to large vessels and signs of circulatory disorders in the limbs, immediate surgical intervention is recommended. After ensuring control of the limb injuries, it is necessary to return to the activities of points C and D.

Systemic response to trauma

To improve the outcomes of treatment of the most severe polytrauma, the Hannover medical school of polytrauma proposed a system of so-called “damage control”, according to which the surgical treatment of injuries of both internal organs and the musculoskeletal system is divided into two stages: short-term operations such as mini-craniotomy for epi- and subdural hematomas, laparotomy with clamps on the spleen pedicle and tamponade of liver rupture, puncture epicystomy, etc., and fractures of large bones, especially the hip, are immobilized with external fixation devices. Then the victim undergoes intensive therapy until the hemodynamic and other indicators of homeostasis are completely stabilized, and after 1-2 days, reconstructive operations on the internal organs are performed, and after 5-7 days and later, minimally invasive osteosynthesis of long bone fractures is performed. Such tactics significantly improved the outcomes of severe polytraumas and made it possible to preserve the life and health of the victims, who were previously considered hopeless. Separate protocols for "damage control" for abdominal, thoracic, craniocerebral, spinal and orthopedic injuries were identified, which received the corresponding abbreviations - for example, DCS (damage control surgery) - "damage control" of the abdominal and thoracic cavity, DCO (damage control orthopedics) - "Damage control" of the musculoskeletal system.

The basis for the introduction of the "damage control" system was the immunological studies carried out in patients with polytrauma. According to the results of these studies, tissue destruction triggers a local inflammatory response with an increase in the total concentration of pro-inflammatory cytokines. Cytokine levels correlate with the degree of soft tissue and bone damage. Local inflammatory response activates polymorphonuclear leukocytes, which attach to capillary endothelial cells and stimulate the release of free oxygen radicals and proteases, resulting in damage to the vessel wall, leading to interstitial edema. All these processes are

known abroad as multiple organ dysfunction syndrome (MODS). The release of pro-inflammatory cytokines and products of damaged cells forms systemic inflammatory changes, which is facilitated by the presence of ischemic ones. dead and infected tissue. This explains the high frequency of infectious complications (primarily pneumonia) in patients with polytrauma of specific complications such as respiratory distress syndrome in adults, early multiple organ failure, etc. In order to apply damage control in practice, three factors must be carefully evaluated:

1) the severity of the initial injury ("first hit" - the first hit);

2) the biological constitution of the patient (age, body weight, concomitant diseases);

3) the number of required trauma operations, their expected duration and trauma (blood loss). These operations are the "second hit" for the seriously injured person.

General principles of treatment

A unified approach to assessing the nature of trauma, understanding the patterns of the course of the pathological process makes it possible to develop coordinated tactics in the provision of resuscitation and surgical care. Timely and adequately provided medical care to victims with polytrauma in the first period of traumatic illness is of particular importance.

It is known that mortality due to polytrauma is divided into three groups. The first group of deaths occurs shortly after injury and is caused by injuries that are incompatible with life (brain, aortic rupture, etc.). The second type of death - from several minutes to several hours after injury - during potentially surgically corrected (subdural and epidural hematomas, ruptures of the spleen, liver, kidneys, etc.), the lack of timely treatment can lead to massive blood loss and death. The third group of deaths is made up of children who develop sepsis or multiple organ failure within a few days after the injury. The concept of "golden hour" is more applicable to patients of the second group, when their existing injuries are still potentially corrected. The “golden hour” represents a critical time window for therapeutic and surgical interventions in children with polytrauma that reduce the incidence of death.

Since most pediatric traumatic injuries occur with a blunt object, several organs are usually damaged as a result of its impact, therefore, children are characterized by a combination of craniocerebral trauma with bone fractures, chest and abdominal injuries. The approach to such polytrauma consists in the implementation of resuscitation measures aimed at restoring the circulating blood volume and optimizing the delivery of oxygen to the tissues. In this case, the small amount of time spent on providing venous access at the scene of the accident, restoring airway patency and axial immobilization for the purpose of hemodynamic and cerebral support, as well as limiting the effect of any unrecognized spinal injury will be justified.

The first stage of providing care in a hospital setting is resuscitation. They carry out activities to preserve life. In parallel with life-saving measures in this phase, clinical trials are carried out aimed at detecting circulatory disorders, breathing disorders, symptoms of brain compression, and spinal cord injury.

In case of combined injuries, bone and abdominal pathology is inextricably linked, therefore Focused Assessment with Sonography for Trauma (FAST protocol) is included in the ATLS recommendations as a mandatory initial diagnostic study of patients with polytrauma or abdominal trauma to detect hemoperitoneum, hemopericardium, hemothorax and pneumothorax (Fig. 4.1). This study allows you to quickly (within 3-3,5 minutes) determine the surgical tactics of further patient management simultaneously with resuscitation measures.

Figure 4.1. FAST (focused assessment with sonography in trauma). Localization of the US sensor

1-subxyphoidal (pericardial space) – heart, major vessels

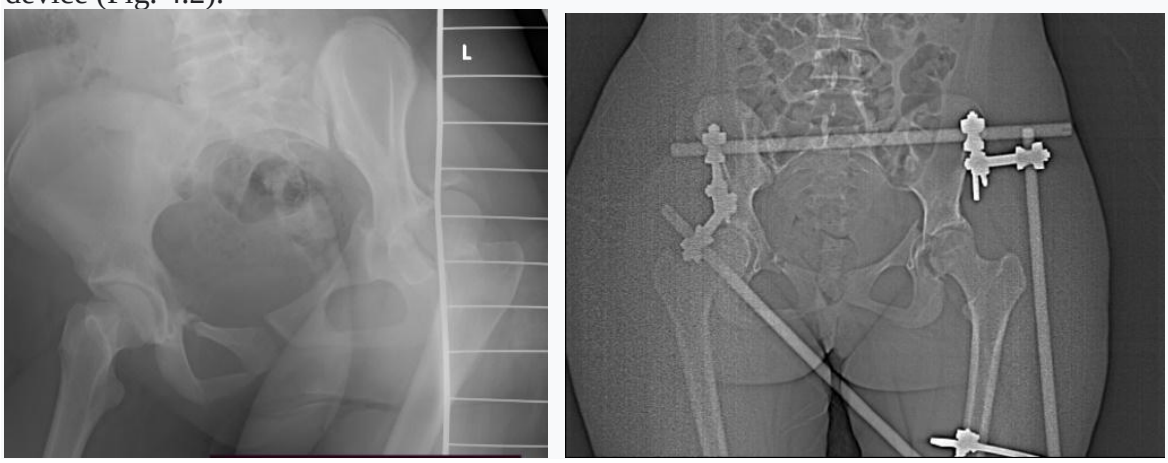
2. hepatorenal pocket (Morrison's bag) – liver, right kidney

3. splenorenal space – spleen, left kidney

4. pelvis (Douglas space) – retroperitoneal space, hollow organs, diaphragm

The manipulations of the first minutes include the operation of thoracic drainage. Indications for this manipulation are considered tension pneumothorax. Priority is given to urgent surgical interventions to stop massive bleeding in the body cavity, decompressive trepanation, elimination of cardiac sac tamponade, that is, operations that save the patient's life.

The second stage is urgent operations. Operations are performed according to vital indications - stopping massive bleeding in case of rupture of the liver or spleen, damage to large thoracic or abdominal vessels, open injury to the pelvis, damage to the great vessels. Simultaneously with operations for health reasons, intensive treatment of shock is carried out. With the guarantee of vital functions, patients with polytrauma receive the necessary amount of surgical care. Stabilization of fractures of long bones, unstable injuries of the spine, pelvic ring, large joints is important. In case of combined abdominal injuries and unstable pelvic injuries, after stopping intra-abdominal bleeding, the pelvic ring is stabilized with an external fixation device (Fig. 4.2).



a b

Figure 4.2 (**a**) AP pelvis of a polytraumatazed child 14 y.o. (after catatrauma) with hemodynamic instability who sustained a traumatic head injury, closed thoracic trauma and a displaced pelvic ring fracture among other fractures. This child's hemodynamic status stabilized after application of the external fixation (apparatus «pelvic-femur») (**b**).

The third stage is stabilization of the state. After performing operations according to vital indications, the patient is given infusion therapy in the conditions of the intensive care unit in order to normalize the functions of respiration, cardiovascular system, blood coagulation system, peripheral tissue metabolism. Depending on the severity of the injury, the stabilization phase lasts from 6-12 hours to several days. The goal of this phase is to stabilize the most important organs and systems and prepare as quickly as possible for the operations of the next stage. In this phase, a computed tomogram of the skull, X-ray of the chest, abdomen, pelvis, and extremities are performed. A trauma management strategy is adopted.

The fourth stage is deferred operations. Stabilizes all diagnosed limb fractures. Fractures of long bones, unstable injuries of the pelvic ring, severe instability of the spine have priority for delayed intervention after anti-shock therapy and stabilization of vital functions. Stabilization of fractures makes it possible to eliminate pain and stress, prevent further tissue trauma, stop bleeding, and treat traumatic brain and chest trauma with an elevated position of the upper half of the body or a free drainage position. The fifth stage is rehabilitation.

ABDOMINAL BLUNT TRAUMA

Liver injuries

The liver is the largest organ of the abdominal cavity, and therefore is easily damaged by injuries of the lower third of the chest and upper third of the abdomen. In children liver is easily injured even with a slight impact on it because its fragile parenchyma contains less connective tissue. Liver thin-walled vessels do not shrink after injury, which increases the intensity and duration of bleeding. Impurities of bile in the spilled blood increase the clotting time.

At slaughter the right share of a liver is torn 6 times more often, than the left, and its diaphragmatic surface is damaged twice more often.

There are several types of liver damage: rupture, crushing, laceration, and burst.

The rupture occurs when applying direct blows to a large area of the upper abdomen or lower sternum. Simple ruptures occur because of crushing, when the compression is not very strong.

Crushing is characterized by multiple cracks with deep ruptures of the parenchyma, which cross the liver tissue.

Laceration is separation, complete or incomplete, of liver tissue pieces. It occurs when falling from a height.

Liver burst is a type of rupture with the formation of multiple star cracks, occurring due to the application of a short and strong blow to a small surface. Sometimes the liver capsule does not rupture, but subcapsular or central ruptures and hematomas of the liver parenchyma occur.

Pain and subsequent bleeding after liver trauma lead to the rapid development of shock. Conservative measures can temporary improve child's condition, but if the bleeding does not stop, hemodynamic disorders progress again. The pain in liver injuries is constant, localized in the right hypochondrium, right half of the abdomen or where the blow was inflicted.

Over time the pain intensifies, that indicates irritation of the peritoneum with blood and bile that flow into the abdominal cavity. Subcapsular and central ruptures are characterized by two-phase clinical manifestations. The general condition of patients with subcapsular and small central gaps deteriorates slightly. The abdomen remains soft, and muscle pain and tension can only be determined at the site of slaughter. Clear clinical manifestations are noted in 1-2 days after injury.

When intrahepatic hematomas lacerate into the biliary system, it is manifested by the development of hemobilia syndrome, which is characterized by the Sandblum triad: pain, bleeding in the intestines and jaundice.

Hemobilia is usually diagnosed late; without surgery, mortality in this pathology is very high up to 32-100%. The diagnosis is confirmed by ultrasound and CT (presence of intrahepatic hematoma), angiography (leakage and accumulation of contrast outside the arterial bed, Fig. 4.3) and fibroegogastroduodenoscopy (bleeding from the vater nipple).

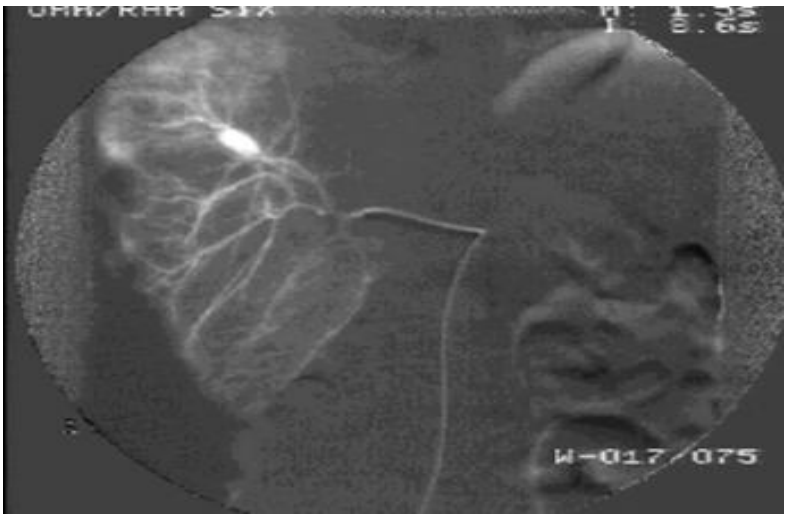


Figure 4.3. Celiacography shows blood pooling in the right lobe (arrow).

In deep liver ruptures sharp anemia, enlarged liver, severe pain and fever are presented; child's condition worsens rapidly.

Massive ruptures of the parenchyma with damage to large vessels and intense intra-abdominal bleeding are characterized by extremely severe condition of children, with a typical manifestation of traumatic shock.

Pain on palpation is noted in all children. There is a characteristic symptom of "navel" - sharp pain when pressing on the navel due to tension of the round ligament of the liver, Kulenkampf's symptom - sharp pain on palpation of the abdomen in the absence of muscle tension, Hedry's symptom - pain in the right costal arch when pressing on the lower chest.

Dynamic blood monitoring with an interval of 30 minutes is of great diagnostic value. Progressive decrease in hemoglobin and erythrocytes, indicates the presence of a catastrophe in the abdominal cavity of the child. A characteristic feature of liver damage is high leukocytosis - up to 15-25 × 109 / l, which appears quickly after injury and increases over the next few hours. Increased transaminase activity is a reliable test for liver damage.

Survey X-ray can provide only additional information, therefore, ultrasound, CT, MRI, angiography, and laparoscopy are performed to confirm the diagnosis.

According to CT results, liver injuries are divided into 6 grades:

Grade 1:

- hematoma: subcapsular, ≤ 10% of the liver surface;
- rupture: rupture of the capsule, ≤1 cm deep.

Grade 2:

- hematoma: subcapsular, 10–50% of the liver surface;
- hematoma: intraparenchymatous, ≤10 cm in diameter;
- rupture: rupture of the capsule, 1–3 cm deep, ≤10 cm long.

Grade 3:

- hematoma: subcapsular, ≥50% of the liver surface, or rupture with active bleeding;
- hematoma: intraparenchymatous, ≥10 cm in diameter;
- rupture: rupture of the capsule, ≥3 cm deep.

Grade 4:

- hematoma: intraparenchymatous with active bleeding;
- rupture: rupture of the parenchyma involving 25–75% of the liver lobes or 1–3 Quino segments (within one lobe).

Grade 5:

- rupture: rupture of the parenchyma involving ≥75% of the liver lobes or ≥3 segments according to Quino;
- vessels: extrahepatic veins detachment (inferior vena cava and hepatic veins).

Grade 6:

- vessels: whole liver detachment

Treatment of liver injuries.

In more than 90% of cases liver injuries without capsule damage are "stabilized" without surgery. It is necessary to follow the waiting tactics along with ongoing anti-shock and hemostatic therapy. Grade 1-2 liver injuries are managed with conservative treatment, Grade 3 ang higher require surgery. During surgery for liver ruptures, non-viable tissues are removed, bleeding vessels are ligated, hepatic suture is applied, hemostatic sponge, omentum, muscle or mesh is tamponade, the liver is sutured to the diaphragm (hepatopexy) or resected.

Early complications and mortality after severe liver injuries are associated with massive blood loss, coagulopathy, and acidosis.

In case of hemobilia, only surgical treatment can stop the bleeding, segmenthectomy or even a hemihepatectomy can be required.

Spleen injuries

Injuries to the spleen account for about 30% of abdominal organs injuries in children. Mortality from multiple and combined injuries of the spleen reaches about 19%.

In isolated one-moment damage to the spleen, the condition of a child) remains satisfactory up to 6-12 hours after injury.

Later characteristic symptoms appear: the forced position on the left side with legs pulled up to the abdomen. Abdominal pain increases when moving.

Examination shows pallor of the skin and visible mucous membranes, dryness and white layers on the tongue, cold extremities and cold sweat, frequent breathing with a decrease in respiratory excursion. At heavy injuries weakening of breath at the left, lag in breath of the left half of a stomach, tachycardia, pulse of small filling is noted.

One of the constant signs of traumatic injury to the spleen is pain: the most intense pain is localized in the spleen, it is exacerbated by deep breathing, radiating to the left upper arm, shoulder and shoulder blade, note a positive "phrenic symptom". Pain syndrome in isolated splenic injury is combined with moderate passive muscle rigidity of the left half of the anterior abdominal wall.

In the first 6 hours after injury, a general blood test reveals leukocytosis with a neutrophilic shift of the formula to the left and moderate lymphopenia. Hemoglobin and erythrocytes are slightly reduced.

Survey X-ray shows high standing of the left dome of a diaphragm with restriction of its mobility during breath, smoothing of a costal-diaphragmatic sinus at the left, homogeneous obscuration in a spleen projection due to hemorrhage and clots, and displacement of the air bubble of the stomach medially or downward.

Following grades of spleen injury are detected according to the ultrasonographic criteria: grade I (mild) - contusion, subcapsular hematoma without violating the integrity of the capsule; grade II (moderate) - traumatic destruction of 25-30% of the parenchyma and 10% of the circulating blood volume or free fluid in the abdominal cavity (blood); grade III (severe) - traumatic destruction of more than 30% of the parenchyma and more than 10% of the circulating blood volume or free fluid in the abdominal cavity (blood) with stable or unstable hemodynamics. CT is is especially informative for the detection of subcapsular and intraspleen hematomas.

Treatment. The main goal of treatment of traumatic spleen injuries is to stop the bleeding. Previously all attempts to preserve the spleen were strongly condemned due to the risk of recurrent profuse bleeding, now spleen-sparing surgery is accepted (Fig. 4.4).

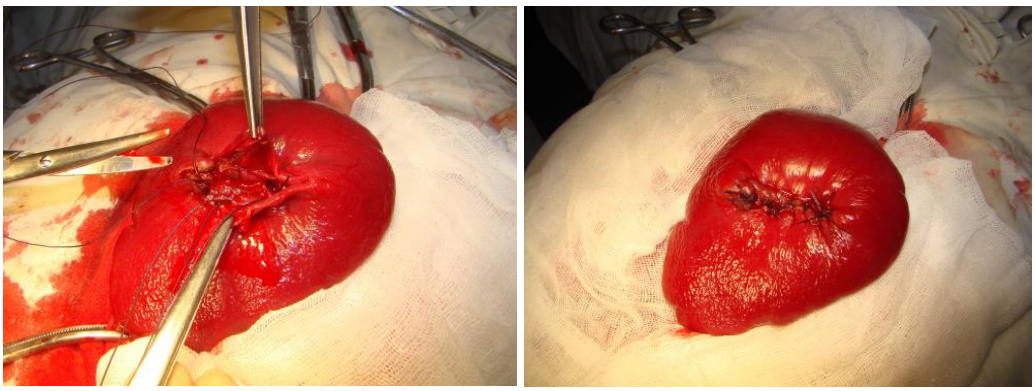


Figure 4.4. Suturing of a spleen rupture (intraoperative photo).

Indications for urgent laparotomy or laparoscopy in spleen injury are following: instability of hemodynamics set against hemostatic therapy; suspicion of crushing or tearing of the spleen; grade III spleen damage according to ultrasonographic criteria in unstable hemodynamics; suspicion of hollow organs damage; lack of blood supplies and infusions; inability to monitor and dynamically control the condition of the abdominal organs.

Indications for splenectomy are following: separation of the spleen from the vascular leg; crushing the body into several fragments; ruptures of the spleen, which pass through its gate; ruptures and decapsulation of the spleen; combined damage to the spleen and hollow organs; open injuries with contamination of the abdominal cavity; the presence of peritonitis; very serious general condition of the injured child.

Injuries of the pancreas

Injuries to the pancreas account for about 4% of internal organs, they are usually difficult to diagnose. Clinical symptoms are epigastric pain, vomiting, intoxication. Routine CT allows to clearly visualize the gland. Magnetic resonance cholangiopancreatography is a highly informative method, but has limited use in the acute phase of polytrauma, for this examination requires a relatively long time and general anesthesia in young children whose vital functions are impired. Elevated serum amylase and lipase levels often accompany abdominal trauma, but do not provide information on the extent of the injury and the need for surgery. An increase in amylase levels is not a specific test for pancreatic injury, as it can also be observed in salivary gland injury, perforation or intestinal obstruction, intracranial hemorrhage, and the like.

Tactics of treatment of severe pancreatic damage is contradictions. Contusion of the gland without rupture of the large ducts heals spontaneously. Pseudocysts are formed in 45-100% of cases of the excretory ducts ruptures in patients are treated conservatively. There is an opinion that early surgery with distal pancreatostomy reduces the duration of parenteral nutrition and hospital stay.

Pancreatic pseudocyst presents like dense-elastic tumor-like mass in the epigastric region in 3-5 days after injury. The diagnosis is initially confirmed by ultrasound,then CT (Fig. 4.5).

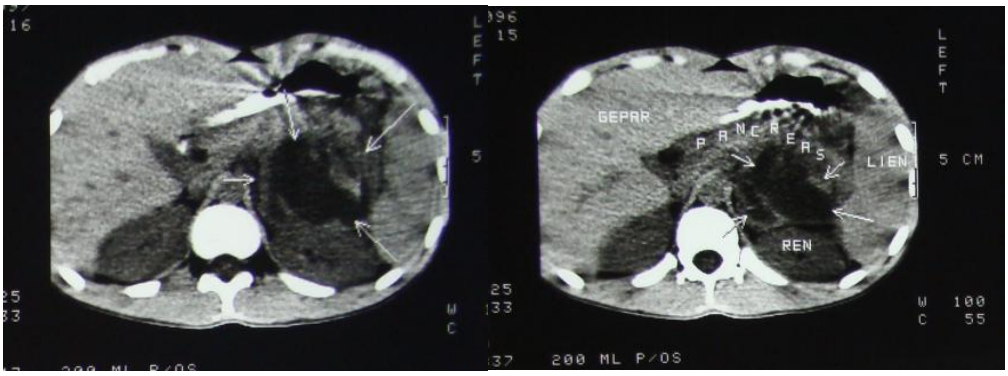


Figure 4.5. CT of the abdominal cavity. Pseudocyst of the pancreas (arrow).

Conservative treatment lasts 4-6 weeks and includes intensive fluid therapy, complete parenteral nutrition, suppression of gastrointestinal peristalsis and secretion, antibacterial therapy. If spontaneous regression does not occur (up to 40% of cases), there is a need for percutaneous puncture and drainage of the cyst.

Injuries of the hollow organs of the abdominal cavity

Closed injuries of the gastrointestinal tract in children account from 9 to 19% of all abdominal injuries, and open penetrating injuries of the hollow organs are very rare. Most often, damage to the hollow organs occurs because of compression (between the anterior abdominal wall and spine), rupture of the mesentery or overstretched intestinal loop.

Closed lesions are penetrating and nonpenetrating. At penetrating damages the subserous or submucosal hematoma is formed, the contents of a hollow organ get into a free abdominal cavity.

Due to anatomical and physiological features, the small intestine till the part of the ileum are injured more often. Isolated penetrating lesions of the stomach and colon are rare.

The clinical picture of nonpenetrating injuries is vague. There is pain, there may be vomiting. At penetrating ruptures, the picture of catastrophe in the abdominal cavity develops. The contents of the stomach cause irritation of the peritoneum, manifested by sharp pain, mainly in the epigastric region, vomiting with blood.

Intestinal damage

The clinical picture of small bowel injuries depends on the nature of the rupture and the time that has elapsed since the injury.

At nonpenetrating damages of intestines, the general condition of children, as a rule, does not worsen. Disturbing abdominal pain, observe a single vomiting of gastric contents. There is no clinic of peritonitis.

At penetrating damages acute peritonitis quickly develops. When the intestinal wound is small or covered, peritonitis develops slowly.

The most common symptom of penetrating damage to the small intestine is pain, sharp, intense, constant. The location of the pain does not always correspond to the location of the injury. The condition of child is usually serious. Expressed symptoms of shock (pallor, frequent weak pulse, tachycardia, tachypnea). The abdomen is symmetrical, not bloated, limited mobility during respiration. Palpation of the abdomen is painful, positive symptoms of peritoneal irritation. Often there is no hepatic dullness, percussion is determined by tympanitis. Intestinal motility is weakened or absent.

Damage to the duodenum in children is rare. With no characteristic symptoms, it is very difficult to diagnose. There are two types of rupture of the duodenum: when the posterior leaf of the peritoneum in involved or not. Clinical signs may include stomach expansion, bile vomiting, and sometimes palpable tumor-like formation in the epigastrium.

To confirm the injury of the hollow organs of the abdominal cavity, it is necessary to perform a survey X-ray in a vertical position. The symptom of a "sickle" - free gas under the dome of the diaphragm - will indicate a violation of the integrity of the wall of the hollow organ (Fig. 4.6).

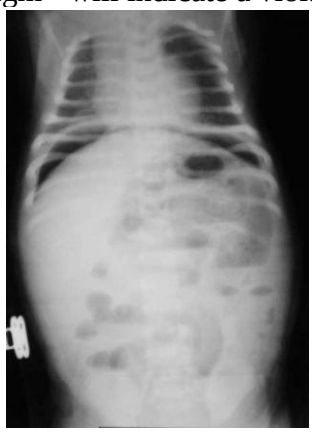
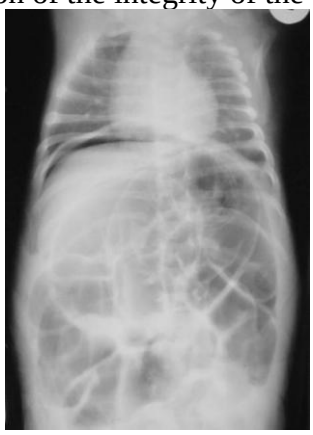
A  B 

Figure 4.6. Survey X-ray in a vertical position. A - free gas under the right and left domes of the diaphragm; B - free gas under the right dome of the diaphragm.

Diagnosis of duodenal injuries is difficult. The diagnosis can be confirmed by contrast study of the gastrointestinal tract, which reveals a "severed" duodenum or its spiral configuration. CT scan can provide valuable information. Damage to the duodenum is often accompanied by pancreatic trauma.

Damage of the colon is rare in children, and in typical cases, the clinical presentation is no different from the clinic of damage to the small intestine.

Treatment. Penetrating injuries of hollow organs are an absolute indication for emergency surgery under general anesthesia. Median laparotomy is the optimal surgical approach. Hemorrhagic exudate with intestinal impurities indicates damage to the hollow organ.

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Situational tasks*:*

1. A child, 9 years, was delivered to hospital by ambulance car. 15 minutes ago a boy was hit by a car. The child is conscious, doesn't answer questions, adynamic, the skin is pale, peripheral pulse is thready, heart rate is 95 beats per minute. AP- 70/0 mm. Examination revealed abrasions in the right subcostum.
    A. What is the diagnosis?
    B. Defme the subsequent tactics of treatment.

2. A boy, 10 years, was hospitalized in 40 minutes after he fell from tree with complaints about pain in the left hip, left forearm, thorax and abdominal cavity. Examination revealed swelling, deformation of the hip and forearm, limitation of extremities movement, abrasions in the left side of the thorax, pulse is 110 beats per minute, arterial pressure is 90/50 mm. Put tentative diagnosis. Make a plan of examination.
    A. What methods may be used to establish diagnosis?
    B. What is your tentative diagnosis?
    C. What urgent measures must be administered?

3. A child of 5 years was injured in the car accident. In 20 minutes after the accident paramedic arrived and found out the following. The child is conscious, extremely agitated, pale, the body is covered with sweat. Breathing, central nervous system, the head, thorax, vertebral column and abdomen are without signs of clinical affection. Pulse is 140 beats per minute, arterial pressure is 90/45 mm. Reaction of filling of subungual capillaries is 3 seconds. Left hip is deformed in its upper part, its palpation causes agitation of the child and crying, active movements in the joints of the left extremity are absent. On the left side of the pelvis there are abrasions and subcutaneous hematoma. Palpation of wing of left ilium bone causes abrupt pain and guarding reaction of the child (tries to draw away doctor's hand). The diagnosis established by paramedics: fracture of the left hip bone with dislocated splinters, fracture of the left ilium bone, contusion of the soft tissues. Traumatic shock (the degree of severity isn't determined).
    A. What are early signs of traumatic shock of children?
    B. If traumatic shock is present, what should the treatment be started with
    C. What complications may develop in a patient?

4. A child, 6,5 years was hospitalized, temperature is 37,6°C, agitation, pain at swallowing and dysphagia. From the anamnesis: the child underwent endoscopic examination for gastritis occasion.
    A. What is the most probable diagnosis?
    B. Recall the diagnostic methods.
    C. What treatment must be administered?

5. A boy, 12 years, was hospitalized with complaints about abdominal pain, weakness and dizziness. The child fell from the second floor. At examination: the child is pale, covered with cold sweat, rapid weak pulse, superficial respiration, decreased arterial pressure. The anterior abdominal wall gaps in breathing, at palpation it is moderately tense, acutely painful. Positive symptom of Shchotkin-Blumberg.
    A. What is your tentative diagnosis?
    B. What methods of examination must be used?
    C. What is the emergency treatment?

Test task for initial level of knowledge

1. A child of 12 years was hit into the abdomen one hour ago. The condition of the child is moderately hard; the child assumes forced position in bed. The skin is pale; pulse-122 beats per second. When pressing onto the left rib arch-pain is experienced. Positive symptoms of Veinert, Coolencamph. Macroscopically the urine is without changes. What is the most probable diagnosis?
    A. Rupture of left kidney, retroperitoneal hematoma;
    B. Rupture of the spleen;
    C. Rupture of the liver, intraperitoneal hematoma;
    D. Rupture of the spleen, intraperitoneal hemorrhage;
    E. Rupture of the hollow organ, peritonitis.
2. A patient was hospitalized with complaints about acute pain in the left part of the thorax, dyspnea. A day ago he fell from 2,5 m. At AP roentgenogram of the thoracic organs doctors determined fractures of the 6^{th}, 7^{th} and 8^{th} ribs, horizontal level of fluid reaches the 4^{th} rib. The diagnosis established is hemopneumothorax. What measures should be performed?
    A. Puncture of the pleural cavity in the 2^{nd} intercostum by the medium-clavicular line on the left;
    B. Puncture of the pleural cavity in the 7^{th} intercostum by the posterior axillary line;

C. Puncture of the pleural cavity and thoracocentesis in the 2nd intercostum by the medium-clavicular line on the left;
D. Puncture of the pleural cavity and thoracocentesis in the 5lh intercostum by the medium axillary line on the left;
E. Puncture of the pleural cavity and thoracocentesis in the 7th intercostum by the posterior axillary line on the left.

3. At examination of the patient after road accident the doctor observed cyanosis, hard breathing. The condition of the patient is severe, right part of the thorax gaps in breathing, intercostal spaces are distended on the right, percussion reveals box sounds, at auscultation breathing is absent. What is your diagnosis?
   A. Open pneumothorax
   B. Pneumoperitoneum
   C. Acute purulent pleuritic
   D. Tense pneumothorax
   E. Total haemothorax on the right
4. A patient, 16 years, is staying in a hospital with severe traumatic shock. Joint thoracic and abdominal trauma. Superficial breathing, arterial pressure is 80/60 mm, heart rate is 115 beats per minute, breathing rate is 42 per minute. What are the urgent measures to correct respiratory disorders?
   A. Narcotic analgesics.
   B. Artificial lung ventilation.
   C. Urgent operation with blood reinfusion.
   D. Mechanical ventilation.
   E. Central analgesics.
5. The key measures to treat the patient in traumatic shock are:
   A. Effective analgesia and cooling the body areas with bums;
   B. Effective analgesia and glucocorticoids;
   C. Effective analgesia and infusion therapy;
   D. Effective analgesia and cardiac glycosides;
   E. Effective analgesia and transport immobilization;
6. A boy, 12 years, after car accident, has: diagnosed comminuted fracture of the diaphysis of the femur, contussion, multiple rib fractures and haemopneumothorax, “scalping” wound of the shank. Which one of the named injuries is dominant?
   A. Closed comminuted fracture of the diaphysis of the femur;
   B. Multiple rib fractures and haemopneumothorax;
   C. Concussion of the brain;
   D. “Scalping wound” of the shank;
   E. All injuries are equal
7. An unconscious patient was hospitalized after the road accident. Arterial pressure is 60/0 mm, pulse is 140 beats per. Objectively: fractured femur in its medium third. Intraperitoneal bleeding. The computed tomogram of the brain shows hemorrhagic contusion of the frontal part. When may femur osteosynthesis be performed?
   A. Immediately after termination of the diagnostic process;
   B. In post-shock condition, after the bleeding stops;
   C. In post-shock condition, not later than on the 3rd day;
   D. After arrest of the hemorrhage;
   E. In post-shock condition
8. A patient in traumatic shock has arterial pressure - 50/0 mm, heart rate is 160beats per minute. What is the shock index?
   A. 2,5
   B. 1,5
   C. 3,2

D. 0,5
E. 0,3

9. A patient, 15 years, after the road accident complains about abrupt dyspnea. Objectively: pale, cyanotic skin. Subcutaneous emphysema in the area of the thorax, abdomen and neck on the right. Auscultation: no breathing on the right, heart rate is 130 beats per minute, arterial pressure makes 80/60 mm, central venous pressure is 140 mm, rate of breathing is 30 per minute, Ht-0,27, Hb- 90 g/1. Subsequent therapy must include such measures:
   A. Urgent artificial lung ventilation;
   B. Massive infusion therapy by crystalloid solutions;
   C. Infusion of dophamine, 2-5 mcg/kg min;
   D. Puncture of the pleural cavity on the right;
   E. Oxygenation by 100 % oxygen.

1. A patient, 17 years, 2 hours ago fell from the ground floor. Stupor condition, pale with multiple scratches of the face, torn wounds on the left forearm. Closed fracture of the left shoulder and hip. Pulse is 110 beats per minute, arterial pressure is 90/40 mm. Blood analysis: erythrocytes - 3,5x10 /1, Hb - 100 g/1. What infusive shouldn't be used for shock treatment?
   A. Solution of crystalloids;
   B. Solutions of gelatine;
   C. 5 % glucose solution;
   D. Solutions of hydroxystarch;
   E. Albumin solution.
2. A patient, 8 years, after being released from under the building debris, was hospitalized. In two hours the injured limb is swollen and cold. The patient's condition is aggravating, blood pressure is 90/40 mm, anuria. The doctor suspects syndrome of position compression. What is pathogenetic mechanism of this condition?
   A. Endogenous intoxication;
   B. Vascular constriction caused by pain syndrome;
   C. Intoxication of the cardio-vascular system;
   D. Necrosis of convoluted tubules of the kidneys;
   E. Lipid degeneration of hepatic cells
3. What is the infusive therapy at performing post-shock management of the patient?
   A. 20 ml/h.
   B. 45 ml/h
   C. 15 ml/h.
   D. 30 ml/h.
   E. 50 ml/h.
4. A child, 4 years, received blunt injury of the thorax (after falling from the ladder onto the soil). Increasing dyspnea and cyanosis. Left side of the thorax gaps in breathing, at auscultation on the left breathing isn't heard, heart sounds are determined in the right part of the thorax. There is subcutaneous crepitation of the skin. Roentgenogram shows left-side hydropneumothorax, pneumomediastinum. The diagnosis established is closed injury of the thorax, hydropneumothorax, hydromediastinum. What are your actions at first aid?
   A. Introduction of analgesics, draining left pleural cavity, antishock therapy;
   B. Analgesics, antishock therapy, suprajugular mediastinomy;
   C. Draining left pleural cavity, antishock therapy, artificial lung ventilation;

D. Antishock therapy, artificial lung ventilation, antibiotics, suprajugular mediastinomy;
E. Antishock therapy, draining left pleural cavity.

5. A child, 8 years, was hospitalized in one hour after abdominal trauma. General condition of the child is severe. Pale. The abdomen is enlarged. At percussion over the abdomen-tympanitis, hepatic dullness isn't determined. Diffuse tenderness over all surface of the abdomen, expressed of muscles of the anterior abdominal wall. What diagnosis is the most probable?
   A. Subcapsular hepatic hematoma;
   B. Rupture of the pancreas, peritonitis;
   C. Rupture of the liver, intraperitoneal hemorrhage;
   D. Retroperitoneal rupture of the bladder;
   E. Injury of the hollow organ, peritonitis.
6. A patient, 10 years, was hospitalized into surgical department after the road accident, with closed trauma of the thorax and fractured ribs on the right. The patient has been diagnosed right-side pneumothorax. The patient is indicated draining of the pleural cavity. What is the place of pleural puncture?
   A. In the 2^{nd} intercostum by the medium-clavicular line;
   B. In the 6^{th} intercostum by the posterior-axillary line;
   C. In the 7^{th} intercostum by the scapular line;
   D. In the pleural sinus projection;
   E. In the place of greatest dullness, determined at percussion.
7. A patient, 12 years, after the road accident obtained polytrauma: closed fractures of the right humerus and bones of left forearm with dislocation of splintered parts, closed blunt abdominal trauma. The patient was brought to hospital in 30 minutes after the trauma. Skin is pale. Arterial pressure is 90/20 mm, in the fractured areas- there is deformation and pain. The abdomen is tense, at palpation there is acute pain, positive symptom of Shchotkin-Blumberg. What measures must be performed first?
   A. Urgent laparotomy;
   B. Infusion therapy to stabilize arterial pressure;
   C. Immobilizing fractures, anesthesia;
   D. Local anesthesia of fractured areas;
   E. Additional examination to establish correct diagnosis.
8. A patient was brought to hospital in one hour after the road accident with complaints about pain in the right part of the thorax, hard breathing. At examination - on the right of the thorax there are superficial scratches, at palpation - fracture of the right 4^{th} and right 5^{th} ribs is determined. At auscultation- breathing on the right isn't heard. At percussion- in the lower areas of the 5^{th} rib there is dullness. Arterial pressure is 100/70 mm, pulse is 106 beats per minute. What is the diagnosis?
   A. Rib fracture, pneumohaemothorax;
   B. Contusion of the thorax, fracture of the ribs;
   C. Contusion of the thorax, injury of the lungs;
   D. Contusion of the thorax, fracture of the ribs, subcutaneous hematoma;
   E. Additional examination to establish correct diagnosis;

THEME 5
OEDEMATOUS SCROTUM SYNDROME. TRAUMATIC INJURIES OF THE URINARY SYSTEM.

Overview

Is stipulated for high occurrence of these conditions in pediatric surgery and urology. Children with oedematous scrotum syndrome make a great per cent among all urgent conditions. Introduction of surgical tactics cardinally improved the results of treatment. Traumatic injuries of the urinary system organs are one of the most difficult parts of urgent urology. The majority of these cases cause disability or death of the patient. Knowledge of this topic allows avoiding severe consequences of such traumas.

Educational aims:

1. To learn the list of diseases causing oedematous scrotum syndrome.
2. To learn the most frequent conditions accompanied by oedematous scrotum syndrome.
3. To master principles of diagnosis and treatment of oedematous scrotum syndrome.
4. To master principles of diagnosis and treatment of traumatic injuries of the urinary system

A student must know :

1. anatomo-physiological information about scrotum and urinary system
2. main clinical manifestations of diseases and injuries of the scrotum.
3. kinds of torsion of the testicle.
4. indications for orchoectomy at torsion of the testicle.
5. criteria of vitality of the testicle at its torsion
6. causes of increase of mass of Morganii's hydatid and its necrosis.
7. surgical tactics of necrosis of Morganii's hydatid
8. what is haemato-testicular barrier
9. surgical tactics of traumas of the testicle
10. general principles of treatment of diseases, accompanied with acute inflammation of testes and scrotum, define indications for operative treatment.
11. mechanism of injury of the urethra.
12. method of diagnostics of trauma of the urethra.
13. danger of catheterization of the urethra at its trauma
14. surgical tactics of trauma of the urethra (primary suturing of the urethra, delayed operation on the urethra)
15. types of ruptures of the bladder, mechanism of development.
16. method of diagnostics of trauma of the bladder
17. surgical tactics of intraperitoneal rupture of the bladder.
18. surgical tactics of retroperitoneal rupture of the bladder.
19. mechanism of renal injuries.
20. classification of traumatic injuries of the kidneys.
21. methods of diagnostics of traumatic injuries of the kidneys..
22. conservative treatment of traumatic injuries of the kidneys.
23. indications for different types of operative interventions of traumatic injuries of the kidneys.
24. kinds of operation of of traumatic injuries of the kidneys.
25. possibility of renal autotransplantation at its severe trauma.
26. treatment tactics at joint traumatic injuries of the kidneys, bladder and urethra.

A student must be able to :

1. to differentiate injuries and diseases of the scrotum according to causes of oedema.

2. to interpret the additional methods of examination of oedematous scrotum syndrome.
3. to offer algorithm of doctor's actions at oedematous scrotum syndrome and determine the treatment tactics.
4. to detect visually affected hydatid, to perform diaphanoscopy;
5. to interpret the additional methods of examination of traumatic injuries of the kidneys, urinary bladder and urethra
6. to offer algorithm of doctor's actions at traumatic injuries of urinary system and determine the treatment tactics.
7. to administer urgent therapy at main injuries of the urogenital system in children, perform the main medical procedures: measuring the arterial pressure, defining the blood group and Rh-factor, catheterization of the bladder or suprapubic puncture.

Terminology

Term	Definition
1. Urolithiasis	Stones in the cavities of the kidneys, ureters, bladder and
2. Urostasis	Disordered haemodynamics
3. Haematuria	Blood in the urine. Erythrocytes more than 1000 in 1 ml – pathology by Nechiporenko.
4. Leucocyturia	Leucocytes in the urine, more than 10 in general urine analysis, more than 4000 in 1 ml of urine by
5. Renal colic	Severe pain in the lumbar area, connected with presence or movement of the calculi or congregations.
6. Dysuria, dysuric disturbances	Painful urination, frequency of urination and vesical tenesmus
7. Anuria	No excreted urine
8. Oedematous scrotum, "acute" scrotum	Several diseases connected by changes of the testicles and scrotum.
9. Urethrorrhage	Blood excreted from the urethra

CONTENTS

Acute diseases of the scrotum constitute a serious pathology of reproductive system in boys and frequently cause disordered reproduction in adults.

For a long time many acute diseases of the scrotum were diagnosed as "non-specific orchitis". The treatment was conservative and standard. Such incorrect tactics caused high percentage (40-77 %) of atrophy of the testis and even disordered function of a healthy one. This problem has recently been solved by S.Y. Doletskiy, A.E. Soloviov, O.V. Tereshchenko, O.B. Okulov, Y. B. Yudin et al. There have been determined acute scrotal diseases requiring surgical intervention. In general there has been changed treatment tactics towards the increase of operative intervention. This has improved treatment of children with oedematous scrotum syndrome.

Torsion of the spermatic cord.

There is no other extremely difficult for diagnosing and treatment disease of the scrotum but torsion of the spermatic cord. It is observed in all age groups, but the most frequently it may develop and thus evidence disproportion of growth (development) in 1-year children and children of puberty age, in those who make up 90% of all patients with this diagnose.

The etiological factor of torsion of the spermatic cord is an abrupt contraction of the muscle-erector of the testis, fibres of which are spirally directed. Usually there develops intravaginal torsion when the cord with a testis is torsed within the vaginal layer of the testis. In young children torsion of the spermatic cord and testis with its layers (extravaginal form) occur mostly in neonatal period. Morphologic immaturity of the spermatic cord and its surrounding tissues play crucial role in etiology and pathogenesis of the extravaginal form of spermatic cord torsion.

The complication of spermatic cord torsion is acute impairment of blood and lymph circulation and development of total haemorrhagic infarction. Irreversible pathological changes in the testis develop very early, necrosis develops in 6-10 hours from the disease onset. Immune

component also plays very important role in prognosing the consequences of the disease. Immunological reactions at spermatic cord torsion under some conditions increase the pathological process and cause complete destruction of the testis.

Clinical picture depends on age, terms of referring to doctor and position of the testis. At first local signs are expressed, then-the general ones. Local changes are characterized by oedema and hyperemia of scrotum's skin, there is hard malformation larger than the testicle by size (Fig. 5.1).

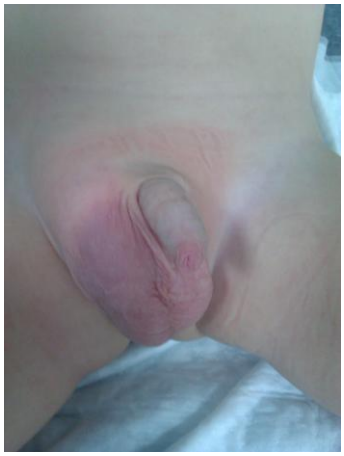

Fig. 5.1. Acute scrotum

The spermatic cord is thickened. It is impossible to palpate the testicle distinctly, diaphanoscopy (transillumination) test is negative. Intravaginal spermatic cord torsion is accompanied with more vividly expressed clinical picture. The first symptom is sudden moderate or severe pain in the testicle. Pain and tenderness are usually localized in the scrotum on the affected side. There may be pain irradiation towards the direction of the spermatic cord.

Local symptoms are often combined with general: dizziness, pale skin, cold sweat, nausea, sometimes unconsciousness. With development of necrosis of the testicle the pain syndrome expression decreases, general condition improves a little, but there develop local symptoms: increased oedema and hyperemia of the scrotum that will further involve the healthy side. The skin has glassy edema shade. The testicle is hard, increased in size, painful. Very rarely the disease has a lesser symptomatic course.

Diagnostics of torsion of the testicle is based on the data of anamnesis, local symptoms: swelling, pain, enlargement and hardening of the testicle, its tension, changed position ((pulled to the root of the scrotum), general symptoms. In establishing the diagnosis the following methods are very important: diaphanoscopy, scanning, ultrasound stethoscoping of the testicle, doppler-examination of vessels of the affected testicle spermatic cord.

Differential diagnostics is performed with acute impairment of hydatides, orchitis, allergic oedema of the scrotum, tumor of the testicle, acute nonspecific epididymitis , idiopathic infarction of the scrotum.

Treatment doesn't include only surgery (Fig 5.2); it must be performed just after establishing the diagnosis or its suspicion. The access is through the scrotum. After detorsion the spermatic cord and performing its block by 0.25-0.5 % procaine solution, 2 % lidocaine solution, it is necessary to check for vitality of the scrotum. To do it warm furacilline compresses are applied for 15-20 min. then the testicle is examined, whether its color has changed or not (blue color turning into usual color), if protein layer is glistening or not.

Fig. 5.2. Torsion of the spermatic cord (intraoperative photo).

The testicle must be removed only when it is completely necrotized. The testicle that is left is fixed in 2-3 points to the internal surface of the scrotum. The operation is finished with draining

the scrotum. In postoperative period anti-inflammatory physio-, heparin- and vitamin-therapy, hyperbaric oxygenation are administered. In case of signs of sensibilization to testicular antigen in blood, immunosuppressive therapy is administered. The children must stay under dispensary surveillance of a urologist for not less than 1 year.

Acute affection of hydatides of the scrotum.

Acute affection of testicular hydatides is observed in children of all age groups. It usually affects children from 4 till 15 years. The hydatides of the scrotum are derivatives of the primary sexual ducts- Wolffian ducts and Muller's ducts. There are 5 types of hydatides. In all cases we deal with disordered blood circulation resulting from twisting of the hydatid around its pedicles. Subsequently there develops vessel thrombosis, infectioning and necrosis of the hydatide. According to the degree of venous stasis during operation it is possible to determine greately enlarged dark or slightly enlarged pale rosy hydatid twisted around the pedicle. At late treatment or no treatment at all the testicle becomes inflamed too. It is very rare that the cyst malformation of the hydatids with their subsequent malignisation may be observed.

Clinical picture depends on the disease stage. Local signs are dominating (pain in the scrotum). There are expressed hyperemia, oedema, skin tenderness and the scrotum asymmetry caused by its enlargement on affected side. Their expression depends on the disease term. At palpation of the scrotum near the upper pole it is possible to palpate the testicles if swelling is absent and we can see affected hydatid as a dark-blue, dark-red or red nodule. Then if there develops acute ascites of the testicle, the last isn't differentiated with the paradidymis. Cremasteric reflex is weak on the affected side.

Diagnostics **is** based on the data of anamnesis, clinical picture, the additional methods: dyaphanoscopy, diagnostic puncture of the testicle layers- are rarely used.

Differential diagnostics is performed with trauma and allergic oedema of the scrotum, torsion of the testicle, acute nonspecific epididymitis, acute parotid orchitis. Very rarely we may differentiate it with strangulated inguinoscrotal hernia and pyoinflammatory diseases of the scrotum.

Treatmen**t** is operation, because conservative treatment may cause atrophy of the testicle on the affected side. The affected hydatid is removed during the operation (Fig. 5.3).

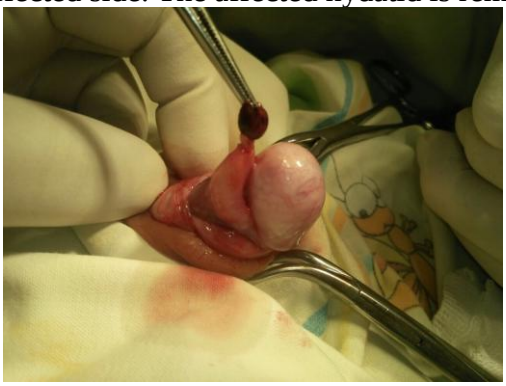

Fig. 5.3. Torsion of the Testicular Appendage

The skin is cut towards the direction of skin folds of the scrotum (the length up to 2 cm). The tissues of the scrotum are cut layer by layer. During cutting of serous cavity of the testicle after removing light haemorrhagic exudate, the affected hydatid is to be found and removed by ligating its pedicle with electrocoagulator or ligature. If uncomplicated hydatides are found, they are also removed. If there are layers of fibrin, the last is removed too. Also a positive result may be reached byintraoperational block of the spermatic cord with 5-10 ml of 0.25 % procaine solution. The postoperative management includes adequate analgesia and anti-inflammatory desensitizing therapy. Sometimes hyperbaric oxygenation is prescribed.

Postoperative complications are rare, they are: haematocele, inflammatory process in the area of the scrotum.

Outcome of surgical treatment is positive and doesn't influence the subsequent development

of the testicle.

Allergic oedema of the scrotum.

Allergic oedema of the scrotum (Quincke's oedema) – is the most common disease of the scrotum that doesn't require surgical treatment. It is almost impossible to determine the etiological causative agent, but such children, as a rule, have unfavorable allergic anamnesis. The disease is seasonal: it develops in autumn or in spring and summer.

Clinical picture consists of the general and local symptoms. The onset is acute. Local changes are characterized by rapid oedema of skin of the scrotum that evenly includes both sides of the scrotum. Sometimes it also includes the adjacent areas and penis. There may develop haemorrhages into the tissue of the scrotum accompanied by the oedema. The skin of the scrotum becomes vividly-red or dark-red. There is itching and pain at palpation. At the same time the testicle and epididymis are almost without changes. All these symptoms develop on the background of gradual temperature increase, headache and general malaise.

Diagnostics **is** based on complaints, data of anamnesis, changes of skin of the scrotum, unchanged testicle, epididymis and spermatic cord, and the characteristic disease development.

Differential diagnostics is performed with other acute diseases of the scrotum (torsion of the spermatic cord, impaired hydatids, orchitis, etc). The diagnosis to be established is the most difficult in the first hours from the disease onset. As the disease proceeds, establishing the diagnosis becomes less difficult.

Treatment of allergic oedema is conservative, it is exclusion of the allergen causing the disease and desensitizing therapy.

Prognosis is favorable.

Orchitis.

Orchitis is an acute inflammation of the testicle resulting from infection, trauma or other factors. There are specific and non-specific orchitis. Non-specific orchitis develops at trauma and spreading inflammation from the adjacent tissues. In the newborns the process is often spread from the umbilical vessels, in older children- from the epididymis in case of its inflammation and some other cases. Recently the nonspecific orchitis morbidity has decreased as the diseases demanding surgical treatment are more frequently determined. Specific orchitis develops at presence of the causative agent tropic to tissue of the testicle. This is observed at epidemic parotitis, tuberculosis and infectious hepatitis.

Infectious agent is the only etiological factor. But as the disease progresses, auto-immune changes develop in the body, and they may lead to deeper pathological changes in both affected and healthy testicles.

Pathogenesis is connected with direct action of the causative agent onto the spermatogenic epithelium. Parenchymatous and interstitial cells are affected. Swelling of tissues of the testicle in case of decreased elasticity of tunica albuginea will lead to even greater ischemization of the organ. All this causes atrophy of the testicle.

Clinical picture depends on main disease after which orchitis has developed. Most often the cause is epidemic parotitis. Orchitis develops in the beginning of or after epidemic parotitis. The disease onset is acute. Local hyperemia with aggravating condition of the child is observed: increasing headache, nausea, vomiting. Local symptoms are pain in the affected testicle, which irradiates into the lumbar area, groins and perineum. The testicle increases in size, it is tender at palpation, later there may develop oedema and hyperemia of the scrotum.

Differential diagnosis is performed with all diseases of the scrotum, first of all with acutely affected hydatids of the testicle. It is necessary to remember that at this condition the testicle stays intact for a long time.

Treatment is the conservative one. It is therapy of the main disease, daily local procaine blocks of the spermatic cord, desensitizing therapy, heat, suspensory. At autoimmune reactions some authors use glucocorticoids, salicylic acid, cyclophosphan. Sometimes in severe cases operation after Solovyov is performed for decompression of the testicle: the tunica albuginea is cut with its subsequent restoration in a few days.

Prognosis isn't always favorable. There may happen atrophy of the testicle, disordered spermatogenesis.

Acute specific epididymitis

Acute specific epididymitis affects persons after 18 years, it is very rare in children. The etiological agent of it is infection. Pathogenesis is determined by the way of infectioning- -the ascending one. That is why acute specific epididymitis is a secondary disease. In pathogenesis of children it is important to pay attention to defects of urinary tract.

Clinical picture local symptoms always dominate- these are pain, oedema, skin hyperemia. Ascites of the testicles is also possible. Palpation: there is always hard, increased in size, tender epididymis. There may be determined thickened and painful spermatic cord. General symptoms are less expressed than local; they dominate in clinical picture of young children. Blood analysis shows leucocytosis, urine analyses show leucocyturia and proteinuria.

Diagnostics is based on anamnestic data (urologic disease), local and general symptoms, laboratory data (leucocytosis, leucocyturia, proeinuria).

Differential diagnostics is performed with torsion of the spermatic cord, acutely affected hydatids and allergic oedema of the scrotum.

Treatment is the conservative one, only when there are no doubts referring to the diagnosis. But as isolated acute specific epididymitis is rare in children, explorative intervention is almost always performed, which is terminated with decompression of the tissues of the scrotum. After acute manifestations subside, urological examination of the child must be performed.

Prognosis depends on time of pathogenetic treatment.

The peculiarities of venous system of the left testicle and high venous hypertension may cause thrombosis of the hydatid vessels and productive dysfunction of the testicle.

Torsions and rotation of Morgagni's hydatid may occur at epididimytis, disordered blood circulation and venous thrombosis, it makes 5% of all patients with acutely affected testicle.

In certain patients we may observe haemorrhagic infarction, as a result of inflammation separate foci of infection are determined.

The development of chronic ascites of the testicle, secondary epididymitis may cause obstruction of the ductus deferens.

One of mechanisms of atrophy of the testicle at acute affection of the testicle may be autoimmune reactions.

Stages of urgent measures at acute disease of the testicle.

Stage	Measure	Methods and medications
I	Preoperative preparation.	Anesthetics
II	Operative intervention	Removal of hydatides, draining of the layers of the testicle. Blocks of the spermatic cord with 0.25 % procaine solution +
III	Postoperative management	1. Introduction of antibiotics at severe inflammation of the testicle. 2. Block of the spermatic cord with hydrocortisone solution + 0.25 % of procaine. 3. Vascular drugs- penthoxyphillin, trental. Vitamins E,A, antioxydants

Trauma of the scrotum.

Scrotal traumas take the leading position among scrotal diseases. There are such kinds of them: contusion, rupture, dislocation and torsion of the testicle.

The etiological agent here is direct trauma. In contusion of the scrotum and testicle there develops oedema of tissues, which is caused by functional impairment of blood circulation. In severe cases there develops haemorrhage, subcapsular or intracapsular haematoma. Rupture of the testicle is the most severe trauma of the scrotum. It is characterized by rupture of the tunica albuginea, this is accompanied by prolapsing of parenchyma of the testicle. There are transverse ruptures, with various localization, depending on affected place. The most severe impairment is crushing of the testicle or its

separation from the spermatic cord. Another kind of trauma of the testicle is its dislocation. Dislocated testicle(s) may be determined under the skin of abdomen, hip, perineum, in the inguinal channel. The open traumas of the scrotum are lacerated, pierced, incised and bite wounds accompanied with trauma of the testicle, paradidymis, spermatic cord and sometimes penis. They are often accompanied with traumas of other organs.

Clinical picture depends on the degree and character of trauma. In closed traumas quite often there are swelling and haemorrhage into the soft tissues, enlargement and tenderness of the testicle and epididymis, thickening and tenderness of the spermatic cord. In case of haemorrhage into the soft tissues we may speak of haematocoele or haematoma of the scrotum. Fluctuation is often determined. The scrotum size may increase 3-5 times. It isn't always possible to palpate the testicle or epididymis because of acute tension. The spermatic cord is often thickened and tender. In open traumas there may be wound of the scrotum, testicle, completely or partially without layers. As a rule, ruptures of the testicle and epididymis are accompanied with traumatic shock of different degrees (from acute pain to comatose condition).

Diagnostics of traumas of the scrotum is based on the data of anamnesis, clinical symptoms, especially pathological changes in area of the genitals. Oedema, swelling, enlargement of the testicle, tenderness at palpation are characteristic for contusion. Haematocele is characteristic for rupture of the testicle. Dislocation of the testicle is characterized by its shift (dislocation), and empty scrotum. If there are signs of wounds, it shows us an open injury of the scrotum.

Differential diagnostics is performed with other acute diseases of the scrotum: torsion of the spermatic cord and hydatids, allergic oedema and orchitis. The majority of these diseases are preceded by intense physical exercises.

Treatment of closed traumas of the scrotum may be conservative and operative. In the first 24 hours bed regimen is administered, suspensory, cold onto the scrotum. In the end of the 2^{nd}-3^{rd} day heat and physiotherapy are administered. If haematoma is present, it is cut and drained, if necessary-haemostasis is performed. In contusion of the testicle procaine block of the spermatic cord is added (0.25-0.5 %procaine solution), if necessary- it is repeated. At rupture of the testicle it is necessary to perform closure of tunica albuginea under general ansthesia with the scrotal access. This should be done as early as possible, on the 1^{st}-3^{rd} day. The operation is terminated by draining the scrotum with a rubber tube for 1-2 days. At crushing of the testicle orchidectomy is indicated. Dislocation of the testicle may be accompanied with its torsion that is why it is better to perform operation. At cryptorchism it is necessary to perform reduction of the testicle. In all types of trauma of the testicle antibiotics are administered as well as vitamin E in age dosages and hyperbaric oxygenation. Open traumas of the scrotum are treated only by surgical method. Primary surgical management and draining of the wound are always performed. At prolapsed scrotal organs they are reduced, the wound is sutured and drained.

The prognosis of treatment of scrotal organs under correct conservative and timely surgical treatment is favorable.

Traumatic injuries of the urinary system

Although traumatic injuries of the urinary system is rarely the cause of death, there is a definite need for accurate diagnosis and treatment, especially when one considers some of the long- term sequelae from genitourinary injuries.

Injury to the male external genitalia – penis, scrotum, and scrotal contents—is relatively uncommon. Penile trauma is frequently iatrogenic from circumcision and can be either an acute problem, such as complete transection, resection of excessive skin, urethral violation, or a complication of healing such as scar phimosis.

Blunt and penetrating injuries to the penis are usually associated with urethral injury. These cases should be evaluated and managed according to the basic principles discussed in the section on urethral trauma.

Many authors have reported cases of penile tourniquet injury from hair, rings, string, rubber bands, and other objects. One should suspect this etiology in most cases of penile swelling. Release of the tourniquet is essential; however, even with release, loss of tissue including skin,

glans, and urethra is common.

Because of the small size and mobile nature of prepubertal descended testes, testicular damage, even with apparently severe scrotal trauma, is infrequent. When testicular damage is suspected, surgical exploration is indicated to repair the torn tunica albuginea, control bleeding, drain any hematocele, and salvage the testis, if possible. Before removal of the injured testis, one must be sure that the contralateral testis is viable. Ultrasound examination has been useful in some cases. Many children with scrotal and testicular trauma often have severe visceral injuries that take precedence in their management.

Relatively minor trauma associated with severe scrotal or testicular pain is often indicative of testicular torsion. Radionuclear scanning, Doppler studies, and ultrasound can all be helpful. If there is any question about the diagnosis, surgical exploration with detorsion and fixation or orchiectomy and contralateral, fixation are essential.

Scrotal hematoma with no testicular injury is usually best treated with analgesics and rest. Penetrating injuries will require debridement if tissue damage is extensive. One can usually mobilize enough skin to cover the testis. If this is unsuccessful, thigh flaps or skin grafting can be utilized as a means of covering a viable testis.

Injury to the fe male external genitalia. Straddle injury in girls may cause severe labial hematoma. Sedation or anesthesia may be needed to assess vaginal and/or urethral damage. Many girls experience urinary retention when hematoma formation is extensive. This is best treated with urethral catheter drainage until the hematoma resolves. If necessary, trocar suprapubic drainage can be performed.

Urethral trauma. Male anterior urethral injury is usually iatrogenic in nature, especially following instrumentation. Inappropriate catheter size (too large), forcible urethral dilation, and inappropriate management of Foley catheters (failure to secure the catheter to the lower abdomen, inappropriate meatal care, etc.) will lead to stricture. Prevention is the key. The bulbar urethra may be crushed against the pubic rami in some cases of straddle trauma, especially in boys in middle childhood, while “jumping” on bikes. This injury might be prevented by the use of a padded crossbar. Frequently no gross blood is noted either in voided urine or at the urethral meatus. Since symptoms may be minimal at the time of injury, these patients often present late with a urethral stricture.

Blood at the meatus after any trauma or in the urine at the time of initial evaluation of a patient with straddle injury warrants retrograde urethrography. This should be done with fluoroscopic control and use of a water-soluble contrast agent. Even if the patient arrives with an indwelling urethral catheter, this procedure can still be accomplished by placing a small Christmas tree adapter, blunt needle, or intravenous catheter next to the indwelling catheter. The meatus is occluded with the instrument and catheter in place while the injection is made. Once the site of anterior urethral injury has been determined, immediate repair is needed if the mucosal laceration is extensive. This may require endoscopic evaluation. If mucosal laceration is minor, an indwelling Silastic Foley catheter of small size should be left for several days. Large mucosal transections are best handled by primary, end-to-end anastomosis with spatulation and mobilization. Again, a Silastic Foley catheter provides adequate drainage and is very helpful in realignment when placed intraoperatively. Drainage for several days is usually adequate.

Penetrating trauma may cause extensive penile urethral damage. These cases cannot be handled with the methods previously discussed because of the chordee created by urethral mobilization and reconstruction. Therefore, it is best to suture the urethral margins to the skin edges and plan for secondary urethroplasty with either a tube graft or patch graft of split thickness non-hair-bearing skin.

Male posterior urethral injury. Although disruption of the prostatomembranous urethra is not as common in children as in adults, it is still frequently seen with pelvic fracture and lower abdominal blunt injury. It should be suspected if there is blood in the urine or at the meatus or if a catheter cannot be passed. As described previously, fluoroscopic retrograde urethrography is essential. An indwelling catheter may be passed through a partial laceration once, but conversion

into a complete laceration is all too easy if that catheter is removed and reinserted.

Once this injury is diagnosed, the management is highly controversial. Primary realignment with or without suprapubic cystostomy has many advocates. Equally well supported is limited exploration with initial suprapubic cystostomy followed by secondary repair of the stricture. Webster and associates have reported their series and reviewed 19 others. Their conclusions, based on the incidence of stricture, impotence, and incontinence associated with the two approaches, favor a conservative approach with primary suprapubic cystotomy and stricture repair 4 to 6 months later. Primary urethral realignment should be reserved for patients with rectal injury, huge pelvic hematoma with ***very*** high-riding bladder, and associated bladder neck injury. Good results in limited pediatric series have been reported using these basic guidelines.

Famale anterior urethral injury. Blunt pelvic trauma with pelvic fracture is the most common cause of female urethral injury. The anterior vaginal wall is frequently lacerated, and the patient should be examined for laceration if passage of a catheter is unsuccessful. Retrograde urethrography has not been reported to be helpful.

Parkhurst and associates have advocated a two-stage repair with primary suprapubic cystostomy and later urethroplasty. Gonzales and Guerriero have favored realignment of the urethral ends over a soft urethral catheter and careful repair of the vaginal tear. The site of injury and extent of damage seem to dictate the complications, which include vaginal stenosis, urethrovaginal fistula, urethral stricture, incontinence, hydrocolpos, vesical stone, and salpingitis.

Bladder injury. Children with blunt or penetrating abdominal trauma, especially those with pelvic fractures, are at risk for bladder injury. Children are more prone to bladder injury than adults because of the more abdominal position of the bladder. Blood in the urine, blood at the meatus, inability to void, or inability to obtain urine with catheter placement should alert the initial evaluator to possible bladder damage. Since bladder injury is frequently associated with injury of other organs, a high mortality has been reported.

When indicated, a retrograde urethrogram, as previously discussed, is best accomplished first. Otherwise, a catheter is placed for bladder drainage. Cystography including oblique films should be obtained through the catheter. False-negative studies have been reported, especially with penetrating wounds.

If a ureteral or renal injury is suspected, it is advisable to obtain an excretory urogram before cystography because extravasation of contrast media may obscure visualization of the kidneys or ureters. Fluoroscopic control and performance of a “limited” cystogram can circumvent this problem. A water-soluble contrast agent is used that is absorbed and excreted with time, but because time is usually of the essence, these aspects of x-ray selection and performance should be considered.

Extraperitoneal rupture is the most common bladder injury. If the rupture is small and extravasation is limited, one can consider long-term bladder drainage alone. This is especially true in girls in whom catheters can be left for the 7- to 14-day period required to treat the rupture. However, if the superior pubic ramus is fractured, a bone spicule may cause continuous penetration. In boys, prolonged catheterization leads to a high incidence of urethral stricture, which can be eliminated by a surgically placed suprapubic catheter at the time of bladder repair.

Intraperitoneal rupture is more common in children than in adults, but it still accounts for only about 20% of all bladder injuries. The larger peritoneal surface of the childhood bladder accounts for this higher incidence. Intraperitoneal mine is usually rapidly absorbed, leading to azotemia and acidosis. These features dictate that intraperitoneal rupture be repaired even if it is minor. The peritoneum should be irrigated and appropriate vesical drainage, suprapubic or urethral, should be initiated promptly.

Ureteral injury, although uncommon, is more frequently seen in children than in adults. The most common injury to the ureter is avulsion at the ureteropelvic junction. It is hypothesized that this avulsion is caused by sudden trunk flexion or by compression of the pelvis and upper ureter against the rib cage or the lumbar transverse process. In cases of severe blunt trauma, the injury may go undetected for several days unless there is a high index of suspicion. An excretory

urogram will show extravasation of contrast media from the renal hilum and no ureteral filling. Hematuria is infrequent or transient and therefore should not be relied on to aid in the diagnosis. A retrograde pyelogram, especially with fluoroscopic control, may prove helpful in defining the exact site of injury.

Once ureteral injury is recognized, prompt surgical intervention is mandatory. The procedure of choice is a tension-free ureteropyelostomy or pyeloplasty. Proximal diversion and retroperitoneal drainage are indicated. Proximal diversion may be omitted in cases without associated gastrointestinal injury, which are quickly recognized and repaired without tension. If pyeloplasty is impossible, ureterocalycostomy has been advocated.

Penetrating injuries are rare. Injuries from low-velocity missiles are best repaired by debridement and spatulated ureterostomy. Because of more extensive microvascular damage secondary to the thermal injury of high- velocity missiles, extensive debridement is often needed. Extensive debridement will mandate a more involved reconstructive procedure, depending on the site of the injury and the amount of viable ureter left. Upper ureteral defects can often be managed by renal mobilization and "reverse" nephropexy. Lower defects allow the use of several techniques including ureteral reimplantation with vesico-psoas hitch, vesical flaps, transureteroureterostomy, renal autotransplantation, or ileal interposition. In childhood, the renal unit and bladder are highly mobile and may permit the use of reverse nephropexy and a bladder mobilization procedure that would not be applicable in an adult.

Renal injury. The child with a kidney injury frequently has multisystem injuries. Victims of blunt trauma usually have head and skeletal injuries, whereas victims of penetrating trauma have abdominal visceral injuries.

Blunt trauma to the torso leads to a higher incidence of renal injury in children than in adults. Several reasons for this observation have been suggested by Kuz- marov et al.:

1. The childhood kidney is larger in proportion to the body.
2. The childhood thoracic cage is less ossified and therefore less protective because it is more flexible.
3. Children have less perirenal fat and less developed muscles of the flank and abdominal wall.
4. Fetal lobulations are more prominent, allowing easier "fracture" along these lobulation lines.

Because of the kidney's mobility within Gerota's fascia, the kidney can be crushed against the ribs or vertebral bodies and suffer a laceration or contusion. Mobility also allows stretching of the vascular pedicle, which can result in vascular avulsion, especially of the vein, arterial spasm, or arterial intimal tears with secondary thrombosis.

Penetrating injuries from high-velocity bullets can cause extensive thermal damage to the kidney and surrounding viscera. Low-velocity bullets and other penetrating objects are not associated with such extensive damage.

Numerous authors have shown that anomalous kidneys are more readily injured and account for between 1% and 23% of all injured renal units. Frequently, the anomalous kidney is damaged by minimal trauma. Hydronephrosis is the most common anomaly that predisposes the kidney to injury with consequent hematuria or rupture.

Guerriero has suggested an excellent system for classification of renal injury:

Minor Renal Injury

1. Contusions : Parenchymal injury without a break of parenchyma or tear of the renal capsule.
2. Shallow cortical laceration: "Superficial" parenchymal laceration without extension to the collecting system and with the capsule intact.
3. Forniceal laceration: Disruption of the junction of the collecting system and the renal parenchyma, although the parenchyma and capsule are intac.

Major Renal Injury

1. Renal pedicle injury: Laceration or thrombosis
2. Deep parenchymal laceration with extension into the collecting system. The capsule may be intact or disrupted.
3. Shattered kidney: Multiple fragments from several deep parenchymal lacerations. The

capsule may be intact or disrupted.

The essential step in managing renal injuries is accurate diagnosis. Gross hematuria, flank or abdominal mass, fractured ribs or lumbar spine, crush injury of the abdomen, chest, or pelvis, severe deceleration, or penetrating wounds of the flank or abdomen are all indications for infused computerized axial tomography. Controversy exists over the need for radiologic evaluation when only microscopic hematuria is present. If the mechanism of injury does not absolutely mandate evaluation, microscopic hematuria may be observed and checked for clearance with serial urine analyses.

As always, a high index of suspicion is needed because renal pedicle injuries and ureteropelvic disruptions may not be associated with hematuria. The initial radiographs may show either a scoliosis with concavity on the side of injury or an obliterated psoas muscle. Computerized tomography with intravenous and oral contrast is done in all individuals thought to have these injuries. Prompt and equal

excretion of contrast media with no extravasation excludes major renal injury. Such findings occur in about 85% of all renal injuries and indicate that the injury can be treated nonoperatively if the contralateral renal unit is satisfactory, the patient is stable, and associated injuries do not warrant an operation. Even if an exploratory examination is performed because of another injury, nonoperative management is still usually best.

In about 15% of cases, the excretory radiographs are abnormal but often not diagnostic. In these instances, further diagnostic studies and the therapeutic approach are controversial. An unstable patient with major injury to other viscera will require exploration. This is the group of patients in whom the "one-shot IVP" is often useful to confirm bilateral renal function before exploration. Concomitantly, the damaged renal unit can be explored through the transabdominal approach. Vascular control is advisable before entering Gerota's fascia because this thick fibrous coat often tamponades renal bleeding.

Patients who are clinically stable but have a major abnormality of the radiographic studies require a more extensive urologic evaluation to determine the exact injury so that management is optimal. Optimal management will preserve renal function, shorten hospitalization, lessen the need for secondary surgery, and minimize long-term complications. In the past, angiography was the procedure of choice to delineate the extent of damage when the excretory radiographs were abnormal. Recently, computerized axial tomographic scanning has generally replaced both angiography and the even older intravenous pyelogram because it is less invasive, can clearly define the details of the injured kidney, may detect other unsuspected injuries, and provides some information even when the vascular supply is disrupted. If a renal unit does not enhance with contrast media administration, then a renal vascular injury is likely and exploration or angiography must follow. Radionuclide scanning may be helpful in defining vascular pedicle injury, but in reality this modality is little used for this indication. If there is vascular perfusion the vascular pedicle is intact. If perfusion is not seen, however, another test is needed to prove the presence of a kidney because renal agenesis would give similar findings. In general, retrograde pyelography has little place in evaluating renal trauma except in differentiating ureteropelvic disruption from other extravasations.

Digital subtraction angiography is a far less invasive study than traditional angiography, but it rarely is helpful in evaluating acute renal trauma because of the actual time and technique required to produce the films. Over time, it may prove to be useful in delineating post-traumatic complications such as arteriovenous fis- lulae.

If the diagnosis of renal artery thrombosis is established, immediate surgical repair of the intimal tear and thrombectomy are mandatory for renal salvage. Autotransplantation and "bench" repair can be considered for cases of arterial or venous avulsion or multivessel injury.

Major extravasations without major parenchymal or vascular injury can be treated expectantly. Falling hematocrit, unstable vital signs, and expansion of the hematoma are signs of delayed bleeding and the need for possible surgical intervention.

The most controversial injuries are those with devascularization of segments of parenchyma,

extensive hemorrhage usually contained by Gerota's fascia, and urinary extravasation. This type of injury occurs in about 10% of cases and accounts for 90% of short-term complications—secondary bleeding, abscesses, uri- nomas—and long-term complications—arteriovenous fistulae, encysted hematomas, and hypertension. Once diagnosed, transabdominal exploration with renal vascular control before the opening of Gerota's fascia is indicated. Devascularized segments of parenchyma are debrided with strict attention to closure of the collecting system with absorbable material, to approximation of the parenchyma by capsular mattress sutures of absorbable material, and to adequate drainage. It is sometimes necessary to bolster the mattress sutures with fat or absorbable hemostatic material. Careful isolation of the kidney from other repaired viscera can be aided by peritoneal closure or vascularized omental flaps.

Finally, renal damage due to blunt trauma to an anomalous or solitary renal unit occurs all too frequently. Children known to have solitary kidneys or ectopic kidneys need to be advised concerning participation in sports or other rough and unusual activities. All children should be raised to use proper vehicular restraint devices because prevention of injury is the first step in treating trauma.

Early, accurate diagnosis and management are the key to renal salvage. Algorithmic demonstration of a suggested plan of attack follows.

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Tests for initial level of knowledge

1. Increasing swelling in the lumbar area is typical for:
   A. doubling of the upper urinary tract
   B. hydronephrosis
   C. nephroptosis
   D. renal rupture
   E. kidneys tuberculosis
2. Bladder catheterization is contraindicated in acute urinary retention caused by:
   A. phimosis
   B. bladder tumor
   C. urethral stone
   D. urethral rupture
   E. bladder stones
3. The main method of diagnosis of bladder injury in children:
   A. excretory urography
   B. cystoscopy
   C. cystography

D. bladder catheterization
E. chromocystoscopy

4. Absolute indication for urgent kidney exposure in injury is:
A. macrohematuria
B. microhematuria
C. increasing swelling in the back area, shock
D. fever, chills, vomiting
E. renal colic

5. The most informative examination method in the diagnosis of ureteral rupture:
A. Plain radiography
B. infusional urography
C. cystoscopy
D. uroflowmetry
E. cystography

6. The most remarkable symptoms for retroperitoneal rupture of the bladder are:
A. hematuria, dysuria, paravesicle hematoma
B. pyuria, hematuria, fever
S. nausea, vomiting, acute urinary retention
D. hematuria, oligoanuria, renal colic
E. pain, nausea, pyuria

7. What genital diseases in children do not require immediate surgery?
A. acute balanoposthitis
B. testicular torsion
C. torsion of testicular hydatid
D. hydrocele testis
E. all of the above

8. The most characteristic symptoms of traumatic injuries of the kidneys?
A. abdominal pain
V. pain in the lumbar region
C. bloating
D. haematuria, pararenal hematoma, shock
E. oligo-, anuria

Keys for tests:

1.D
2.D
3.C
4.C
5.B
6.A
7.D
8.D

Tests for final level of knowledge

1. In examination of the boy with trauma fracture of the pelvis and bladder injury were discovered. What contributes to retroperitoneal bladder rupture?
A. Overflow of the urine bladder and pelvic fractures in the form of a "butterfly".
B. Overflow with bowel contents.
C. Body commotio while injury.
D. Reflex spasm of the bladder sphincter.
E. None of the answer is not correct

2. Which of the following most informative examination method in kidney injury?
A. cystoscopy and chromocystoscopy
B. plain radiography

C. excretory urography
D. ultrasound of the kidneys
E. retrograde pyelography

3. At what pathology urine retention is reflex?
A. phimosis
B. balanoposthitis
S. bladder tumor
D. ureteral stones
E. urethral rupture

4. The hospital admitted teenager with torsion of the left testicle. Disease lasts 3 days. While examination changes in the right half of scrotum were revealed. Why does contralateral testis suffers in torsion of the spermatic cord?
A. A reflex spasm of blood vessels (testiculo-testicular reflex).
B. Due to autoimmune process through breach of the hemato-testicular blood-barrier
S. Due to local rise of temperature.
D. Due to psychological overload.
A. Due to operative injury in testicle distorsy.

5. What is early symptom of ureteral injury?
A. constant hematuria
B. one-time hematuria
S. acute urinary retention
D. abdominal pain
E. all of the above

6. While conducting the surgery testicular torsion for 540^{o} was revealed. How can we assess the viability of testicle in spermatic cord torsion?
A. due to painful palpation.
B. due pains in the corresponding half of scrotum and inguinal area.
C. due to the color and intensity of testicular bleeding while cut of tunica albuginea.
D. due to specific smell.
E. due to blood test.

7. The boy the fall on a bicycle frame and got urethra injury - its rupture. 2 hours since the injury, the child in a specialized hospital. What is the emergency aid?
A. restricting fluid intake.
V. appointment of drugs that prevent erection.
C. applying the primary suture of the urethra.
D. epicystostomy.
E. an attempt to set a constant flow of urine by means of Foley catheter.

Keys for tests:

1.A
2.C
3.E
4.A
5.B
6.C
7.D

Tasks for final level of knowledge

1. Doctor on duty examined the11-yeas old boy with hyperemic, swollen scrotum, complains last for 2 days. While examination dark well-defined mass was detected on the anterior surface of

the testicle in its upper third, testicle is painful in palpation, mobile.
1. What is the most likely diagnosis in this patient?
2. What methods of diagnostics you confirm the diagnosis?

Key Answer

3. Acute affection of hydatides of the scrotum.
4. Dyaphanoscopy, ultrasound diagnostic

2. Parents of teenager with complains of intense pain in the left half of the scrotum appealed to surgeon. One day of disease. While examination, the left half of the scrotum is swollen, hyperemic, testicle is firm in palpation, pulled upwards, sharply painful. In ultrasound examination the lack of blood flow, stasis in the vessels left testicle is defined.
1. What disease may be suspected?
3. What is the tactics of the doctor on duty?

Key Answer

1. Torsion of the spermatic cord.
2. Urgent surgical treatment.

3. 12 years old girl was admitted to the surgical department with complaints of pain in the right iliac region, disuric urination disorders. The examination revealed pain on palpation of the right kidney, irradiating down and in the inguinal region. In urine examination - erythrocythuriya, leukocyturia. In ultrasound examination dilatation of pyelocaliceal system of right kidney and upper ureter.
1. What is your preliminary diagnosis?
3. What method will help you to make out diagnosis?

Key Answer

1. urolithiasis.
2. excretory urography

THEME 6

Benign and malignant embryonal tumors. Hepatic tumors. Vascular anomalies.

Overview:

Childhood cancer was almost always fatal before 1970. Today, 80% of children diagnosed with cancer will survive at least 5 years; 70% will be cured. However, cancer continues to be the leading cause of nonaccident-related deaths in children.

The healing process in children and adolescents with oncological diseases depends greatly on the knowledge and experience of all those involved in the patients' care. Progress in pediatric oncology has antedated even some of the more recent accomplishments in adult oncology. Considerable advances have been made in the treatment of children and adolescents with leukemia, lymphoma, sarcoma, and other cancers, and this success has resulted in a significant improvement in the overall survival of these young patients.

Educational aims:

After completing this part of module students should be able to:

1. Recognise the main features of tumors in children
2. Counsel such patients and if younger patients, both the child and parents, about the likely diagnoses and management
3. Know what the possible complications are
4. Know the indications for urgent treatment or referral to a specialist centre
5. Refer a patient with tumors to an appropriate specialist

A student must know:

1. Peculiarities of pediatric tumors
2. Clinical presentations and diagnosis of mediastinal tumors
3. Clinical presentations, diagnosis and treatment of the Wilms' tumor
4. Pathology and clinical manifestation of the neuroblastoma due to localization
5. Diagnostic evaluations and treatment of the neuroblastoma
6. Embryology and pathology of the teratomas
7. Diagnosis, classification, management of the Sacrococcygeal teratomas
8. Pathology, diagnosis and treatment of the dermoid cysts
9. Rhabdomyosarcoma: pathology, diagnosis, main principles of treatment
10. Classification and clinical features of the nevi
11. Predisposing factors, clinical signs and treatment of the melanoma
12. Pathology and classification of the vascular anomalies
13. Pathology, clinical manifestations and imaging of the infantile hemangioma
14. Treatment of the infantile hemangiomas
15. Vascular malformations: clinical manifestations, imaging and treatment

A student must be able to:

1. Make the physical exam of children
2. Evaluate the CT and MRI investigations
3. Take care for children with tracheostomy tube
4. Analyze the blood tests to evaluate the type of coagulopathy.
5. Appoint additional investigations in children with abdominal, mediastinal

Terminology

Term	Definition
Teratomas	Tumors comprising more than a single cell type derived from more than one germ layer
Dermoid cysts	Congenital cysts that are lined by skin with fully mature pilosebaceous structures
Wilms' tumor	Malignant tumor containing metanephric blastema, stromal and epithelial derivatives

Neuroblastoma	Tumour of neural crest origin which may occur in the adrenal medulla or anywhere along the sympathetic ganglion chain, namely in the neck, thorax, abdomen, and pelvis.
Hepatoblastoma	Malignant liver neoplasm occurring in infants and children and composed of tissue resembling fetal liver cells, mature liver cells, or bile duct cells.
Hemangiomas	Benign, and usually a self-involuting tumor, (swelling or growth) of the endothelial cells that line blood vessels and is characterised by increased number of normal or abnormal vessels filled with blood.
Vascular malformations	Congenital lesions of vascular dysmorphogenesis that can be local or diffuse
Lymphatic malformations	Wide spectrum from localized masses to areas of diffuse infiltration to chylous fluid accumulations in various body cavities
Venous malformations	Slow-flow lesions consisting of venous channels that can develop anywhere in the body.
Arteriovenous malformations	Fast-flow vascular malformations characterized by abnormal connections or shunts between feeding arteries and draining veins without an intervening capillary bed.

CONTENT

Incidence and Management of Childhood Cancer

Cancer in children is uncommon; it represents only about 2% of all cancer cases. Nevertheless, after trauma, it is the second mostcommon cause of death in children older than 1 year. Every year 130–140 children per million under the age of 16 years, or around 1 out of 500 children, are diagnosed with childhood cancer. The incidence within the first 5 years of life is twice as high as from 6 to 15 years of age.

Survival probability has considerably changed within the past 30 years. Clinical research by cooperating groups of pediatric oncology centers has progressively increased the long-term survival rate from <20% before 1975 to >70–80% depending on the specific disease, in the new millennium. International cooperation contributes to quality assurance because the majority of children with an oncologic disease are treated according to standard protocols.

While in adults about 80% of cancerous diseases pertain to the respiratory, gastrointestinal, and reproductive organs, only <5% of cancerous diseases of children are manifested in these organs. Furthermore, the histopathology of pediatric neoplasia differs markedly from that of adults: in children, embryonal and immature cells can be found at very different stages of development, which perpetually proliferate and rarely mature.

Diagnosis and therapy must be adjusted to the individual child according to the clinical manifestation and the extent of the tumor. Treatment normally requires 1–3 years, followed by check-ups for the following 5–7 years. The child newly diagnosed with cancer is critically ill during the first 2–6 months; after that his or her life continues similar to that of a healthy child, except that periodical treatment adjustment and check-ups are necessary.

Unlike adult malignancies, which are primarily carcinomas, fewer than 10% of solid tumors of childhood are epithelial malignancies. Although the spectrum of malignancies in childhood is more limited than in adults.

These very primitive or embryonal malignancies often lack morphologically distinguishing characteristics. The exact diagnosis in pediatric cancer patients is crucial because chemotherapy for childhood malignancies is carefully tailored to each specific tumor type.

Whereas light microscopy remains the primary tool of pathologists, they can now rely also on immunohistology, electron microscopy, DNA content of tumor, cytogenetic abnormalities, and specific tumor gene expression to establish a diagnosis.

Cancer is a genetic event. Constitutional gene mutations that are hereditary (i,e., passed from parent to child) or nonhereditary (i.e., de novo mutations in the sperm or oocyte before fertilization) contribute to an estimated 10% to 15% of pediatric cancers. Constitutional chromosomal abnormalities are the result of an abnormal number or structural rearrangement of the normal 46 chromosomes and may be associated with a predisposition to cancer. Examples are the predisposition to leukemia seen with trisomi 21 (Down syndrome) and to germ cell tumors with Klinefelter syndrome (47XXY).

Along with an understanding of the molecular basis of hereditary childhood cancer has come the opportunity to identify children who are at high risk of malignancy and, in some cases, to intervene before the cancer develops or when it is still curable. Two examples include familial adenomatous polyposis and familial thyroid cancer.

Teratomas

Teratomas (from the Greek word *teraton*, meaning monster) are tumors deriving from more than one germ layer and comprised more than a single cell type.

Sacrococcygeal teratoma

Sacrococcygeal teratoma (SCT) is the most common tumor found in newborns and infants with the incidence of 1 per 20,000 ~ 40,000 live births.

Classification The Altman classification divides tumors in function of their anatomical extension into four groups:

Type I predominantly external tumor with minimal extension into pelvis (30 – 35 %);

Type II: external tumor with significant intrapelvic extension (25 %);

Type III external tumor with intra-abdominal extension (15–20%);

Type IV 20%: intrapelvic tumor only, not visible externally (20 %).

Clinical Presentation**.** Prenatal ultrasound graphic examination is useful in the diagnosis of sacrococcygeal teratomas and the main manifestation of fetal sacrococcygeal teratoma is sacral mass. In-utero shunting can lead to fetal hydrops, which is associated with high mortality.

Newborns tupically present with a mass protruding from the sacral region. SCT are commonly very large (average 8-10 cm), well encapsulated and grossly lobulated. These masses possess both cystic and solid components. SCT is nearly always arising from the tip of the coccyx (Fig. 6.1).

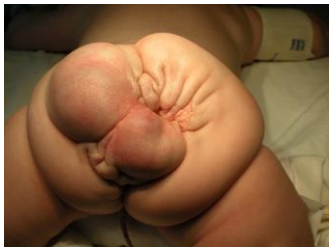
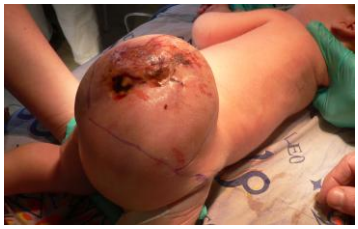
A.  B.  C. 

Fig. 6.1. Type I Sacrococcygeal teratomas (A, B), C- type I Sacrococcygeal teratoma with ulceration.

The diagnosis of purely intrapelvic teratomas is often delayed. Children develop constipation, urinary retention, an abdominal mass, or symptoms of malignancy, such as failure to thrive.

AFP levels, which can be normally elevated in newborns, should be obtained and then followed to ensure that they return to normal by 9 months of age. An association of the triad of presacral teratoma, anal stenosis, and sacral defects was first reported by Ashcraft and Holder, who also confirmed the autosomal dominant nature of the condition. Currarino proposed that adhesions between the endoderm and ectoderm form, causing a split notochord that results in the syndrome, and the triad now bears his name.

In neonates presenting with large external masses, the degree of pelvic and abdominal involvement should be assessed preoperatively with either US scanning, CO, or MRI.

Complications: mass effects of the tumor, recurrence (10-20% initially benign tumor, and 50% of these are malignant recurrences), intraoperative and postoperative bleeding; wound infection.

Differential Diagnosis**:** meningomyelocele, rectal abscess, dermoid cyst, lipoma, neurogenic tumor, pilonidal cyst.

Treatment. For SCT diagnosed at birth, complete surgical excision en bloc with the os coccyx is the golden standard and sole treatment.

In fetuses with tumors larger than 5 cm, cesarean delivery should be considered to prevent dystocia or tumor rupture. In most cases, sacrococcygeal teratomas should be resected electively in the first week of life, since long delays may be associated with a higher rate of malignancy.

For large tumors, this is best achieved through an inverted V incision; smaller tumors can be managed by a posterior sagittal incision. When the intraabdominal tumoral component is important (Type III), the abdominal component is first dissected using a Pfannenstiel incision or laparoscopic abdominal exploration, followed by the perineal approach, allowing for en-bloc removal of the entire tumor together with the coccyx. Newborns will rarely have malignant tumors, and therefore seldom need adjuvant chemotherapy.

Follow-up of these patients should include serial AFP levels to ensure return to normal by 9 months of age and rectal examination every 3 month until 3 years of age. because the latest reported recurrence has been reported at 33 months of age.

Prognosis:

Benign - disease survival is greater than 90%; malignant - significant mortality, although good progress has been made recently in treatment of these tumors.

Time of diagnoses is key:

- < 2 months of age, only 7-10% are malignant
- Age 1 year, 37% malignant
- Age 2 years, 50% malignant

Ovarian teratoma

Mature cystic teratomas account for 10-20% of all ovarian neoplasms. Not only are they the most common ovarian germ cell tumor but also the most common ovarian neoplasm in patients younger than 20 years. They are bilateral in 8-15% of cases.

Mature cystic teratomas of the ovary often are discovered as incidental findings on physical examination, during radiographic studies, or during abdominal surgery performed for other indications. When symptoms are present, they usually consist of abdominal pain (47.6%), abdominal mass or swelling (15.4%), and abnormal uterine bleeding (15.1%). Bladder symptoms, gastrointestinal disturbances, and back pain are less frequent. When abdominal pain is present, it usually is constant and ranges from slight to moderate in intensity. Torsion and acute rupture commonly are associated with severe pain.

Complications of ovarian teratomas include torsion, rupture, infection, hemolytic anemia, and malignant degeneration.

Torsion is by far the most significant cause of morbidity, occurring in 3.2-16% of cases. Rupture of a cystic teratoma may be spontaneous or associated with torsion and occurs in approximately 1-4% of cases. Rupture may occur suddenly, leading to shock or hemorrhage with acute chemical peritonitis. Chronic leakage also may occur, with resultant granulomatous peritonitis. Prognosis after rupture usually is favorable, but the rupture often results in formation of dense adhesive disease.

Infection is uncommon and occurs in only 1% of cases. Coliform bacteria are the organisms most commonly implicated.

Autoimmune hemolytic anemia has been associated with mature cystic teratomas. In several series, removal of the tumor resulted in complete resolution of symptoms. In this context, radiologic imaging of the pelvis may be indicated in cases of refractory hemolytic anemia.

In its pure form, mature cystic teratoma of the ovary always is benign, but in approximately 2% of cases, it may undergo malignant transformation into one of its elements.

Mature cystic teratomas of the ovaries should be removed by simple cystectomy rather than salpingo-oophorectomy. Laparoscopy is an acceptable approach in teratomas resection. Benefits include reductions in postoperative pain, blood loss, and hospital stay. Although the rate of malignant degeneration is only 1%, the cyst should be removed in its entirety, and if immature elements are found, the patient should undergo a standard staging procedure.

Testicular teratoma

The incidence of all testicular tumors in men is 2.1-2.5 per 100,000. Germ cell tumors represent 95% of testicular tumors after puberty, but pure benign teratomas of the testis are rare, accounting for only 3-5% of germ cell tumors.

Childhood testicular teratomas are uniformly benign, with no documented cases of retroperitoneal or lung metastasis in differentiated lesions.

Pure teratomas of the testis are rare. During and after puberty, however, even mature teratomas (composed of entirely mature histologic elements) can metastasize to retroperitoneal lymph nodes or systemically, regardless of their histologic appearance. Morbidity is associated with growth of the tumor, which may invade or obstruct local structures and become unresectable. Malignant transformation is significantly more common in testicular teratomas than in their ovarian counterparts, and recurrence risk is around 20% in both mature and immature testicular teratomas.

Testicular teratomas most often present as a painless scrotal mass, except in the case of torsion. The masses are firm or hard in 83% of cases. Most are not tender and do not transilluminate. Testicular pain and scrotal swelling occasionally are reported with teratomas, but this is nonspecific and simply indicates torsion until proven otherwise. Hydrocele frequently is associated with teratoma in childhood. On examination, the testis is diffusely enlarged, rather than nodular, although a discreet nodule in the upper or lower pole sometimes can be appreciated.

Testicular teratomas traditionally have been treated by simple or radical orchiectomy. More recently, conservative excision by enucleation also has been recommended for prepubertal teratomas of the testis.

The risk of malignancy increases with maturation of the testes, and this is a significant concern in children at or near puberty. In this group, areas of normal surrounding testicular tissue should be excised and sent for frozen section. If frozen section reveals areas of maturity, proceeding to orchiectomy is recommended. Enucleation or partial orchiectomy for teratoma in pubertal or adult males is not recommended.

Cervical teratoma

This is a rare tumor (5-6% of teratomas). Ultrasound features include a unilateral and well-demarcated partly solid and cystic, or multiloculated mass, calcifications (in about 50% of cases), and polyhydramnios (in about 30% of cases due to esophageal obstruction). The prognosis is very poor and the intrauterine or neonatal mortality rate (due to airway obstruction) is about 80%. Some children with airway compression need ex utero intrapartum treatment (EXIT procedure) with intubation, or tracheostomy and resection on placental support. Survival after surgery is more than

80% but, since these tumors tend to be large, extensive neck dissection and multiple additional procedures are necessary to achieve complete resection of the tumor with acceptable functional and cosmetic results.

Dermoid Cysts

Dermoid cysts are congenital cysts that are lined by skin with fully mature pilosebaceous structures. They are the result of sequestration of skin along lines of embryonic closure. The head and neck are the sites of predilection, but these lesions have been described in other midline sites, including the sacral area, perineal raphe, scrotum, and presternal area.

Dermoids are usually round, soft, and often fixed to deep tissues or to bone. They usually present as a painless mass of 1 or 2 cm in diameter but can grow up to 4 cm or more if untreated. Some are associated with a sinus tract, especially those on the nose. This site is also typical for intracranial extension and a familial occurrence.

Dermoid cysts should be excised because they tend to grow and may rupture or become infected, resulting in a more difficult excision and a higher risk of recurrence.

Nephroblastoma (Wilms' tumor)

Nephroblastoma, also referred to renal embryoma, is named after Carl Max Wilhelm Wilms, a German pathologist and surgeon. He was one of the first to propose that tumor cells originate during the development of the embryo. It is the malignant renal tumor that develops from the metanephrogenic tissue.

Wilms' tumor may be sporadic, familial, or associated with specific genetic disorders or recognizable syndromes. Example is WAGR syndrome (Wilms' tumor, aniridia, genitourinary abnormalities, end mental retardation).

Wilms' tumor is the most common primary malignant renal tumor of childhood and comprises 65 of all pediatric tumars.. The boys are affected as frequently as the girls; usually the tumor is diagnosed before 3 years. It is rarely at nephroblastoma that the bilateral affection of the kidneys is observed. The tumor grows in the renal capsule for a long time, but metastases may form even in the initial period. Metastases frequently affect the lungs, liver, bones and retroperitoneal lymphatic nodes.

Clinical picture. Wilms' tumor is generally silent and is commonly detected as an abdominal mass during a well-baby clinic visit or by a parent while bathing or otherwise caring for the infant or child. The tumor lies deep in the flank and is frequently smooth and not tender. Palpation should be gentle and minimal to avoid rupturing the renal capsule and disseminating the disease. Nonpalpable tumors are typically discovered by ultrasonography during evaluation for abdominal pain. Tumor is usually painless; however, when pain occurs, it is often associated with hemorrhage in the tumor and, occasionally, hematuria. Fever, anorexia, and weight loss may occur, although rarely. Hypertension is present in approximately 25% of patients; the incidence of hematuria is similar.

Wilms' tumor also occurs as bilateral disease in 3% to 5% of children, and it is occasionally reported in up to 10% of children.

Ultrasonography (US) is a good screening examination of a mass to determine its site of origin. If the mass is suspected to be a Wilms' tumor or neuroblastoma, computed tomography (CT) of the abdomen with contrast should be considered for further delineation. Also requested are chest radiography and radionuclide bone scan if the tumor appears to be a clear-cell sarcoma of the kidney. A combination of these studies may be required.

CT with intravenous contrast is the gold standard used to evaluate the extent of the abdominal tumor and the integrity of the contralateral kidney. Although MRI avoids radiation exposure, it has not been shown to be superior to CT scanning in standard assessments. MRI may be the preferred method to follow children with bilateral Wilms' tumor after resection.

Differential diagnosis of nephroblastoma is performed with the malformations of kidneys (hydronephrosis, renal polycystosis, renal doubling, etc), retroperitoneal tumors (nephroblastoma, rhabdomyoblastoma, teratoma), sometimes with liver tumors.

National Wilms Tumor Study Group staging system

Stage Description

I Tumor confined to the kidney and completely resected. No penetration of the renal capsule or involvement of renal sinus vessels

II Tumor extends beyond the kidney but is completely resected (none at margins; no lymph nodes). At least one of the following has occurred:

(a) penetration of the renal capsule,

(b) invasion of the renal sinus vessels,

(c) biopsy of tumor before removal,

(d) spillage of tumor locally during removal

III Gross or microscopic residual tumor remains postoperatively, including inoperable tumor, tumor at surgical margins, tumor spillage involving peritoneal surfaces, regional lymph node metastases, or transected tumor thrombus

IV Hematogenous metastases or lymph node metastases outside the abdomen (e.g., lung, liver, bone, brain)

V Bilateral renal Wilms tumors at onset

Treatment. The treatment of Wilms' tumor has been the model for the multidisciplinary management of a pediatric solid tumor. Treatment involves the 3 major modes of cancer therapy: surgery, chemotherapy, and radiotherapy. Surgical removal is attempted in all cases of Wilms' tumor.

The standard surgical management of all patients with a unilateral renal tumor includes radical nephro-ureterectomy and retroperitoneal lymph node sampling. If preoperative imaging is satisfactory and does not suggest a bilateral process, routine exploration of the contralateral kidney is not necessary.

Inoperable and bilateral Wilms' tumors are generally treated with preoperative chemotherapy, which is administered to render the tumor resectable and, in the case of bilateral tumors, to conserve as much renal tissue as possible.

Prognosis. Approximately 80-90% of diagnosed children survive with current multimodality therapy. Synchronous bilateral cases have a 70-80% survival rate, while those with metachronous tumors have a 45-50% survival rate. The prognosis for patients who relapse is not good, with only 30-40% expected to survive after retrieval therapy.

Neuroblastoma

Neuroblastoma has varying presentations and a survival rate that ranges from 0% to 100% depending on the tumor biology. For patients with high-risk disease, aggressive and novel treatments are needed.

Neuroblastoma is a tumor of neural crest origin which may occur in the adrenal medulla or anywhere along the sympathetic ganglion chain, namely in the neck, thorax, abdomen, and pelvis. Seventy-five per cent of tumors occur in the abdomen (adrenal medulla 50 %, paraspinal ganglia 25 %), 20 % occur in the thorax, and 5 % occur in the neck and the pelvis.

Macroscopically, the tumor appears as a very vascular, nodular, friable, solid mass which grows rapidly and invades the adjacent tissues. Areas of hemorrhage, necrosis, and cystic degeneration are not uncommon.

Microscopically, neuroblastomas include a spectrum of tumors with varying degrees of neural differentiation, ranging from undifferentiated small round cells to tumors containing mature ganglion cells (ganglioneuroblastoma or ganglioneuroma).

Frequency. The incidence of neuroblastoma is 9.6 per million populations. Of all patients, 50 % are younger than 2 years and 90 % are younger than 8 years; the peak incidence occurs at 18 months of age.

Clinical. Neuroblastoma can mimic several other disorders and thus is sometimes difficult to diagnose. It may develop at any site of sympathetic nervous system tissue and can be accompanied by nonspecific systemic symptoms. However, the signs and symptoms of neuroblastoma are usually characteristic of the tumor site and the extent of disease (Table 9.1).

More than 70% of neuroblastomas arise in the abdomen, either in the adrenal gland or in retroperitoneal sympathetic ganglia.

Table 9.1. Special clinical presentations of neuroblastoma (by Joann L. Ater and Laura L. Worth)

Tumor Type	*Signs and Symptoms*
Mass	Cervical or supraclavicular swelling or mass ± Horner's syndrome; may be thoracic extension of paraspinal mass or metastasis from abdomen Nodular, firm midline abdominal, or flank mass Posterior mediastinal mass found incidentally because of respiratory or other symptoms
Metastatic	Fever, irritability, failure to thrive, bone pain, anemia, bleeding, hepatomegaly, subcutaneous nodules, orbital proptosis ± periorbital ecchymoses
Neurologic	Opsoclonus-myoclonus syndrome and ataxia due to paraneoplastic syndrome without brain metastasis Weakness or paralysis due to spinal invasion of paraspinal tumor Horner's syndrome due to tumor involvement of superior cervical ganglion
Catecholamine related	Hypertension, flushing, and increased sweating; secretory diarrhea due to vasoactive intestinal peptide

A lot of signs and symptoms accompany metastatic neuroblastoma, including fever, irritability, failure to thrive, bone pain, bluish subcutaneous nodules, orbital proptosis, and periorbital ecchymosis. Orbital proptosis and periorbital ecchymosis can be pathognomonic for neuroblastoma and, if present, may prompt the initiation of additional studies to confirm the diagnosis and determine the extent of disease. The most common sites of metastasis are the long bones and skull, bone marrow, liver, lymph nodes, and skin.

Neurologic signs and symptoms at presentation are less common. Involvement of the superior cervical ganglion can result in Horner's syndrome. Paraspinal neuroblastoma can invade the neural foramina and produce symptoms of spinal cord and nerve root compression.

Some neuroblastomas produce catecholamines, which can cause increased sweating and hypertension, and some release vasoactive intestinal peptide, which causes secretory diarrhea. These unusual presentations, along with spinal cord compression, are found more commonly with thoracic neuroblastoma than with abdominal neuroblastoma.

Diagnosis. On plain radiographs, CT scans, or magnetic resonance imaging (MRI) scans of the chest and abdomen, the primary neuroblastoma tumor is generally discovered as a mass or multiple masses (Fig. 9.2).

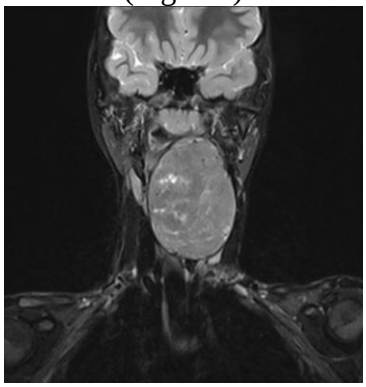


Fig. 6.2. MRI; neurogenic neck tumor (arrow).

Tumor markers (i.e., homovanillic acid (HVA) and vanillylmandelic acid (VMA) in urine) are elevated in 95% of cases. Tests for these tumor markers help to confirm the diagnosis of neuroblastoma. A pathologic diagnosis is made using tumor tissue obtained by biopsy or tumor resection.

Staging. Various staging schemes for neuroblastoma were used in the past. In 1988, an international staging system was devised, establishing a common set of criteria that could be used world- wide and would permit the accrual of large numbers of cases and allow valid comparisons of data (Table 9.2)

Table 9.2. International Neuroblastoma Staging System

Stage	Description
I	Localized tumor confined to area of origin; complete excision, with or without microscopic residual disease; ipsilateral and contralateral lymph nodes negative (nodes attached to primary tumor and removed en bloc with it may be positive)
IIA	Unilateral tumor with incomplete gross excision; ipsilateral and contralateral lymph nodes negative
IIB	Unilateral tumor with complete or incomplete excision; positive ipsilateral, nonadherent regional lymph nodes; contralateral lymph nodes negative
III	Tumor infiltrating across the midline with or without lymph node involvement; or unilateral tumor with contralateral lymph node involvement; or midline tumor with bilateral lymph node involvement or bilateral infiltration (unresectable)
IV	Dissemination of tumor to distant lymph nodes, bone, bone marrow, liver, or other organs
IV-S	Localized primary tumor as defined for stage I or II with dissemination limited to liver, skin, or bone marrow (limited to infants younger than 1 yr)

Treatment. Patients are classified by risk group when determining the best treatment approach. The most important clinical and biologic prognostic factors currently used to determine risk group are patient age at diagnosis, tumor stage, Shimada histology, and ploidy in infants.

The treatment recommendation for low-risk neuroblastoma for patients with stage I or II disease is usually surgical removal. Children with spinal cord compression at diagnosis may require urgent chemotherapy, surgery, or radiotherapy to avoid neurologic damage. Infants younger than 2 months old with stage IV-S neuroblastoma are at higher risk if they have massive liver involvement and respiratory compromise. In this patient group, low-dose cyclophosphamide or very low-dose hepatic radiation may be recommended to alleviate symptoms.

Treatment of intermediate-risk neuroblastoma includes surgery, chemotherapy, and in some cases radiation.

Treatment of high-risk neuroblastoma usually consists of induction chemotherapy followed by high-dose chemotherapy and autologous bone marrow or peripheral blood stem cell transplant.

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common type of soft tissue sarcoma diagnosed during the first 2 decades of life, accounting for 4.5% of all cases of childhood cancer.

Histology. Rhabdomyosarcoma is a malignant tumor of mesenchymal origin. The two major histologic subtypes of tumor are embryonal and alveolar. Embryonal rhabdomyosarcoma occurs more often, affecting two thirds of all patients with disease. In addition to occurring in younger patients, this type of tumor has a favorable survival rate of 60%. Tumors occur more frequently in the head and neck region as compared with the extremities. Spindle-cell histology is common in paratesticular lesions, whereas botryoid lesions are generally polypoid masses filling the lumen of hollow viscus, such as the vagina, bladder, and extrahepatic bile ducts. Alveolar rhabdomyosarcoma occurs in older children, and tumors are most commonly located on the trunk or extremities. These lesions are composed of small, round, densely packed cells arranged around spaces resembling pulmonary alveoli. Prognosis is worse, with a 5-year survival rate of 54%.

Clinical picture. Rhabdomyosarcoma typically presents as an asymptomatic mass. Specific symptoms are generally related to mass effect, compression of adjacent organs or complications of the tumor.

Diagnosis. Standard laboratory work, including complete blood counts, electrolytes, renal фтв liver function tests, and urinalysis should be performed. In addition, imaging studies of the primary tumor should be performed with computer tomography (CT) or magnetic resonance imaging (MRI). CT is advanta- geous for the evaluation of bone erosion and abdominal adenopathy, whereas MRI provides better definition of the tumor and surrounding structures.

Open biopsy of a mass suspected to be rhabdomyosarcoma should be performed to confirm the diagnosis.

Treatment.Surgical biopsy of a primary lesion is often performed prior to a definitive surgical resection.

The main goal of surgical intervention is wide and complete resection of the primary tumor with a surrounding rim of normal tissue. A circumferential margin of 0.5 cm is considered adequate. Such a margin may be unobtainable, however, especially with head and neck tumors. In addition to surgical treatment, all patients with rhabdomyosarcoma receive some form of chemotherapy.

Hepatic tumors

Hepatic tumors in children are relatively rare. The most common malignant neoplasms in the liver are not primary hepatic tumors but rather metastatic lesions such as Wilms' tumor, lymphoma, and neuroblastoma. Primary liver tumors comprise between 1% and 4% of all solid tumors in children.

Hepatoblastoma

Hepatoblastoma accounts for about 80% of the malignant liver tumors in children. It comprises 1% of all pediatric malignancies and affects mostly young children between 6 months and 3 years old, but cases in neonates and school-age children are also seen.

Histology. Hepatoblastoma is defined as a malignant tumor with divergent patterns of differentiation, ranging from fetal epithelial hepatocytes, to embryonal cells, and with differentiated tissues, including osteoid-like material, fibrous connective tissue, and striated muscle fibers. The morphology of hepatoblastoma seems to reflect distinctive phases of hepatogenesis, from endoderm to mature liver cells.

Clinical picture. As mentioned above, the most common presenting sign of a hepatoblastoma is an asymptomatic abdominal mass. The child is often in good health and the

tumor mass is discovered incidentally. A small minority may have other symptoms such as pain, irritability, minor gastrointestinal disturbances, fever, pallor, failure to thrive and even tumor rupture. Patients with the anaplastic variant of hepatoblastoma who often have distant metastases at diagnoses are more frequently symptomatic.

Diagnosis. Measurement of the serum α-fetoprotein is well established as an initial tumor marker. The first imaging study is usually an abdominal ultrasound and, if duplex technique is employed, tumor vascularity can be gauged and the hepatic veins assessed. This is usually followed by computerized axial tomography or magnetic resonance imaging.

Treatment. Complete surgical resection remains the cornerstone of curative therapy of hepatoblastoma. However, more than half of the patients present with either an initial unresectable tumor or with distant metastases. When resection at presentation is not feasible, neoadjuvant chemotherapy can shrink the tumor size extensively and enable safer and complete resection at a later stage. Hepatic transplantation for unresectable primary lesions can be effective for tumors confined to the liver. Aggressive resections, less than total hepatectomy, may be successful in select patients with tumor encroachment on the vena cava or main portal vein.

The prognosis for a patient with recurrent or progressive hepatoblastoma depends on many factors, including the site of recurrence, prior treatment, and individual patient considerations.

Hepatocellular Carcinoma (or Hepatoma)

Hepatocellular carcinoma occurs in 23% of all primary pediatric liver tumors, most often in school-age children and adolescents, very few cases are diagnosed in children less than 5 years old.

Hepatocellular carcinoma occurs predominantly in the setting of underlying liver disease and cirrhosis. Compared with adults, in children cirrhosis is less commonly part of the antecedent process, while congenital disorders of the liver, such as metabolic disease, are common.

Clinical picture. Unlike children with hepatoblastoma, children with hepatocellular carcinoma are usually symptomatic at diagnosis. Pain is common and may even occur in the absence of an obvious mass. But most of these symptoms are nonspecific and include anorexia, fatigue, nausea, and vomiting and weight loss. Up to 10% may present with tumor rupture with signs and symptoms of a hemoperitoneum.

Diagnosis. The α-fetoprotein is elevated in approximately 85% of patients with most levels more than 1,000 ng/mL. Though elevated, these levels are usually lower than those measured in hepatoblastoma patients.

Treatment. complete resection of the primary tumor is essential for long-term survival. Hepatocellular carcinoma is relatively chemotherapy resistant and therefore carries a poor prognosis with a dismal rate of cure

Complete surgical resection or hepatectomy and transplantation for tumor localized to the liver is often the only hope.

Unfortunately, cure is rarely possible in the setting of metastatic disease. Even with aggressive attempts at surgical resection, tumor relapse is common and tumor-free survival rates of not more than 25% to 30% can be achieved.

Benign tumors of the liver

Benign tumors of the liver that occur in childhood include hemangiomas or vascular tumors, hepatocytic adenomas, focal nodular hyperplasia, mesenchymal hamartomas, and various types of cysts or cystic disease.

Hemangiomas are benign vascular tumors of infancy that may involve either the skin or viscera, including the liver.

Hepatic hemangioma is the most common benign tumor of the liver in infancy. These lesions share the same patterns of growth and involution as their more common cutaneous

counterparts. It is probable that most liver hemangiomas remain undetected because they are clinically silent.

Hepatic hemangiomas are true vascular tumors. They must be differentiated from misnamed hepatic hemangiomas observed in adulthood, which are actually venous malformatins.

Diagnosis can be fortuitous on a routine ultrasonography or when presented with an abdominal mass. Multiple hemispherical cutaneous hemangiomas may be present and warn the physician of possible visceral lesions. MRI or CT with contrast is needed to confirm diagnosis (Fig. 8.8). Large lesions may cause congestive heart failure; medical treatment (Propranolol) is indicated in some cases.

Hemangioendothelioma. These are highly proliferative cellular lesions of variable malignant potential. The Kasabach-Merritt syndrome, a platelet-trapping coagulopathy, has also been observed. These lesions may appear very cellular but do not metastasize. If a primary lesion produces symptoms, resection is indicated for relief.

Mesenchymal hamartomas present as an enlarging abdominal mass or hepatomegaly, and are usually not symptomatic. They can grow to great sizes causing respiratory distress or evidence of caval obstruction. Often an open biopsy is necessary to make the diagnosis. Anatomical resection is the recommended treatment for large lesions.

Focal nodular hyperplasia in children presents as an irregularly shaped, nontender liver mass. It is frequently found incidentally at laparotomy for another cause or on radiographic studies performed for another indication. On abdominal sonography, the lesions may be isoechoic, hypoechoic, or hyperechoic compared with normal liver parenchyma. The classic central scar may not be seen on ultrasonography. CT typically shows a hypervascular lesion with a dense stellate central scar.

Nonparasitic cysts of the liver are rare and occur more commonly in adults than in children. Although they may be present and symptomatic at birth, most are asymptomatic and are identified incidentally at autopsy or laparotomy. Symptoms are related to abdominal distention or displacement of adjacent structures.

Congenital Melanocytic Nevus and Giant Hairy Nevus

Congenital nevi are composed of large clusters of pigment producing cells of melanocyte lineage that are clustered in the dermis and produce pigment at birth. When compared with the more common acquired nevi, congenital nevi are larger, have increased cellularity, and lie deeper in the dermis.

The incidence of congenital nevi has been estimated to be 1 in 100 births for small lesions and 1 in 20,000 for large lesions. After birth it is difficult to distinguish congenital from acquired nevi even by histology. Congenital melanocytic nevi are classified according to size: small, less than 1.5 cm in diameter; medium, 1.5 to 19.9 cm; and large (giant), greater than 20 cm.

The potential for congenital nevi to transform into melanoma is the prime reason that these lesions come to surgical attention. However, considerable uncertainty surrounds estimation of the true risk for malignant degeneration.

Giant hairy nevi are a special subset of congenital nevi. These melanocytic cutaneous lesions are extensive and encompass entire anatomic regions that may exceed 20% of the body surface area (Fig.6.3). These nevi are often hairy with benign surface nodularity consisting of focal growths of neuroectodermal tissue. When the head or upper part of the trunk is involved, neurologic abnormalities have been observed in up to 20% of patients.

Some neonates with giant hairy nevi have been found to have malignant melanoma shortly after birth, thus justifying early surgical ablation. Serial resection of lesions with the use of subcutaneous tissue expanders is indicated to improve aesthetic result.

Fig. 6.3. Congenital giant hairy nevus of the trunk

Nevus Sebaceous of Jadassohn

Sebaceous nevus is a solitary, well-circumscribed, alopecic plaque of epidermal origin. SN is commonly found on the scalp, temple, or preauricular region and is sometimes diagnosed at birth. Histologic analysis in children reveals epidermal hyperplasia and hypoplastic hair follicles and sebaceous glands. At puberty the sebaceous glands mature, and progression of the epithelial hyperplasia occurs (Fig.6.4). Progression to basal cell carcinoma has been reported in up to 6% to 22% of adults.

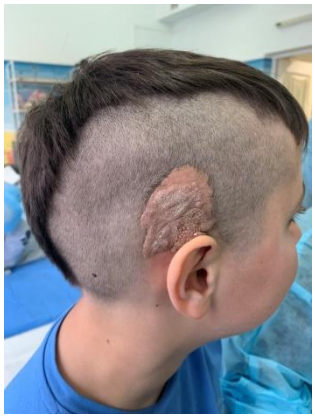


Fig. 6.4. Nevus sebaceous

The management of choice remains excision, but delay until later in adolescence is reasonable.

Melanoma

Melanoma rarely occurs in pediatric patients, so when a child has a suspicious skin lesion, melanoma is usually not considered. However, as in adults, when the lesion is irregularly colored, changes color, rapidly increases in size, or has irregular borders, removal should be considered. The pathologic evaluations of the skin lesions is needed, because melanomas in children and adolescents present with histologic features distinct from those found in melanomas in adults.

Once the diagnosis of melanoma is confirmed, patient is evaluated clinically for evidence of satellite, in-transit, or lymph node metastasis; a chest radiograph is done; and serum lactate dehydrogenase levels are measured.

Treatment for pediatric patients with melanoma have to performed by multidisciplinary group, with input from pathologists, melanoma surgeons, medical and pediatric oncologists, and radiation oncologists.

VASCULAR ANOMALIES

Vascular anomalies (vascular tumors and vascular malformations), often named "angioma" or hemangioma, in fact represent a broad spectrum of disorders from a simple "birthmark" to life-threatening entities, which affect mainly infants, children, and young adults.

Better understanding of vascular anomalies has come in the past several decades with improved knowledge about the growth of blood vessels (angiogenesis) and the development of a more logical classification system.

Based on their biologic and clinical behavior, vascular anomalies can be broadly divided into two groups: vascular tumors and vascular malformations.

Vascular tumors, of which the infantile hemangioma is the most common example, are true neoplasms that arise from endothelial hyperplasia. Conversely, vascular malformations are congenital lesions of vascular dysmorphogenesis that arise because of errors of embryonic development. These lesions exhibit normal endothelial cell turnover.

A biologic classification system of vascular anomalies has been devised based on physical characteristics, natural history, and cellular features. This system was accepted in 1996 by the International Society for the Study of Vascular Anomalies (ISSVA). However, since then, knowledge about these disorders has increased considerably. The ISSVA Classification of Vascular Anomalies was recently updated in Amsterdam, Netherlands (May, 2018). The interactive document is available at www.issva.org.

Table 8.2. ISSVA* Classification of vascular anomalies

Tumours	Vascular malformation			
benign, locally aggressive, malignant	Simple	Combined (defined as two or more vascular malformations found in one lesion)	Of major named vessels	Associated with other anomalies
	Capillary malformation (CM)	CVM		
	Lymphatic malformation(LM)	CLVM		
	Venous malformation (VM)	LVM		
	Arterio-venous malformation (AVM)	CAVM		
		CLAVM		

•*ISSVA, International Society for the Study of Vascular Anomalies

VASCULAR TUMORS

Infantile Hemangiomas

Infantile hemangiomas are the most common tumor of infancy, occurring in the skin in up to 4% to 10% of white infants, with a female-to-male ratio of 3 to 5:1. The incidence may be significantly higher in premature infants.

Infantile hemangiomas have a unique and characteristic life cycle of rapid growth in the first year of life (proliferative phase) followed by spontaneous slow regression from ages 1 to 7 years (involuting phase). Once involuted, they never recur.

Hemangiomas first appear in the neonatal period, with a median age at onset of 2 weeks. Hemangiomas are most often single cutaneous lesions and have an anatomic predilection for the head and neck region (60%). Internal hemangiomas can be found in the liver, gastrointestinal tract, mediastinum, and airway. Tumors that involve the superficial dermis are first seen as a red, raised lesion. Superficial tumors that are larger or that exhibit more rapid growth can cause ulceration of the skin and bleeding.

Tumors in the lower dermis, subcutaneous tissue, or muscle appear bluish with slightly raised overlying skin and have frequently incorrectly been called “cavernous” hemangiomas.

With experience, history and physical examination can establish an accurate diagnosis for 90% of these tumors.

During the involuting phase of hemangiomas tumor slowly regresses, although it may grow in proportion with the child. Both the deep color and the bulk of the tumor show continued gradual improvement until the regression is entirely complete by age 10 to 12 years. In the final involuted phase of the tumor, 50% of patients have nearly normal skin in the area of the prior lesion (Fig.6.5.). Patients that had larger tumors may have lax or redundant skin and yellowish discoloration.

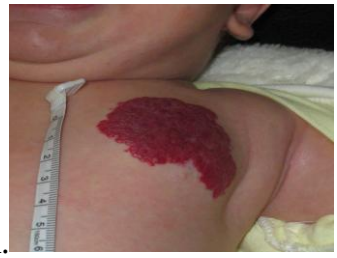
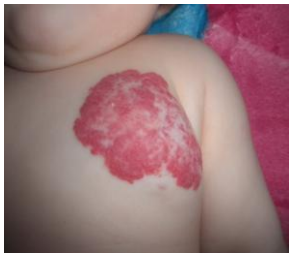
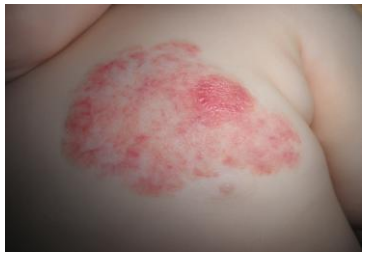
A.  B.  C. 

Fig. 6.5. Infantile hemangiomas. A - proliferative phase, B,C – involuting phase.

The differential diagnosis of cutaneous hemangiomas consists primarily of other vascular anomalies and some malignant tumors.

The majority of infantile hemangiomas do not require any specific treatment other than observation and reassurance of the parents.

Reasons for treatment or referral to a vascular anomalies center are the following: dangerous location (impinging on a vital structure such as the airway or eye), unusually large size or rapid growth, and local or endangering complications (skin ulceration or high-output heart failure).

Because hemangiomas are tumors of pure angiogenesis, pharmacologic therapy involves angiogenesis inhibition. Since 2008 the first-line antiangiogenic therapy for hemangiomas are beta-blockers (Propranolol) (Fig.6.6).

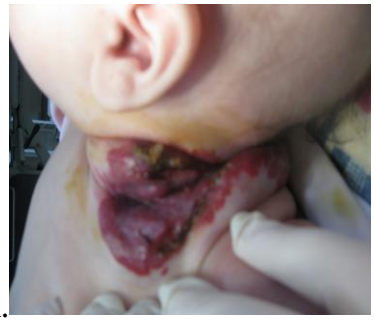
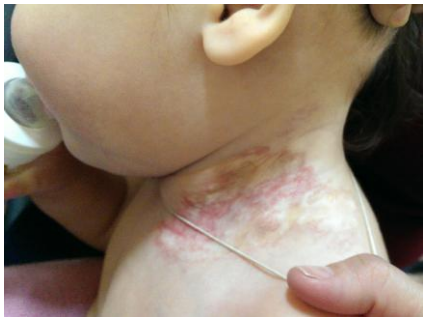
A.  B. 

Fig. 6.6. Complicated neck infantile hemangiomas. A – before treatment, B – after treatment with Propranolol.

After complete involution of the hemangioma, cosmetic distortion is the primary indication for operative management.

For the difficult-to-treat and life-threatening large hemangiomas, especially in the liver, angiographic embolization may be required to manage high-output cardiac failure.

Congenital hemangiomas are less common. They are present and are fully grown at birth. They often regress rapidly, before 1 year of age, or can remain stable or partially involute. On the basis of this natural history, these 3 types are named, respectively, rapidly involuting congenital hemangiomas (RICH) (Fig.6.7.), noninvoluting congenital hemangiomas (NICH) and partially involuting congenital hemangiomas. Rapidly involuting congenital hemangiomas may be associated with transient thrombocytopenia and consumption coagulopathy.

Other clinically significant benign or locally aggressive and borderline vascular tumors are tufted angiomas and kaposiform hemangioendotheliomas.

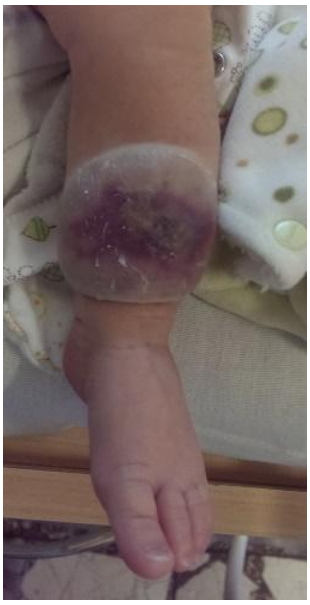
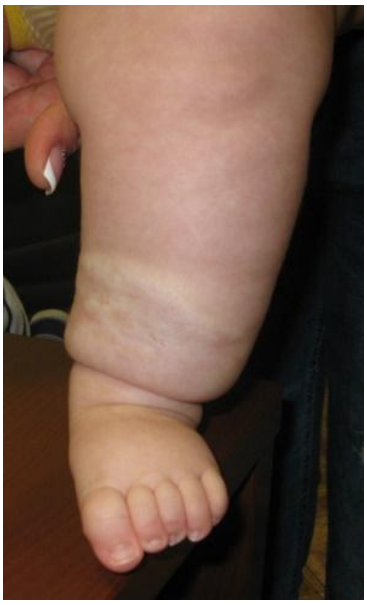
A.  B. 

Fig. 6.7. Rapidly involuting congenital hemangiomas (RICH): A – newborn, B – 9 month.

Tufted angiomas appear as erythematous or brown plaques or macules in children and young adults. Ultrasound reveals separate lobular structures with vessels along the edges.

Kaposiform hemangioendothelioma may affect the skin and subcutis tissue but often involves the deep tissues, presenting as a locally aggressive tumor. In most cases, however, the skin covering the tumor does not show any signs of infection, but a tough infiltration with local pain (“wasp sting symptom”) (Fig.6.8.).

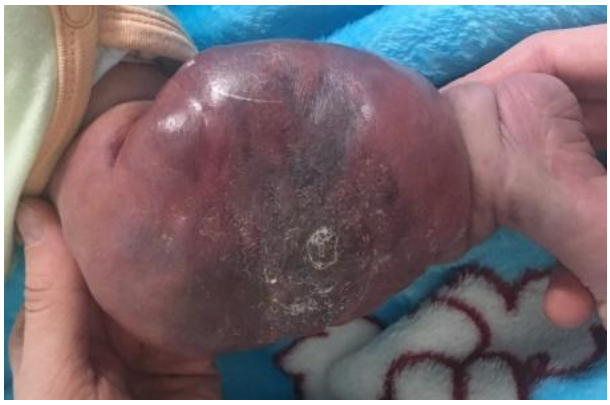


Fig. 6.8. Kaposiform hemangioendothelioma with skin infiltration (“wasp sting symptom”)

Color duplex sonography clearly shows an increased microcirculation which is not located in the center, as in active infantile hemangiomas, but at the lobuli divided by septa.

Kasabach-Merritt Phenomenon

Kasabach-Merritt phenomenon was first reported in 1940 as a case of profound thrombocytopenia, petechiae, and bleeding in conjunction with a "giant hemangioma". The platelet count with this disorder is typically less than 10,000/mm^3 and may be accompanied by decreased fibrinogen levels and a mildly elevated prothrombin time (PT) and partial thromboplastin time (PTT). Bleeding can result from this platelet-trapping coagulopathy at many sites.

It is now established that this thrombocytopenic coagulopathy is not caused by infantile hemangioma. Kasabach-Merritt phenomenon occurs with the uncommon, often more aggressive, vascular tumors, kaposiform hemangioendothelioma and tufted angiomas.

Resection, when feasible, corrects the hematological anomalies within a day; however, this is rarely possible. Pharmacological therapy should be given promptly, and, if effective, residual tumor and sequelae are minimized.

VASCULAR MALFORMATIONS

Vascular malformations are a subtype of vascular anomaly, composed by dysplastic vessels lined by normal endothelium, originated from abnormal morphogenesis of the vascular tissue. They are present at birth, though sometimes they are not detected until later in life.

Vascular malformations are divided into 4 groups: simple malformations, combined malformations, malformations of major named vessels, and malformations associated with other anomalies. "Malformations of major named vessels" was the name chosen for those malformations named "truncular" in the Hamburg classification.

Most simple malformations are composed mainly of only 1 type of vessel (capillaries, lymphatics, or veins), with the exception of arteriovenous malformation, which contains arteries, veins, and capillaries (ie, not a combined venous malformation with an arterial malformation but rather a separate disease composed of several types of vessels). Similarly, nonacquired arteriovenous fistula is not considered a combined malformation.

Capillary malformations *(*CMs*)*, which have been previously referred to as "port-wine stains," are present at birth as permanent, flat, pink-red cutaneous lesions (Fig. 6.9). The most common location is the head and neck region .

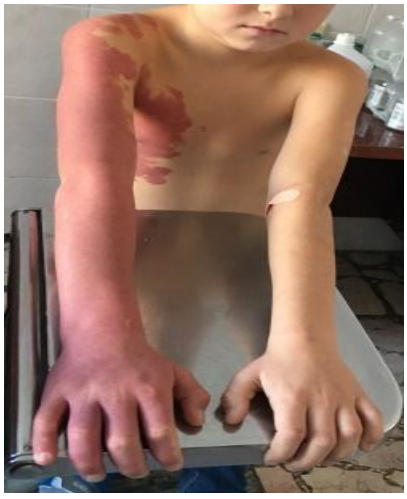


Fig. 6.9. Capillary malformations of the left hand.

Capillary malformations can be associated with underlying soft tissue and skeletal overgrowth as well as other internal abnormalities. These lesions often take part in other clinical syndromes and anomalies, such as Sturge-Weber syndrome (associated to facial lesions), Proteus

syndrome, Cobb syndrome, CLOVES syndrome, and spina bifida occulta. This should be taken into account when evaluating these patients.

Clinically, capillary malformations appear as pink to violaceous macular stains that can be extensive and involve any area of the body. Facial capillary malformations tend to become darker, more violaceous, thicker, develop blebs and sometimes become hyperkeratotic as the patient matures. Capillary malformations in the extremities may be associated with hypertrophy of underlying tissues.

Unlike other vascular malformations, the diagnosis of CMs is usually based on clinical features and no imaging studies are necessary.

Treatment of CMs is indicated primarily for cosmetic purposes. Flash-lamp pulsed-dye laser therapy causing photothermolysis of the CM will improve the appearance by lightening the color of the lesion. Surgery is reserved for the correction of underlying tissue deformities.

Lymphatic malformations (LM) LMs are the wide range of diseases, different in their clinical presentation, course, and prognosis.

According ISSVA 2012 updated classification, LMs are divided into following types:

- **a.** common (cystic) lymphatic malformation
- **b.** generalized lymphatic anomaly (GLA)
- **c.** Kaposiform lymphangiomatosis (KLA)
- **d.** lymphatic malformation in Gorham-Stout disease (GSD)
- **e.** channel-type lymphatic malformation (CTLM)

Complex LMs such as GLA, KLA, channel-type LMs, and LMs in Gorham-Stout disease are frequently diagnosed at a later stage, as have no specific clinical symptoms. They are extremely rare for general practitioners to have intimate knowledge of them. Commonly these diseases are diagnosed at specialized centers. Clinical examination and visualization (ultrasound, MRI) are sufficient to make a diagnosis of cystic LMs. Careful analysis and reconciliation of clinical presentation, hematological changes, radiological aspects, and histopathology are required to make the diagnosis of other LMs types.

GLA is multisystemic lysosomal disorder, that affects visceral organs, skin, and bone. Bone affection is specific for cortical layer preservation and minimal progressing.

KLA is characterized by intrathoracic and extrathoracic lesions, including chylothorax, chylopericardium, cystic LMs, with bone and spleen involved.

Channel-type LM is a complex pathology of lymphatic system that is characterized by dilatation of lymphatic vessels (lymphangiectasis), their dysmotility, and distal part obstruction, that results into lymph inadequate flow, stasis, and lymphatic reflux.

Cystic LMs are the most common. According to the cysts' size, they are divided into macrocystic, microcystic, mixed cystic. Such division is a practical consideration, as macrocystic LMs has better prognosis for good outcome, as they can be treated successfully both surgically and by sclerotherapy. Microcystic LMs' infiltrative nature embarrasses or renders effective treatment

Approximately 75% of these cystic LMs occur in the head and neck. Other anatomic sites are the axilla and thorax, mediastinum, retroperitoneum, buttock, and anogenital regions. Roughly half are detected at birth, with 80% to 90% detected by the age of 2 years; some LMs can be seen prenatally by fetal ultrasonography. From mild cosmetic deformity to life-threatening airway obstruction, these malformations have myriad clinical presentations.

According to previous terminology, macrocystic LMs were referred to as "cystic hygromas" and microcystic LMs as "lymphangiomas." However, these names should be abandoned.

LMs appear as soft compressible masses, and may have a bluish hue. Involvement of the dermis (lymphangioma cutis) may produce puckering of the skin or vesicles that weep clear yellowish fluid.

Diffuse infiltration of the subcutaneous tissue can produce extensive lymphedema that also falls within the spectrum of LMs. One unique factor among the vascular anomalies is that LMs are at risk for infection that can lead to cellulitis or even systemic illness. Similarly, infections located elsewhere in the body or viral illnesses can cause increased size and tension of LMs.

Well-localized and cystic LMs are easily characterized by ultrasonography. MRI, however, provides the most reliable diagnosis and is superior in documenting the full extent of more complex LMs as well as their macrocystic and microcystic components (Fig.6.10-6.12). LMs are hyperintense on T2-weighted sequences because of their high water content.

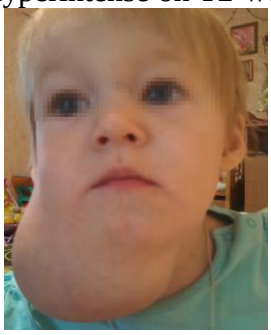
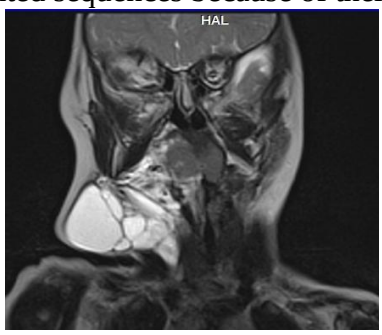
A.  B. 

Fig. 6.10. Macrocystic LM, A – photo, B – MRI.

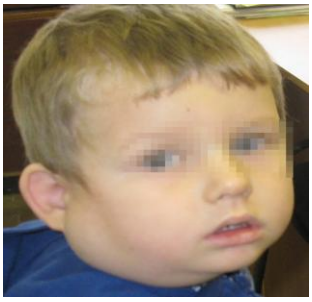
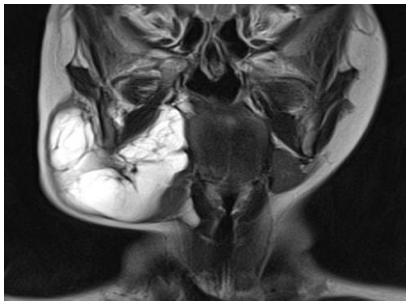
A.  B. 

Fig.6.11 – mixed LM, A – photo, B – MRI.

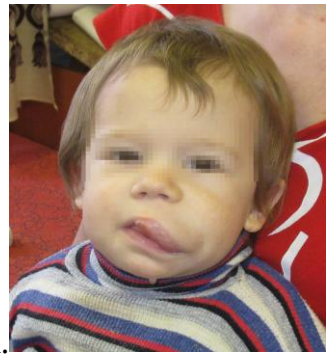
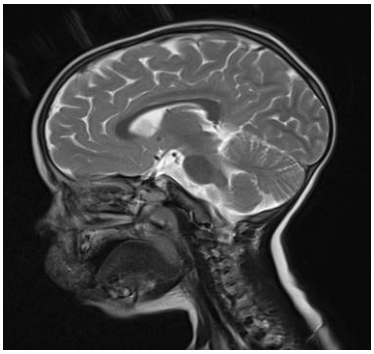
A.  B. 

Fig. 6.12. Microcystic LM. A – photo, B – MRI.

Traditionally management involves surgical therapy, with the goal being removal of involved tissue without sacrificing vital structures. Due to the anatomical relationships in the head and neck, extirpation can be cosmetically and/or functionally morbid. Further, these lesions tend to recur, partially due to the fact that complete excision is frequently impossible. These factors have been the driving force in the search for nonsurgical therapies for the management of LM. Multiple nonsurgical therapies have been proposed and include diathermy, cryotherapy, radiation therapy, fibrin glue, and percutaneous sclerosants. Sclerosants that have been used include doxycyline, bleomicyn, and OK-432. Outcomes are excellent, with minimal morbidity in the majority of cases. (Fig. 6.13).

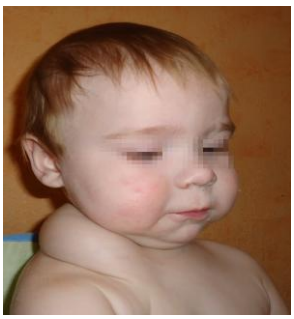
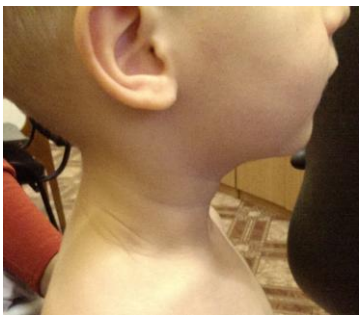
A.  B. 

Fig. 8.11. Neck LM. A – before treatment, B – after treatment with OK-432.

Venous malformations (VMs) often incorrectly called “cavernous hemangiomas,” are slow-flow lesions consisting of venous channels that can develop anywhere in the body. They are most common in the skin and soft tissues. VMs may be seen at birth or become apparent later, depending on the anatomic location. They tend to enlarge slowly with normal growth of the patient but can dilate and become symptomatic at any time. As with other vascular malformations, the proportional growth that occurs may become exaggerated during puberty. On examination, these soft, bluish, compressible lesions will expand with dependent position and Valsalva maneuver (Fig. 6.14).

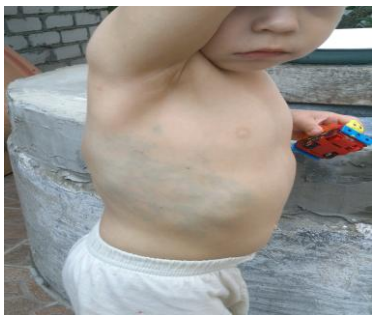


Fig. 6.14. Venous malformations of the chest wall

Episodes of phlebothrombosis secondary to stasis may lead to acute pain and swelling. Associated local overgrowth and limb-length discrepancy are not uncommon. Involvement of bones and joints may lead to pathologic fractures, hemarthroses, and subsequent arthritis.

Large VMs also can be complicated by localized intravascular coagulopathy caused by stasis and stagnation of blood within the malformation, leading to consumption of coagulation factors. The clotting profile consists of prolonged PT, decreased fibrinogen, and elevated D-dimers. The PTT is often normal.

Radiologic modalities useful for the diagnosis of VMs include ultrasonography, MRI, and venography. MRI is most informative and demonstrates hyperintense lesions with T2-weighted sequences.

The indications for treatment of VMs are appearance, pain, loss of function, and bleeding. Unfortunately, cure for VMs, as with LMs, is difficult to achieve for all but the most localized and therefore less problematic lesions.

Intralesional sclerotherapy is the mainstay of treatment for most VMs. Sclerosing agents cause direct endothelial damage, thrombosis, and scarring. VMs have a propensity for recanalization and, therefore, re-enlargement. Cure with sclerotherapy is rare. Results from treatment are often stated in terms of patient satisfaction from decreased pain and appearance, given that recurrence is so prevalent. Surgical resection is typically reserved for well-localized lesions but is marked by procedural morbidity and recurrence.

Arteriovenous malformations (AVMs) are fast-flow vascular malformations characterized by abnormal connections or shunts between feeding arteries and draining veins

without an intervening capillary bed. AVMs are familiar to neurosurgeons as one of the more common vascular anomalies that occur in the central nervous system. Indeed, intracranial AVMs are more frequent than AVMs of the skin and soft tissues within the head and neck region. Other common sites of involvement are the extremities, trunk, and viscera. At birth, they appear as a pink cutaneous blemish that can be confused with both CMs and the premonitory sign of an infantile hemangioma. However, fast flow across arteriovenous shunts is present beneath the innocent-appearing surface. This fast flow becomes more evident in childhood, and the lesion develops into a mass. AVMs grow in proportion with the child, but puberty, pregnancy, or local trauma tends to trigger more rapid expansion. The lesion becomes more obviously warm to touch and may develop a bruit. Because of expansion and local steal phenomenon, skin ischemia can develop, leading to pain, ulceration, and bleeding. For large AVMs, high-output cardiac failure can result.

Ultrasonography and Doppler imaging are excellent tools to elucidate the fast flow of these lesions and to distinguish them from VMs.

The mainstays of treatment for ANMs are angiographic embolization alone or in combination with surgical excision and sometimes with sclerotherapy.

Combined vascular malformations associate 2 or more vascular malformations in one lesion. These may be simple malformations, malformations of major named vessels, or a combination of both types.

Vascular malformations (simple and/or of major named vessels) may be associated with anomalies of bone, soft tissue, or viscera. These nonvascular anomalies are often overgrowth of soft tissue and/or bone or, rarely, undergrowth. Most of these associations are eponymous syndromes.

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2. Evidence-based pediatric oncology / edited by Ross Pinkerton, A.G. Shankar, Katherine Matthay. — 2nd ed., Blackwell Publishing, 2007 – 565 p.
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Tests for initial level of knowledge

1. Regarding haemangiomas, which of the following statements is correct?
   A. They are more common in preterm infants
   B. They are more common in boys
   C. They are generally present at birth
   D. They are common in dark skinned infants
   E. Most haemangiomas completely resolve by the age of 3
2. Regarding complications of vascular anomalies, which of the following statements is correct?
   A. Haemangiomas are malignant tumours
   B. Multiple haemangiomas are a cause of cardiac failure in infants
   C. Port wine stains can cause a coagulopathy
3. Regarding vascular lesions, which of the following statements is correct?
   A. Pulsed dye laser is not a suitable treatment for port wine stains
   B. Pyogenic granulomas are caused by localised infection
   C. Large venous malformations are associated with sudden death
4. What is the main clinical sign of Wilm's tumor?
   A. Abdominal mass
   B. Abdominal pain
   C. Hematuria
   D. Vomiting
   E. Constipations
5. What modality can you use in the management of a port wine stain?
   A. Topical tacrolimus
   B. Oral steroids
   C. Pulsed dye laser
   D. Surgical excision
6. Which of the following is true of a tumor suppressor gene?
   A. Allows unrestricted cell growth and proliferation.
   B. Promotes different phases of the cell cycle.
   C. Produces proteins that block the activity of cyclins.
   D. Regulates cell growth and repair.
7. Which of the following has the most negative prognostic outcome in a patient diagnosed with Neuroblastoma?
   A. High-TrkA expression of the tumor
   B. The patient is considered to be Stage 2 according to the INSS
   C. The patient is six months of age
   D. The tumor expresses N-myc amplification.
8. What is the most common form of pediatric cancer?
   A. Brain tumors
   B. Leukemias
   C. Lymphomas
   D. Neuroblastoma
   E. Retinoblastoma
9. What is the most common localization of teratoma?
   A. Head
   B. Neck
   C. Sacricoccygeal region
   D. Thorax
   E. Abdomen

Keys for tests

1. A, E
2. B
3. B
4. A
5. C
6. C: Progress through each phase of the cell cycle is controlled tightly by a group of proteins called cyclins. Tumor suppressor genes produce proteins that block the activity of cyclins, thereby inhibiting cell growth and division.
7. D: N-myc amplification regardless of age or stage is associated with advanced disease, rapid tumor progression, and a poor prognosis.
8. B
9. C

Tests for final level of knowledge

1. A mother takes her 11 year old son to the emergency department because of a lesion on the back of his outer ear. It appeared suddenly and rapidly enlarged over the course of a week. The lesion is uncomfortable and has bled on minor trauma. What do you think this lesion is?
    A. An embryological ear remnat
    B. An infantile heamangioma
    C. An abscess
    D. A keratin horn
    E. A pyogenic granuloma
2. This 4 year old child was born with an extensive bluish birthmark covering 80% of her upper limb. On examination, it is compressible and can be drained against gravity. What abnormality do you think this child has?
    A. Syndactyly
    B. A venous malformation
    C. An arteriovenous malformation
    D. A haemangioma
    E. Klippel-Trenauny syndrome
3. A two year female has recently been diagnosed with a Wilms tumor. Her presenting signs and symptoms likely included:
    A. A soft mass in the left lower quadrant, pain, dysuria and peri-umbilical petechiae
    B. An abdominal mass, hematuria, hypertension and fever
    C. Enlarged abdomen detected by her parents, decreased ambulation, fatigue, and hyperglycemia
    D. A firm left lower quadrant mass, hyponatremia, constipation, and polyuria
4. You see a 3 year old girl in the emergency department. Her parents have noticed that she has become increasingly irritable over the last few weeks and that she now has a “big tummy.” On examination, a palpable mass is noted in her left flank and confirmed on ultrasound. Blood tests reveal a normal full blood count, normal liver and renal function, but elevated urinary homovanillic (HVA) and vanillylmandelic (VMA) acids. Which of the following is the most likely diagnosis?
    A. Nephroblastoma (Wilm’s tumour)
    B. Lymphoma
    C. Neuroblastoma
    D. Teratoma
    E. Sarcoma
5. A 15 year old girl presents to the emergency department with a tonic-clonic seizure. She has been complaining of headaches over the last two months that have been getting progressively worse. Three years ago she had a primary melanoma with a Breslow

thickness of 4 mm removed from her back. On examination she has a mild left sided hemiparesis. What is the next investigation?

A. MRI scan of brain
B. MRI scan of thorax
C. Full blood tests
D. Immulogical investigations

6. Mother brings her 6 week old doter to see pediatric surgeon. She is concerned about a rapidly enlarging red mark on his nose. It appeared two weeks ago and initially looked like a bruise. It is bright red with an irregular surface. What do you think this lesion is?
   A. A congenital haemangioma
   B. An infantile haemangioma
   C. A salmon patch
   D. A pyogenic granuloma
7. The baby's mother is concerned because she has read on the internet that haemangiomas are tumours and this means that her child has cancer. She is concerned that this needs immediate treatment. What would you tell her?
   A. Although haemangiomas are classified as tumours, they are benign and the majority do not need treatment
   B. She has been misinformed - haemangiomas are not tumours because they regress and only need treatment if there are complications
   C. The haemangioma may be cancerous and an expert needs to examine it
   D. Although the lesion is not cancerous she may want to consider surgery early in order to halt its progression
8. A mother brings her 4 year old daughter with a known venous malformation affecting her left arm to your GP surgery with a sudden painful swelling in her left elbow. Her mother tells you that she has recently been refusing to wear her compression stocking. On examination, she has a large venous malformation affecting her left arm. Her left elbow joint is tensely swollen and painful to touch and is slightly warmer than the right side. She does not complain of any bruising or bleeding and is apyrexial. What do you think is the most likely diagnosis?
   A. Deep vein thrombosis
   B. Haemarthrosis
   C. Cellulitis
   D. Dependent oedema
9. You are investigating the 14 years old girl with a large venous malformation affecting her trunk. The patient's mother tells you that her daughter has also been suffering from nosebleeds as well as easy bruising over the last couple of days. You refer her daughter to hospital to be admitted. Her blood results are as follows:
   - Platelet count: 100 x 109/l (normal range 150-400 x 109/l)
   - D-dimers: 4000 μg/l (normal range 0-372 μg/l)
   - Fibrinogen: 0.8 g/l (normal range 1.7-4.0 g/l)
   - Activated partial thromboplastin time: 28 s (normal range 26.0-38.0 s)
   - Prothrombin time: 10 s (normal range 9.9-12.5 s).

   What do you think the underlying diagnosis is in this patient?
   A. Localised intravascular coagulopathy
   B. Kasabach-Merritt phenomenon
   C. Disseminated intravascular coagulopathy
   D. Thrombocytopenia
10. Mother brings her 4 week old son to see you. She is concerned about a rapidly enlarging red mark on his upper eyelid. It appeared two weeks ago and initially looked like a bruise. It is bright red with an irregular surface and is causing the baby's eyelid to droop. What do you think this lesion is?

A. A congenital haemangioma
B. An infantile haemangioma
C. A salmon patch
D. A pyogenic granuloma

11. You are investigated a 2 month old child with infantile hemangioma of the upper eyelid. Having reassured the mother that this tumor is benign and will regress by itself, what would you do next? D-dimers: 4000 μg/l (normal range 0-372 μg/l)
    A. Refer the baby to your local dermatology or plastic surgery department non-urgently
    B. Send them away, asking the mother to return if she has any other questions
    C. Arrange an urgent ophthalmology review for the baby
    D. Arrange to see the baby again in two months' time

Keys for tests

No 1

A. An embryological ear remnant
These are skin formations, which are not characterized by bleeding
B. An infantile haemangioma
An infantile haemangioma usually regress before age of 3 years
C. An abscess
An abscess is characterized by hyperthermia, local signs of inflammation without bleeding
D. A keratin horn
A cutaneous horn looks like a growth on the outside of the skin. It can appear as a large bump, cone, spike, or horn. The growth may be the same color as the skin
E. A pyogenic granuloma
Pyogenic granulomas can grow rapidly and will often bleed profusely with little or no trauma. This is the most likely diagnosis.

No 2

A. Syndactyly -
is a condition wherein two or more digits are fused together.
B. A venous malformation
This is the most likely diagnosis, because venous malformation tend to get bigger if increase pressure on venous system. If the venous malformation is superficial, it will be discolored blue and may appear in different areas of your body
C. An arteriovenous malformation
the lesion is usually fast-flow, warm, or pulsatile.
D. An infantile hemangioma
At the age 4 years infantile hemangioma usually involutes
E. Klippel-Trenauny syndrome
It’s complex vascular anomalies that usually affected low extremities

No 3

A. C: Lorazepam is indicated for the prevention of anticipatory nausea and vomiting
B. B: A firm flank mass is present that can be seen as an abdominal enlargement. Hematuria, malaise, fever, and hypertension are often present. Wilms tumor presentation may also include nausea and vomiting, anorexia, anemia, and hypotension
C. A: Aniridia is the congenital absence of the iris of the eye and has been identified as a congenital malformation associated with Wilms tumor. Hemihypertrophy and “horsehoe” kidney have also been linked with Wilms tumor.
D. C
E. MRI scan of brain to reveal brain metastasis

No 4

A. A soft mass in the left lower quadrant, pain, dysuria and peri-umbilical petechiae The mas is firm, peri-umbilical petechiae are not common
B. An abdominal mass, hematuria, hypertension and feverA firm flank mass is present that can be seen as an abdominal enlargement. Hematuria, malaise, fever, and hypertension are often present. Wilms tumor presentation may also include nausea and vomiting, anorexia, anemia, and hypotension
B. Enlarged abdomen detected by her parents, decreased ambulation, fatigue, and hyperglycemia symptoms are characteristic of pancreatic defeat
C. A firm left lower quadrant mass, hyponatremia, constipation, and polyuria
D. The mas is localized in upper abdomen, polyuria is not

No 5

A. A congenital haemangioma. This lesion appeared when the boy was 2 weeks old. Congenital haemangiomas are rare tumours that are present at birth and can be divided into two main groups - rapidly involuting congential haemangioma and non-involuting congential haemangioma.
B. An infantile haemangioma. This is a typical history of an infantile haemangioma. It is an extremely common lesion occurring in about 10% of Caucasian births. They have been called strawberry naevi or capillary haemangioma in the past. These terms should now be avoided because strawberry nevus has sometimes been used to describe a pyogenic granuloma; and capillary haemangioma can be confused with capillary malformation (port wine stain).
C. A salmon patch. Salmon patch is the most common vascular lesion of infancy. It occurs in about 40% of infants at birth. These lesions are flat and pale and they do not proliferate. Typically they are found in the central portion of the forehead, the glabella, and upper eyelids and disappear in two to four years. This lesion is raised, dark red, and enlarging and therefore not a salmon patch.
D. A pyogenic granuloma. Pyogenic granuloma is part of the differential diagnosis for infantile haemangioma. In this baby, the lesion is more likely to be an infantile haemangioma because pyogenic granulomas normally present later in life and have a more pedunculated appearance than this diffuse lesion.

No 6

A. Although haemangiomas are classified as tumours, they are benign and the majority do not need treatment An infantile haemangioma is classified as a tumour by the International Society for the Study of Vascular Anomalies. Haemangiomas are benign and the majority do not need treatment.
B. She has been misinformed - haemangiomas are not tumours because they regress and only need treatment if there are complications. Haemangiomas are vascular tumours. Most will regress although treatment may be required if there are complications.
C. The haemangioma may be cancerous and an expert needs to examine it. Infantile haemangiomas are the most common benign tumours in children. They have no features of malignancy. There are some exceedingly rare vascular tumours that can occur in children but the appearance and history will differentiate them from the common infantile haemangioma.
D. Although the lesion is not cancerous she may want to consider surgery early in order to halt its progression Surgery is not indicated for an uncomplicated infantile haemangioma as they are known to completely resolve.

No 7

A. Deep vein thrombosis. Most patients with large venous malformations suffer from painful episodes of thrombophlebitis affecting the superficial veins. The use of compression garments combined with low dose aspirin therapy can reduce the

occurrence of thrombophlebitis. Deep vein thrombosis is less common although there is an increased incidence of sudden death due to pulmonary embolism in patients with large venous malformations.

B. Haemarthrosis. The history of a sudden tense swelling that is painful, in a joint, is suggestive of haemarthrosis. This may be because the malformation is affecting the synovium of the joint. If this is the case, generally haemarthrosis following minor trauma will occur before the age of 10. Haemarthrosis can be an indicator of a serious coagulopathy. So you must take a detailed history with regards to bleeding and check coagulation urgently.

C. Cellulitis. Cellulitis can be a serious and recurrent problem in patients with large venous and lymphatic malformations. This is due to minor skin trauma from itching and dryness, which introduces organisms, coupled with slow flow of blood through the lesion. The patient can progress quickly to septic shock and it is important that you start intravenous antibiotics early. Ideally you should take blood cultures before starting antibiotics. Patients usually complain of increased pain and sometimes only minor swelling within the lesion. It may be difficult to detect erythema within the lesion due to the skin discoloration, although generally the area is hot and the patient may be febrile. In this clinical scenario the swelling is not due to cellulitis because the patient is systemically well. Although cellulitis can occur in the absence of systemic symptoms, in a lesion of this size infection usually progresses rapidly, and you would expect the child would be to be severely unwell.

D. Dependent oedema. Although venous malformations enlarge when dependent - that is, when gravity acts to pool blood within the abnormal vessels, this is different from dependent oedema, where excess fluid moves into the tissues. Wearing compression garments during the day can greatly improve this swelling although they can be hot and uncomfortable. This patient does not have dependent oedema because the swelling is mostly concentrated around the elbow joint. If due to dependent oedema you would expect to find swelling in the most dependent part of the limb, that is, the hand.

No 8

A. Localised intravascular coagulopathy. Localised intravascular coagulopathy is common in patients with large venous malformations. This can become chronic disseminated intravascular coagulopathy. Patients tend to present after childhood with painful but compressible swelling.

B. Kasabach-Merritt phenomenon. Kasabach-Merritt phenomenon is not associated with venous malformations.

C. Disseminated intravascular coagulopathy. Disseminated intravascular coagulopathy is a consumptive coagulopathy that can develop in patients with large venous malformations. It can be life threatening. The clinical presentation can range from slight bruising to thromboembolic events. Blood tests show low fibrinogen, platelets, and clotting factors with raised D-dimer or fibrinogen degradation products and prolonged activated partial thromboplastin time and prothrombin time. This needs urgent specialist treatment by a vascular anomalie specialist and a haematologist. This is not disseminated intravascular coagulopathy because the activated partial thromboplastin time and prothrombin time are normal.

D. Thrombocytopenia. These blood tests do show a low platelet count. Since the low platelets are part of a global disturbance in coagulation, you cannot explain these results by an isolated thrombocytopenia.

No 9

A. A congenital haemangioma. This lesion appeared when the boy was 2 weeks old. Congenital haemangiomas are rare tumours that are present at birth and can be

divided into two main groups - rapidly involuting congential haemangioma and non-involuting congential haemangioma.

B. An infantile haemangioma. This is a typical history of an infantile haemangioma. It is an extremely common lesion occurring in about 10% of Caucasian births. They have been called strawberry naevi or capillary haemangioma in the past. These terms should now be avoided because strawberry nevus has sometimes been used to describe a pyogenic granuloma; and capillary haemangioma can be confused with capillary malformation (port wine stain).
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D. A pyogenic granuloma. Pyogenic granuloma is part of the differential diagnosis for infantile haemangioma. In this baby, the lesion is more likely to be an infantile haemangioma because pyogenic granulomas normally present later in life and have a more pedunculated appearance than this diffuse lesion.

No 10

A. Refer the baby to your local dermatology or plastic surgery department non-urgently. Uncomplicated infantile haemangiomas should be left to regress spontaneously.
B. Send them away, asking the mother to return if she has any other questions. This would be reasonable management in a small uncomplicated infantile haemangioma but this child is at risk of amblyopia.
C. Arrange an urgent ophthalmology review for the baby. This child is at risk of amblyopia. Amblyopia is reduced vision in an eye that has not received adequate use in childhood. If vision in one eye is obstructed for a significant period of time (for example, by a large eyelid tumour) there will be central suppression of the image from that eye leading eventually to a non-recoverable deficit. He needs to be seen by a paediatric dermatologist, preferably with a multidisciplinary team and ophthalmologist at the earliest opportunity in order to assess his vision and if necessary start treatment. Possible treatment options include intralesional steroid injections, systemic steroid treatment, embolisation, vincristine therapy,[6] propanolol[7] (only recently recognised as a treatment option), and surgical debulking. Treatment is always started and supervised by a specialist such as a dermatologist or plastic surgeon with an interest in vascular lesions.
D. Arrange to see the baby again in two months' time. This would be too late. By this time the sight in the baby's affected eye may be permanently damaged.

Tasks for final level of knowledge

Task 1. A GP refers a 7 year old girl who is unwell with a venous malformation covering 60% of her body to you. She has a sore throat. Her temperature is 38.5°C. On examination she has widespread bruising and petechiae. Her blood results are as follows: Platelets: 20 x 10^9/l (normal range 150-400 x 109/l), D-dimer: Greater than 5000 µg/l (normal range 0-372 µg/l), Prothrombin time: 15 s (normal range 9.9-12.5 s)Fibrinogen: 0.8 g/l (normal range 1.7-4.9 g/l). Which of the following coagulopathies could the child have?

Answer Key. Localised intravascular coagulation

Task 2. Mother brings her 4 week old son to see you. She is concerned about a rapidly enlarging red mark on his upper eyelid. It appeared two weeks ago and initially looked like a bruise. It is bright red with an irregular surface and is causing the baby's eyelid to droop.

1. What do you think this lesion is?
2. What is the treatment option?

Answer Key. 1.This is a typical history of an infantile haemangioma. It is an extremely common lesion occurring in about 10% of Caucasian births. They have been called strawberry naevi or capillary haemangioma in the past. These terms should now be avoided because strawberry nevus has sometimes been used to describe a pyogenic granuloma; and capillary haemangioma can be confused with capillary malformation (port wine stain). Oral Propranolol to prevent visual disturbance

Task 3. A 3 year old boy is brought to the emergency department with a large, painless abdominal mass. The child's parents inform you that the child has been generally unwell for three weeks. You are concerned that the mass may be a tumour. Which investigations would be best to confirm your suspicion?

Answer Key.

- Abdominal ultrasound
- Full blood count
- Liver function tests
- Urinary homovanillic (HVA) and vanillylmandelic (VMA) acids
- MRI

Task 4. You are called to the emergency department to see a 14 year old boy. He presented with chest pain and a two day history of gradually increasing swelling and plethora of the face and right arm, and distended neck veins. Based on clinical examination and his chest x ray (demonstrating a large mediastinal mass), your working diagnosis is that of superior vena cava obstruction. You arrange a CT scan which shows a large tumour, likely lymphoma, encapsulating the superior vena cava and infiltrating the myocardium. What is your immediate management of this patient?

Answer Key. Urgent hospitalization, incisional biopsy to make the exactly diagnosis and begin the treatment.

Materials for self-study of the students

Main tasks	Notes (instruction)
Repeat: Anatomy of mediastinum, abdominal organs, blood and lymphatic vessels Physiology of vasculogenesis Pathogenesis of embryonic tumors	To sketch out the anatomy of mediastinal organs Top represent the methods of diagnosis of diagnosis of tumors and vascular anomalies
Study: Pathogenesis of childhood cancer The diagnostic possibilities of US, CT, MRI, PET	To make differential diagnosis of childhood cancer To make the indications to surgical treatment of tumors and vascular anomalies To know minimally invasive method of vascular anomalies treatment

THEME 7

ACUTE HAEMATOGENIC OSTEOMYELITIS. TB - OSTEITIS.

Topic actuality:

Pyo-inflammatory diseases make up a serious problem in children. Clinical picture of purulent diseases has changed, this is connected with spread of microorganisms resistant to antibiotics. So, structure and development of pyo-inflammatory diseases have changed. Topic urgency is stipulated for wide distribution of these diseases, their complications and consequences that may cause early disability.

Aims:

1. To learn the list of pyoinflammatory diseases of bones, joints and soft tissues.
2. To know main clinical manifestations of pyoinflammatory diseases of bones, joints and soft tissues.
3. To differentiate pyoinflammatory diseases of bones, joints and soft tissues according to their cause and localization.
4. To discuss the additional methods of examination:ultrasound diagnostics, roentgenological, laboratory and biochemical analyses, haemodynamic data (AP, Ps), t^o.
5. To show puncture of the joint, abscess opening.
6. To identify the peculiarities of development of pyoinflammatory diseases of bones, joints and soft tissues.
7. To explain and establish the tentative clinical diagnosis.
8. To offer algorithm of doctor's actions at pyoinflammatory diseases of bones, joints and soft tissues, tactics of treatment.
9. To discuss general principles of treatment of pyoinflammatory diseases of bones, joints and soft tissues, determine indications for the surgical treatment.

Basic skills and knowledge necessary for the topic (interdisciplinary integration):

Subjects	Skills
Propaedeutics of pediatric diseases. Faculty pediatrics Roentgenology Surgical diseases, operative surgery and topographic anatomy Pharmacology Physiotherapy and curative physical training Traumatology	To describe case history of children with pyoinflammatory diseases of bones, joints and soft tissues. To use additional methods of examination necessary to establish the diagnosis, to discuss the data obtained. Roentgenological methods of examination of bones and joints. To show schematically joints and bones. Technique of joint puncture. To determine key priorities of mini-invasive methods of examination. Administration of pathogenetic and symptomatic therapy. Administration of physiotherapy and curative physical training. To be able to perform immobilization of the extremities.

4. Tasks for individual self-preparation for the lesson.

4.1. The list of key terms and characteristics a student must know :

№	Term	Definition
1.	Acute haematogenous osteomyelitis	Severe pyoseptic bone disease that may develop on the background of changed microorganism reactivity, which may be connected with growth, and is accompanied with considerable impairment of homeostasis.
2.	Lymphadenitis	Purulent inflammation of the lymph nodes.
3.	Adenophelgmon	Purulent inflammation of the group of the lymph nodes with subcutaneous cellular tissue.
4.	Abscess	Purulent inflammation of soft tissues demarcated with a capsule.
5.	Phlegmon	Purulent inflammation of soft tissues without definite borders.

4.2. Theoretical questions:

1. Etiological structure of pyoinflammatory diseases of bones, joints and soft tissues in children.
2. Pathogenesis of pyoinflammatory diseases of bones, joints and soft tissues, anatomic-physiological peculiarities and adjacent factors that may contribute to the process generalization.
3.To know clinical picture of phlegmon, abscess, lymphadenitis, hematogenous osteomyelitis.
4. To know principles of additional methods of examination and interpreting the obtained data at this pathology.
5. Trends of complex therapy of pyoinflammatory diseases of bones, joints and soft tissues in children.
6. Possibility of surgical intervention to influence the local focus in complex therapy of pyoinflammatory diseases.

7. Tactics of treatment after haematogenous osteomyelitis.

4.3. Pracical tasks (level 3)

1. To collect the anamnestic data, including information about pregnancy and delivery development.
2. To examine an ill child, to perform palpation, auscultation.
3. To describe the objective status and define clinical and roentgenological symptoms of pyo-inflammatory diseases- phlegmon, abscess, lymphadenitis, haematogenous osteomyelitis and their complications.
4. To explain and make plan of examination and treatment.
5. To define indications for operative treatment, peculiarities of postoperative period.
6. To perform bandaging, technique of incisio, puncture of the bone and joints

CONTENTS

Bone and Joint Infection (BJI) - Osteomyelitis (OM) and septic arthritis (SA) are relatively rare infections in children. Depending on the age of the patients, the clinical picture may vary from predominantly general symptoms and pseudoparalysis in infancy to localized pain and fever in older children and adolescents. OM and SA can be followed by hyperthermia of unknown origin and are difficult to diagnose.

For practical reasons, by the nature of the course of the disease, "acute", "subacute" and "chronic" cases are cases with a history of < 2 weeks, 2 weeks - 3 months and > 3 months, respectively.

The epidemiology of BJI in children changes in the era of resistant organisms, as well as congenital and acquired immunodeficiency. Improvements in microbiological diagnostics, especially new molecular techniques, have increased the rate of detection of demanding and slow-growing microorganisms. Recent epidemiological studies have shown a fairly stable incidence of about 0.13–0.22 per 1000 children per year. There is an increase in the incidence rate compared to previous decades. Children under the age of five are most often affected, and between the ages of one and four, boys are twice as likely to get sick as girls. In the first year of life, there are no gender differences in morbidity.

In studies conducted before the conventional molecular diagnostic tests were available, the most commonly excreted organism was methicillin-susceptible Staphylococcus aureus (MSSA). However, the incidence of certain microorganisms, such as S. aureus, containing the Panton-Valentine's leukocidin virulence gene (PVL), and methicillin-resistant S. aureus (MRSA), has increased. The incidence of Haemophilus influenzae type b has decreased, while the incidence of others, such as Kingella kingae, has increased. There is a pronounced variability of pathogens by geography.

TB infection is important to consider in risk groups in all age groups, although it primarily affects older children and, as a rule, in countries with endemic tuberculosis (TB). TB OM often affects the spine and is an important differential diagnosis in children with back pain. Children with primary immunodeficiency, sickle cell disease and human immunodeficiency virus (HIV), as well as premature infants, are at increased risk of osteoarticular infection caused by a different set of microorganisms compared to children without underlying medical conditions.

Pathophysiology

In most cases, OM in children is acute and hematogenous in origin. The putative port of entry or primary lesion can be identified in about 44% of cases, most often due to ear, nose, and throat infections. The child develops bacteremia, and the microorganism enters the bone through the feeding artery. In children, the metaphyseal area is most often affected, since it has dense capillary loops that allow bacteria to precipitate and form an infectious focus, where virulence factors promote adhesion and cause bone lysis. Localized necrosis occurs and the inflammatory exudate causes bone destruction, which promotes the spread of infection. The Havers and Volkmann canals contract and collapse, and if left untreated, infection and necrosis spread to the periosteum. This causes capillary compression and ischemia, further necrosis and ultimately sequestration. A

critical level of bacteremia is required for OM to occur, although host immunity is likely to play a significant role in individual susceptibility.

Subacute OM with no previous clinical picture or bacteremia is sometimes noted. This is usually a direct infection of a bone or joint as a result of puncture wounds, trauma, or surgery. The infection is more insidious, and Brody's abscess can develop in the bone without damaging the periosteum.

SA affects the joints and the synovium and joint space. In children, the joint capsule extends to the metaphysis in most joints, so septic arthritis can develop as a secondary infection from the adjacent OM. Conversely, OM can develop when bacteria break through the periosteum from adjacent primary septic arthritis. Adjacent infection is more likely in young children because there are transphyseal vessels that cross the growth plate into the pineal gland, providing additional transmission routes between the joint and bone. In older children, the growth plate, on the other hand, can serve as a barrier to the spread of infection in the joint. This barrier disappears when the cartilage of the growth plate ossifies.

Discitis is an infection / inflammation of the intervertebral disc or vertebral endplate that is rare in children. It is difficult to diagnose, especially in non-verbal toddlers. Unlike adults, the intervertebral discs in children under the age of 8 are vascularized, since these are areas of growth. This contributes to the hematogenous spread of the infection to the intervertebral disc. Sometimes microbes can be identified by biopsy, which is often done to rule out malignancy. There is usually a clinical response to antimicrobial therapy.

Clinical presentation, diagnostics and microbiology

Newborns: 2 months and younger

Babies may have non-specific symptoms or signs, such as refusal to eat, drowsiness, fever, or pseudoparalysis of the limbs. Parents sometimes report that their child is in pain when changing a diaper or bandage. Differential diagnosis in case of damage to the upper limb - Erb's palsy or fracture of the clavicle associated with birth trauma. Babies can also be irritable, edema, or erythema.

Risk factors, as with any neonatal sepsis, include prolonged rupture of the membranes, maternal hyperthermia, and known maternal carriage of group B Streptococcus.

The most common organisms causing osteoarticular infections are group B Streptococcus and S. aureus.

Infants in the intensive care unit are at increased risk of bacteremia and therefore acute hematogenous osteoarticular infection due to frequent invasive procedures, ventilation, central venous access, parenteral nutrition, and immature immunity. This increases the risk of developing coagulase-negative staphylococci, group B streptococci, S. aureus and Candida spp. Long bones and large joints, especially the thighs, are usually affected. Bone-articular infections are likely to be multifocal in this age group, and all potential sites should be assessed if suspected.

Premature babies are at particular risk of bacterial dissimilation into the blood from the intestines, especially with necrotizing enterocolitis. Therefore, they are more likely to be infected with enterococci or gram-negative organisms such as Escherichia coli and Klebsiella pneumoniae. This age group is also at a higher risk of contracting meningitis due to bacteremia. Infants with central venous access and complete parenteral nutrition are susceptible to fungal sepsis. Cephalohematoma with secondary skull infection and meningitis have been reported, as well as heel OM after heel injection for screening Guthrie.

All infants under 3 months of age with fever greater than 38 ° C or feeling unwell due to risk factors for sepsis should be screened for sepsis, including blood culture, lumbar puncture, urine culture, and chest x-ray. Typically, infants are then empirically treated with broad-spectrum intravenous (IV) antibiotics until culture results and/or symptoms resolve.

Some newborns do not have septic fiver, but there is localized edema and / or pseudoparalysis. It is most common in term infants who have been discharged from the hospital. It can be a soft tissue abscess or erythema.

White blood cell (leukocyte) counts, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) are not informative diagnostic indicators in the age group of newborns, and normal values cannot rule out serious infection.

A conventional x-ray is usually the first examination of a bone lesion because it is fast, affordable, and allows fractures and dislocations to be ruled out.

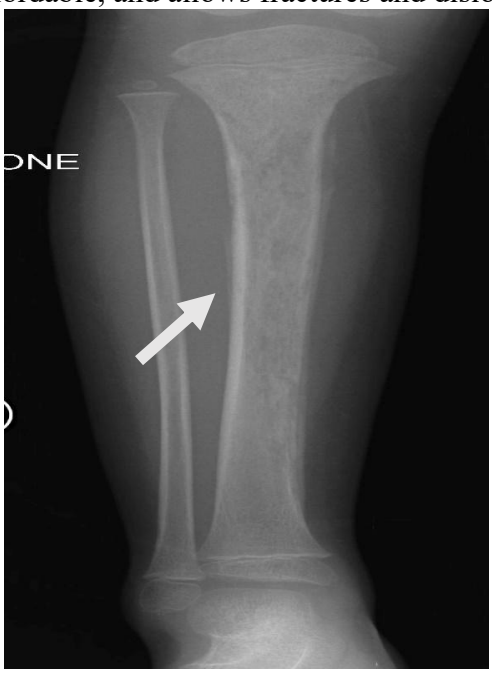


Fig. 7.1 Acute ostomyelitis of the tibia. Linear periostitis and macular osteoporosis on an X-ray in frontal projection..

With confirmed osteoarticular infection, up to 90% of primary radiographs are without pathological changes. For infections of bones and joints in all age groups, magnetic resonance imaging (MRI) is the preferred imaging method. In newborns, this can be done without general anesthesia ("feed and wrap").

Ultrasound scans are especially useful for premature infants who cannot be removed from the incubator and may reveal a bone abscess or joint effusion. Effusions, abscesses, and joint involvement are easy to see, and ultrasound is also useful as an aspiration assistance method to confirm the diagnosis and identify the pathogen.

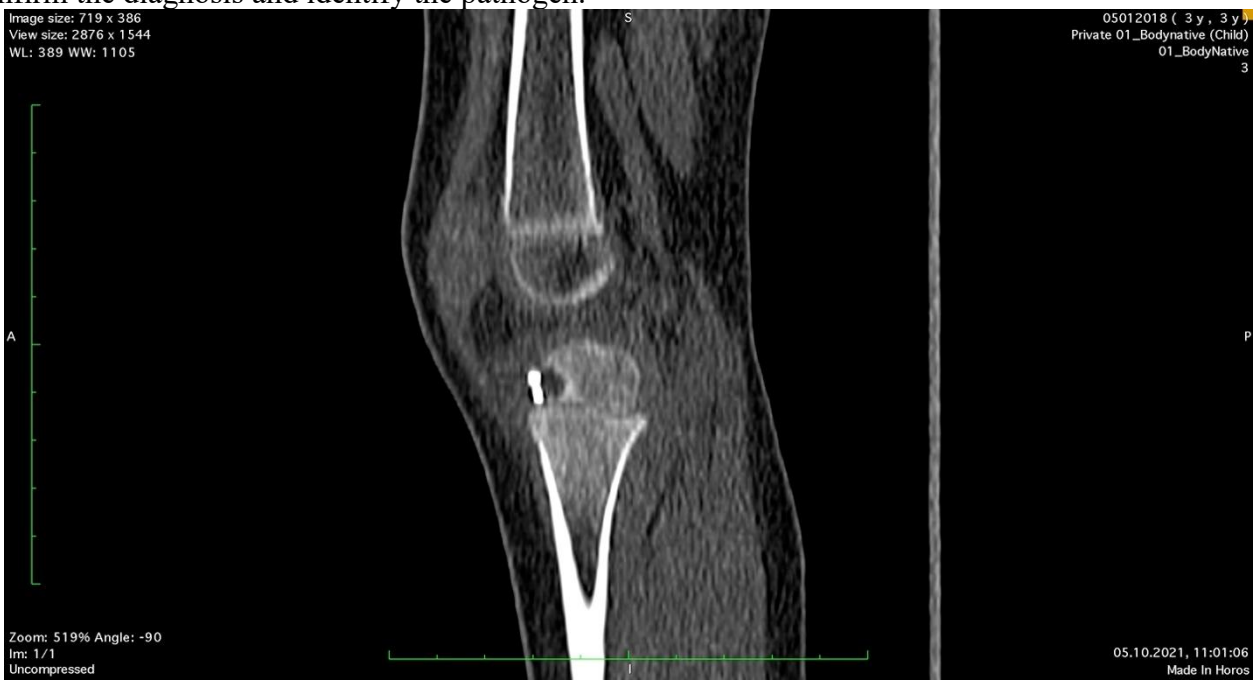


Fig. 6.2 A 3-year-old child has a focus of osteolysis in the proximal epiphysis of the tibia. Osteomyelitis developed after an infected wound with the presence of foreign bodies, which are visualized on a CT.

Children: 3 months to 5 years

After three months, it is likely that the infection was from the environment and not perinatally. The most common pathogens are S. aureus, Streptococcus pyogenes (group A streptococci), and Streptococcus pneumoniae.

Kingella kingae is becoming an increasingly prominent pathogen in this age group as a result of improved diagnostic tools. The use of polymerase chain reaction (PCR) for diagnosis has shown that this gram-negative organism is more common in this age group than S. aureus. It is finicky and therefore not grown using traditional cultivation methods, although it may be more successfully grown from broth. It has been consistently shown to be the causative agent of previously negative culture-negative bone-joint infections.

Invasive K. kingae infection probably occurs in young children after pharyngeal carriage. Children with osteoarticular infection with K. kingae are more prone to afebrile and are more likely to have normal CRP and white blood cells than children with infection with S. aureus and other gram-positive organisms. This can delay the diagnosis, and a small number of children will become seriously ill with K. kingae osteoarticular infection.

Children have any combination of pseudoparalysis, pain, lethargy, bear weight loss, and sometimes hyperthermia of unknown origin. There may be deformity, erythema, pain, swelling, or limited range of motion. The differential diagnosis of a lame child or a child with a painful limb includes transient synovitis or "irritation of the hip", trauma (“child abuse” syndrome must be considered), leukemia, and autoimmune arthritis such as juvenile idiopathic arthritis. The latter remains the most common cause of monoarthritis in children. However, if a child has monoarthritis, the most important diagnosis to rule out is septic arthritis, even in children with a known diagnosis of juvenile idiopathic arthritis.

CRP and ESR are sensitive diagnostic tools in all children older than three months. Although CRP may be a more reliable test than ESR, normal values do not rule out culture-negative osteoarticular infection. Subacute OM in outpatient children may have more subtle symptoms of pain and limited movement without fever. These children may have normal CRP and ESR. The differential diagnosis is Ewing's sarcoma or other bone tumor, so imaging and biopsy may be indicated.

Serum procalcitonin is sensitive and reported to be more specific, but not more susceptible to bone and joint infections than CRP or ESR, especially when the lower cut-off value of 0.4 ng / mL is used. Modern molecular tests, such as PCR, can improve the effectiveness of the diagnosis of osteoarticular infections even after the initiation of antibiotic therapy. However, these tests are usually not available at many centers.

If BJI is suspected, an adequate blood culture should always be obtained. However, since blood culture is positive in only 10–70% of children with osteoarticular infection, a negative predictive value is low.

X-rays are usually done to rule out fracture and conditions such as Perthes disease and dislocation of the femoral epiphysis when the femur is affected. If the infection persists for 10 days or more, a simple x-ray can show a reaction in the periosteum or lytic changes in the bone.

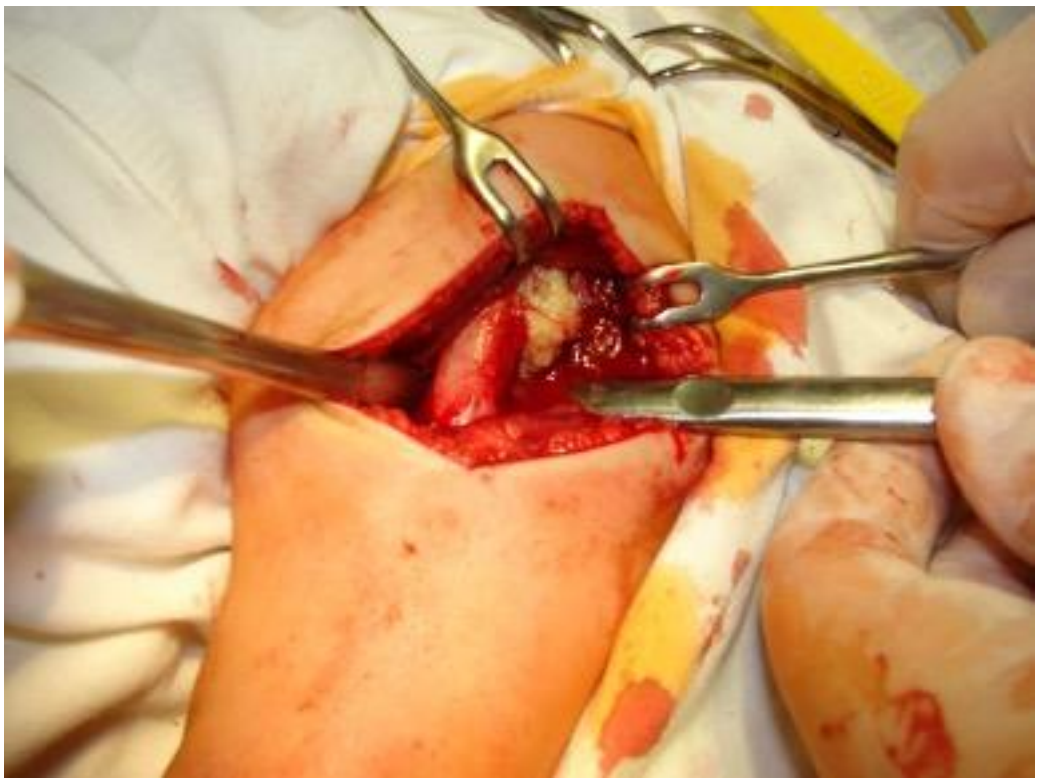


Fig. 6.3 Surgical wound in a 5-year-old child with tbc osteitis of the proximal shoulder.

Computed tomography (CT) can be helpful for young children, but is not recommended due to the dose of ionizing radiation. However, it shows more subtle changes and takes less time than an MRI, so the child does not need to be sedated or anesthetized. In addition, computed tomography can be useful for guided biopsies, especially if the spine is affected. Technetium bone scans are now used less frequently due to the associated radiation exposure. However, at times, it can be quite helpful in multifocal disease.

MRI is the imaging technique of choice for diagnosing osteoarticular infection. It is sensitive and specific, and is especially useful for the simultaneous assessment of bones, joints and soft tissue without ionizing radiation. Acute OM usually presents on MRI as decreased bone marrow intensity on T1-weighted images without an increase in gadolinium. If gadolinium is used, abnormal amplification is observed when sequencing with T1-weighted saturated fat. Increased bone marrow intensity is observed on T2-weighted and short tau inversion recovery (STIR) sequences. STIR sequences are fluid sensitive and can be useful for localizing an infection to a specific site in difficult cases. MRI is also useful for differentiating subacute OM or Brodie's abscess and bone tumors.

In young children, the pineal gland and growth plate are more likely to be affected, and bone growth may be impaired, causing deformity. MRI with gadolinium shows hypo-amplifying lesions in the growth cartilage if there is a lesion. This is not visible in the unimproved images; therefore, gadolinium reinforcement is recommended in this group.

If S. aureus is identified as the causative agent, consideration should be given to the presence of MRSA, MRSA communities (CA-MRSA) and PVL strains, especially in severe disease.

Children who have recently had a chickenpox infection are at risk of developing invasive group A streptococcus (GAS). GAS also colonizes the oropharynx and causes tonsillitis and pharyngitis in young children. Bone-articular GAS infections are almost always hematogenous. This is associated with toxic shock syndrome, and children with invasive GAS infection may be shocked when presented, requiring aggressive resuscitation and treatment with intravenous immunoglobulin and antimicrobial drugs with antitoxin activity.

Children over 5 years old

BJI with S. aureus are most common in older children. This is usually the MSSA. However, S. aureus MRSA and PVL (PVL-SA) should always be considered in severe cases, for which prompt treatment with appropriate antibiotics is essential. Children with MRSA or PVL-SA typically have greater elevations in CRP and white blood cells, more persistent hyperthermia, and more severe disease. Although these biochemical parameters should increase the MRSA or PVL-SA infection suspicion index, they may be normal and should not be used to differentiate the infectious agent. In both MRSA and PVL-SA, osteoarticular infection is much more likely to be complicated by deep vein thrombosis and associated embolic sequelae than MSSA disease, pathological fractures, and pneumonia.

Older children and adolescents are the group most likely to develop subacute osteoarticular infection, in part because of an increased likelihood of puncture wounds and a decrease in bacteremia. It is important to consider Pseudomonas aeruginosa in this group, especially if there is a history of stab wounds to the foot.

Rarer organisms include non-tuberculous mycobacteria, Pantoea agglomerants caused by palm thorn injury, Fusobacterium necrophorum associated with chronic sinusitis, Bartonella henselae associated with cat scratches, brucellosis in endemic regions, and Candida spp. associated with paronychia, corticosteroid prescription, or primary immunodeficiency.

Tuberculous (TB) bone infection

This is usually seen in the adolescent age group. Half of all children with probable tuberculosis do not have an active pulmonary focus. In children, pulmonary tuberculosis is often asymptomatic. The initial infection may be missed, and the subsequent infection of the bones and joints may appear much later, when local symptoms appear.

The pathogenesis of TB bone infection is hematogenous or sometimes direct spread from a nearby lymph node. Tuberculous OM is characterized by necrotic (caseating) granulomatous inflammation. Diagnosis includes Mantoux test, interferon gamma release assay (IGRA), imaging and biopsy of suspected lesions, as these may be difficult to distinguish from neoplasms.

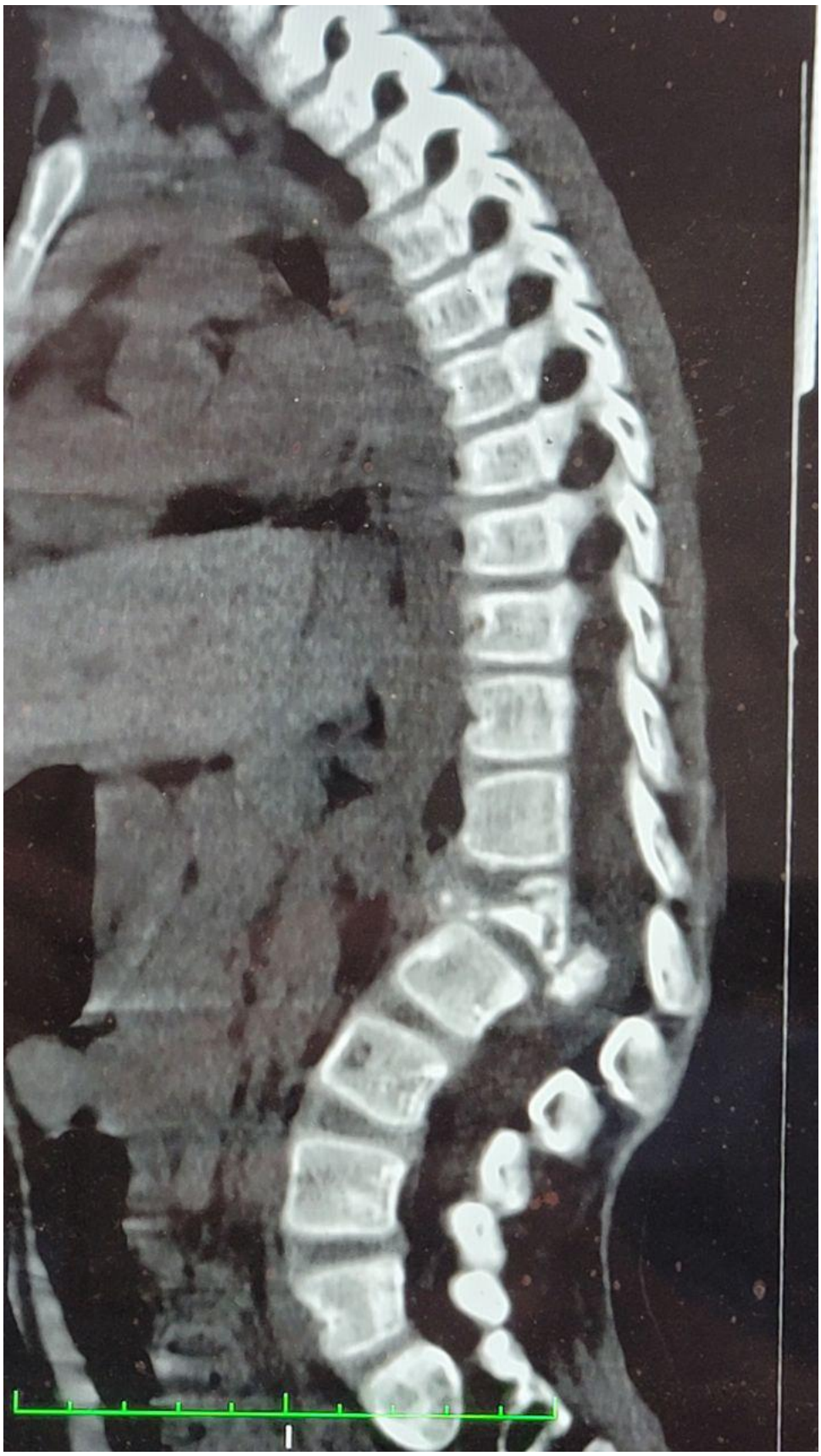


Fig. 7.4 CT 6y. patient with L1 bone destruction by TB infection.

At least half of all TB infections of the bones involve the spine, usually the lower thoracic and lumbar spine. The rest have infections of the long bones and synovial joints. After the spine, the hips, knees, ankles, and elbows are most commonly affected.

Pott's disease is a vertebral OM with an adjacent paravertebral abscess, later associated with extensive bone destruction and collapse of the spine. Children with Pott's disease are at risk of spinal cord compression. They may need internal or orthopedic stabilization of the spine. Deformity occurs when there are multiple levels of collapse in the later stages of the disease.

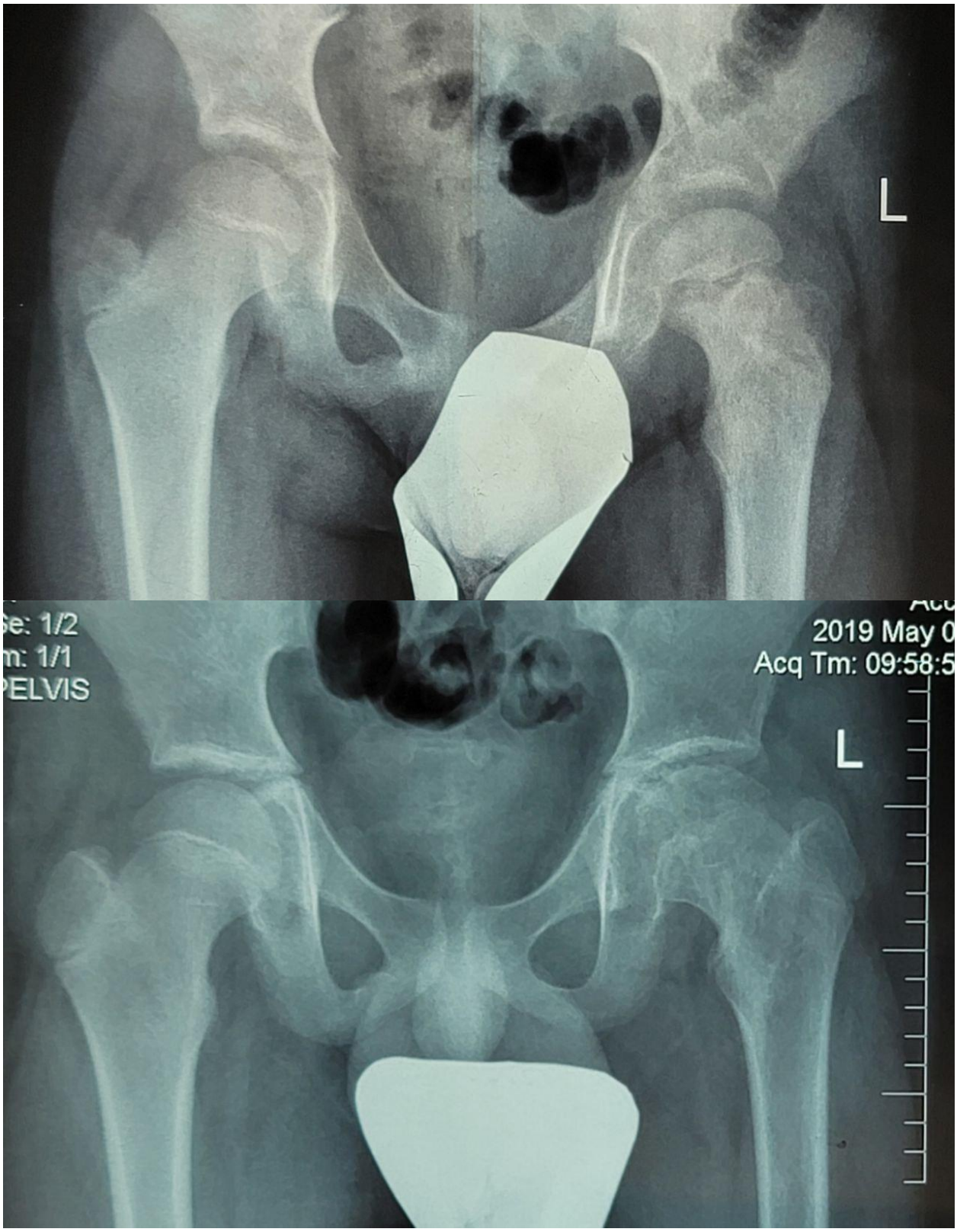


Fig. 7.5-7.6 X-ray of a child with septic arthritis of the hip joint. Osteolysis is visible in the area of the growth zone. The X-ray taken a 2 years later shows a partial closure of the growth zone and a relative shortening of the left femoral neck.

Tuberculous dactylitis is primarily a disorder of young children (<10 years of age) and may go unrecognized for a long time. The hands are usually affected, but the feet can also be affected.

Initial treatment includes adequate drainage of pus, collection of culture specimens and other microbiological investigations, including antibiotic susceptibility testing, and prompt initiation of empiric antibiotic therapy.

The choice of empirical antimicrobial therapy is based on the most likely pathogen based on patient age, immune status, underlying disease, Gram stain of the aspirate, and other clinical and epidemiological considerations, including the prevalence of MRSA in the community.

Suggested treatment for uncomplicated (rapid resolution of fever and other symptoms) BJI in childhood may include short intravenous therapy followed by high-dose oral antibiotics.

The total duration of treatment is 3-4 weeks for OM and 2-3 weeks for SA.

Treatment less than 3 (OM) and 2 (SA) weeks is not recommended.

Antibiotic treatment

Regimens for the first empirical and then targeted antibiotic treatment should take into account local, regional and national characteristics of bacterial contamination and antibiotic recommendations.

Newborns: 2 months and younger

Antibiotic treatment for acute hematogenous OM usually begins empirically and focuses on treating all common infectious organisms in every age group.

In the neonatal period, treatment should include broad-spectrum antibiotics, including gram-positive and gram-negative microorganisms. However, since most cases are caused by group B streptococci, treatment should be quickly optimized once confirmed. Ideally, there should be adequate penetration of the blood-brain barrier because of the higher risk of meningitis after bacteremia in this group. Third-generation cephalosporins or a combination of beta-lactam and gentamicin are often used.

In addition, antibiotics for resistant organisms such as MRSA (usually vancomycin) and extended-spectrum beta-lactamase (ESBL) producing organisms should be considered in preterm infants and children with significant perinatal morbidity admitted to the intensive care unit. Antibiotics are widely used in neonatal wards and there is a high risk of nosocomial infections, especially with extended stays. Antifungal drugs (usually fluconazole, liposomal amphotericin B, or caspofungin) should be considered if there is no response after 24 to 48 hours of antibiotic treatment.

Intravenous antibiotic therapy is recommended for newborns between two and six weeks.

Children aged 3 months to 5 years

Children between three months and five years of age should receive initial intravenous antibiotic therapy that covers both S. aureus and K. kingae. K. kingae is inherently resistant to glycopeptide antibiotics and also has low sensitivity to clindamycin. K. kingae isolates from the oropharynx that are increasingly producing beta-lactamases. Therefore, the antibiotic of choice for intravenous administration should be a second- or third-generation cephalosporin or other appropriate drug recommended by local or regional microbiologists.

It is not known how long children with OM should be treated with intravenous antibiotics. Long courses of up to six weeks are historically preferred. However, more recent randomized controlled trials indicate that a short course of intravenous antibiotics (2-7 days) followed by oral therapy for one to two weeks is sufficient.

The transition to enteral antibiotics is especially important for young children, since compliance with the regimen depends on the taste of the selected drug. At this age, children tend to take liquid antibiotics. Some antibiotics, such as flucloxacillin and clindamycin, are tasteless and poorly tolerated. Erythromycin is often used in children who are allergic to penicillin. However, due to side effects from the gastrointestinal tract, the oral course is often not completed. For

flucloxacillin, clindamycin and erythromycin, the need to take the drug every 6 hours prevents most parents from successfully administering adequate antimicrobial therapy to their children. Oral cephalosporins and amoxicillin / clavulanic acid taste good, can be administered three times daily, and usually do not cause serious side effects. Children who are genuinely allergic to penicillin rarely cross-allergic to cephalosporins. However, clindamycin should be used in patients with immediate hypersensitivity if macrolide resistance is not suspected.

Children over 5 years old

In regions with low incidence of MRSA, older children should first be treated with high-dose intravenous flucloxacillin (50 mg / kg four times daily) unless there is a history of multiple hospitalizations or other risk factors, such as recent travel to areas where MRSA is common. If there is no clinical response after 24–48 hours of treatment, consideration should be given to switching to an alternative antibiotic covering resistant organisms. In addition, it is necessary to suspect and investigate the formation of an abscess.

If MRSA is suspected, it is advisable to start treatment early because of the higher incidence of serious complications. Glycopeptides are usually first-line, often vancomycin. Linezolid and daptomycin are also increasingly used in difficult cases after consultation with a microbiologist. If PVL-SA is suspected, empirical regimens for initiation of treatment often include anti-MRSA therapy plus additional antistaphylococcal agents (such as linezolid and clindamycin). Although there is limited data on specific treatment regimens for PVL MSSA and PVL MRSA bone and joint infections, consult a specialist and focus on antibiotics once the full antibiotic susceptibility profile is known. Patients with MRSA or PVL-SA OM should be closely monitored for abscess formation and deep vein thrombosis. Intravenous treatment may be extended.

The transition to enteral antibiotics for MSSA infection may consist in the use of flucloxacillin or amoxicillin / clavulanic acid tablets or cephalexin suspension if the tablets cannot be taken. If flucloxacillin is prescribed, the 6-hour regimen must be strictly adhered to. For uncomplicated MRSA infection, clindamycin (also a 6-hour dosage) and linezolid are suitable drugs, although linezolid should be used for resistant organisms or recurrent intractable infections. The clindamycin suspension has an unpleasant taste and may not be well tolerated; therefore, tablets are recommended whenever possible. It should be noted that Clostridium difficile enterocolitis is rare in children receiving clindamycin.

In subacute cases where P. aeruginosa infection is suspected, antipseudomonal agents such as ceftazidime, tobramycin, or ciprofloxacin should be included. For children with sickle cell disease, second- and third-generation cephalosporins can be used where MRSA is not suspected, as they cover both Salmonella and S. aureus spp. If Salmonella is identified as the causative agent, ciprofloxacin can also be used, although susceptibility must be confirmed.

Long-term courses of intravenous antibiotics are indicated for immunocompromised patients and usually requires a percutaneous intravenous central catheter (PICC). These patients often require broad spectrum Gram-positive and Gram-negative drug coverage at high doses. More than one antibiotic is commonly used. Sequential imaging of these cases may be warranted to monitor the resolution or progression of the disease. In the case of fungal OM, intravenous antifungals such as fluconazole, voriconazole, liposomal amphotericin B, and caspofungin are used.

In case of tuberculous OM, four-component therapy (rifampicin, isoniazid, pyrazinamide and ethambutol) is first prescribed for 2 months, followed by therapy with two drugs (rifampicin and isoniazid) for 4-10 months. Diagnosis usually includes biopsy and drainage of any lesion.

Treatment of all children with osteoarticular infection is interdisciplinary and should include the involvement of pediatric, orthopedic, radiological and microbiological specialists. Orthopedic devices and physical therapy may also be required if the spine is affected or if the function is severely impaired.

Sclerosing osteomyelitis is also called a new bone. The most frequently affected are diaphysis and metaphysis of the femur and tibia. The clinical signs are bone thickening and hardening of muscular tissues. The roentgenological peculiarities of this pathology are sudden constriction

of the marrow canal or its obliteration, bone thickening after dense homogenous sclerosis, sometimes there are foci of destruction with small sequesters.

Albuminous osteomyelitis — is a rare disease that is characterized by feeling of discomfort in the extremity, then- continuous pain, in 1-2 months there appear swelling, sometimes hyperemia. Local temperature reaction is usually slight. The most frequent localization is distal metaphysic of the femur or proximal or the tibia. Roentgenogram shows cavity of irregular shape or circular cavity with periostal layers. The diagnosis may be easily confirmed by puncture of the focus if albuminous fluid is obtained. Histological examination of the focus determine plasmic-cellular granulation tissue (Russell's body).

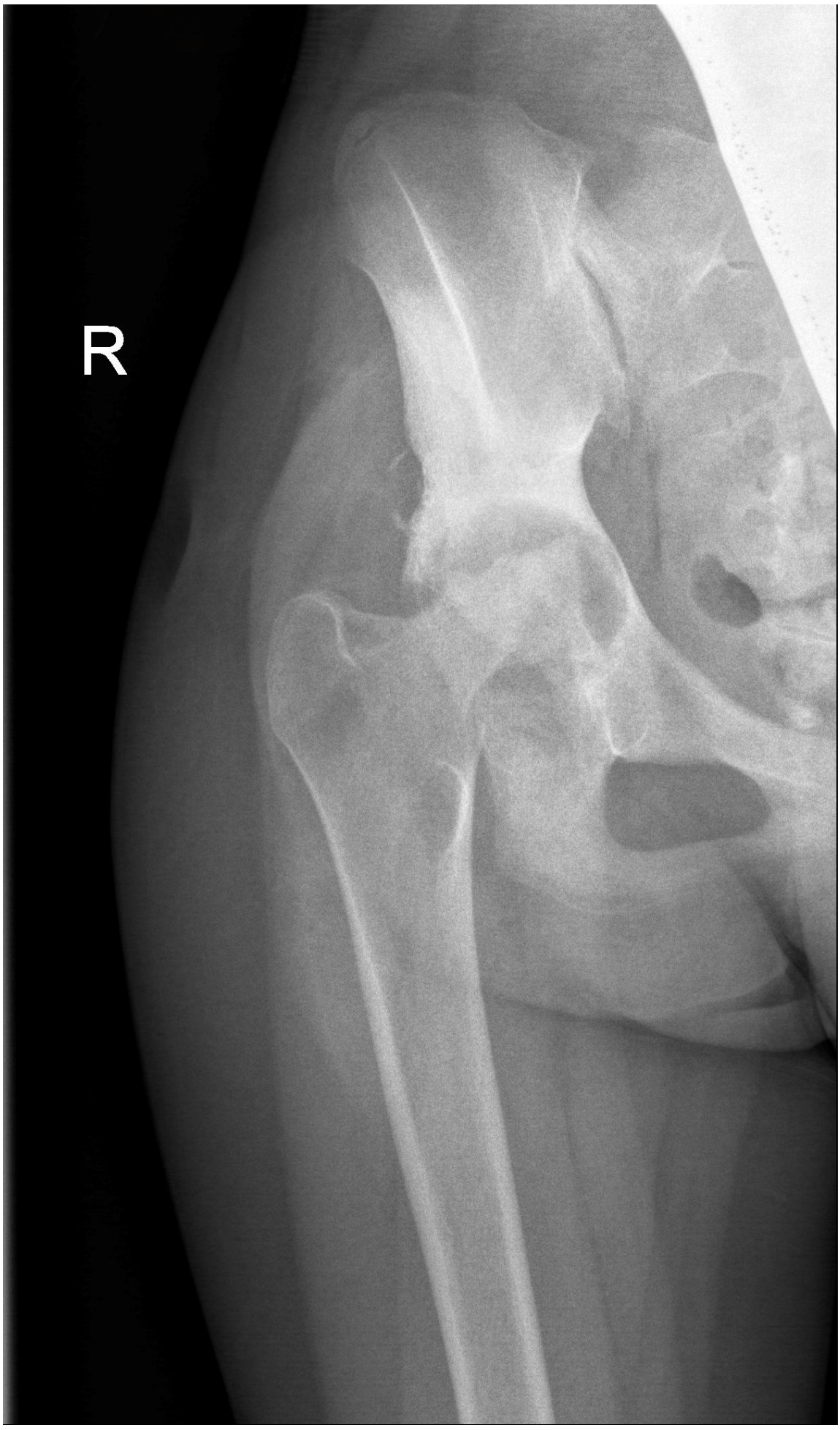
Abscess *of Brodi* is localized in metaphysis of long tubular bones (femur, tibia). It is characterized by limb dysfunctioning, pain in the focal area, nocturnal pain, swelling, frequently with arthritis. Roentgenological picture shows oval or circular cavity with sclerotizing capsule, it may include sequesters and is situated on the border of metaphysis and epiphysis.

Surgery

Surgical treatment of OM is rarely required if osteomyelitis is quickly diagnosed and treated. If congestion in the periosteum, soft tissue, or cyst is visible on imaging, surgery may be indicated for diagnostic (microbial identification) and therapeutic purposes. Bone biopsy, decompression drill, or intralesional curettage may be performed if antibiotic therapy does not respond within the first 72 hours, although this may also indicate the presence of resistant organisms or a fungal infection.

In the case of postoperative complications such as an infected implant, all necrotic tissue must be removed. This may require a combined procedure involving plastic surgeons as soft tissue coverage with split skin graft (SSG) or musculocutaneous flaps may be required. If there is an infected implant, all metal parts should be removed if bone stability permits. If the osteotomy or fracture has not yet recovered, if an infection is present, longer antibiotic suppressive therapy may be required while the hardware remains in place.

If there is subacute or chronic OM with lytic or cystic bone changes, cavities, or sequesters, they can also be removed and drained. This can be done with an ultrasound or CT scan and can be very helpful in excluding malignant neoplasms.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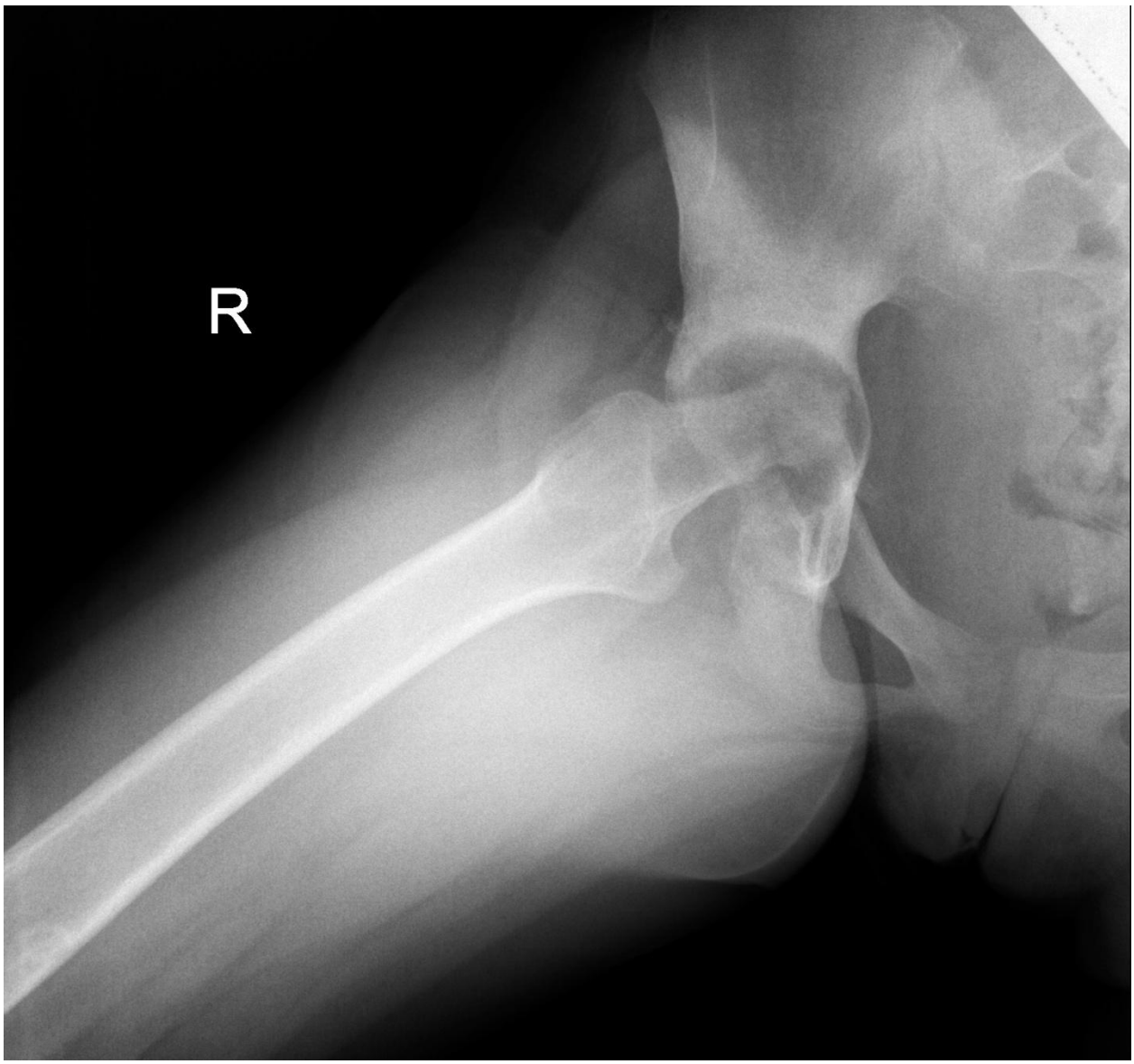

Fig 7.7-7.8 Complete lysis of the femoral head, as a complication of previous osteomyelitis.

Complications

Complications of bone and joint infections can be serious and are usually preventable with well-timed treatment. When infection penetrates the growth zone, infection and / or debridement of the wound can damage the metaepiphysis and stunt growth. This can lead to progressive angular deformity or limb length mismatch throughout the child's remaining height. Surgical epiphysiodesis, corrective osteotomy, or external fixation devices may be required to correct subsequent deformity.

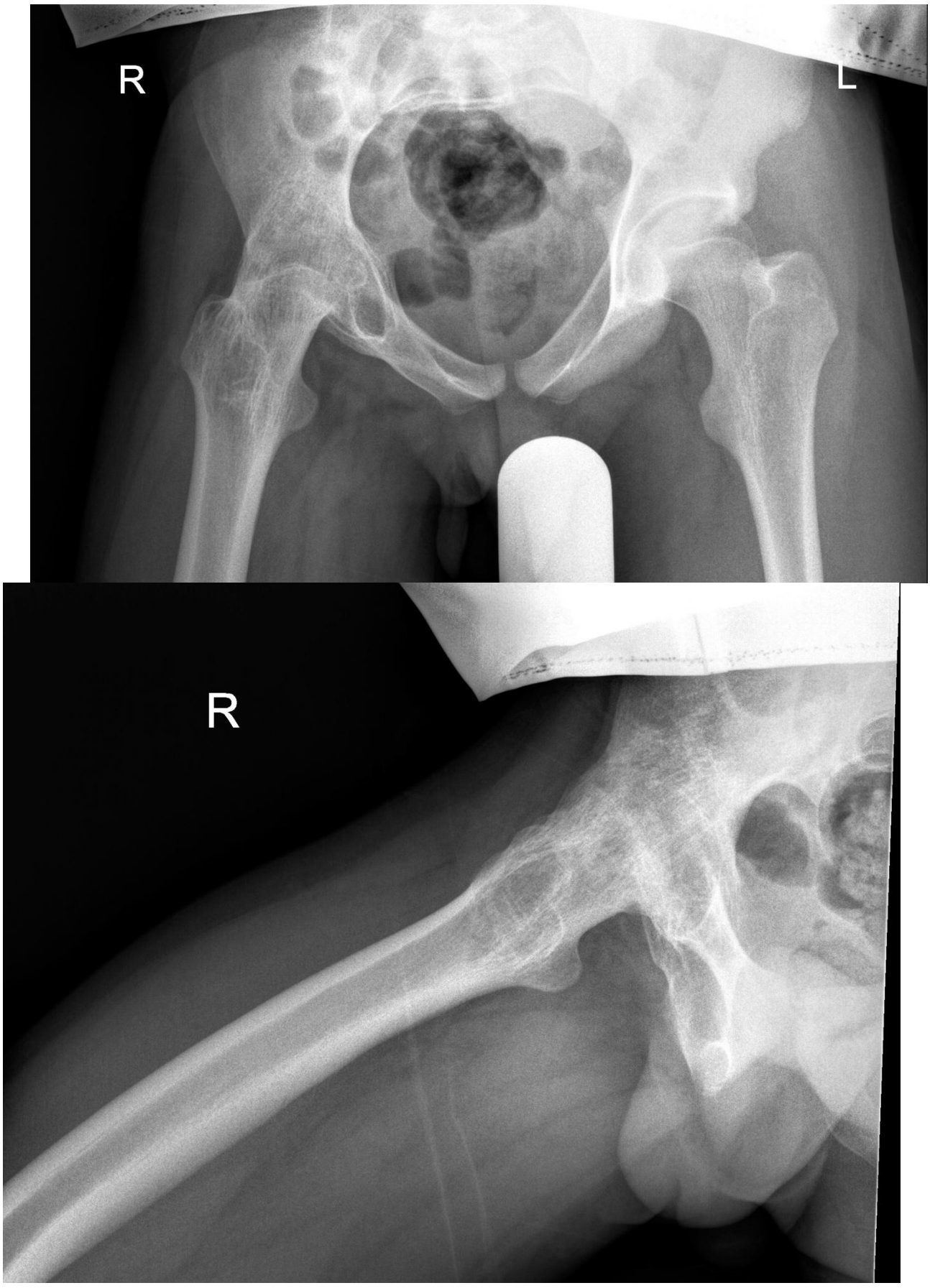

Fig. 6.8-6.9 Ankylosis of the right hip joint after osteomyelitis

Deep vein thrombosis and thromboembolic complications are more common where the causative agent is MRSA or PVL-SA.

Key practice points, indicating the level of evidence for each

1. BJI is more likely to affect children under 5 years of age, and the infection is more likely to affect the joints of the lower extremities. [IIA]
2. Staphylococcus aureus is the most common microorganism causing BJI in children of all ages. In addition, Kingella kingae is a common causative agent in children under 5 years of age. [IIA]
3. C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) have a high sensitivity for the diagnosis of BJI, which increases slightly when the two tests are combined, while the specificity is low. [IIB]
4. Ultrasound has a high sensitivity for diagnosing septic arthritis, whereas magnetic resonance imaging (MRI) is the most reliable imaging test for diagnosing general BJI. [IIA]
5. Isolation of a microorganism from bone, joint or blood in combination with clinical manifestations or radiological data, according to the BJI clinic, is the gold standard for diagnosis in children. [IIA]
6. Empiric antibiotic therapy should be initiated as soon as possible after collection of appropriate microbiological specimens for suspected BJI in children. [IIA]
7. Empiric therapy should include appropriately coated antibiotics against methicillin-susceptible Staphylococcus aureus (MSSA) and methicillin-resistant Staphylococcus aureus (MRSA) in geographic areas with a prevalence of this bacterium of more than 10-15%. [IIA]
8. Empiric therapy for young children should include appropriate K. kingae coverage in the appropriate areas. [IIA]
9. First generation cephalosporins, antistaphylococcal penicillins and clindamycin are the antibiotics most studied for BJI in children. [IIA]
10. If MRSA infection is suspected and the patient is not critically ill, empiric therapy should be considered including clindamycin if S. aureus clindamycin resistance is less than 10-15%. A glycopeptide or other suitable antibiotic for MRSA, such as linezolid, should be included if the incidence of topically clindamycin-resistant MRSA is high. [IIIB]
11. Septic arthritis (SA) in children should be treated with joint drainage by arthrocentesis, arthrotomy, or arthroscopy, depending on the preference and experience of the pediatrician and surgeon. Arthrocentesis may be appropriate as the only invasive procedure for uncomplicated SA in children. [IIB]
12. Brief intravenous therapy followed by oral therapy is appropriate for most children with uncomplicated BJI, based on the absence of complications and a favorable outcome. [IA]
13. Subsequent oral antibiotic therapy should be guided by the patient's antibiotic susceptibility to bacteria, if isolated; if susceptible, first-generation antibiotics, cephalosporins and clindamycin, are chosen. [IIA]
14. The minimum total duration of antibiotic therapy for septic arthritis should be 2-3 weeks and 3-4 weeks for osteomyelitis. [IA]
15. Complicated BJI or high risk of BJI, eg caused by Salmonella, MRSA or Panton-Valentin-leukocidin (PVL) -positive strains developing in young children or with slow onset of clinical improvement, may require longer administration as intravenous (IV), and oral therapy. [IIB]
16. Risk factors associated with sequelae include infants and newborns, infections with MRSA or PVL-positive strains, longer duration of symptoms prior to initiation of therapy, and hip involvement. Thus, children with BJI who have any of these risk factors should be treated more carefully and for a longer time to avoid complications. [IIB]
17. The multidisciplinary team should monitor children with BJI until osteoarticular function is restored and the consequences have been corrected. If bone growth is the only problem, an orthopedic specialist is enough. Babies with BJI in the hip or growth zone in another area should be followed up for an extended period of time. [IIB]

Tests for final level of knowledge, keys for tests:

1. 9 years old child feels pain in the upper third of the right ankle, the temperature increased up to 39°C, the child cannot stend on the leg. In anamnesis: patient had the injured shin and had a sore throat. Which disease do you most likely have to deal with?

A. Fracture of bone
B. Acute hematogenous osteomyelitis
C. Rheumatic fever
D. Tuberculous osteomyelitis
E. Malignant tumor

2. after earlier suffering a purulent otitis one-year old boy has been felling pain in the upper third of the left thigh, increased body temperature up to 39°C. Objectively: swelling of the hip in the top third and flatness of inguinal fold. The limb is in half-bended position. Active and passive movements are impossible because of the acute pain. What is the most likely diagnosis?

A. Acute coxitis
B. Intermuscular flegmona
C. Osteosarcoma
D. Acute hematogenous osteomyelitis
E. Brodie Abscess

3. 12 years old child has been sick for 2 days. Patient complains about hyperthermia, pain in the lower third of the right hip and knee joint. In anamnesis-got injured 3 days ago. When researching a moderate increase of local temperature, a slight infiltration of soft tissue. When percussion at this section the local pain intensifies. Preliminary diagnosis: acute hematogenous osteomyelitis of bottom third of right hip. What caused pain on condition of osteomyelitis in the early days of the disease?

A. Increased internally articular pressure
B. Injury
C. Contracture of knee joint
D. Exfoliation of periosteum
E. Increased intraosseous pressure

4. Two months old child is hospitalized in the surgery with a temperature of 38.5°C, edema, hyperemia and the lack of movement in the area of the left shoulder joint. In anamnesis: omphalitis, pseudo furunculosis. What is the most likely diagnosis?

A. Erba Paralysis
B. Plexitis
C. Flegmona of newborn
D. Shoulder injury
E. Meta-arthrosteitis

5. 13 years old boy feels pain in the upper third of the left thigh, increased body temperature to 39°C. Thigh swelling was detected in upper third and flatness of inguinal fold. The extremity was in half-bended position. Active and passive movements are impossible because of the acute pain. What is the most likely diagnosis?

A. Acute hematogenous osteomyelitis
B. Acute coxitis
C. Intermuscular flegmona
D. Osteosarcoma
E. Brodie Abscess

6. In the unit there is a 26 days old child with a diagnosis: navel sepsis. During bypassing the presence of swelling of the right shoulder was detected, the lack of active movements in a humeral joint, dangling of the right hand. Mother notes impairment of child for the last 24 hours, raising the body temperature to 38.8°C. What is the preliminary diagnosis?

A. Fracture of humerus
B. Meta-arthrosteitis of the humerus
C. Traumatic brachio plexitis

D. Shoulder flegmona
E. Fracture of collarbone

7. in the clinic of Pediatric Surgery a boy 12 years old was delivered with complaints about two fistuls in the bottom third of the left thigh, increased body temperature, general weakness. 6 months ago patient came through acute hematogenous osteomyelitis of left hip bone. On x-ray picture of the left thigh - total sequestration 12x3 cm. What is the preliminary diagnosis?

A. Primary chronic osteomyelitis
B. Tuberculosis
C. Ewing's Sarcoma
D. Osteoid-OsteomaSecondary chronic osteomyelitis

8. In 36 hours after surgical treatment (making incisions) of a 3-year-old child with phlegmona of the hand is observed the spreading hyperemia and infiltration onto the forearm by the type of "flame tip" is observed. What is the complication?

A. Abscess of the forearm
B. Lymphangitis
C. Adenophlegmona of the forearm
D. Cellulitis
E. Streptococcal impetigo

9. A 3-week child is restless, the body temperature has increased up to 38.3^0C, there is enlargement and hyperemia of the right mammary gland, pain at palpation. The diagnosis is mastitis. How should the local treatment be performed not to have complications in the mammary gland?

A. Radial incisions up to the aureole
B. Retromammary introduction of the antibiotics
C. Incisions by the "chess order"
D. Arch-like incision
E. Puncture

10. in the clinic of Pediatric Surgery a 10 years old boy was delivered, with complaints about fistula in the bottom third of right hip, increased body temperature, general weakness. 8 months ago patient came through an acute hematogenous osteomyelitis of right femur. On x-ray picture of the right hip there is a sequestration of the femur size 1.5 x 3 cm. What is the tactic of patient treatment?

A. Conservative treatment
B. Operative intervention in the period of remission
C. Courses of conservative treatment after 3-6 months
D. Immediate surgical intervention
E. Dynamic surveillance after achieving remission

Tasks for final level of knowledge

1. Two months old child hospitalized to the surgical unit. Mother complains about increased body temperature up to 38.5°C, edema, hyperemia and the lack of movement in the area of the left shoulder joint. In history: omphalitis, pseudo furunculosis.

1. What is the most likely diagnosis?
2. The doctor's Tactics when detecting the disease.
3. The Etiology and pathogenesis of disease.
4. Name the main ways of treatment.
5. What are the features of the dispensary supervision of the child after recovery?

2. 12 years old child sick 2 days. Has complains about hyperemia, the pain in the lower third of the right hip and knee joint. In history: 3 days ago there was a trauma. Objectively: moderate increase of local skin temperature, a slight infiltration of soft tissue. During percussion of this section the local pain intensifies. It has been stalled a preliminary diagnosis: acute hematogenous osteomyelitis of bottom third of right hip. The pain is caused by osteomyelitis in the early days of the disease.

1. Your diagnosis.
2. What steps should be performed to prevent disease?
3. What actions should make the surgeon first.
4. What caused painful sensations during the percussion of the thigh-bone?
5. What conservative measures should be used during treatment of the disease?

3. 2 years old child entered after 2 days of the beginning of the disease. Condition worsened, hyperthermia, restlessness appeared. There was an edema in the middle third of the shoulder, skin hyperemia with clear contours. Palpation detected an acute pain, and softening of the skin in the center of formation, fluctuation.

1. Your diagnosis.
2. The doctor's Tactics when detecting the disease.
3. The Etiology and pathogenesis of disease.
4. Name the main ways of treatment.
5. What preventive measures should be held after recovery?

4. 13 years old boy. Patient feels pain in the upper third of the left thigh, increased body temperature to 39°C. Thigh edema was detected in upper third, flatness of inguinal fold, hyperemia of skin coverings, the increase of their density. Extremity was in half-bended position. Active and passive movements were impossible because of the acute pain.

1. What is the most likely diagnosis?
2. What steps should be performed to prevent disease?
3. What actions should make the surgeon first.
4. What caused painful sensations during the percussion of the femur?
5. What conservative measures should be used during treatment of the disease?

5. 9 years old child feels pain of the right hand, increased temperature to 38 ° C, the child cannot bend the fingers of the hand. In anamnesis: had injured hands: three days ago cut his palm. Objectively: swelling of the hand, hyperemia with no clear borders, distributed on forearms, palpation sharply painful.

1. Which disease do you most likely have to deal?
2. The doctor's tactics when detecting the disease.
3. The etiology and pathogenesis of disease.
4. Name the main ways of treatment.
5. What are positions of the incisions at this disease.

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THEME 8

CONGENITAL MALFORMATIONS OF RESPIRATORY SYSTEM IN CHILDREN. CONGENITAL MALFORMATIONS OF ESOPHAGUS. CONGENITAL DIAPHRAGMATIC HERNIA.

Overview:

Congenital anomalies account for one third of infant deaths and are one of the leading causes of death in this age group in most developed countries. Congenital malformations of the respiratory system now rank second, behind those of the cardiovascular system, as a cause of infant mortality. With a rate of 0.25 death per 1000 live births, they have surpassed those of the nervous system (0.23 per 1000) in the past decade in the United States. However, congenital malformations of the lungs and airways include a wide spectrum of developmental anomalies, some of which remain asymptomatic and are discovered incidentally on imaging studies. Their frequency has been reported to range from 7.5% to 18.7% of all congenital malformations, 4,5 but their exact overall incidence is difficult to ascertain. They may be part of more complex syndromes and are often associated with other congenital anomalies, particularly those involving the heart and great vessels.

In the structure of morbidity of the population of Ukraine respiratory diseases traditionally occupy 1st place in children, 3rd place in infant mortality that comprise about 12%, and in the structure of disability they take 4th place. Anomalies of the tracheobronchial tree and lungs found in about 10% of children with the syndrome of airway obstruction.

1. **Aims of the lesson:**

The aim of this part of module is to provide help in identifying those children with congenital malformations of airway and lungs and to provide guidance on the diagnosis, differential diagnosis, defining indications for surgery and choice of optimal surgical treatment.

1.To repeat relevant anatomy of the airway, lungs and mediastinum in children.

2.To recall basic embryology of respiratory system

3.To learn nomenclature of malformations presenting with respiratory insufficiency

4.To learn main clinical signs characterizing the malformations of airway, lungs, esophagus, diaphragm that present with respiratory distress.

5.To collect complaints and history in patients suspicious for malformations respiratory system and their parents.

6.To determine the clinical signs of respiratory failure

7.To know diagnostic modalities used in infants and children with malformations of respiratory system

8.To know main radiological signs of individual congenital malformations of airway, lungs, esophagus, diaphragm.

9.To know main endoscopic signs of individual congenital malformations of airway.

10.To be able to carry out differential diagnosis of individual congenital lesions of airway, lungs, esophageal atresia and congenital diaphragmatic hernia.

11.To know types of congenital airway stenosis, their clinical presentation, diagnosis and treatment.

12.To know variants of vascular rings causing compression of the airway, their clinical presentation, diagnosis and treatment.

13.To learn clinical presentation, diagnosis and management of bronchogenic cysts.

14.To learn clinical presentation, diagnosis and management of pulmonary aplasia, agenesis and hypoplasia.

15.To learn clinical presentation, diagnosis and management of congenital emphysema.

16.To learn clinical presentation, diagnosis and management of cystic lung lesions.

17.To learn clinical presentation, diagnosis and management of pulmonary sequestrations.

18.To learn clinical presentation, diagnosis, prenatal diagnosis and management of congenital diaphragmatic hernia.

19.To learn clinical presentation, diagnosis and management of esophageal atresia.

20.To prescribe laboratory and instrumental examination of patients with symptoms of respiratory disorders.

21.To evaluate the results of X-ray observation in patients with malformations of respiratory system.

22.To describe the technique esophagogram, determine the indications for its use and evaluate the results.

23.To evaluate the results of computed tomography in patients with congenital malformations of respiratory system, determine the indications for its use.

24.To describe the technique of bronchoscopy and its features and value in patients with malformations of respiratory system.

25.To evaluate the results of prenatal ultrasound diagnosis of malformations of the respiratory system of the fetus.

3. Basic skills and knowledge, necessary to know: (interdisciplinary integration)

Subjects	Skills obtained
1. Anatomy, topographic anatomy	Define the boundaries of lungs, heart and mediastinum in different age periods.
2. Physiopathology	Clinical interpretation of laboratory analyses.
3. Morbid anatomy	To compare morphological changes, peculiar for certain variants of congenital malformations of airway, lungs, great vessels, esophagus, and diaphragm.
4.Microbiology	To master technique of collecting material for investigation. To interpret the results.
5. Pediatric diseases propaedeutic	To collect complaints, medical history, to examine the child, to use additional methods of examination in congenital malformations of thoracic organs.
6. Topographic anatomy and surgery	To master methodics (schemes) of operative access and methods of operative intervention on thoracic cavity organs in children of different age groups.
7. General surgery	To display methods of patient preparation for treatment, diagnostics and operation.
8. Hospital surgery	To carry out differential diagnosis, to ground methods of diagnosis and treatment of congenital malformations of airway, lungs, great vessels, esophagus, and diaphragm in children.
9. Clinical pharmacology	To classify medications necessary for treatment of surgical diseases, remember about pharmakinetics and methods of action of medications in children of different age groups.

4. Tasks for individual self-preparation for the lesson

4.1. The list of key terms and characteristics a student must know:

Term	Definition
Norma	complete absence of anatomic divergence from the classic description of organ's structure with full function of all of the structural units
Anomaly	anatomic changes of an organ, that under ordinary conditions do not cause violation of its function, but with the development of pathological process, it may cause influence on its function
Malformation	rough morphological changes in an organ or system with violation of their function
Respiratory insufficiency	The pathological condition in which normal level of blood gases (Pa O2 - 92 - 100 mm Hg .; Pa CO2 - 35 - 45 mmHg) is provided (or not provided) due to exertion of compensatory mechanisms that significantly affects the patient's condition
Stridor	noisy breathing, which occurs when the airway lumen is narrowed by 2/3 or more at the level of the larynx, trachea or main bronchi
Respiratory distress-syndrome	In a narrow sense - respiratory distress syndrome in newborns is associated with surfactant deficiency; In a wider sense it means respiratory disorders and respiratory failure associated with pulmonary injury or violation of airway patency

CONTENT

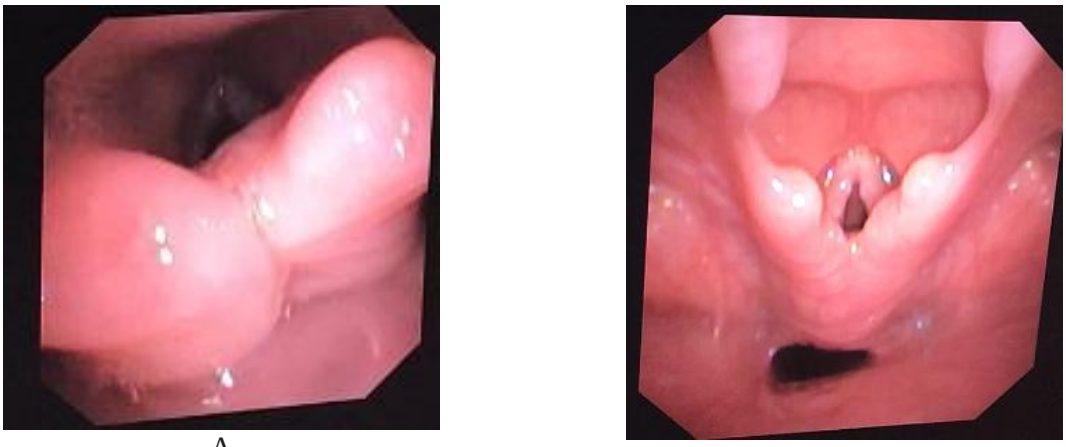
Congenital airway malformations

The group of congenital malformations of the airway, that presents with respiratory distress include large amount of following entities: choanal atresia, laryngomalacia, laryngeal web, atresia and stenosis, Subglottic hemangioma, tracheal stenosis, bronchogenic an enterogenic cysts, etc. Most of them cause airway narrowing and obstruction followed by respiratory failure even since the first days of life. Traditionally, defects of the upper respiratory tract (the level of the beginning of trachea) are within the competence of ENT doctors, but in terms of diagnosis, differential diagnosis and timely medical care they certainly are interesting for the doctor of any specialty.

Choanal atresia is a congenital obstruction of posterior nasal passages, may be membranous or osteal, unilateral or bilateral. Unilateral choanal atresia. In the case of bidirectional choanal atresia nasal breathing becomes impossible, the child breathes through the mouth. Apnoic attacs occur during sleep or feeding. Diagnosis is made by the attempts of passing the catheter through the nose into the nasopharynx. Contrast study and nasopharyngoscopy may be useful. The baby is fed through the nasogastric tube. The treatment is surgical. The early correction is recommended. In the cases of impossibility of adequate ventilation the surgery is indicated in the first days of life. The cutoff of septum is made through the palatal or nasal approach depending on age of the child.

Laryngomalacia is the most common cause of congenital stridor in infants, accounting for approximately 60% (range 50–75%) of all congenital laryngeal anomalies. Boys are affected twice as often as girls. Laryngomalacia is an enigmatic disease of unknown etiology. It is believed to be due to a delay in maturation of the supporting laryngeal cartilages, causing an inward collapse of the supraglottic structures on inspiration. A high-pitched fluttering inspiratory stridor is the hallmark of laryngomalacia. Typically, the stridor worsens during increased airway demands, such as crying, feeding, or the child's being in the supine position. Usually, the course of the disease is self-limiting, with onset around the age of 2–4 weeks, progression to a culminating point at around 6–8 months, and resolution occurring by 2 years of age. Feeding difficulties are related to gastro-esophageal reflux in up to 80% of cases. Regurgitation, recurrent vomiting, occasional coughing, or choking are seen in moderate to severe cases. Aspiration is often due to uncoordinated breathing

and swallowing during deglutition. In 5 - 10% of cases laryngomalacia can cause apnea, feeding difficulties and cor pulmonale. These patients require surgical correction. Mild cases of laryngomalacia, seen in 80–90% of infants, require only diagnostic confirmation conducted in the outpatient clinic by awake transnasal fiberoptic laryngoscopy (Fig. 8.1).



A B

Fig. 8.1. Endophoto of laryngomalacia causing supraglottic obstruction: A – hypertrophy of arytenoid cartilages and their mucosa, B – shortening of aryepiglottic folds

In most cases such patients do not require special treatment. A symptomatic therapy is administrated. Prednisone (2 mg/kg) is assigned for 10 - 14 days. Patients with severe and progressive respiratory insufficiency may require endotracheal intubation or tracheostomy. Over the past 20 years endoscopic intervention, namely epiglottopexy or CO_2 laser supraglottoplasty have become spread widely. The goal of these procedures is to widen the supraglottic lumen and thus to overcome the inspiratory obstruction. The operation includes the vaporization of excessive mucosa of arytenoid folds and trimming the epiglottis. Efficiency of supraglottoplasty varies between 80 - 100% according to various studies.

Congenital laryngeal atresia, webs, and stenosis. *Laryngeal atresia* is a life-threatening malformation. The lesion results from failure of recanalization of the epithelial septum, which forms around 6 weeks of gestation and separates the developing esophagus from the tracheal bud. In the past, affected babies would often die at birth or have severe neurologic sequelae because of the failure to establish an airway. In recent years, routine prenatal ultrasound examinations have allowed the diagnosis to be made before birth, resulting in a higher survival. On ultrasound, the lungs appear more echogenic and enlarged, causing eversion of the diaphragms; a distended trachea can also be observed. Fetal MRI confirms the increased fluid content of the lung, with a hyperintense signal on T2-weighed images. Near term, the fetus have to be delivered using the EXIT procedure (EX utero Intrapartum Treatment). The cesarean section is done, the fetal head and neck are exposed and the airway is secured usually by means of tracheostomy, while oxygenation is maintained through the placenta and working umbilical cord.

The prognosis is better with *laryngeal webs*, which can present with openings of varying sizes. Of these, 75% are glottic, with the rest supraglottic or subglottic in location. Complete webs present like atresias, whereas partial webs present with stridor and a hoarse or weak cry and may cause varying degrees of respiratory difficulty depending on the degree of obstruction. Partial webs may also present as a difficult to intubate neonate who is placed under general anesthesia for repair of another malformation such as esophageal atresia. The diagnosis of laryngeal webs is confirmed at endoscopy and treatment is excision or laser ablation, although some smaller subglottic webs may respond to dilation.

The **congenital subglottic stenosis** is the results of defective recanalization of the larynx. The defect involves cricoid cartilage and usually occurs at the level of approximately 2 - 3 mm below the glottis. The most common presenting symptom is stridor, which is worsened by

increased respiratory effort and upper respiratory tract infection. In fact, in milder forms the stridor may only be noticed during intercurrent upper respiratory tract infections and be misdiagnosed as croup. Therefore, recurrent "croup" in infancy should always raise the possibility of a fixed upper airway narrowing such as subglottic stenosis, particularly when the course of the illness and response to treatment are atypical. The gold standard of diagnosis of congenital subglottic stenosis is flexible and rigid videolaryngotracheobronchoscopy (Fig. 8.2).

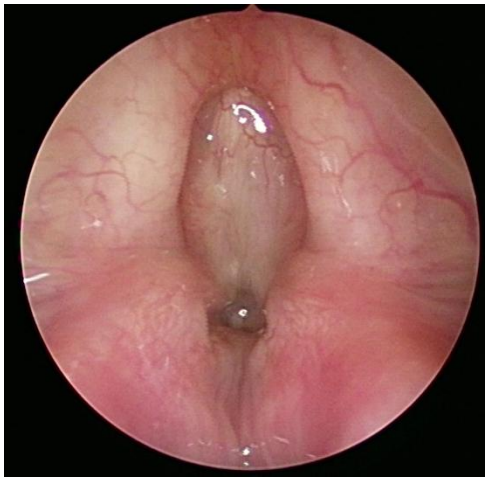
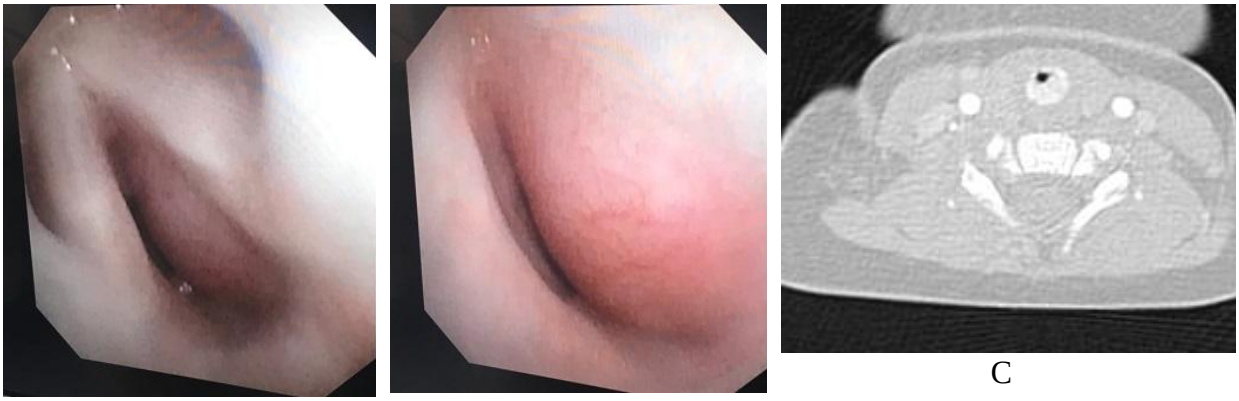


Fig. 8.2. Endoscopic picture of congenital grade IV subglottic stenosis. The stenotic zone is localized slightly below the vocal cords, the lumen of the larynx below glottis is almost absent

Bronchoscopy is the definitive diagnostic tool and is usually necessary to exclude other causes of narrowing in this region, particularly subglottic hemangioma. Acquired subglottic stenosis is much more common than the congenital form and has the history of laryngeal trauma, the most common of which would be endotracheal intubation. This is particularly true when intubation is prolonged, as in the premature neonate. In mild cases of congenital subglottic stenosis a conservative approach using supportive care, particularly during intermittent episodes of "croup," should be used because the subglottic narrowing may improve with laryngeal growth. Surgery should be reserved for patients who fail to cope with this conservative management, and have moderate to severe airway narrowing. Tracheostomy or cricoid split procedure are often the first line surgical treatment. Most tracheostomized patients can be successfully decannulated within 2 to 3 years, although stridor and varying degrees of breathing difficulties may persist for many years. Costal cartilage graft laryngotracheal reconstruction or partial cricotracheal resection are the surgeries used for severe forms of congenital subglottic stenosis.

Subglottic hemangioma is a rare benign tumor of the airway, accounting for only 1.5% of all congenital laryngeal anomalies. Subglottic hemangioma is more common in female than in male patients with a ratio of 2–3:1 and are potentially life-threatening in the absence of treatment. They are benign tumors consisted of hyperplastic endothelial cells, mast cells, pericytes, fibroblasts and macrophages. The severity of the disease depends on the size of the tumor. Respiratory viral infections enhance respiratory disorders. Understanding of evolution of subglottic hemangioma is essential in making a decision about the best treatment option. They undergo a rapid proliferative phase lasting a few months, followed by a period of stabilisation, and finally, a slow involution phase of several years. According to A.L. Bruckner (2006), complete resolution occurs in 50% of the cases by the age of 5 years, 70% by the age of 7 years and 100% by the age of 10–12 years. During the first weeks of life, the infant is asymptomatic. Usually, symptoms of inspiratory stridor followed by biphasic stridor with barking cough and slight hoarseness start at around 2–4 months, manifesting in all infants by the age of 6 months. Symptoms of respiratory distress with suprasternal and chest retractions, feeding difficulties and failure to thrive depend on the severity of the airway obstruction. The progression of symptoms reaches a plateau between the ages of 10 and 12 months, and the symptoms then decrease slowly and finally

disappear around the age of 2 years, although complete resolution of the tumor may take as long as 5–10 years. This presentation must be differentiated from a subglottic cyst, stenosis or papilloma. The mainstay of the diagnosis is laryngotracheoscopy, both fiber optic and rigid under general anesthesia (Fig. 8.3A, B). The subglottic hemangioma appears as a reddish smooth mass, mostly located in the subglottis and extending cranially to the under surface of the vocal cord. The tumor mass is spongy and compressible, allowing for easy intubation with an ET tube. An extensive or therapy-resistant large hemangioma requires a contrast-enhanced MRI or CT scan to identify extension into the neck or mediastinum (Fig. 8.3C).



A B C

Fig. 8.3. Videolaryngoscopy (A, B) and CT with contrast enhancement (C) showing subglottic hemangioma in an infant

Knowing that subglottic hemangioma is a self-limiting disease with spontaneous resolution, treatment must be aimed at maintaining the airway without tracheotomy, while avoiding any long-term sequelae. Treatment modalities include observation, medical treatment (systemic steroids, propranolol), endoscopic treatment (intralesional steroid injections, laser resection) and open surgery (tracheostomy, excision through a laryngofissure or tracheotomy, tracheal resection with anastomosis). Observation is appropriate for children with mild symptoms as well as children older than 1 year having reached the phase of spontaneous tumor regression. Systemic steroids (prednisolone 4-5 mg/kg per day) should be used as adjuvant therapy or as curative treatment only for a short period. Clinical efficiency of this treatment if administered alone is approximately only 25%. Boston protocols do not recommend systemic steroid treatment for longer than 3 weeks if the symptoms do not markedly improve. Potential long-term side effects such as failure to thrive, osteoporosis, adrenal suppression and Cushing syndrome should not be underestimated. Administration of propranolol is rather new and efficient method of treatment of hemangiomas, and subglottic or tracheal hemangioma in particular. Possible explanations for the therapeutic effects of propranolol include vasoconstriction, decreased expression of VEGF and βFGF genes via the downregulation of the RAF-mitogen-activated protein kinase pathways, as well as the triggering of apoptosis of capillary endothelial cells. Propranolol is used at 2 mg/kg of bodyweight per day during at least 6 months. Endoscopic intralesional steroid injections is reported to have success rates exceeding 75%, this treatment modality is much more efficacious than systemic steroids. Laser resection is appropriate for slow-growing tumors that become symptomatic at the age of 4–6 months. Open surgery was found to be the most efficient treatment, albeit with potential complications. It is mostly suitable for fast-growing tumors or bilateral tumors. In the future, propranolol might well supersede all other treatment modalities for children who do not have a contra-indication to the prescription of b-blocking agents.

Laryngeal cysts. These are usually supraglottic and generally present in the neonatal period (although possibly much later) with hoarse or muffled voice or even aphonia, stridor, and respiratory difficulty. A lateral neck radiograph may show a rounded supraglottic swelling, and at

laryngoscopy, a bluish fluid-filled cyst is found, usually in the epiglottic folds. Aspiration can relieve acute symptoms, but resection is ultimately necessary to prevent recurrence.

Congenital tracheal stenosis has different morphological reasons and distributed to intrinsic (organic) tracheal stenosis caused by complete cartilaginous rings, compression stenosis (due to the anomalies of great vessels - congenital vascular ring or mediastinal tumors and cysts) and functional stenosis (tracheomalacia). Complete cartilaginous tracheal rings tracheal stenosis is also characterized by absence of tracheal membrane. These two factors cause narrowing of tracheal lumen varying in lengths. A number of types have been recognized. Cantrell J.R. and Guild H.G. (1964) were the first who identified three types of congenital tracheal stenosis based on 24 observations of morphological variants: generalized hypoplasia (30%), funnel-shape stenosis (20%), segmental stenosis (50%). This classification was later modified by H.C. Grillo and the 4th type was described.

Type I: generalised tracheal hypoplasia. Almost the entire trachea is stenotic while only the first to third cranial rings are normal.

Type II: funnel-shaped tracheal narrowing. The abnormal tracheal segment varies by location and length, but always has a funnel configuration that is shaped from the cranial to caudal direction.

Type III: segmental tracheal stenosis. This type is characterised by a short-segment stenosis located at different levels of the trachea, at times below an anomalous right upper lobe bronchus.

Type IV: bridge bronchus stenosis. In this variant of Type III, the anomalous right upper lobe bronchus is in the proximity of the carina, and via horizontally branching bronchi, the stenotic bridge bronchus connects the proximal trachea to the rest of the lungs. (Fig. 8.4).

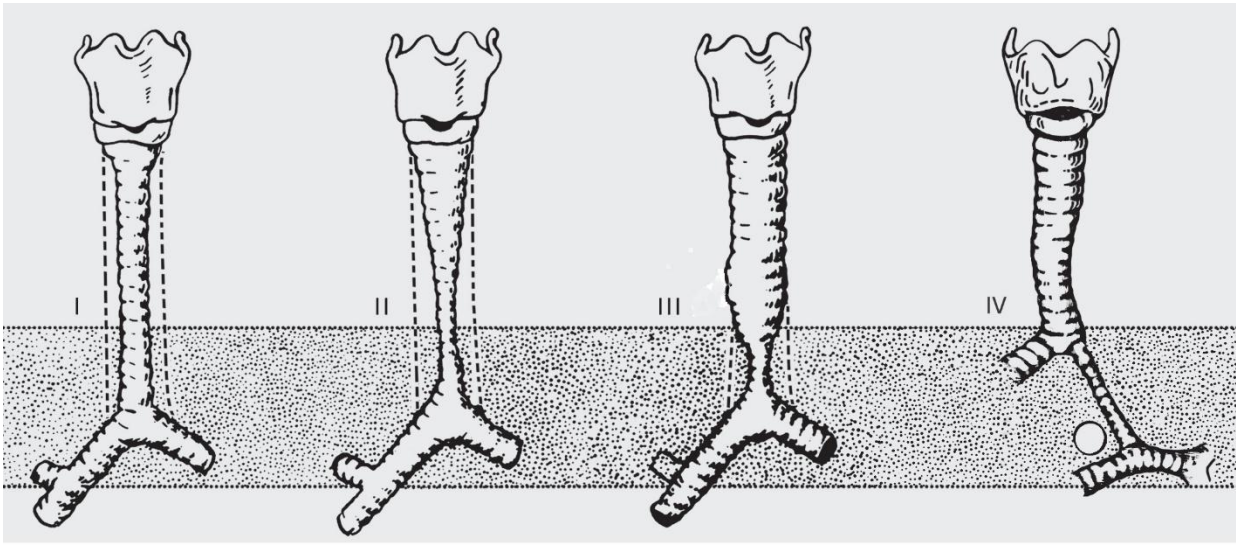
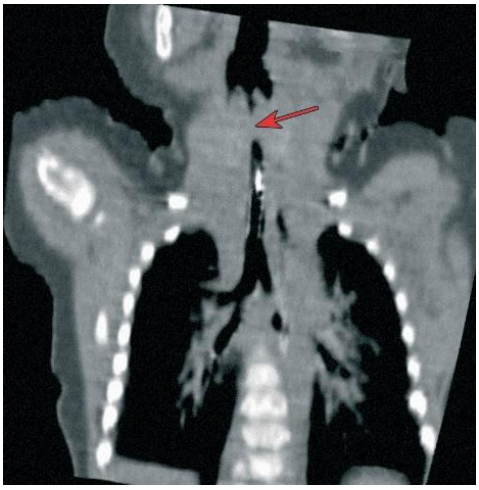


Fig. 8.4. Types of congenital tracheal stenosis (Cantrell J.R., 1964, Grillo H.C., 2004)

In general, the onset of symptoms starts a few months after birth when the baby's activities lead to an increase in respiratory demands. The length is less critical than the degree of the stenosis in producing symptoms. Main symptoms are biphasic stridor with predominant wheezing, chest retractions, cyanotic attacks and recurrent bronchitis or pneumonia. When the narrowing is severe, breath sounds may be accompanied by significant respiratory distress, whereas in mild cases they may be noticeable only when respiratory load is increased, as with exercise or infection. Depending on the degree of stenosis a defect may be compatible with long life. However, in such patients inflammatory complications in the lung, bronchiectasis, pulmonary hypertension and pneumofibrosis join over time accompanied with progressing of respiratory and cardiovascular failure. Radiographs of the airway may identify a narrowed segment, although CT with contrast enhancement is necessary to define the extent of the lesion and to search for an extrinsic cause

(Fig. 8.5A, B, C). CT scan with 3D reconstructions is the examination of first choice for achieving an accurate diagnosis. This diagnostic method allows assessing the anatomy of the mediastinum, relationship between the airway and mediastinal cardiovascular anomalies if present. Bronchoscopy along with CT is necessary tool of diagnosis and planning treatment (Fig. 8.5D). It should be performed very carefully because of probable decompensation of the compromised airway due to mucosal edema which may be a result of an even slightest mucosal injury is likely to, provoking an acute airway crisis. In the absence of infection, tracheal cartilages are readily visible during endoscopy and must be precisely counted. Normal rings are easily distinguished from circular rings. Respiratory function tests show evidence of fixed obstruction with characteristic flattening of the inspiratory and expiratory portions of the flow-volume loop. Management is difficult. In some patients the stenosis improves with tracheal growth, and conservative symptomatic treatment and support should be the recommended approach when possible. Dilation techniques with prolonged iendotracheal intubation may be used in newborns and infants for temporal stabilization of respiratory status while achieving optimal age for radical surgery. Endoscopic stenting has been used in some centers. This should be avoided when there is an associated vascular ring or sling because of the risk of erosion with arterial fistulization. There are few main open surgical approaches have been used: resection with end-to-end anastomosis, enlargement patch tracheoplasties (pericardial, cartilage), tracheal autograft technique, and slide tracheoplasty. Primary resection with end-to-end anastomosis is suitable for short segment stenoses not exceeding one-third of the tracheal length. Slide tracheoplasty is now the technique of choice for long-segment tracheal stenosis as it matches the basic requirements for adequate airway reconstruction, that is steady cartilaginous support and a fully mucosalised inner lumen. The basic principle of this surgical technique consists of increasing the tracheal circumference at the level of the stenotic segment. This is achieved by transecting the stenosis horizontally at its midpoint, slitting the upper and lower stenotic segments anteriorly and posteriorly, respectively, and then sliding the two segments over one another. Use of cardiopulmonary bypass provides better conditions for the operation as it secure stable gas exchange at the period of opened airway. Tissue engineering for trachea is the perspective field of investigation and may provide a useful alternative for tracheal reconstruction in the future.



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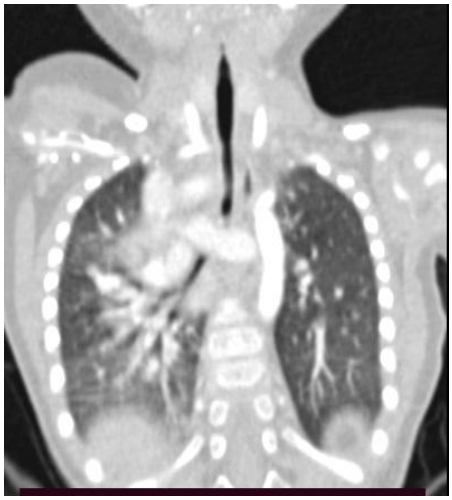

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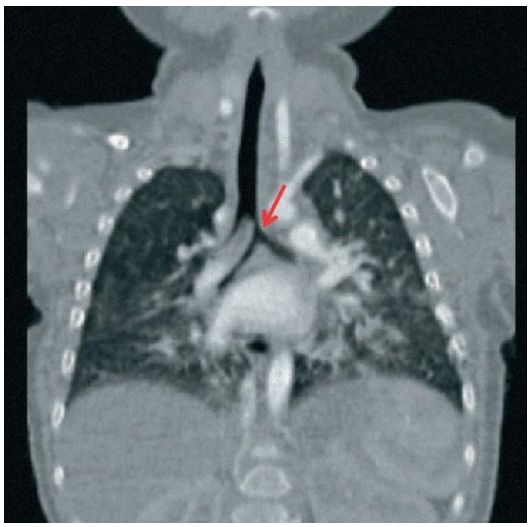

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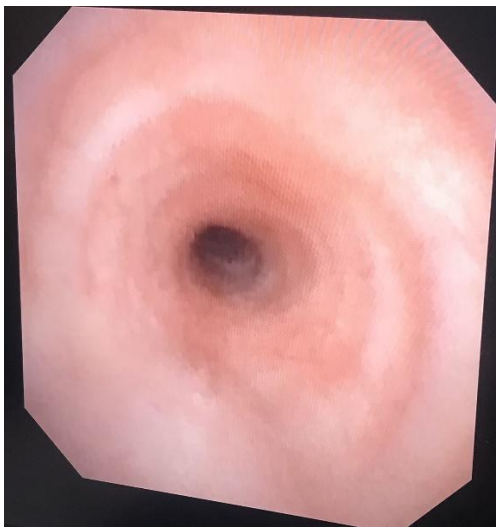

D

Fig. 8.5. A - CT frontal reconstructions in: (A) 3-month-old patient with congenital stenosis of the cervical trachea, absence of the lumen in the subglottis and proximal cervical segment of the trachea (arrow); (B) 8-months-old girl with long segment complete rings stenosis of thoracic segment of the trachea; C - male infant aged 1 year with bridging bronchus stenosis. Right tracheal bronchus, congenital stenosis of suprabifurcation segment of the trachea as "bridging bronchus" (arrow). D – Endophoto of complete rings segmental tracheal stenosis: tracheal rings are of "O" shape, tracheal membrane is absent in the area of defect

Compression tracheal stenosis may be caused by malformations of the aortic arch and its vessels, pulmonary artery (vascular rings and loops), and congenital cysts and tumors of the mediastinum (teratoma, hemangioma, bronchogenic cysts).

Vascular rings and slings

Abnormally developed aortic arch and its branches and pulmonary artery form complete or incomplete vascular ring around the trachea and esophagus and cause their compression and severe airway obstruction resulting in respiratory disorders that may be life-threatening. Vascular rings do not cause cardiovascular symptoms, so usually drop out of sight of cardiac surgeons. Their clinical significance relates entirely to the effects of compression of the trachea and esophagus. Prolonged inadequate treatment of children in somatic hospitals without timely surgical care threatens death from asphyxia or chronic inflammation in the airways and lungs of patients with disability.

Vascular rings are uncommon malformations. Anomalies of the aortic arch and its branches are 0.5 - 3.5% of all congenital cardiovascular system. Most vascular ring is found in isolated form. There is an association with patent ductus arteriosus or septal defects of the heart, Fallot's tetralogy, pulmonary atresia, and transposition of the great arteries.

Most vascular rings are associated with varying degrees of persistence of bilateral fourth aortic arches during early development. The model of hypothetic double aortic arch by Edwards (1948) consists of right and left aortic arches, two ductus arteriosus and two upper descending aortas. The normal aortic arch is the result of regression of the right upper descending aorta and the right ductus arteriosus.

Anatomical variants of vascular rings and slings are following (Fig. 8.6):

1 - double aortic arch;

2 – right aortic arch with left ligamentum arteriosum

3 - aberrant right subclavian artery;

4 - pulmonary artery sling;

5 – innominate artery compression

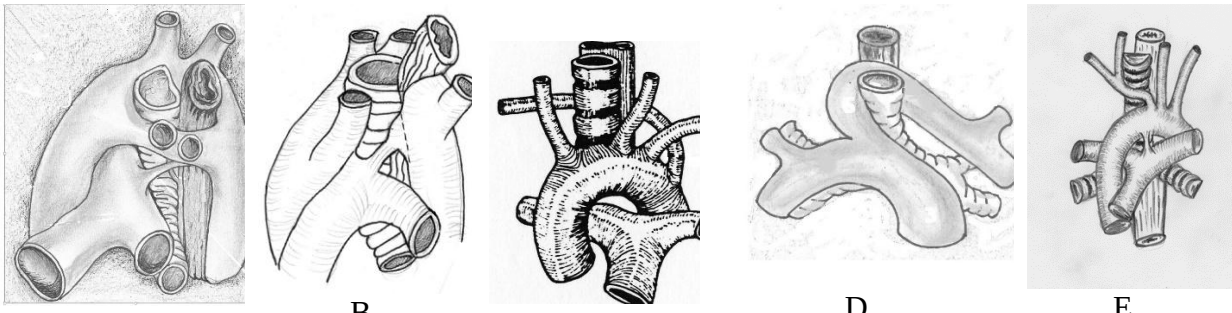

Fig. 8.6. Schematic drawing of the types of vascular rings: A - double aortic arch, B – right aortic arch with left ligamentum arteriosum, C – aberrant right subclavian artery, D – pulmonary artery sling, E – anomalous brachiocephalic trunk (innominate artery). Described in the text below

Various malformations of this group are united by common embryogenetic basis and tracheoesophageal compression syndrome.

Double aortic arch (DAA) (Fig. 10.6A) is the most common variant of the vascular ring and occurs in about 50% of cases. It is caused by the persistence of the embryological fourth aortic arch creating a complete ring around the trachea and esophagus. Each arch gives rise to a carotid and subclavian artery and the descending thoracic aorta may be located leftward or rightward side of the vertebral column or take midline position. In 45–77% of cases, the aorta descends on the left side. Based on the patency of aortic arches the balanced arches, right arch dominant and left arch dominant forms of anomaly are distinguished. In the majority of cases, the right arch is the larger. Rarely, there is an atretic arch (left arch in most cases). The separate branching of brachiocephalic vessels is common without brachiocephalic trunk formation.

The first report on DAA was made by Hommel in 1737, in 1932 Maude Abbott described 5 postmortem cases of DAA considering the possibilities of surgical treatment. In 1939, Wolman reported the syndrome of esophageal and tracheal compression caused by a double aortic arch and a few years later, in 1945, Gross had successfully repaired this type of lesion, followed by the correction of many other forms of vascular rings. In Ukraine prof. D.U. Krivchenya was the first who diagnosed and successfully surgically corrected DAA in 1 year-old girl in 1982.

Right aortic arch with left-sided ductus or ligament (Fig. 10.6B). This is the second commonest vascular ring (25–30%). The characteristic features of anomaly are following. (1) Ascending aorta takes its normal position, aortic arch is located to the right side of trachea and esophagus and passes over the right main bronchus; (2) descending aorta may take either right-sided or left sided position; (3) ductal ligament which connects left pulmonary artery and aortic isthmus closes complete vascular ring that compresses the trachea and esophagus. The aortal diverticulum (Kommerell's) may be present at the site of aortal end of ligamentum arteriosum which more often seen in the cases of right-sided descending aorta and enhances the posterior compression of esophagus and trachea. The aberrant left subclavian artery often arise from this diverticulum.

There are two main variants of the malformation: right aortic arch with aberrant left subclavian artery and right aortic arch with mirror image branching of brachiocephalic vessels. In the last type left common carotid and left subclavian arteries originate from the aortic arch together forming left brachiocephalic trunk.

Thus, the complete vascular ring in this anomaly is formed by the aortic arch on the right side and on back, including Kommerell's diverticulum, ligamentum arteriosum on the left side and pulmonary artery on front.

Double aortic arch and right aortic arch are most commonly found isolately, but association with patent ductus arteriosus is possible. The rare cases of combination with coarctation of aorta and pulmonary artery sling are reported. According to A. Hastreiter et al (1966), right aortic arch

with mirror image branching of brachiocephalic vessels is always combined with congenital heart defects.

Aberrant right subclavian artery (Fig. 10.6C) is the most common anomaly of the aortic arch, appearing in 0.5–0.7% of autopsies. The right subclavian artery arises from the descending aorta and crosses to the right side, mostly behind the esophagus. This anomaly results in the regression of a segment of the 'hypothetic double aortic arch' between the right common carotid artery and the right subclavian artery. In this anomaly vascular ring is incomplete (semi-ring or sling). There are four vessels arising from the aortic arch separately: right and left common carotid arteries, left subclavian artery and, at last, right subclavian artery which arises from the distal part of aortic arch. The incomplete ring around the trachea and esophagus is formed by aortic arch on front and aberrant right subclavian artery on back, which on its way to right arm causes posterior compression of esophagus and trachea.

Pulmonary artery sling (Fig. 10.6D) is characterized by abnormal development of bifurcation of the pulmonary artery trunk. The sling is created by anomalous origin of the left pulmonary artery, which arises as the first branch of the right pulmonary artery. The vessel runs between the trachea and esophagus toward the left hilum. The trachea is trapped in the sling created by the left pulmonary artery. A ductal ligament running from the main pulmonary artery completes the ring. This looping of the aberrant vessel around the carina usually has a strangling effect, resulting in variable degrees of compression and localized narrowing. Thus, the compression stenosis of the trachea just above bifurcation is formed.

There are two forms of anomaly: with normal anatomy of tracheobronchial anatomy and with airway malformation (abnormal bronchial branching at bifurcation in the area, tracheomalacia and complete cartilaginous rings). Combination of pulmonary artery sling with tracheal stenosis due to complete cartilaginous rings was named as "ring-sling complex". Its frequency is up to 50 – 60%. The complex morphological features of the malformation are complemented by pathology of lung vessels. The disorders of lung ventilation is enhanced by compromised blood circulation in the lungs.

Description for the first case of pulmonary artery sling was made by Glaevecke and Doehle in 1897, as an autopsy finding in 7-month baby suffering from severe respiratory distress. The malformation is rare and highly lethal. If untreated 90% of children die within first months of life. Postoperative mortality varies between 50% in early reports and 5% nowadays. Since the first report in the literature by 1995, about 150 cases of anomaly have been reported in the literature and about half of them were clinical observation, the other half autopsies. Associated severe cardiovascular defects occur in 30-50% of cases.

Abnormal brachiocephalic trunk (Fig. 10.6E) is the most common cause of vascular compression of the trachea. It is also known as innominate artery compression syndrome. Tracheal compression occurs due to a relatively distal branching of the innominate artery, which then crosses the trachea causing its anterior compression in association with a narrow thoracic inlet. First description was given by R.E. Gross and E.B.D. Neuhauser in 1948. Another anatomic variant of the anomaly is trifurcation of brachiocephalic trunk when it divides into three vessels: the right subclavian, right and left common carotid arteries. In this case brachiocephalic trunk has a larger diameter and also originates more distally from the aortic arch, causing anterior tracheal compression. Prolonged compression of tracheal wall results in the disruption of the cartilage structure of airway - secondary tracheomalacia.

The **clinical presentation** of compression tracheal stenosis depends on the degree of narrowing of the trachea and esophagus and presents with respiratory and esophageal groups of symptoms. The existence and severity of symptoms are related to the tightness and completeness of the vascular ring. Respiratory signs include inspiratory stridor with or without an association with an expiratory wheeze and tachypnea. The commonest presenting symptom of a vascular ring is stridor. Around 50–70% of symptomatic cases present during the first year of life. Various positions, such as lying on the back, may aggravate the stridor, while it may be relieved by extension of the neck. Other signs are noisy breathing, a hoarse cry and persistence of a barking

cough. In severe cases there are episodes of apnea with cyanosis and unconsciousness. Recurrent respiratory infections aggravate the condition and are more frequent in vascular rings. In mild tracheal compression, symptoms may be present only at the time of respiratory infections. As a result of esophageal compression, the baby feeds poorly and its growth may be retarded. There is difficulty in swallowing liquids and solids. Regurgitation and choking, with aggravation of respiratory symptoms, are common in severe cases, whereas in mild cases of obstruction, dysphagia may be the only symptom.

The natural course vascular rings. Symptoms can be severe from the first days of life and may become critical in the first few months. Following the violation of ventilation due to tracheal compression airway infection, delay sputum evacuation, pneumonia and lung hyperinflation occur. With the increase in the volume and density of food the aspiration syndrome develops due to compression of the esophagus. In this vicious circle there is no hope of survival of patient without surgery. Medical therapy is usually ineffective, provides temporary relief of symptoms. In most cases lethal outcome occurs at age 1 - 2 years from pulmonary complications.

Diagnosis. The examination begins with plain chest X-ray, although the method does not show direct signs of compression. A chest radiograph is mandatory in any symptomatic child. The plain X-ray shows prominence of the mediastinum to the right side in double aortic arch, right aortic arch with aberrant left subclavian artery, right aortic arch with mirror-image branching, and left ductus arteriosus. The aortic knob is on the left side in cases of left aortic arch with aberrant innominate artery, left aortic arch with aberrant right subclavian artery (in this case the aberrant vessel may be seen above the aortic knob). Indirect signs of obstruction is emphysema (single or bilateral), atelectasis and secondary inflammatory changes in the lungs.

The most useful first-line investigation is esophagography (Fig. 8.7), i.e. contrast radiography of the esophagus in the anteroposterior and lateral projections. This is informative, easy and safe carrying study, which does not compromise the respiratory tract exhausted in terms of compensation. The presence of characteristic filling defects of esophagus points to a compression of the esophagus and the trachea. Frontal esophagogram shows impression both on the right and left sides in cases of double aortic arch, right aortic arch with mirror-image branching and left ductus arteriosus. Oblique filling defect from the right, going upwards to the left, is seen in aberrant left subclavian artery with a right aortic arch. In pulmonary artery sling, the esophagus may be deviated to the left. Lateral esophagogram shows posterior compression in cases of double aortic arch, aberrant right subclavian artery, right aortic arch with mirror-image branching and left ductus arteriosus. In pulmonary aortic sling, the esophagus is compressed from the front. In anomalous origin of innominate artery there is no compression on the esophagus but the study is advisable to exclude other variants of vascular ring.

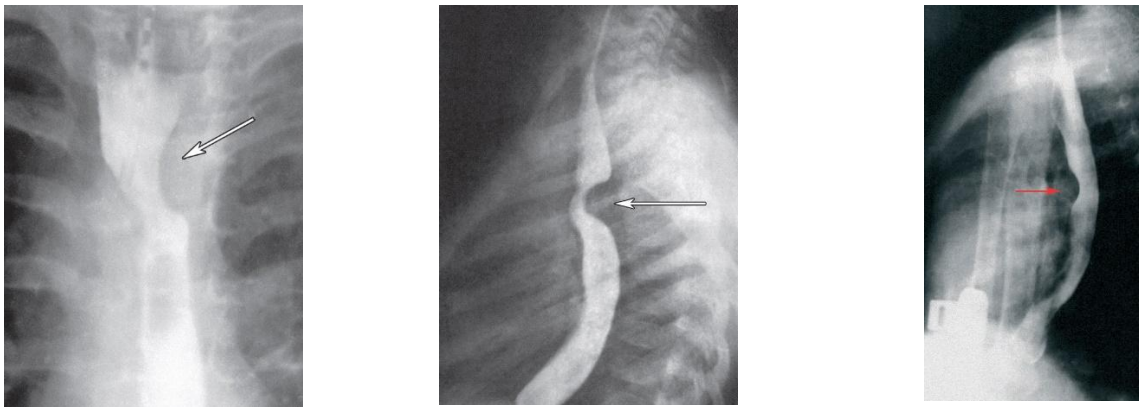

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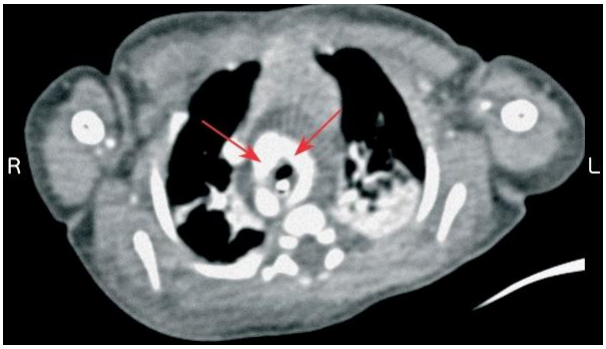
Fig. 8.7. Esophagographs in patients with vascular ring and sling. A, B - Double aortic arch. Anteroposterior (A) and lateral (B) view. Longitudinal esophageal axis is interrupted, suprastenotic esophageal dilation. Indention of posterior esophageal wall due to vascular compression (arrow).

C - pulmonary artery sling, lateral view: compression deformity of anterior esophageal wall (red arrow)

Computed tomography with contrast enhancement is now an essential tool for diagnosis of vascular rings. It is indicated to perform in every child with stridorous breathing especially if compression origin of airway stenosis is suspected. CT allows defining the vascular anatomy of mediastinum in detail as well as relationship of great vessels and trachea (Fig. 8.8). In double aortic arch, CT scans show both of arches forming vascular ring that encircles the trachea and esophagus and causing their compression at the same time. Each aortic arch has two branches of brachiocephalic arteries – subclavian and carotid, this is a characteristic feature of vascular ring. 3D reconstruction improves visualization and helps in preoperative planning. However, ligamentous structures (i.e., a ductal ligament or the ligamentous remnant of a double arch) cannot be seen on either echocardiography or CT scan but their position can be inferred from the nature of the ring.



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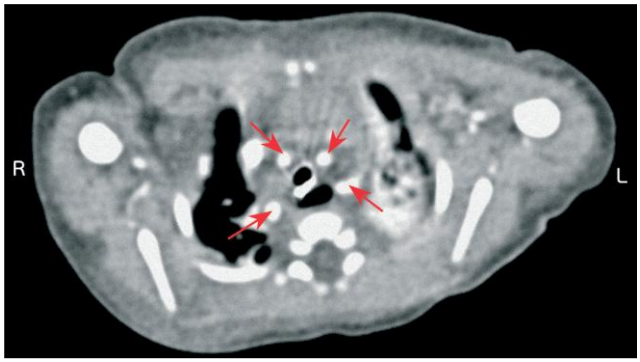

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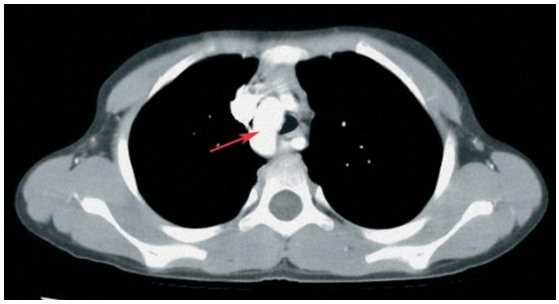

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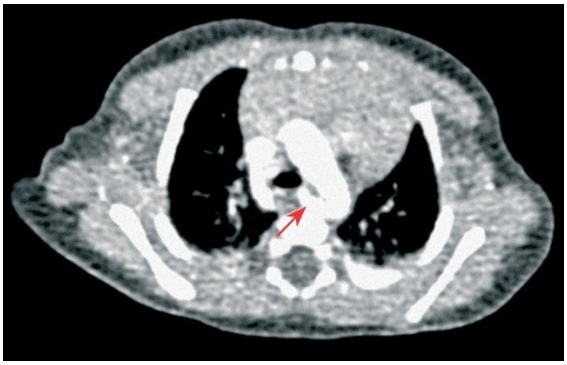

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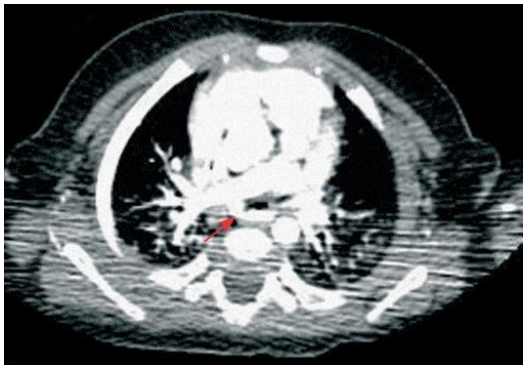

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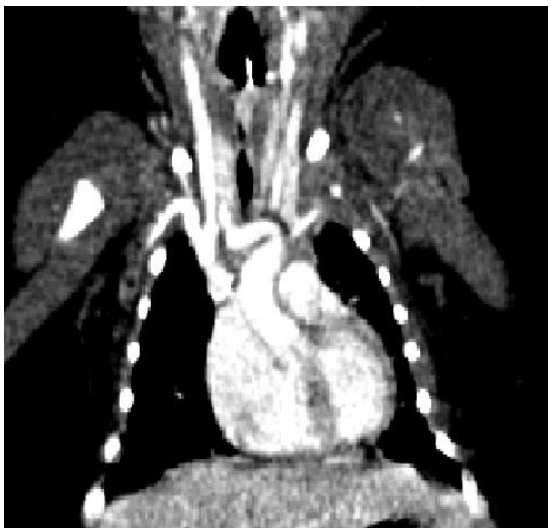

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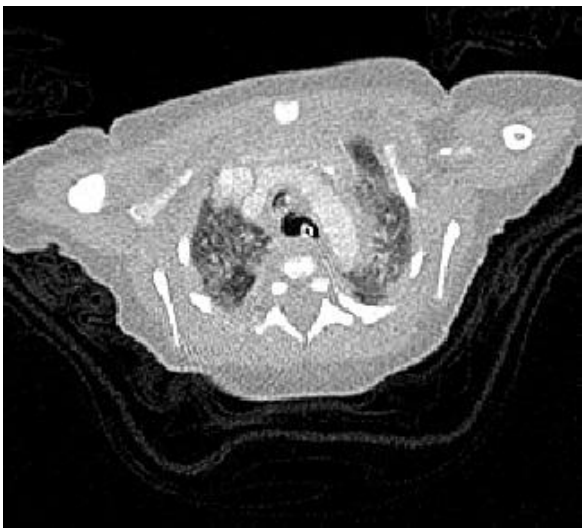
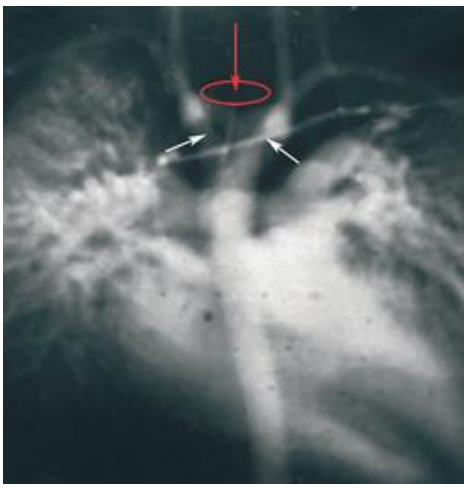


H

Fig. 8.8. Multispiral CT with contrast enhancement and 3D reconstruction. A, B – Double aortic arch (arrows) encircling trachea and esophagus; each aortic arch has two branches of brachiocephalic arteries (B). C – 3D-reconstruction of double aortic arch. Anterior view. D - Right aortic arch with left ligamentum arteriosum causing compression and deformation of the trachea (arrow). E – aberrant right subclavian artery which artery originates from the descending aorta (arrow). F - Pulmonary artery sling. Left pulmonary artery (arrow) originates from the right one, forms the sling around the trachea thus causes its compression. G, H - Brachiocephalic trunk causing anterior compression of the trachea: (G) CT, frontal reconstruction showing abnormal course of the vessel; (H) axial scan, compressed trachea has a slit-like shape

Digital subtraction angiography (DSA) is highly informative special investigation, which allows verifying anatomic and hemodynamic features of variants of vascular rings such as double aortic arch, right aortic arch with left ligamentum, aberrant right subclavian artery and abnormalities of brachiocephalic trunk (Fig. 8.9) along with associated intracardiac defects.. Selective pulmonary arteriograghy is optimal for diagnosis of pulmonary artery sling.



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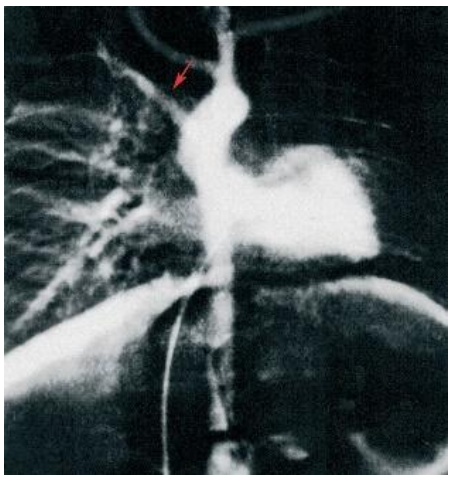

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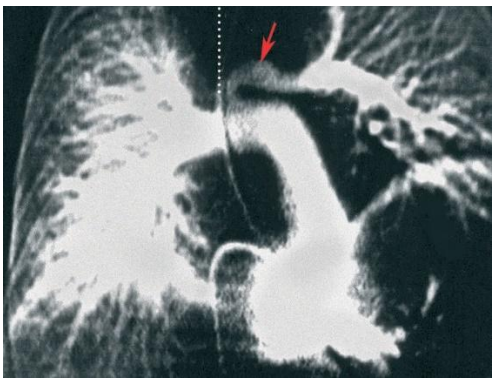

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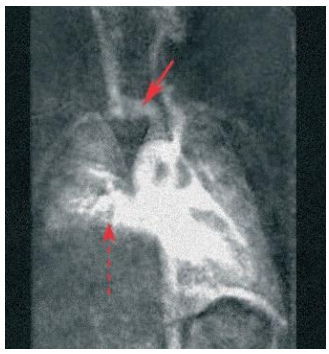
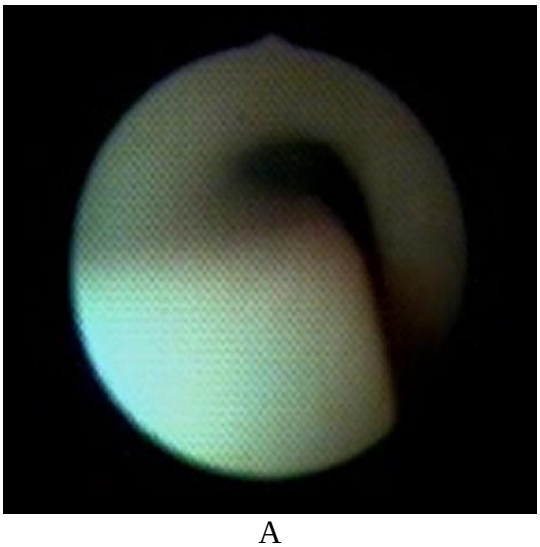


D

Fig. 8.9. Digital subtraction angiography. A – Double aortic arch with balanced arches. Each aortic arch has two brachiocephalic vessels (subclavian and carotid artery). B - Aberrant right subclavian artery, which arises from descending aorta and pulls it to the right (arrow). C - Pulmonary artery sling. Left pulmonary artery (arrow) branches from the right one, has lesser diameter, and forms the sling on its way to left lung hilus. D – Abnormal (distal) branching of the brachiocephalic trunk (arrow). Associated disease — relaxation of the right diaphragm cupola (dotted arrow)

Echocardiography should also be performed to exclude associated cardiac anomalies and to complete the diagnosis.

Bronchoscopy is not essential as a part of routine assessment of a vascular ring unless a pulmonary artery sling is suspected. However, some infants and children with vascular rings will have had airway endoscopy because they present with stridor. The endoscopic appearances are characteristic, with pulsatile extrinsic compression of the airway. Tracheal cartilage cannot be often seen in the area of compression, which is the sign of secondary tracheomalacia (Fig. 8.10). Bronchoscopy may cause injury to mucosa and worsen the obstruction due to swelling of the mucous membranes, so it should be performed immediately before surgery. Airway examination by bronchoscopy does form an important part of the assessment of a pulmonary artery sling because of the common association with tracheal stenosis.



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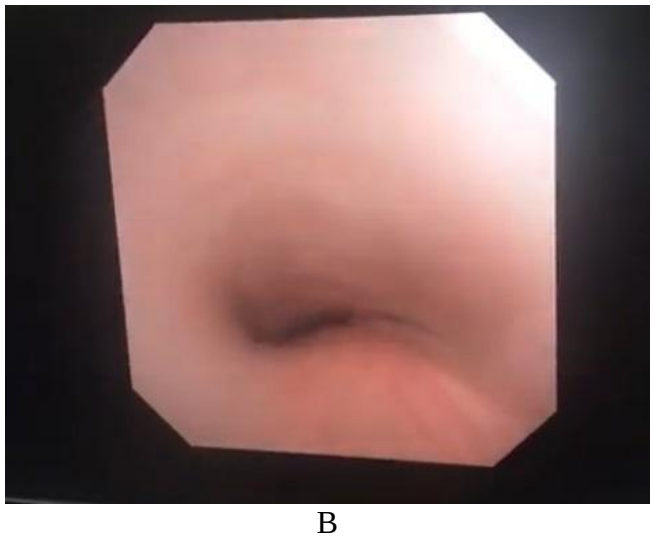


B

Fig. 8.10. Endoscopic photo of tracheal compression due to double aortic arch (A) and abnormal position of brachiochephalic trunc (B). Tracheal walls are deformed and prolapses inside the lumen causing its narrowing. Tracheal cartilaginous rings are not visible due to secondary tracheomalacia

Differential diagnosis**.** Because most children with vascular rings present with stridor, the differential diagnosis includes other causes of stridor. The history can provide important clues – for example, the acute presentation of laryngotracheobronchitis or epiglottitis is totally different from the chronic history of a vascular ring. Laryngomalacia is a common cause of stridor in infancy. The stridor is high-pitched, musical, and exclusively inspiratory. If there is doubt about the diagnosis airway endoscopy and/or a barium swallow will clarify matters. Subglottic stenosis is usually acquired as a consequence of prolonged endotracheal intubation and, therefore, more common in ex-premature infants. The diagnosis is made by airway endoscopy.

It is necessary to conduct differential diagnostics of compression syndrome caused by tumors and cysts of the mediastinum, including bronchogenic and enterogenic cysts, esophageal diverticulum.

Differential diagnosis between vascular ring variants is important to select the optimal method of surgery.

Treatment of vascular compression stenosis of the trachea is only surgical. The essence of the operation consists of division of vascular ring and depends on anatomic variant of the anomaly. In the case of double aortic arch functionally less significant or atretic aortic arch is divided as well as ductal ligament (or patent ductus arteriosus). In the case of secondary tracheomalacia the operation is complemented with anterior or posterior aortopexy to stabilize softened trachea. Correction of right aortic arch with left ligament include division of the ductal ligament and, if necessary, posterior aortopexy or resection of Kommerell's diverticulum. Compression caused by aberrant right subclavian artery is corrected by means of division of this vessel at the site of its branching from the aortic arch. Circulation of the upper limb is kept by the presence of natural collaterals. However, some surgeons recommend reimplantation of the right subclavian artery into the right common carotid artery, thus forming the normal brachiocephalic trunk.

Correction of pulmonary artery sling is a complex reconstructive surgery that may require cardiopulmonary bypass. The left pulmonary artery is dissected as far as possible and freed from the adjacent structures while the ligamentum arteriosum is divided. The pulmonary artery is divided between clamps and the proximal stump closed. The left pulmonary artery is then retrieved from behind the trachea and brought forward to assume the anatomical position next to the main pulmonary artery. The left pulmonary artery is anastomosed to the main pulmonary artery and to the side by using a side-biting clamp. If the pulmonary artery sling is associated with tracheal stenosis this should be repaired at the same time. Localized stenoses can be treated with resection and reanastomosis. A sliding tracheoplasty may be required for longer stenoses. More extensive stenoses with complete tracheal rings can require complex reconstruction on cardiopulmonary bypass and should only be undertaken by specialist teams.

Anomalous origin of the innominate artery is usually treated by suspension of the proximal part of the artery and the aorta to the sternum. The innominate artery can be approached via mid-sternotomy, right anterior thoracotomy or left anterior thoracotomy. The anterior ligament aortopexy first proposed by prof. D.U. Krivchenya allows avoiding such complications as suture tearing, bleeding and recurrence of airway compression.

Surgical correction of all variants of vascular rings for the first time in Ukraine developed and implemented by prof. D.U. Krivchenya during 1982 – 1989.

Bronchogenic and enterogenic cysts

Benign congenital cystic lesions have been referred to by various names, including bronchogenic cyst, esophageal duplication cyst, enteric cyst, and neuroenteric cyst. They are all believed to constitute congenital abnormalities of division of the embryonic primitive foregut.

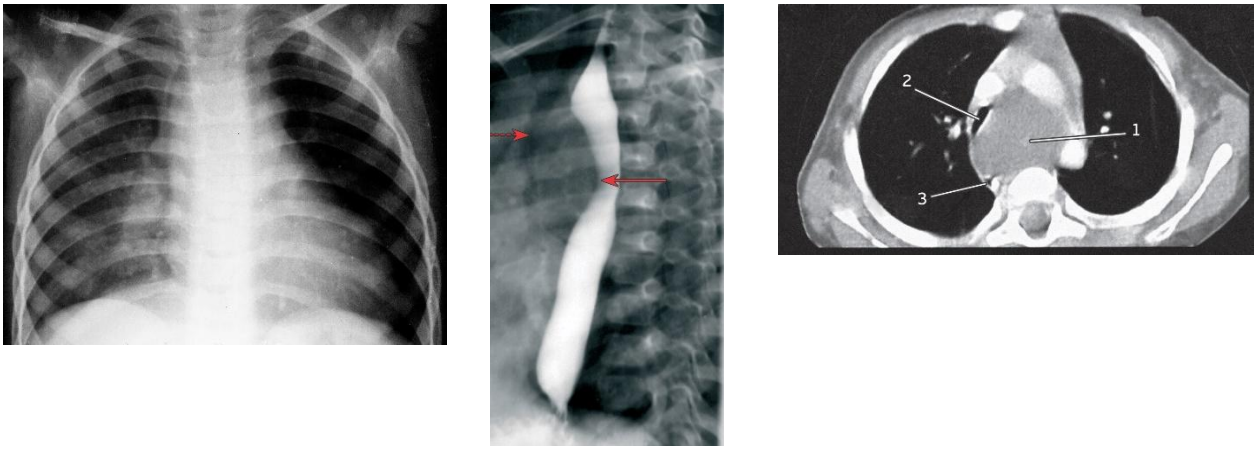
Bronchogenic cysts are the most common of the bronchopulmonary malformations encountered in children. These lesions are usually unilocular, are lined by ciliated columnar epithelium and contain fragments of cartilage and smooth muscle within the wall. Bronchogenic cysts usually demonstrate no patent communication to the airway but are almost always closely attached to the major airways or to the esophagus by dense fibrous tissue.

Enteric cysts of the mediastinum contain gastric mucosa and may or may not communicate with the gastrointestinal tract below the diaphragm. These cysts may be associated with vertebral anomalies, but there exist no intervertebral communication.

Neuroenteric cysts contain both endodermal and ectodermal, or neurogenic, elements. Characteristically, a neuroenteric cyst is connected by a stalk to the meninges and spinal cord. They are associated with spinal deformities, including congenital scoliosis, hemivertebrae, and spina bifida.

In contrast to the situation in adults where these lesions are incidental findings, in children these cysts are usually symptomatic and sometimes life-threatening. Symptoms include persistent cough, progressive dyspnea, wheeze, stridor, and cyanosis. Additionally, gastroenteric cysts might be associated with pain or melena from peptic complications secondary to the activity of the gastric tissue.

Plain chest x-ray is the single most useful diagnostic tool in establishing the diagnosis of these foregut cysts. It demonstrates the cyst in the mediastinum displacing or compressing the trachea, bronchi or esophagus. There may also be evidence of hyperinflation, atelectasis or infection in either lung. Useful adjuncts include esophagography, CT scan (Fig. 8.11), bronchoscopy, and echocardiography.



A B C

Fig. 8.11. Bronchogenic cyst of mediastinum. A – Plain chest radiograph, ovoid shaped mass in the middle mediastinum, deviation of the trachea to the right, hyperlucent left lung (obstructive emphysema). B - right lateral esophagograph: compressive deformation of the esophagus (solid arrow), tracheal deviation (dotted arrow). B — CT with contrast enhancement, axial scan: compression and narrowing of the trachea, the cyst (1) squeezes and “pushes” the trachea (2) and esophagus (3).

Treatment of foregut cysts is always complete surgical excision, traditionally through a thoracotomy. This provides relief of symptoms and definitive treatment. If the entire cyst is excised, there exists no risk of recurrence. If part of the epithelial lining is left behind, these children are at risk for recurrence and must be monitored with serial films.

Tracheomalacia

Tracheomalacia is generalized or local structural anomaly of the tracheal wall with deficiency or weakness of its cartilaginous framework and widening and dysplasia of tracheal membrane resulting in excessive narrowing of the trachea during expiration or in any increase intrathoracic pressure. There are following synonyms of tracheomalacia found in the literature: expiratory tracheal stenosis, tracheal dyskinesia, expiratory prolapse, collapse, expiratory invagination.

Morphological basis of the anomaly is the defect of elastic fibers and smooth muscle in the walls of the trachea and bronchi, shortening and degeneration of cartilage rings, widening and

dysplasia of membranous wall. Cartilage rings were often incomplete, cartilage to muscle ratio was reduced, and there was an increase in the length of the membranous portion of the trachea which is the weakest part of the tracheal wall. In severe cases, the normal cartilage-to-membranous wall ratio of 4–5:1 can decrease to 2:1. These structural features cause the instability of the trachea during the respiration, especially forced breathing and cough. At exhalation the intrathoracic pressure increases and tracheal wall prolapses into the lumen that results in its narrowing down to a complete collapse. Narrowing of the cervical segment of trachea occurs, on the contrary, during forced inhalation. Besides the prolapse of walls of the trachea there is also a convergence of the free ends of cartilage rings that enhances stenosis. This "functional" tracheal stenosis leads to disruption of ventilation and impedes normal expectoration (discharge of secretions). The result is respiratory failure and joining of airway and lung infections.

There are two distinct forms of tracheomalacia – primary (idiopathic and associated with esophageal atresia) and secondary (due to compression or infection). Secondary TM can be congenital and acquired. The disease also is classified into diffuse and localized (up to 1/3 of tracheal length) forms. The morphological signs of tracheomalacia are found in almost all infants operated on esophageal atresia with tracheoesophageal fistula, and 25% of them may require surgical correction. Secondary tracheomalacia may be acquired or possibly associated with another congenital anomaly which compresses the airway wall during its development or growth. A localized area of a soft tracheal cartilaginous wall which develops because of pressure from a vascular ring or a parabronchial tumor or cyst may cause symptoms after the primary lesion is diminished. In infants and children, secondary tracheomalacia may present during the treatment of acquired or congenital severe pulmonary disease such as bronchopulmonary dysplasia, recurrent bronchitis and cystic fibrosis. Tracheomalacia is classified into three degrees by narrowing of the lumen: I - ½, II – ½ -2/3, III - from 2/3 to complete obstruction.

Clinical symptoms of tracheomalacia depend on the location, the length of the abnormal airway segment and the severity of the structural abnormality. Stridor is often insidious during the first few weeks of life. In 60% of the cases, symptoms appear before the age of 3 months, whereas in the remaining cases, the symptoms appear by 1 year of age. The hallmarks of this condition are: prolonged expiratory phase with wheezing, harsh barking cough, attacks of cyanosis, apneic spells and recurrent airway infections. Another characteristic of this dynamic condition is variability in both the symptoms and their intensity. Cyanosis and apneic spells often occur during feeding, coughing and crying. They are typically interspersed with normal periods of quiet sleep. The apneic spells are usually the most prominent clinical features of the disease because of their life-threatening nature. Severe obstruction is accompanied by a forced position as neck hyperextention leading to ease of breathing due to straighten and a certain stabilization of the trachea. Stridor occurs if obstruction is 2/3 of lumen and more, can be a non regular and increases when respiratory infection joins. Inability to extubate may be the symptom of severe tracheomalacia in infants operated on for esophageal atresia. These babies can easily breath through the tracheal tube but any attempt of extubation lead to severe respiratory failure. Endotracheal tube in such cases acts as a tracheal stent. More typically, infants operated on for esophageal atresia correction present with respiratory symptoms in 2 - 3 months after operation when their physical activity increases. Later disability caused by morphological changes in the respiratory system (pulmonary emphysema, recurrent pneumonia).

Comprehensive approach considers videotracheobronchoscopy and chest CT with contrast enhancement as main and first line diagnostic methods in case of tracheomalacia. Bronchoscopy should be performed on spontaneous respiration without use of muscle relaxants, thus the effect of respiratory phases and coughing can be properly assessed. Significant collapse of the trachea and changes of the shape of tracheal lumen are seen only on forced expiration or during coughing (Fig. 8.12).

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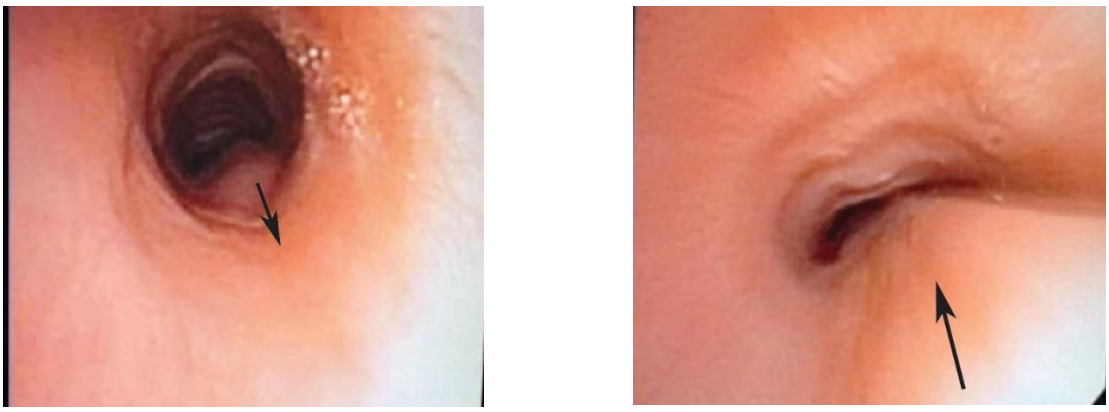
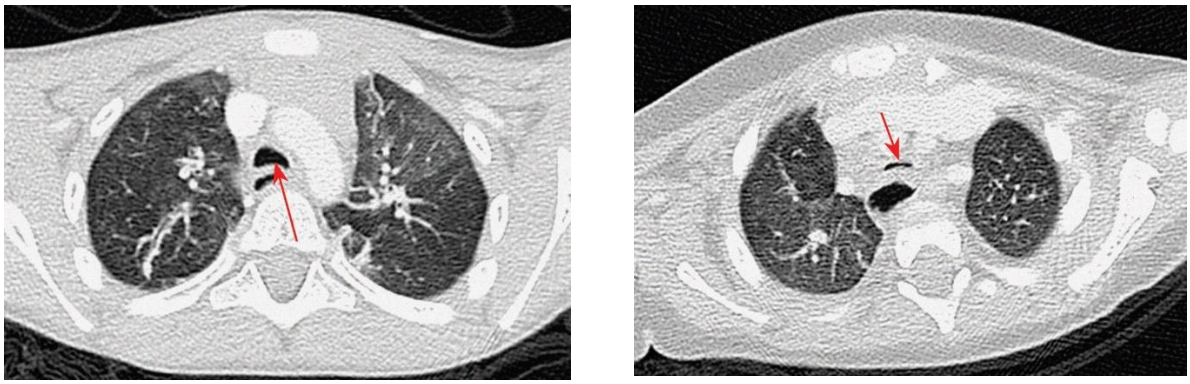


A B

Fig. 8.12. Tracheoscopy in patient with tracheomalacia associated with esophageal atresia. A – Inspiratory phase: tracheal lumen is widened due to the outward move of membranous wall. B – expiratory phase: narrowing of the tracheal lumen due to inward prolapse of membranous wall of the trachea

Today, CT with contrast enhancement is one of the most informative and safe methods of examination in patients with defects of the respiratory system. In case of tracheomalacia, CT shows the narrowing of tracheal cross-sectional area, crescent-shaped, teardrop-shaped or slit-like deformity of tracheal lumen in the narrowed zone (Fig. 8.13). The assessment of the relationship of the trachea with surrounding structures: great vessels, thymus, esophagus is of major significance and facilitates recognition of the origin of tracheomalacia and planning of treatment. The new CT tools such as 3D-reconstruction, virtual bronchoscopy and Cine CT-ultrafast facilitates the identification of functional abnormalities of the trachea in infants and children.



A B

Fig. 8.13. Computed tomography in patients with different variants of tracheomalacia. A – idiopathic tracheomalacia, patient's age 2 years and 4 months. Obstruction of the trachea caused by inward prolapse of the widened membranous wall (arrow). B – tracheomalacia associated with esophageal atresia, patient's age 4 months. Slit-like shape of tracheal lumen. Tracheal obstruction is caused by closing of the anterior and posterior walls due to loss of cartilage rigidity and widening of the membranous wall

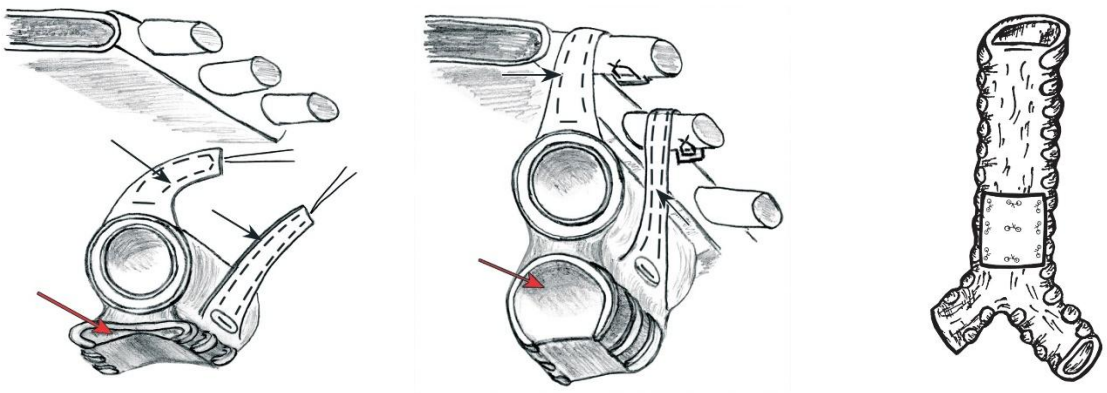
For the diagnostic approach, simple imaging studies, beginning with anteroposterior and lateral chest X-rays, and an esophagram using water-soluble contrast medium may be performed. Chest X-ray films can reveal a variety of anomalies, such as pneumonia, atelectasis from mucostasis or obstructing emphysema. In elder children lateral fluoroscopic view of the trachea is helpful to assess the caliber of the air-filled trachea at the different phases of respiration. Esophagography will rule out recurrent tracheo-esophageal fistula, anastomotic stricture, gastro-esophageal reflux or compression stenosis of the trachea due to vascular rings.

Differential diagnosis is performed with infectious and allergic respiratory diseases (recurrent obstructive bronchitis, recurrent pneumonia, asthma). It should be remembered that these diseases may "mask" tracheomalacia. Because wheezing is a common symptom in these patients, many are mistakenly treated for asthma for long periods. It is now believed that the incidence of asthma is not higher in patients with tracheomalacia than in the rest of the population, so this diagnosis should always be reconsidered in these patients when they show no convincing response to an adequate trial of bronchodilators. In children after correction of esophageal atresia tracheomalacia can be combined with gastroesophageal reflux and recurrent fistula.

Treatment of tracheomalacia depends on the severity of the disease. In children with a minor degree of collapse and mild or moderate symptoms, surgery is not necessary. Usually by 1 year of age, mild respiratory symptoms will resolve because of an increase in the size and rigidity of the trachea. Medical management is directed to the treatment of respiratory infection. Respiratory physiotherapy, bronchodilators and in-hospital observation may be useful.

A surgical approach is required for patients with severe tracheomalacia. Indications for surgery are stridor, tracheal collapse of 2/3 or more, apnea (even a single episode), recurrent pneumonia and bronchitis, inability to extubate the trachea. Choice of operations defined by mechanism of airway obstruction. The narrowing of the tracheal lumen mostly due to prolapse of anterior wall is the indicatuion for aortopexy (Fig. 8.14A, B). Aortopexy has proved to be a safe, expedient method of treating tracheomalacia in most patients with severe tracheomalacia. The aim of aortopexy is to change the cross-sectional profile of the trachea from an elipse to a circle so that its walls will not appose during ventilation and coughing, and at the same time move the trachea away from the esophagus so that when full it cannot compress the airway. The suspension of the aorta vascular structure from the under-surface of the sternum is done without disrupting the fascia between it and the adjacent trachea which acts as a suspension ligament to change the configuration of the trachea.

If the leading factor of expiratory stenosis is the prolapse of tracheal membrane, tracheoplasty is performed. The surgery implies reinforcement of posterior tracheal wall by means of grafts of different materials: autopericardial, fascial or artificial patch (Fig. 8.14C).



A B C

Fig. 8.14. Anterior ligament aortopexy. A – Pericardial patches are formed and reinforced with nonabsorbable thread. B - Pericardial patches are conducted through the intercostal space; the aorta is tightened to the sternum. The black arrows indicate pericardial patches; the red arrows indicate the tracheal lumen, widened due to the procedure. C – Schematic representation of tracheoplasty with pleuro-facial or pericardial patch

The combination of both factors of airway obstruction and a significant degree of stenosis requiring a combination of aortopexy and tracheoplasty.

Prognosis of surgical treatment of tracheomalacia at the correct indications and mode of operation is favorable, positive results achieved in about 95% of cases.

Tracheal diverticulum. This is an extremely rare anomaly, usually arising from the right posterolateral surface of the trachea, that may give rise to symptoms only late in adult life when it becomes infected. Because of this, it has been suggested that it may be an acquired rather than a congenital lesion, but a few reports of congenital diverticula exist. Some consider this malformation as a blind-ending tracheal bud, i.e., the lesser form of an accessory tracheal bronchus.

Esophageal atresia (EA) is the malformations related to defective septation of the primitive foregut. This is the most common among other foregut malformations, with an incidence of 1 in 3000 to 5000 live births. In 80% of cases, the esophageal atresia is accompanied by a tracheoesophageal fistula (TEF) between the lower trachea or carina and distal esophagus.

A classification based on the anatomy of the malformation and the frequency of each variant is of the greatest practical value to the neonatal surgeon (Fig. 8.15).

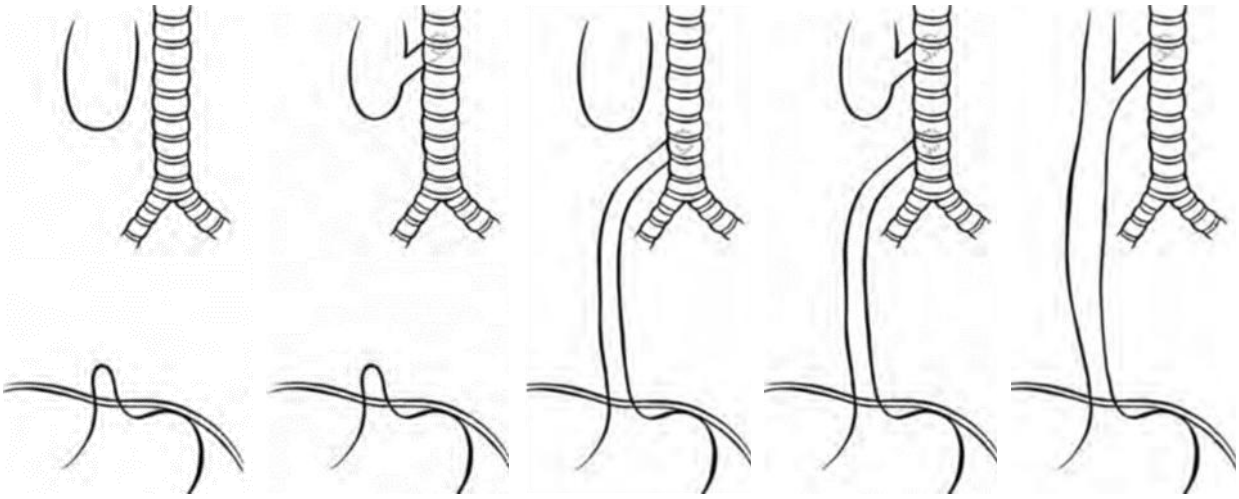


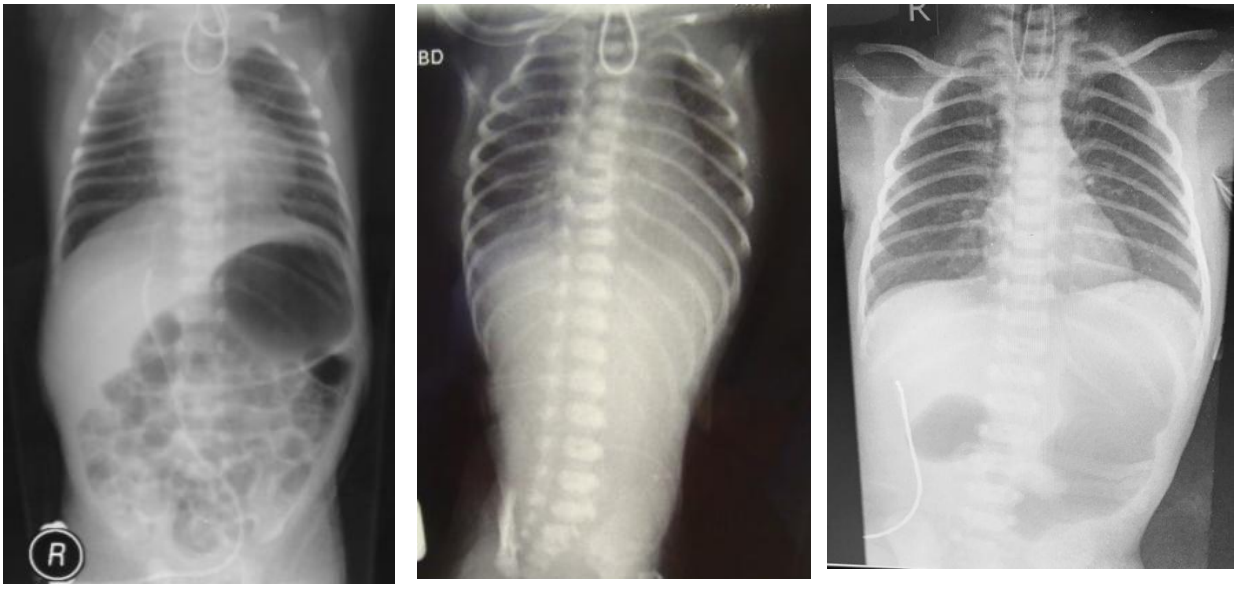
Fig. 8.15. Classification of EA and/or TEF according to R.Gross: A - Esophageal atresia without fistula (8.4%); B - Esophageal atresia with proximal fistula (2.1%); C - Esophageal atresia with distal tracheoesophageal fistula (82.8%); D - Esophageal atresia with proximal and distal fistulas (3.4%); E - Tracheoesophageal fistula (H-type) without atresia (3.3%)

The sporadic reports of familial cases of EA and TEF suggest a polygenic hereditary etiology. The estimated risk of recurrence for parents of a single affected child is 0.5–2.0%. If another sibling is born with EA the risk increases up to 20%. A 10% incidence of non-specific chromosomal abnormalities (translocations, deletions and duplications) has been noted. However, only trisomies 18 and 21 show any definite association with EA and TEF.

Associated anomalies are seen in over half of all newborns presenting with EA and TEF. Congenital heart disease (27%) is the commonest comorbid condition and has the greatest impact upon survival. Recently aortic arch anomalies have been shown to occur frequently in association with long-gap EA and TEF. Other common associated abnormalities include urogenital (18%), skeletal (12%), anorectal (12%), and other gastrointestinal conditions (9%), most notably duodenal atresia. Several phenotypic variants have been reported in association with EA and TEF. The first to be described was the VATER association, which is now encompassed by the VACTERL acronym. The presence of three or more of the features is essential to define the association. The CHARGE association (coloboma, heart disease, atresia choanae, retarded development, genital hypoplasia, and ear deformities with deafness), is another constellation of phenotypes associated with EA and TEF. EA and TEF are also recognized in the SCHISIS association (exomphalos, neural tube defects, cleft lip and palate, and genital hypoplasia).

Clinical presentation and diagnosis. A newborn with EA is often noted to have difficulty clearing saliva. Episodes of coughing, choking and even transient cyanosis may be observed shortly after birth. These signs are frequently overlooked and attempts to feed the infant result in

immediate respiratory distress. The diagnosis is readily confirmed by the failure of passage of a firm nasogastric tube. A characteristic resistance is felt at the blind ending upper esophageal pouch, and the tube cannot be introduced into the stomach. A plain X-ray, which should include the chest and abdomen, demonstrates the nasogastric tube coiled in the upper pouch. An associated TEF is confirmed by the presence of gas-filled intestinal loops below the diaphragm (Fig. 8.16A). In isolated or pure EA, a featureless gasless abdominal X-ray is observed (Fig. 8.16B). The presence of a double bubble on the abdominal film suggests associated duodenal atresia (Fig. 8.16C). A careful search for associated abnormalities is mandatory, specifically checking for patency of the anus. The cardiovascular system should be examined to exclude a major congenital heart defect whose treatment may take priority over correction of the EA.



A B C

Fig. 8.16. A - Chest X-ray in a neonate with EA and distal TEF. The tube is coiled in the blind-ending upper esophagus. Air outlining intestinal loops below the diaphragm confirms the existence of a distal TEF. **B -** "Pure" EA. The nasogastric tube is coiled in the upper esophageal pouch. Absence of air-filled abdominal intestinal loops suggests that there is no distal TEF. **C -** EA and TEF with duodenal atresia. Nasogastric tube coiled in upper pouch. 'Double-bubble' appearance confirms duodenal atresia

Antenatal diagnosis. Fetal diagnosis is now possible in cases of EA and TEF. The classical ultrasonographic features of EA and TEF in the fetus are absence of the stomach bubble and associated polyhydramnios. However, prenatal detection rates remain low (9–24%), and there is a high false-positive rate, with over half of all cases proven not to have EA after birth.

Management**.** Having established the diagnosis, intravenous fluids are started, endotracheal intubation performed and secretions are aspirated intermittently from the upper pouch. Broad-spectrum antibiotics and Vitamin K should be administered. The baby is transferred to the neonatal surgical unit. An echocardiogram performed prior to surgery will alert the surgeon and anesthetist to an underlying cardiac defect that may adversely influence prognosis, and may influence the operative approach by identifying the side of the aortic arch. Surgery is ideally performed within the first 24 hours in an otherwise healthy newborn.

The infant is positioned for right thoracotomy in a lateral or postero-lateral position. Some surgeons choose the surgical access to be on the opposite side of artic arch. The chest is opened through the fourth interspace. The pleura is carefully separated from the ribs to commence an extra-pleural approach towards the fistula. The azygos vein is mobilized and controlled with suture slings. The distal esophagus is identified and the TEF is dissected and divided and the defect of tracheal wall is sutured. Then upper esophageal pouch is identified and mobilized to diminish the

gap. An upper pouch fistula may be identified at this stage, and should be repaired. In most cases of EA with distal TEF, a primary anastomosis is possible, although occasionally considerable tension is required to complete the repair. Chest drain is placed with the tip near the anastomosis. The chest drain should be attached to under-water drainage.

Should additional surgical pathology be present, such as duodenal atresia or imperforate anus, these should be dealt with accordingly under the same anesthetic in the stable infant.

Surgical management of neonates with "pure" EA is challenging and controversial. The gap between the blind end of proximal and distal esophagus is very long in most cases making the primary anastomosis impossible. The majority of pediatric surgeons consider delayed primary anastomosis of the native esophagus, the optimum approach. The infant with isolated EA is initially managed by feeding gastrostomy and continuous suctioning of the upper pouch. After a period of approximately 3 weeks, the extent of the gap can be assessed by fluoroscopy. A distance of less than two vertebral bodies separating upper and lower pouches is ideal to attempt delayed primary anastomosis. A spiral, circular or staged myotomy or upper pouch flap may be required to achieve an anastomosis. Extra length can be obtained on the lower pouch if necessary by proceeding to laparotomy and performing either a Scharli lesser curve myotomy or a Collis gastroplasty. If a primary anastomosis is still not possible the option is esophageal replacement (esophagocoloplasty) at the age of 10-12 months.

Congenital isolated tracheoesophageal fistula (H-fistula). An isolated tracheoesophageal fistula is a rare malformation with prevalence about 1:100000 newborns and seen in about 5% of all esophageal malformations. American researcher D.S. Lamb was the first to describe the defect as early as 1873. The symptoms are recurrent coughing and choking, especially during feedings. The diagnosis is often delayed, resulting in recurrent pneumonia. A distended abdomen with air-filled loops may also be noted. When the diagnosis is suspected, fluoroscopy is performed in the prone position with contrast injection through a nasoesophageal tube starting in the mid-esophagus and gradually moving up. False negative studies may occur. Rigid bronchoscopy is essential to confirm the diagnosis and it facilitates the operation. The fistula, usually located in the lower cervical area, is cannulated during bronchoscopy. Diagnostic studies (Figh. 8.17): esophagography, fistula cannulation followed by chest X-ray, tracheobronchscopy confirm the diagnosis suspected on clinical symptoms. Fistula catheterization facilitate intraoperative identification and allows minimizing the dissection of trachea and esophagus, thereby decreasing the risk of recurrent laryngeal nerve injury. The division of the fistula through a cervical approach usually give excellent results, however the recurrence rate is reported about 5 to 10 %. The defects in esophageal and tracheal walls should be precisely closed. The techniques of fascia interposition or trachea reinforcement with demucosated wall of fistula have been used to reduce the recurrence rate. Attempts at endoscopic occlusion of the fistula with glue, laser, or other means is associated with a high recurrence rate and do not appear warranted in H-type fistulas that can be divided through a cervical approach. These techniques may have their place in the management of recurrent TEF. About 10% of H-fistulas are too low to be divided through a cervical approach and require thoracotomy or thoracoscopy.

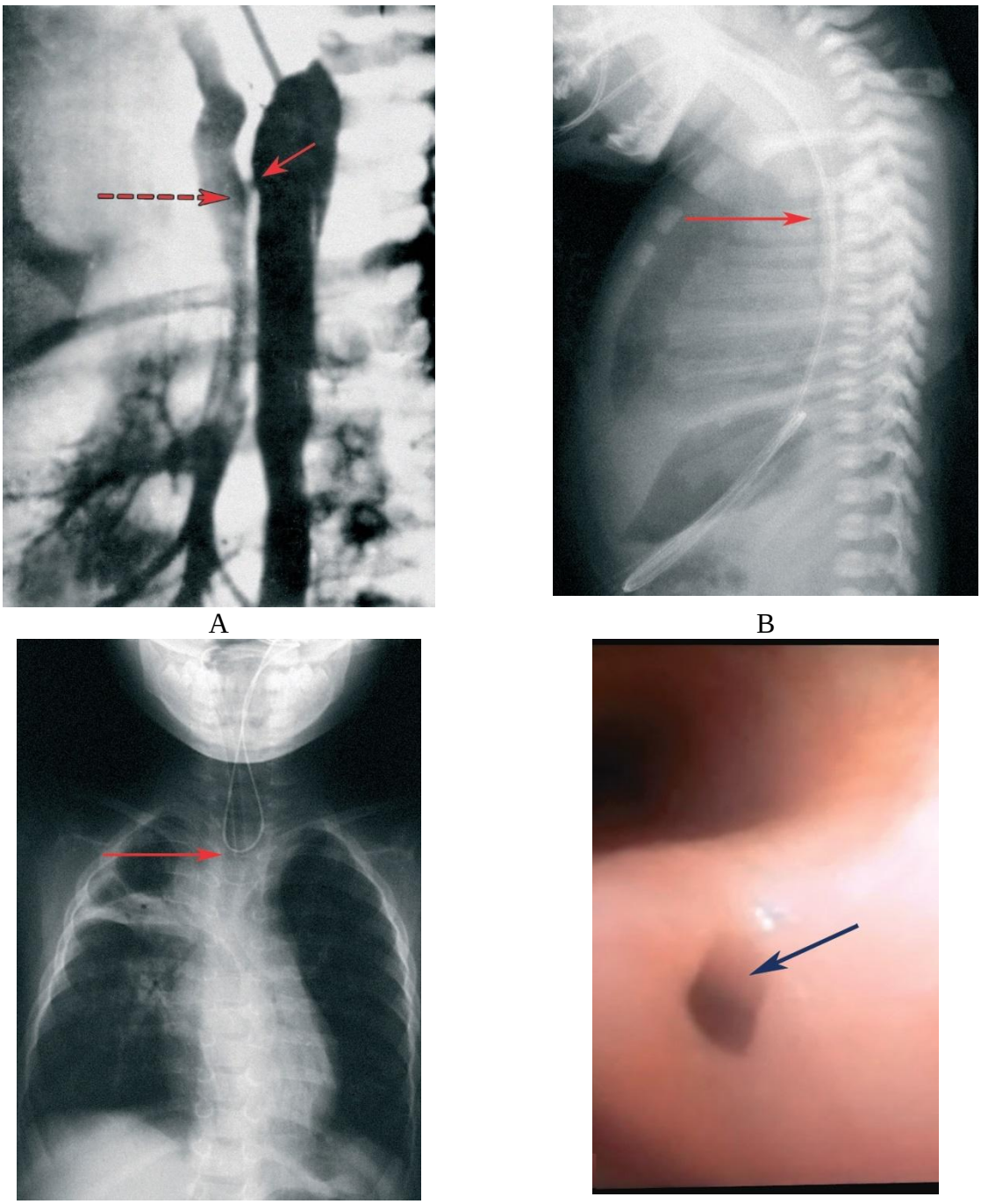


Fig. 8.17. X-Ray and endoscopic methods of diagnosis of TEF. A - esophagograph with water-soluble contrast, H-type fistula between the esophagus and trachea (arrow), trachea and bronchi are filled with contrast (dotted arrow). B - fistula catheterization, a thin probe introduced through the trachea, passed through the fistula into esophagus (arrow) during tracheoscopy. C - Reversion of catheter introduced through the fistula during tracheoscopy and esophagoscopy. Catheter formed loop at the fistula (arrow). D — tracheoscopy. Fistula orifice on the posterior wall of the trachea (arrow)

Congenital malformations of the lungs

Congenital malformations of the lung are uncommon but extraordinarily diverse in their presentation and important to all physicians and surgeons who care for infants and children. The spectrum of presentation includes antenatal diagnosis of malformations to presentation in adulthood and ranges from asymptomatic to immediately life threatening.

Formation of the bronchopulmonary system begins at 3 - 4 weeks of gestation from the caudal foregut segment, i.e. from one tube, which is then delimited on the trachea and esophagus.

Not by chance, some authors consider the malformations of the esophagus (atresia, fistulas) as defects of the respiratory system. On 7 – 8th month of gestation in the process of encoded intensive development of embryo alveolar ducts give rise of acini, and capillaries deepen under an epithelium forming alveoli with aero-hematic barrier as a gas exchange organ.

Respiratory and vascular components of lungs complete their development, mainly before 26 weeks of gestation, but for the valuable functioning and postnatal life-support 30 weeks is needed.

After birth during 1 - 4 months an air-vascular organ continues its development, the number of alveoli increases as well as the lumen of arterioles, blood pressure decreases (since 10th day) with the reduction of fetal circulation. Lungs as an organ complete their formation by 7 - 8 years and continue its growth to 25 - 27 years.

Malformations of the respiratory system occur for unknown reasons. However, endogenous and exogenous teratogenic factors are distinguished among them. First include heredity, chromosome abnormalities, endocrine diseases, biological inferiority of the germ cells, including the pregnancy in late reproductive period of mother and father in old age and the last involve physical (mechanical, electromagnetic, radiation), chemical (any poisons, hormonal, defects of feed), biological (bacteria, viruses).

Classification. The first classification of malformations of lung was developed by Schneider and Schwalbe in 1912, which distinguished between three types of underdevelopment of the lung tissue: agenesis (complete absence of one or both lungs), aplasia (absence of the lung with the presence of rudimentary main bronchus) and hypoplasia (abortive development of one or two lungs). In Ukraine the classification of A.M. Sazonov et al (1981) is rather popular, which, in particular, underlines vascular anomalies in the spectrum of defects of the respiratory system. This classification is as follows.

First group consists of anomalies caused by combined disorders of several structures: agenesis, aplasia, hypoplasia of lungs or lobe (simple and cystic, including polycystic lung) and additional lung lobes.

Second group includes anomalies caused by predominant violation of broncho-epithelial branching: tracheobronchomegaly and tracheobronchomalacia; stenosis of the trachea and bronchi; lobar emphysema; diverticula of trachea and bronchi; broncho-esophageal fistula; bronchogenic cyst; congenital bronchiectasis; hamartohondromas.

Third group includes anomalies of the pulmonary vessels: aplasia and hypoplasia of vessels; aneurysms and arteriovenous fistulas; anomalies of pulmonary venous return.

Fourth group involves anomalies of systemic vessels: pulmonary sequestration (extrapulmonary, intrapulmonary); abnormalities of bronchial vessels (aplasia, hypoplasia) and azygos vein lobe.

And, finally, the fifths group comprises anomalies of other tissues and organs : dermoid cysts; teratomas; mucoviscidosis.

Pulmonary aplasia and agenesis

Pulmonary agenesis and aplasia are rare malformations. Their incidence has been estimated to be 1 in 10 000–15000 autopsies. The malformation have been known since 1762, when the Italian anatomist Morgagni first described it. Since then, about 300 cases have been mentioned in the literature with 2–4 cases per report. Lung agenesis and lung aplasia should be viewed as separate malformations. The mortality of children with lung aplasia is 33% during the first year of life and 50% in the first 5 years of life. Lethal outcomes are usually the result of infection of the single lung and concomitant severe heart and vessel malformations. The course of pulmonary agenesis is more favorable.

Lung aplasia is described as an absence of pulmonary parenchyma and pulmonary artery but with a rudimentary main bronchus and a well-formed tracheal bifurcation. If present, the rudimentary pulmonary parenchyma is hypoplastic and atelectic.

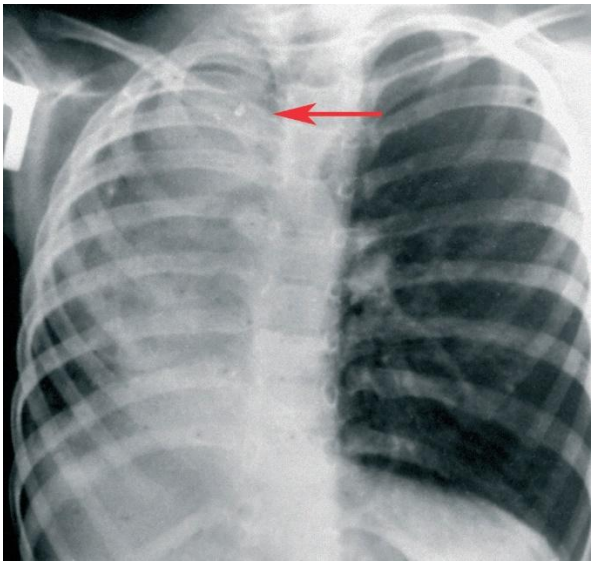
Relevant anatomy and pathophysiology of lung aplasia. The presence of the bifurcation and carina is essential for the pathogenesis of lung aplasia as these provide fixation of the trachea

to the pericardium and aortal arch, which results in a shift of the trachea following the heart and great vessels. Tracheal kinking right above the bifurcation and at the upper thoracic aperture narrows the trachea even further. Such a tracheal shift and double kinking does not happen in lung agenesis where the tracheal bifurcation and its ligaments are absent. The superior vena cava is kinked as well. Tracheal kinking and narrowing in the presence of the main bronchus stump (which may be rather long up to 3 cm) transforms the laminar airflow into a turbulent flow. It also increases resistance and dead space and promotes the piling up of infected bronchial secretions. Normal diaphragmatic movements promote a mediastinal shift into the negative pressured empty pleural cavity with lung herniation and emphysema of herniated segments or lobe due to kinking of the respective bronchi. The perfusion in herniated segments is almost absent. Together with the pressure of the brachiocephalic vessels on the upper trachea, such herniation results in a further worsening of tracheal compression and obstruction. The hemodynamic changes include kinking of the superior vena cava and innominate vein resulting in a decrease of brain venous outflow. Left-to-right blood shunting due to an opened foramen ovale or patent ductus arteriosus results in hypervolemia of the single lung. Such changes cause early and progressive respiratory distress with infection of the single lung in children with lung aplasia.

Clinical presentation and diagnosis. The signs of respiratory failure develop together with the increase in physical activity by the 3rd month of life or after respiratory infections. The anatomical peculiarities of the malformation expedite such complications. If the patients overcome these complications in their early years, respiratory distress develops at the age of 3–4 years and progresses, limiting normal physical activity, especially during the cold season. The main symptoms are shortness of breath, coughing, wheezing in the lungs, noisy breathing, low physical exercise tolerance, frequent respiratory infections, delay in physical growth, headaches, acrocyanosis, chest pains and palpitations. Most of the manifest or enhanced with increased physical activity of children in the first 2 - 3 months of life.

On physical examination one can reveal shift of the heart to affected side, deformity, asymmetry, retraction of the chest on the side of the missing lung.

Plain chest radiograph shows mediastinal shift to the side of the lesion, the affected hemithorax is opaque, while the opposite hemithorax is of increased transparency and enlarged (Fig. 8.18). Lateral X-ray shows the anterior mediastinum enlightenment due to prolapse of single lung. Verification of the diagnosis requires the use of cardioangiopulmonography, CT and MRI. Digital subtraction angiography reliably establish the absence of lung, agenesis of the pulmonary artery, kinking of venous trunks, the position of the aorta and major vessels in relation to the airway (Fig. 8.19).



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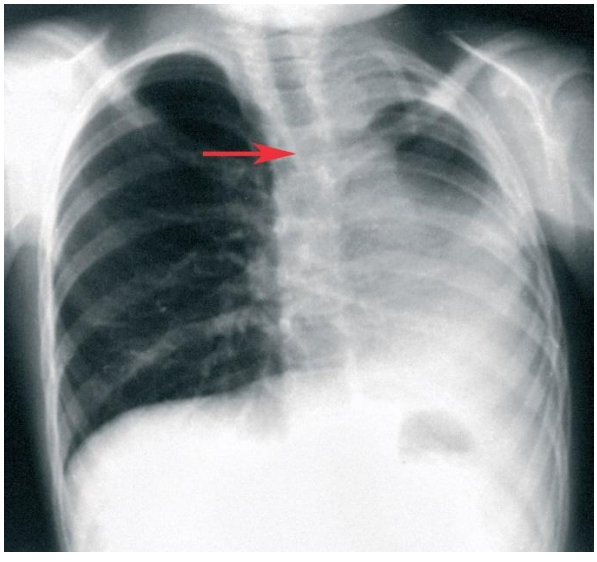
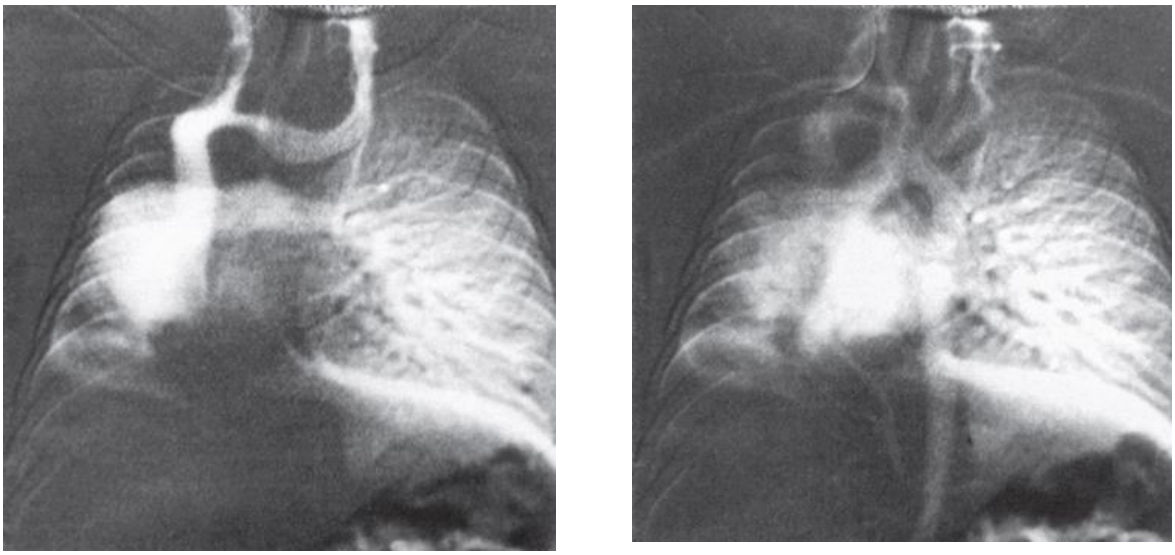


B

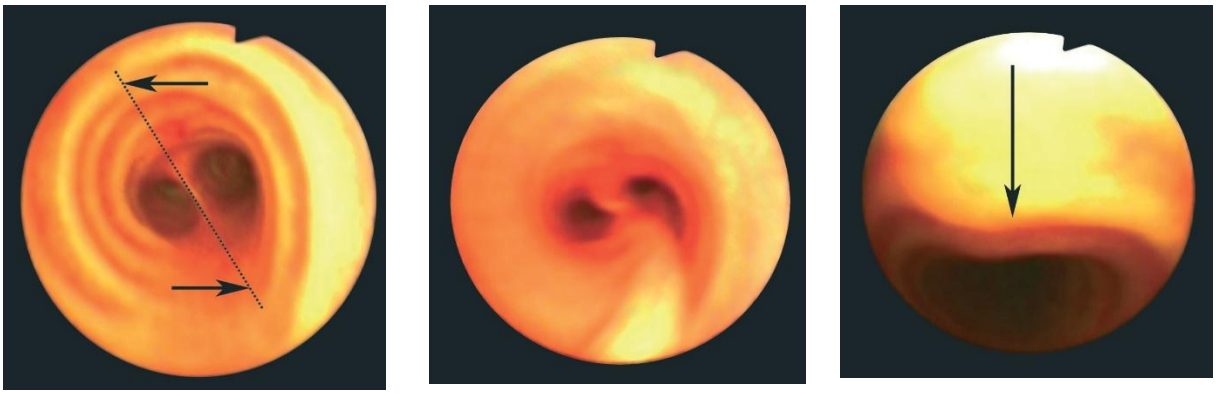
Fig. 8.18 Plain chest radiographs in a female patient, age 4 years, with right lung aplasia and a female patient (A), age 7 years, with left lung aplasia (B). The affected hemithorax is opaque, mediastinal shift to the side of the disease, the single lung is of increased transparency, kinking of the cervico-thoracic tracheal segment (arrows)



A B

Fig. 8.19 Digital subtraction angiography in a female patient with right lung aplasia (age — 3 months). A — venous phase, contrasting of the right heart chambers and pulmonary artery. Mediastinal shift and rotation to the right. Kinking of the superior caval vein. The innominate vein is dilated. B — arterial phase, anteroposterior projection. The heart and aorta turn right and should look like a lateral projection

Trachoebronchscopy shows rotation and anterior vascular compression of the trachea (Fig. 8.20). Tracheobronchography is risky because of the threat of enhance of respiratory failure.



A **B** **C**

Fig. 8.20. Endoscopic photographs of the trachea in a patient with aplasia of the left lung (age 9 years). A — counterclockwise rotation of the trachea and carina. B — prolapse of the tracheal membrane during coughing, twisting of the trachea at its bifurcation. C — anterior vascular compression of the trachea (arrow)

Differential diagnosis should be made with pulmonary agenesis, diaphragmatic hernia, atelectasis of the lung, congenital emphysema, chylothorax and pleuritis, bronchial obstruction due to foreign body.

Treatment. For many years it was believed that patients with lung aplasia subject only conservative treatment of respiratory infections and observation. Recent studies point to the possibility and the need for surgical treatment of respiratory disorders in these patients (C.

Dohlemann at al, 1990, F. Becmeur et al, 1995, D. Krivchenya et al, 1995, 2000, 2007). Considering that the major pathogenetic factors of respiratory distress in the lung aplasia are kinks and compression of airway, displacement, rotation and kinking of great vessels and heart, instability of mediastinum and emphysema of single lung, the following surgical techniques offered:

- cephalad diaphragmatic cupola translocation on the affected side. The operation results in a decrease of the affected hemithorax volume and of the ipsilateral diaphragmatic cupola function by approximately 30%, with normalization of the mediastinal position and mediastinal stabilization. This prevents kinking of the tracheal and large vessels and vascular airway compression. It helps to stop the progression of emphysema in the single lung and promotes tolerance of physical exercise.
- anterior aortopexy to relieve airway compression caused by displacement of cardiovascular complex;
- combination of these operations accompanied by possible resection (amputation) stump bronchus

Pulmonary agenesis. The carina, bronchi, pulmonary vessels and parenchyma are absent in this malformation. In lung agenesis, the single main bronchus is a direct continuation of the trachea. The tracheal bifurcation, the carina and the contralateral main bronchus are all absent. The trachea and main bronchus are less closely connected with the large vessels, which is why their shift to the affected side is less marked, and therefore there is no tracheal and bronchial kinking and stenosis. However, there is a possibility of tracheal compression by the large vessels, especially in the case of right lung agenesis.

Symptoms may appear from the first years of life as a susceptibility to respiratory infections. The chest is asymmetric with diminution of affected hemithorax. The mild-symptom course is possible, and sometimes the malformation is a random finding. Patients presenting late usually have readily detectable flattening and reduced movement of the chest wall on the affected side, with reduced air entry on auscultation, although this often sounds surprisingly better than expected. There may be some breathlessness on exertion and the chest wall deformity may be quite pronounced, with an associated secondary scoliosis.

Imaging techniques combined with rigid bronchoscopy allow a precise diagnosis to be made. On the chest radiograph, there is considerable mediastinal shift and the involved hemithorax is small, with narrowed intercostal spaces. Absence of the ipsilateral mainstem bronchus or the pulmonary artery are definitive diagnostic findings and this can be established by endoscopy, angiography, echocfrdiography or axial imaging techniques. A single lung is of increased transparency, enlarged and hyperinfated, thus may simulate congenital emphysema (risk of erroneous operation on a single lung!).

Management is initially limited to supportive treatment (including oxygen if necessary), correcting associated malformations, and the prevention and treatment of respiratory infections. In some cases, tracheal stenosis or vascular compression may coexist and require treatment. Some patients with severe respiratory distress due to mediastinal shift, tracheal kinking and its vascular compression may require surgery. Cephalad diaphragmatic cupola translocation on affected side and anterior aortopexy may be considered as effective procedures.

Pulmonary hypoplasia is defined as an underdevelopment of all structural elements of the lung (bronchi, vessels and parenchyma). The malformation is believed to be caused by derangement of development of primary lung buds.

Pulmonary hypoplasia is almost always accompanied by hypoplasia of the corresponding pulmonary vessel. This follows obviously from the second rule of lung embryogenesis, in which the vasculature follows the bronchial development.

Hypoplasia as an isolated phenomenon is rare. More commonly, pulmonary hypoplasia is associated with conditions that interfere with lung growth including following: conditions leading to an egress of lung fluid (severe oligohydramnios, compression of the thoracic cage and abdominal contents by the uterus, laryngotracheoesophageal cleft and large tracheoesophageal

fistula); space-occupying lesions (congenital diaphragmatic hernia, eventration, lung malformations, thoracic tumors, pleural effusion, chylothorax, abdominal conditions pressing on the diaphragm); thoracic cage anomalies (Jeune syndrome, achondroplasia, scoliosis); conditions preventing normal fetal breathing movements (anencephaly, phrenic nerve agenesis).

Pathogenesis of pulmonary hypoplasia involves derangement of bronchial patency, which sustains hypoxia of the lung tissue and desolation of pulmonary vessels, pneumosclerosis and of obstructive emphysema.

Hypoplasia of the lungs may be combined with other defects, such as diaphragmatic hernia, malformations of the skeletal system, urinary tract, heart, central nervous system and the digestive system.

There are two major types of lung hypoplasia distinguished: simple and cystic hypoplasia.

Clinically, patients with pulmonary hypoplasia may present in early infancy with respiratory distress ranging from mild to severe, depending on the degree of hypoplasia. Commonly, it is the associated anomalies that draw the attention. In severe bilateral hypoplasia the thoracic cage is obviously reduced in size and characteristically bell shaped, with the base of the chest widening at diaphragmatic level to a normalsized abdomen. The patient is tachypneic, with restricted chest wall movement, and in respiratory distress. Less severe degrees of hypoplasia—unilateral or bilateral—may present later with persistent tachypnea or disproportionate shortness of breath with exercise.

Occasionally, the abnormality is coincidentally noticed on examination or chest radiograph when the patient presents with an intercurrent infection. In unilateral hypoplasia, the chest cage appears asymmetric, with diminished air entry and chest expansion on the side of the lesion together with mediastinal shift to that side. The chest radiograph confirms mediastinal deviation and a lung that often appears hyperlucent. Isotope scanning usually reveals a greater impairment of perfusion than ventilation on the side of the lesion.

The diagnosis and treatment of pulmonary hypoplasia is dependent on the underlying etiology. For example, severe respiratory impairment can be seen with pulmonary hypoplasia from a CDH. In these cases, stabilization of the underlying physiologic impairment is the priority.

Treatment is initially directed at any associated anomalies, with particular focus on congenital cardiac lesions. If an operation is needed to remove a nonfunctioning lobe or lung, every effort is made to preserve functional parenchyma.

The absolute indication for surgical treatment is local and mosaic forms of lesion with involving up to 12 lung segments with progressive and recurrent type of inflammatory course.

The method of surgical treatment depends on the amount of affected lung. Typical anatomical resection of the affected lung zones is the operation of choice for the lesion localized within one or two lobes or whole lung.

Segmental resection of the lung is indicated in the cases of confirmed cavities, both within the bronchus and parenchyma of the segment.

Sparing (atypical) resection of the affected area is indicated for local and mosaic lesion.

Combined resection are advisable to carry out in patients who have cavitary lesions in adjacent areas of the lungs.

At sparing and combined operations pneumatization of remaining lung tissue plays a positive role in the prevention of bronchial kinking and fulfilling the remaining volume of the chest cavity.

In the absence of associated lesions, unilateral pulmonary hypoplasia is compatible with normal growth, development, and survival. Long-term complications include reduced exercise tolerance, recurrent infections and sometimes a worsening chest deformity with scoliosis.

Congenital emphysema is a rare malformation that is characterized by hypertrophy, hyperinflation and overdistention of the lung parenchyma. Although usually described as affecting lobes it may affect individual segments causing an increase in parenchymal lung volume and a decrease in blood flow and potentially compromising ventilation. It may present early with respiratory distress in infants and hence require urgent surgery. Although “congenital lobar

emphysema" is the commonest phrase used in the literature, it does not necessarily correspond with the anatomic location of the affected zone, especially if the superior division (S1, S2, S3) bronchopulmonary segments of the left lung are affected.

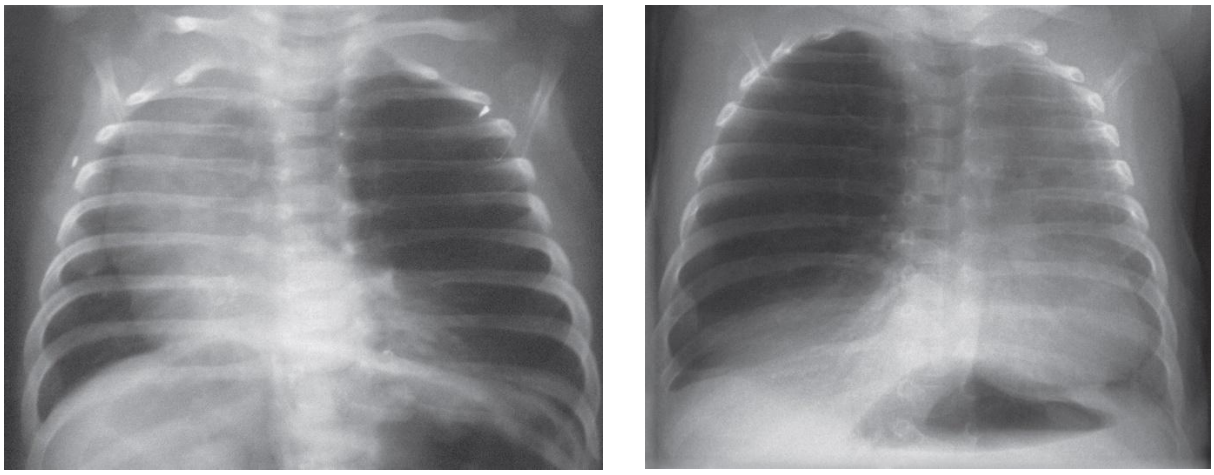
The following causes have been described as a cause of emphysema: dysplasia and lack of bronchial cartilage (bronchomalacia); folds of mucous membrane creating a ball-valve-type effect; obstruction by mucus plug; bronchial stenosis; bronchial atresia; external compression of the bronchus caused by abnormal vessels; enlarged lymph nodes. A polyalveolar lobe has also been found in association with some cases of congenital emphysema. Nevertheless it has been suggested that in up to half of all cases the exact cause of congenital emphysema remains unknown.

The diagnosis of congenital emphysema is based on clinical findings, chest radiography, digital subtraction pulmonary angiography (DSA) and CT with intravenous contrast enhancement.

The differential diagnosis for such cases included pneumonia; respiratory tract foreign bodies; bronchial obstruction by aberrant blood vessels; pulmonary agenesis, aplasia, hypoplasia and dysplasia; and agenesis of pulmonary artery and its branches. Bronchoscopy was used in some cases to exclude lung aplasia or foreign bodies. Bronchography is considered dangerous particularly in those patients with an acute presentation.

The main radiographic signs of with congenital emphysema are following: (1) increase of the lung transparency on the affected side with decreased vascular pattern; (2) shift of the mediastinum to the contralateral side (mediastinal herniation); (3) pseudodextrocardia in left-sided cases; (4) ipsilateral diaphragm flattening; (5) ipsilateral expansion of intercostal spaces; (6) reduction of size of contralateral lung field and decrease of its transparency (Fig. 8.21)

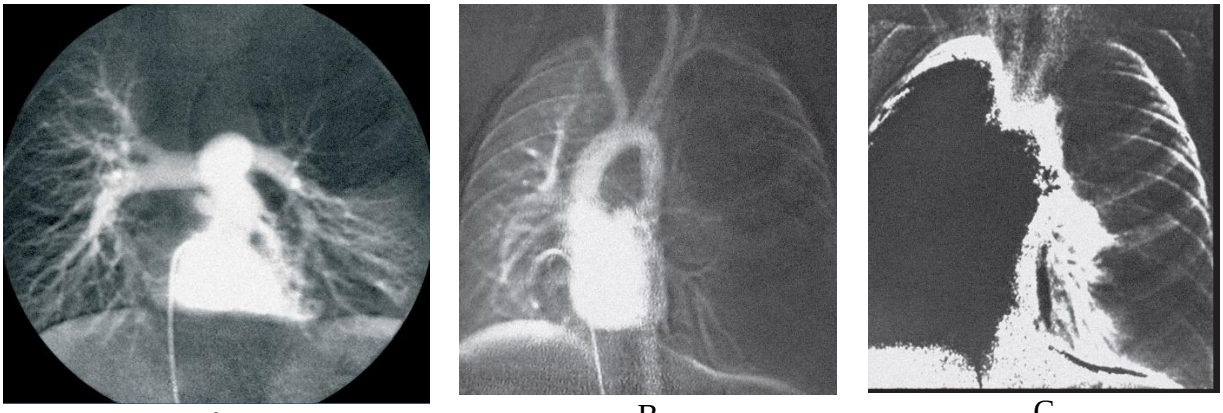
There are three types of clinical presentations of congenital emphysema are distinguished: decompensated, sub-compensated, and compensated. Thus, typical symptoms in the decompensated form were acute respiratory insufficiency with shortness of breath at rest, skin pallor, and in critical cases, cyanosis, asphyxia and convulsions. In the sub-compensated form, children had symptoms of shortness of breath, coughing, and sweating on participation in minor physical activity.



A B

Fig. 8.21. Chest radiographs showing congenital emphysema of superior (S1–S3) segments of left upper lobe (A) and congenital emphysema of right upper lobe (B). There is increased translucency of the lung field on the affected side, and a diminished vascular pattern with shift of mediastinum to the opposite side—pseudodextrocardia (A). There is also ipsilateral flattening of the diaphragm cupola and widening of the intercostal spaces. The contralateral lung field is reduced and has diminished translucency.

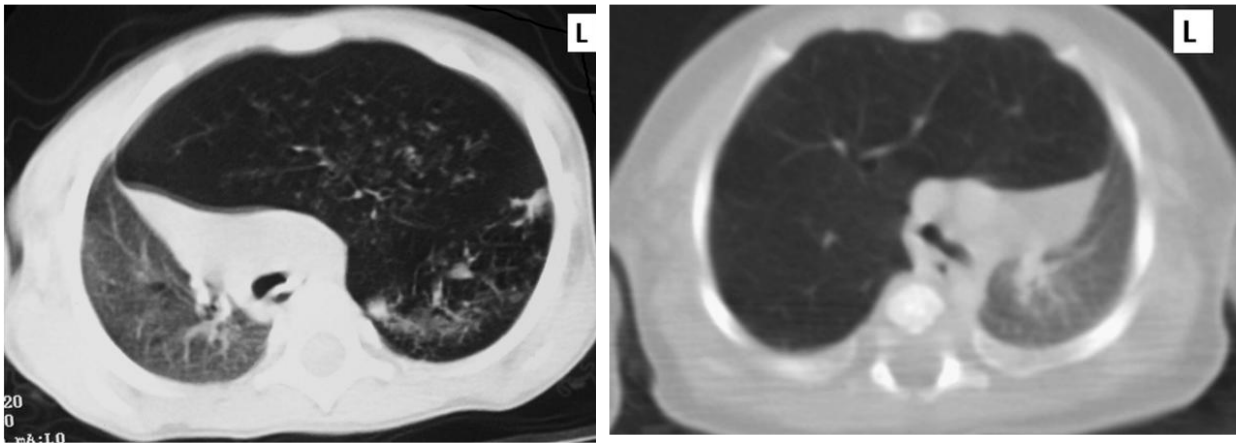
Digital subtraction angiography shows an absence of blood perfusion in the affected segments of the lung; however, the lingual (S_4 and S_5) segments in left-sided disease usually are not affected. Fig. 8.22 illustrates the typical vascular pattern of affected lung parenchyma.



A B C

Fig. 8.22. Digital subtraction angiography (DSA) in a 2-year-old child with congenital emphysema of S_1–S_3 of left upper lobe. The catheter is in the right ventricle. (A) Pulmonary arterial phase showing virtual absence of vascular pattern in the affected left-sided segments, with arteries of the left lower lobe and lingual (S_4 and S_5) segments shifted downward. (B) Delayed contrast within the right pulmonary veins, heart and aorta. The heart is shifted to the right with a rotated aortic arch, visualization of right-side pulmonary veins but absence of left-sided perfusion. (C) DSA volume blood flow measuring mode (videodensitometry). Perfusion can be seen mainly in the right lung ("black picture") with much reduction on the affected left side—only seen in the collapsed left lower lobe and lingual (S_4 and S_5) segments.

Fig. 8.23 illustrates typical CT scan findings including tracheal deviation.



A B

Fig. 8.23. Computed tomograms showing congenital emphysema of S1–S3 of the left upper lobe (A) and right upper lobe (B). Affected lung tissue is overinflated, with a reduced vascular pattern, cardiac shift to the contralateral side and a large mediastinal hernia. The trachea is narrowed and displaced above its bifurcation.

Surgery is performed through a lateral fourth space thoracotomy. Lung-sparing surgery removing only abnormal lung parenchyma is the aim and implies segmental resection in most left-sided cases. Division of a patent ductus arteriosus or short ligamentum arteriosum is performed where necessary. Preservation of lingual (S_4 and S_5) segments of the left lung normalizes position of the thoracic organs, prevents over-distention of remaining lung parenchyma and prevents the development of a large residual postoperative pleural cavity.

Lobectomy is used for cases with whole lobe impairment that is more characteristic for right-sided lesion.

Congenital lung cysts

Congenital lung cysts are intrapulmonary cavities filled with fluid or air, inside lined with epithelium. Some of these may actually be intrapulmonary bronchogenic cysts or unilocular variants of congenital pulmonary airway malformations (CPAM/CCAM). Congenital lung cysts, in contrast to bronchgenic cysts, arise from more distal airways, including the alveoli, or they may be pleural in origin. The incidence of congenital lung cysts ranged from 3.5 to 5.5% among children with chronic nonspecific lung diseases.

The solitary and multiple cysts are distinguished.

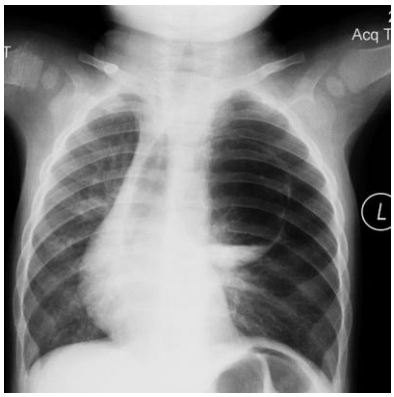
The main clinical symptoms of uncomplicated lung cysts are following: coughing especially in wet cold weather, shortness of breath, frequent recurrences of respiratory infections (bronchitis, obstructive bronchitis, pneumonia), and reduced tolerance to physical activity.

Complications of lung cysts are tension, suppuration and a breakthrough into the pleural cavity. Complicated cases present with acute symptoms including respiratory distress, signs if pulmonary inflammation and suppuration, chest pain, shortness of breath, increase of body temperature, discharge of purulent sputum as "full mouth."

The diagnosis of lung cysts is made by using general clinical and instrumental examination: chest radiography, conventional tomography, CT.

The X-ray signs of cysts are: thin-walled round zone of transparency or opacification (depending on content) omitted by pulmonary vascular pattern (Fig. 8.24). Tension lung cyst causes mediastinal a shift in the opposite direction. Cyst complicated by suppuration that drained into the bronchi, has a fluid level inside. Congenital solitary cysts have one cavity, otherwise multiple lungs cysts consist of many cavities.

Any localized cystic lung lesion that appears congenital and is confirmed on CT (Fig 8.24 B-D) scans should be excised because of the potential risks of infection, erosion into an adjacent bronchus, pneumothorax and the inability to distinguish it from the type I pleuropulmonary blastoma.



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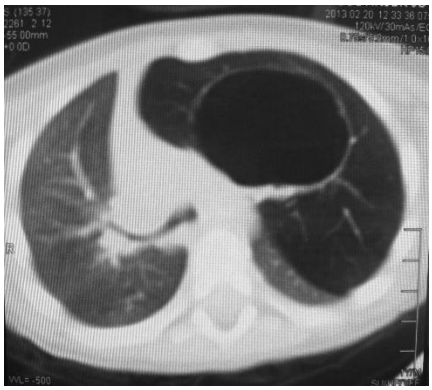

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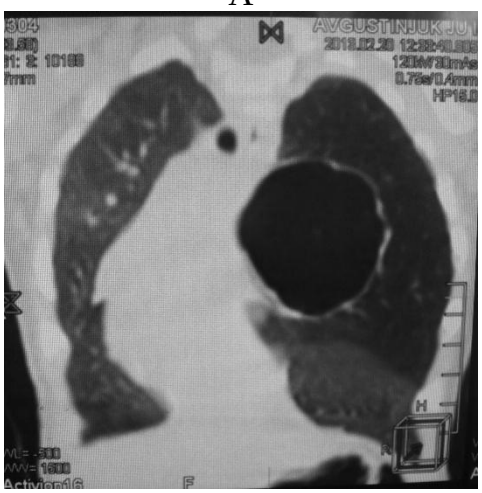
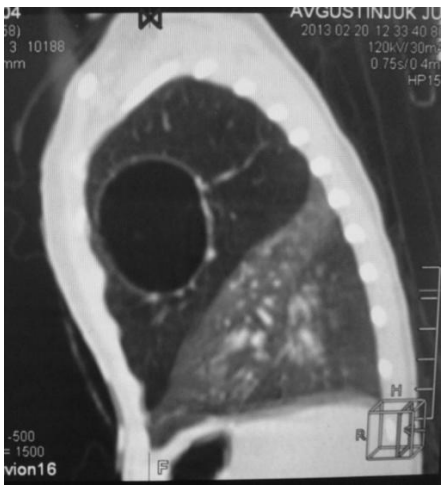




C D

Fig. 8.24. Diagnostic studies in patient with complicated congenital cyst of the left lung (age 10 months). A – Chest radiograph. B, C, D – Chest CT, multiplanar reconstruction. Large cyst of the left lung with fluid level. Emphysema of the upper lobe of the left lung. Displacement of the mediastinum to the right

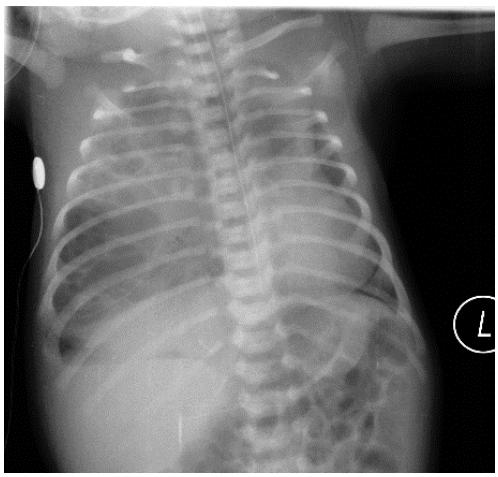
Operations are carried out regardless of the age of the child in compliance with the principle of organ preservation and maximum functional lung tissue.

Congenital Pulmonary Airway Malformations (congenital cystic adenomatoid malformation)

Congenital pulmonary airway malformations (CPAM), previously termed congenital cystic adenomatoid malformations (CCAM), considered a hamartomatous lesion of the bronchial tree by some, whereas others favor a localized arrest in the development of the fetal bronchial tree as the etiology. The incidence of CCAM/CPAM has been estimated at 1 per 25,000 to 35,000 or even 100,000 pregnancies. While relatively uncommon, CPAM constitute 10–30% of congenital lung malformations in most series, with a slight male predominance. Histologically, CPAM are composed of disorganized cysts lined with ciliated cuboidal or columnar epithelium. Involvement is generally unilobar with a slight predilection for the lower lobes, with right and left sides affected equally. Typically, they have normal pulmonary arterial and venous blood supply and communicate with the tracheobronchial tree.

Clinical Features. In the antenatal and immediate postnatal period, the predominant physiologic consequences of a CPAM result from compression of the mediastinum and adjacent normal lung by the mass lesion. Because of the mass effect, polyhydraminos, mediastinal shift, pleural effusions, and fetal hydrops can be identified in the antenatal period. In contrast, the mortality approaches zero for a fetus with a CPAM but without hydrops. In addition, spontaneous resolution in utero of an antenatally diagnosed CPAM is reported to occur in up to 15% of patients. Following delivery, the major risk to the newborn with a CPAM is respiratory distress as a consequence of compression of the adjacent normal lung and mediastinum by the CPAM. Far more commonly, infants born with a CPAM either remain asymptomatic or have persistent mildly increased work in breathing that interferes with feeding.

Diagnosis. *Prenatal diagnosis* by sonography is relatively common, which demonstrates an echogenic pulmonary mass with displacement of adjacent structures. Using antenatal sonography, CPAM is defined as either macrocystic (greater than 5-mm diameter cysts) or microcystic (solid or less than 5-mm diameter cysts), but the natural history of CPAM is determined more by overall size and degree of compression of adjacent structure than by their gross appearance. The prenatal differential diagnosis includes congenital diaphragmatic hernia, pulmonary sequestration, and bronchogenic cyst. Sonographic findings associated with CPAM include polyhydramnios, mediastinal shift, pleural effusions, and fetal hydrops. *Postnatal diagnosis* can often be made by plain chest radiographs; however, in a stable patient, CT scans are often helpful to define anatomy and particularly to identify aberrant systemic blood supply that is more suggestive of a pulmonary sequestration. A CT scan is more accurate than plain chest radiography in confirming complete resolution of an antenatally diagnosed congenital pulmonary lesion that is not seen on postnatal plain chest radiography (Fig. 8.25).



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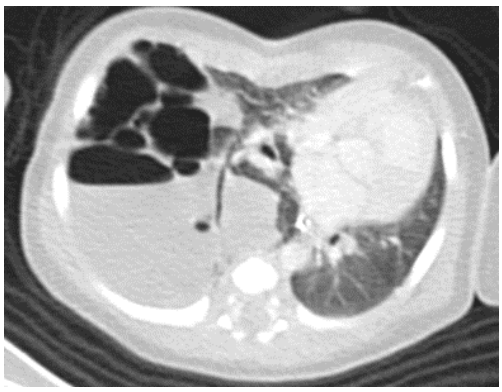

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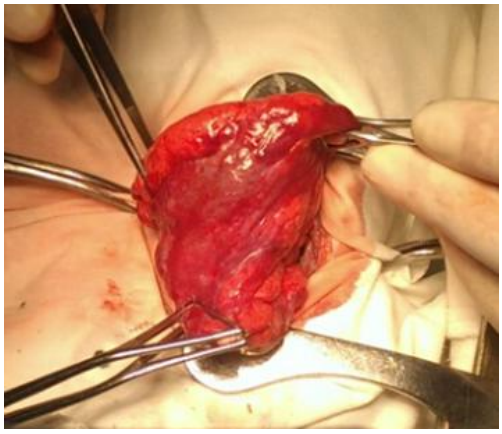

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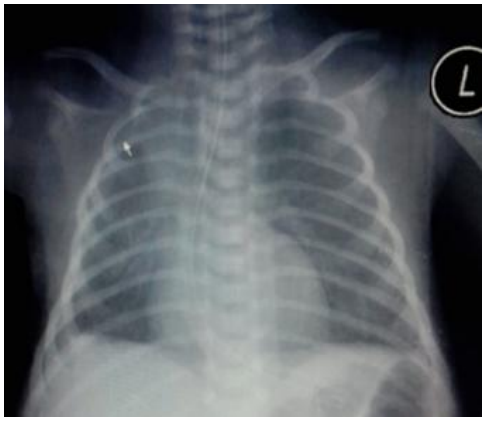


D

Fig. 8.25. Diagnostic tests and operational photo of newborn baby with tension polycystic lower lobe of the right lung. A – plain chest radiograph (age 3 days): the right lung is of increased volume, hyperinflated, has multiple cellular foci transparency, forms mediastinal hernia; mediastinum is shifted to the left. B – CT with contrast enhancement (age 4 days), axial scan at the level of s6 bronchus branching (arrow): multiple cavities in right lower lobe partially filled with fluid. C – operative photo: right lower lobe with multiple cysts and islets of normal parenchyma (age 6 days), operation – right lower lobectomy. D – plain chest radiograph 2 weeks after surgery: right upper and middle lobes expanded to fulfill the hemithorax, mediastinum is at near normal position.

Management. *Fetal therapy.* For congenital cystic pulmonary lesions, it is clear from the literature that provided the fetus does not experience hydrops or physiologic distress, treatment should be expectant management with term delivery and postnatal evaluation. Administration maternal steroids to any affected fetus less than the estimated gestational age of 34 weeks to help fetal lung maturation is reasonable.In fetuses with ultrasonographic evidence of deterioration due to fetal hydrops, intervention should be considered. If the fetus is in the second trimester, several options exist for fetal intervention. Ultrasound-guided intrauterine thoracoamniotic shunting for a macrocystic CPAM with a large cyst has the best outcome with the lowest fetal and maternal risk. If hydrops develops during the third trimester, an EXIT procedure with thoracotomy and lobectomy using placental bypass can permit safe resection and avoid respiratory collapse.

Postnatal therapy. Prompt surgical resection should be performed in any symptomatic newborn. In those patients who remain asymptomatic in the newborn period, delaying resection until infancy is reasonable and allows somatic growth that may facilitate the ease of pulmonary resection. Asymptomatic but persistent CPAM should be resected during infancy to prevent complications of recurrent infections or malignant degeneration. Resection often includes formal lobectomy; however, for small CPAM, nonanatomical resection is reasonable. Traditionally,

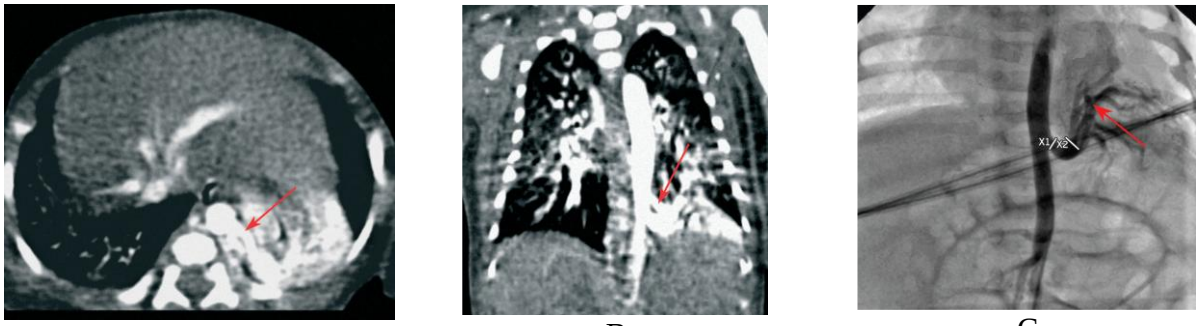
resection was performed through open thoracotomy but recent reports have demonstrated excellent outcomes for minimally invasive pulmonary resection.

Bronchopulmonary Sequestrations

Bronchopulmonary sequestrations are defined as either an intrathoracic or subdiaphragmatic mass of nonfunctional pulmonary tissue that lacks communication with the tracheo-bronchial tree of the normal lung. Histologically, pulmonary sequestrations demonstrate immature lung development, often resembling more peripheral lung parenchyma. Intrathoracic sequestrations are further categorized as either extralobar or intralobar, depending on whether the lesion is invested by its own pleura or that of the adjacent normal lobe, respectively. The arterial blood supply is anomalous, most commonly arising from the descending thoracic aorta. Venous drainage for intralobar sequestrations is most commonly via the pulmonary veins, whereas venous drainage for extralobar sequestrations is typically systemic via either the azygous or hemiazygous veins, inferior vena cava, or directly into the atrium.

Clinical features**.** Both intralobar and extralobar sequestrations can present in the newborn period with either respiratory distress due to mass effect or congestive heart failure because of arteriovenous shunting within the sequestration. Most intralobar sequestrations present later in childhood due to recurrent pulmonary infections from inadequate tracheobronchial drainage, or less commonly with hemorrhage or hemoptysis. The abnormally developed lung tissue is ineffective in gas exchange and therefore is not of benefit to the patient.

The diagnosis of lung sequestration is made by angiography or contrast enhanced chest CT (Fig. 8.26).



A B C

Fig. 8.26. Contrast enhanced chest CT (A, B) and angiography (C) in newborn patients with intralobar sequestration of the left lung (age 7 days and 1 month respectively). A — CT, axial scan. B — frontal reconstruction. Arrows indicate aberrant vessels originating from the thoracic aorta and extending to the left lung sequester. B — angiography. The big arterial vessel (arrow) originates from the abdominal aorta and provides blood supply of the sequester of the left lung

Management of pulmonary sequestrations is surgical. The goal of surgical resection is to remove only the abnormal portion of lung with obvious division of the aberrant artery supplying the sequestration. Care should be given to the division of this artery because it is big in diameter, originates directly from thoracic aorta, has high (the same with aorta) blood pressure inside and may contract after division hiding within the mediastinum and causing uncontrolled arterial bleeding been inadequately ligated. For intralobar sequestration, particularly if previously infected, this often necessitates lobar resection.

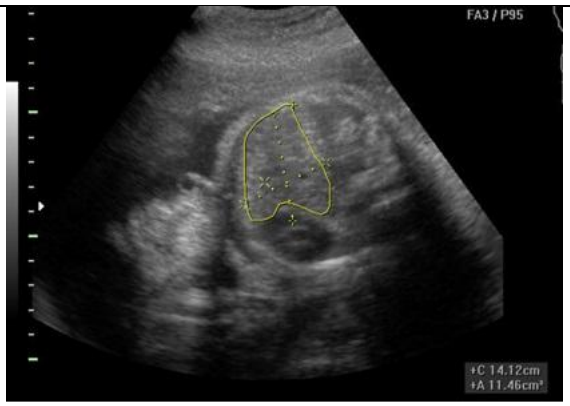
Congenital Diaphragmatic Hernia

Congenital diaphragmatic hernia (CDH) is a developmental abnormality of the diaphragm that allows the abdominal viscera to enter the thoracic cavity. It occurs with an overall incidence ranging from 1 in 2000 to 4000 live births. There is a slight male preponderance with a ratio of 1.5 : 1. Most CDHs occur through a posterolateral defect (Bochdalek hernia), with 90% of these on the left side, 10% on the right and less than 1% bilateral. Less than 5% of CDHs are located anteriorly (Morgagni hernia). A number of syndromes are associated with CDH. In some syndromes, such as Fryns syndrome and Donnai-Barrow syndrome, CDH is present in a high

percentage of affected individuals. Others include Beckwith-Wiedemanch, Simpson-Golabi-Behmel, Coffin-Siris, and Denys-Drash syndromes. CDH has also been reported with trisomies 9, 13, 18, 21, and 22.

Embryogenesis of CDH has been described as a failure of the closure of the pleuroperitoneal canal, which occurs during gestational week 8. Consequently, the abdominal viscera herniats into the thorax, which is thought to cause pulmonary hypoplasia by compression of the growing lung. Furthermore, a toxicological nitrofen model of CDH has shown that abnormalities in the contralateral lung as well as the ipsilateral side are present even before the diaphragm starts to develop.

The *prenatal diagnosis* of CDH has been greatly facilitated by the improved technology and interpretation of prenatal imaging studies (Fig. 8.27). CDH is readily identified on routine prenatal ultrasonography as early as 18 weeks of gestational age. Displacement of the mediastinum, the absence of a stomach bubble in the abdomen, and the presence of abdominal organs in the chest are the signs of fetal diaphragmatic hernia. A right-sided CDH is more difficult to identify because the echogenicity of the fetal liver is similar to that of the lung. Identifying the gallbladder in the fetal chest may be the most reliable sign in these cases. Prenatal magnetic resonance imaging has been shown to be effective in confirming the diagnosis of CDH and detecting additional information that may affect prognosis. The two most commonly used prenatal predictors of postnatal morbidity in infants with CDH are liver position and lung-to-head ratio (LHR). An intrathoracic location of the liver and a low LHR (<1.0) predicts a poor prognosis.



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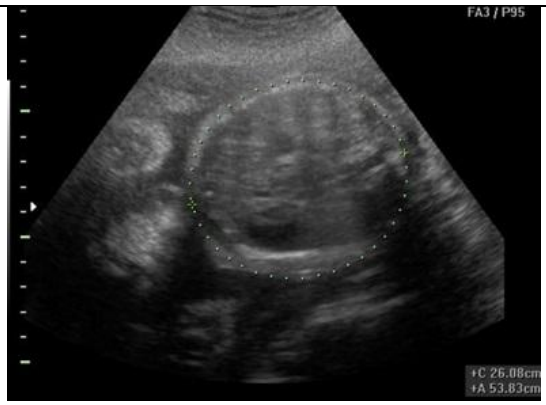

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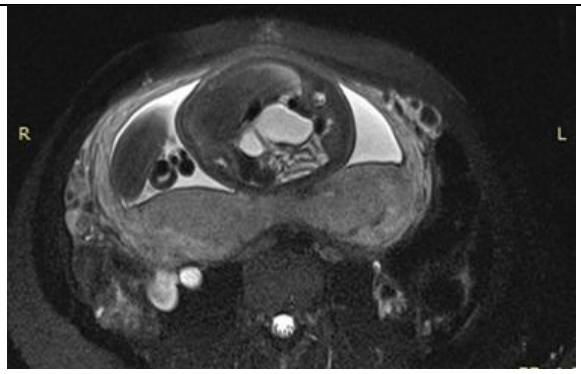

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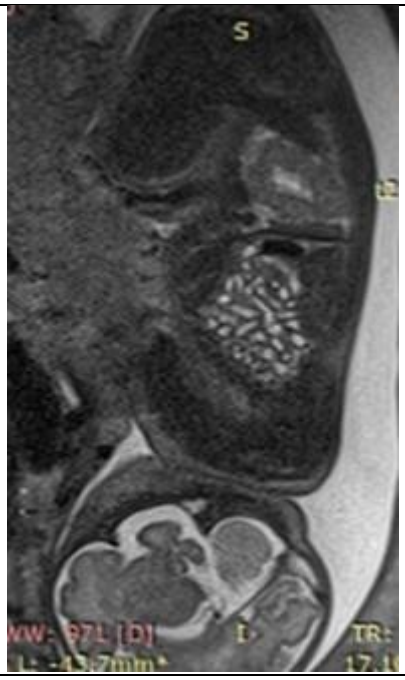
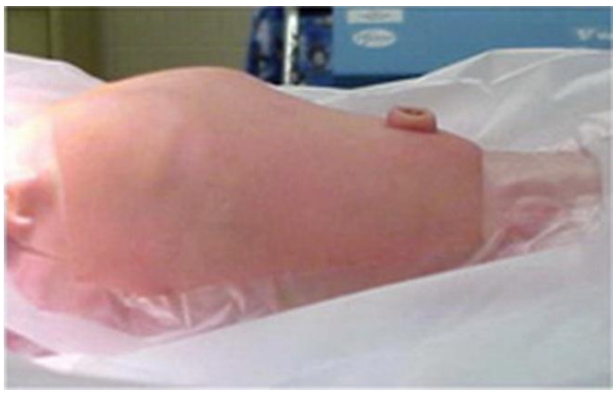


	D
Fig. 8.27. Prenatal diagnosis of CDH: A, B – measuring of lung-to-head ratio on prenatal ultrasonography. C, D, - fetal magnetic resonance imaging showing herniation of liver and intestine into the right hemithirax	

Postnatal presentation and diagnosis. The onset and severity of symptom depends on the amount of abdominal viscera in the chest and the degree of pulmonary hypoplasia. The most severely affected infants present with respiratory distress at birth. Other infants with CDH develop cyanosis, tachypnea, and grunting respirations within minutes or hours after birth. Physical examination reveals a scaphoid abdomen (Fig. 8.28), an increased anteroposterior diameter of the thorax, and mediastinal shift. Breath sounds are absent on the affected side. The definitive diagnosis of CDH is made postnatally by plain radiography of the chest and abdomen by demonstration of air-filled loops of the bowel in the chest and a paucity of gas in the abdomen (Figs. 8.29). The diaphragmatic margin is absent, there is a mediastinal shift to the opposite side, and only a small portion of lung may be seen on the ipsilateral side.



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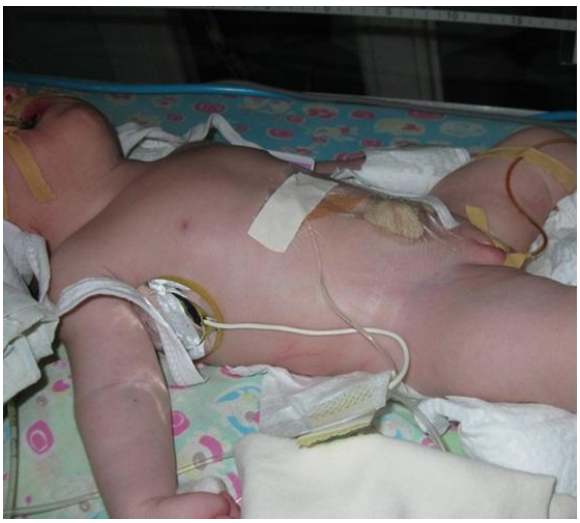
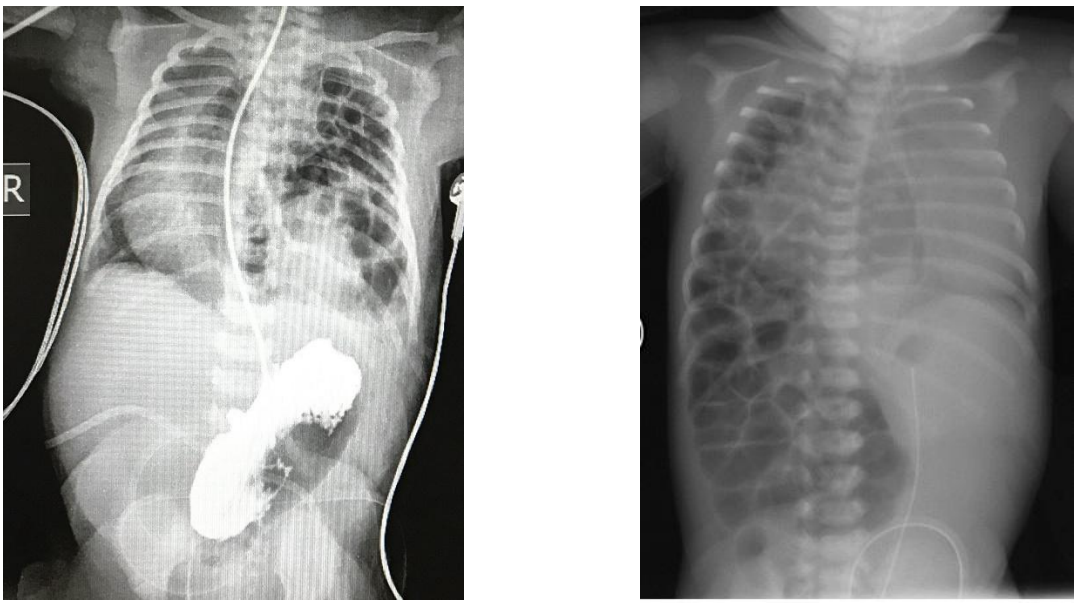


B

Fig. 8.28. Photos of the newborns (A, B) with CDH showing scaphoid abdomen and increased thorax in comparison with abdomen

Although most CDH infants present in the first 24 h of life, 10–20% of the affected infants present later. The symptoms and signs of those patients are nonspecific and include recurrent chest infections, vomiting, abdominal pain, diarrhea, anorexia, failure to thrive, or an abdominal chest X-ray in an asymptomatic patient. Some children present acutely with volvulus or strangulation or acute respiratory distress. Chest X-ray with an in situ nasogastric tube is reliable for the diagnosis, however contrast insertion through NG tube facilitates defining the position of the stomach and help in preoperative planning.



A B

Fig. 8.29. A - Plain chest radiograph in a newborn showing left-sided CDH. The air-filled loops of intestine are I the left hemithorax, mediastinum is shifted to the right. B - X-ray of the chest and upper abdomen showing right-sided diaphragmatic hernia

Differential diagnosis includes cystic lung disease and mediastinal cystic lesions (e.g. cystic teratoma, neuroenteric, bronchogenic, and thymic cysts). The diagnosis of CDH can also be confused with other congenital thoracic conditions, such as eventration of the diaphragm, anterior diaphragmatic hernia of Morgagni, congenital esophageal hiatal hernia, and primary agenesis of the lung.

Management. *Preoperative management.* An infant with respiratory distress requires endotracheal ventilatory support. Bag-and-mask ventilation should be avoided to minimize gaseous distention of the stomach and intestines, which would further compromise lung function. Prompt endotracheal intubation and limitation of ventilation pressures are essential. The concept of "gentle" ventilation and permissive hypercapnia was proposed to minimize barotraumas by strictly limiting the peak inflation pressure. A nasogastric tube is passed and placed on suction and a chest radiograph is done. The basic tenets of management for the infant with CDH are based on the support of the cardiorespiratory system. First, the patient is placed on monitoring equipment, intravenous access established and pre- and postductal oxygen saturations recorded with pulse oximetry. Postductal saturations provide caregivers with an indication of the severity of pulmonary hypertension and right-to-left shunting. High-frequency oscillatory ventilation (HFOV) is a mode of ventilation that can be effectively used in CDH infants to maintain preductal oxygenation while avoiding hyperventilation and barotraumas. Infants who fail to respond to optimal therapy may be placed on extracorporal membrane oxygenation (ECMO). Inotropic support to improve the circulatory status of the patient may also be considered for patients with low mean arterial pressure and derangements in systemic perfusion. Excessive amounts of intravenous fluid should also be avoided because pulmonary edema with worsening pulmonary hypertension and gas exchange may ensue.

Surgical correction. The philosophy regarding the timing of surgical repair has generally shifted from emergent repair to a delayed approach after stabilization of the infant. Although the abdominal approach is widespread among surgeons as it offers good exposure, easy reduction of the abdominal viscera, and recognition and correction of associated gastrointestinal anomalies, we have recently proposed approach through thoracotomy in both right and left-sided CDH. This is based on the concept of thoracalization of abdominal cavity with patch diaphragmatic defect repair. This allows keeping dome-shaped diaphragm, preventing the increase of intraabdominal

pressure and decreasing the big volume of “empty” hemithorax on affected side thus protecting the hypoplastic lung from hyperinflation and barotrauma.

Small diaphragmatic defects can be sutured by direct sutures of the edges of the defect after the contents of hernia are gently reduced in the abdomen.

If the defect is large, it may not be possible to repair it using direct sutures. Various techniques have been described and include the use of prerenal fascia, rib structures, the lastissimus dorsi muscle, rotational muscle flaps from the thoraco-abdominal wall, and prosthetic patches. The operations involving muscle flaps are too long and complex for critically ill patients and can lead to unsightly chest deformities. Prosthetic materials have been advocated for repair of large defects. The most commonly used prosthetic material is PTFE-graft (Fig. 8.30).

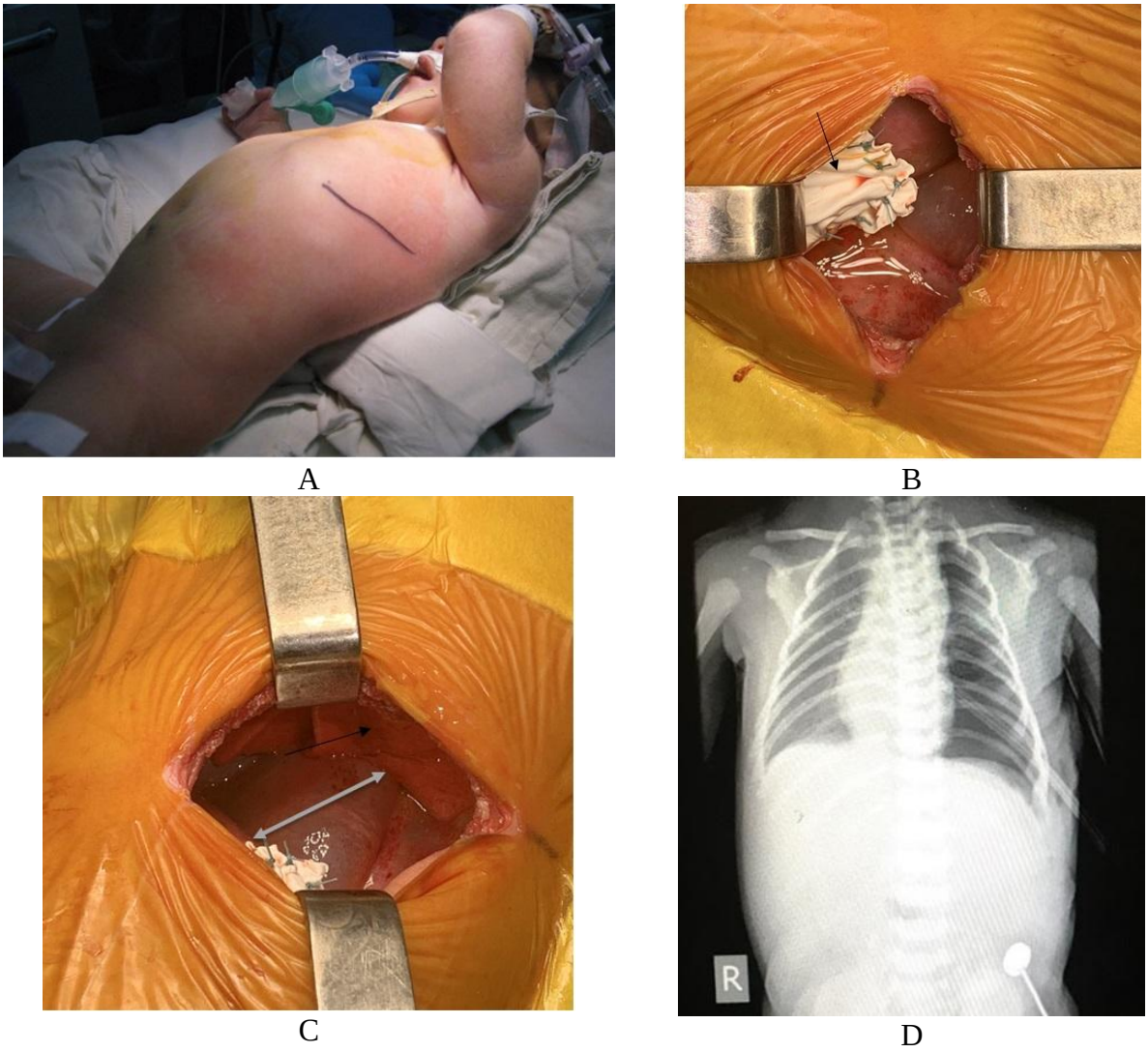


Fig. 8.30. A – thoracotomy as a surgical approach; B – correction of the defect of diaphragm with PTFE patch (arrow); C – “empty” hemithorax and hypoplastic left lung; D – postoperative chest X-ray

Francois Becmeur was the first who used thoracoscopic approach in CDH correction in 2001. This technique has becoming more popular nowadays even in newborns. Stable patients are selected for this surgery and it is recommended to use it for small defects closure without patches (Fig. 8.31).

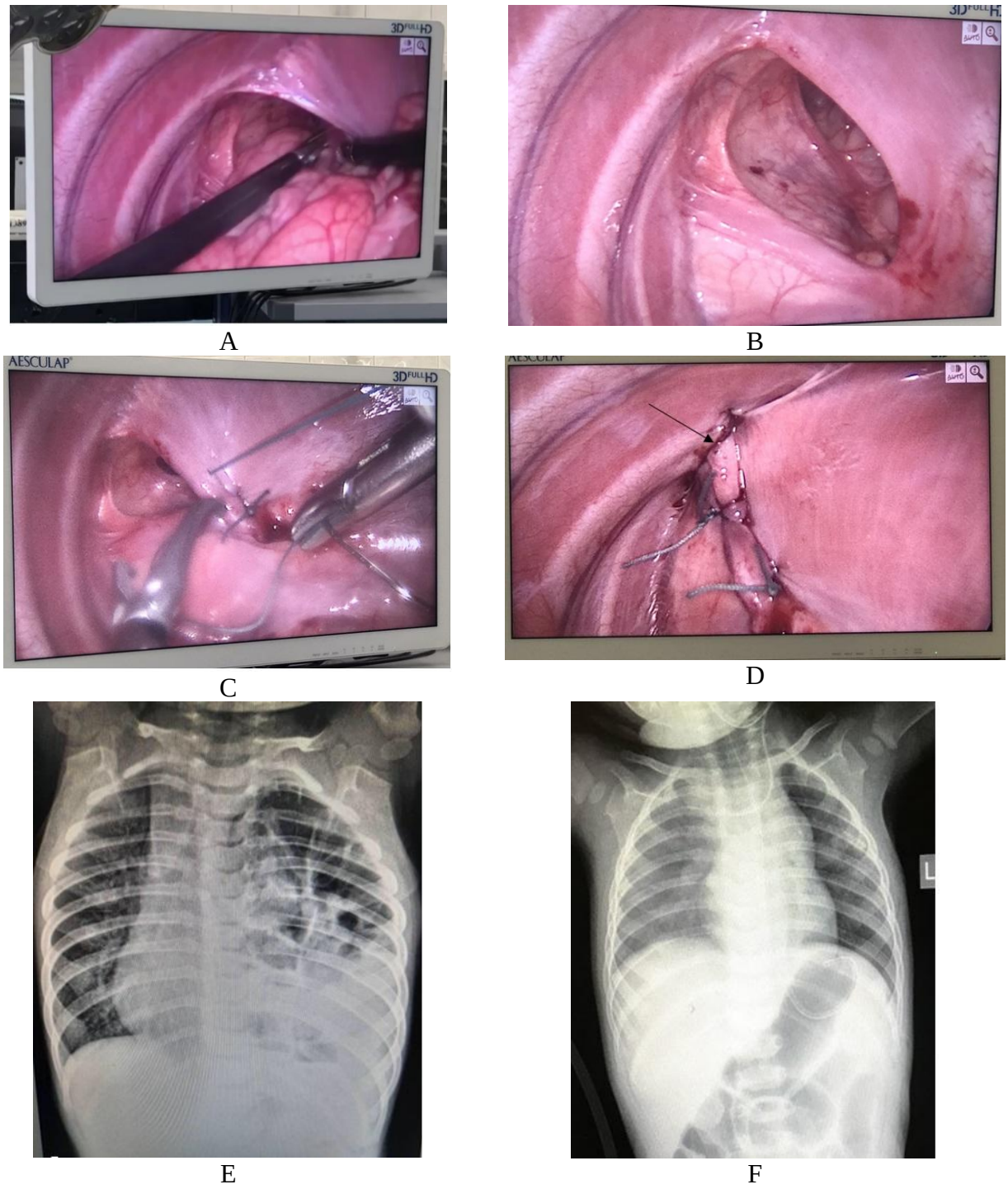


Fig. 8.31. Thoracoscopic CDH closure: A – reduction of herniated intestine, B – defect in the diaphragm, C, D – suturing of the defect using non-absorbable threads and pericostal sutures, E – preoperative chest X-ray, F – posop radiograph

Postoperative care. Ventilatory support should be continued postoperatively. The vital signs are monitored closely with regular blood gas analyses and monitoring of preductal and postductal oxygenation. Some infants show improvement in oxygenation in the immediate postoperative period, the so-called honeymoon period, but deteriorate 6–24 h later. This deterioration is due to pulmonary hypertension and persistent fetal circulation with an increase in pulmonary artery resistance, elevated pulmonary artery pressure, and right-to-left ductal and preductal shunting leading to hypoxemia.

Despite advances in care, intervention, and technology, CDH still carries a high mortality rate due to associated anomalies, particularly cardiac, chromosomal, and neurologic, with prematurity and low birth weight also affecting prognosis. Several recent reviews have documented an improved overall outcome for infants with CDH compared to 20 years ago with survival rates as high as 90%.

Fetal interventions. A clinical trial evaluating the efficacy of open surgical repair for fetuses with CDH demonstrated that the outcome of these infants was no better than infants cared for by conventional means. Thus, the optimism for open fetal surgical repair for CDH diminished, and other modalities were investigated.

An ongoing, prospective study from Europe (FETO group) evaluating fetal tracheal occlusion by inflatable balloon has presented preliminary results that are encouraging, but its limitations are the lack of a randomized design as well as standardized postnatal care.

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Tests for initial level of knowledge, keys for tests

1. What is the cause of stridor?
   A. Narrowing of the airway
   B. Shift of the mediastinum

C. Surfactant deficiency
D. Tension pneumothorax
E. Aspiration

2. What congenital malformation presents with stridor?
    A. Cyst of the lung
    B. Pulmonary hypoplasia
    C. Congenital tracheal stenosis
    D. Extralobar pulmonary sequestration
    E. Esophageal atresia
3. Lesions listed below present with stridor except for:
    A. Congenital tracheal stenosis
    B. Laryngomalacia
    C. Double aortic arch
    D. Subglottic hemangioma
    E. Congenital isolated tracheoesophageal fistula
4. What type of tracheal stenosis is caused by vascular ring?
    A. Congenital tracheal stenosis
    B. Compression tracheal stenosis
    C. Functional tracheal stenosis
    D. Intrinsic tracheal stenosisAcquired tracheal stenosis
5. What are the characteristic features of tracheomalacia?
    A. Increase of entire tracheal perimeter, widening of tracheal membrane, inward prolapse of tracheal walls
    B. Fixed narrowing of the trachea, thickening of tracheal wall
    C. Pulsatile anterior tracheal compression, mucus retention
    D. Narrowing of the trachea, absence of tracheal membrane
    E. Fistula between trachea and esophagus
6. What congenital malformation appears with contralateral mediastinal shift?
    A. Aplasia of the lung
    B. Pulmonary sequestration
    C. Pulmonary artery sling
    D. Congenital emphysema
    E. Esophageal atresia
7. All entities of listed below are the types of congenital tracheal stenosis, except for:
    A. Subglottic hemangioma
    B. Generalized hypoplasia of the trachea
    C. Funnel-shaped tracheal stenosis
    D. Segmental tracheal stenosis
    E. “Bridging bronchus” stenosis
8. X-ray sign of congenital emphysema is:
    A. Ipsilateral shift of mediastinum
    B. Increased lung transparency
    C. Opacity of the affected lung
    D. Multiple air-filled loops of intestine in the hemithorax
    E. Free air in the pleural cavity on affected side
9. What are the X-ray signs of esophageal atresia with distal tracheoesophageal fistula?
    A. Coiled nasogastric tube in the oral esophageal segment and air-filled loops of intestine in the abdomen
    B. Coiled nasogastric tube in the oral esophageal segment and gasless stomach and intestine
    C. The fistula cannot be visualized by means of X-ray study

D. Bronchogram after insertion of water-soluble contrast into the oral segment of esophagus
E. Direct visualization of fistula

10. What diagnostic modality helps to differentiate the lung aplasia and pulmonary agenesis?
    A. Plain chest radiography
    B. Perfusion scintigraphy
    C. Bronchscopy
    D. Ultrasonography of lungs
    E. Digital subtraction angiography

Keys for tests

1. A
2. C
3. E
4. B
5. A
6. D
7. A
8. B
9. A
10. C

Tests for final level of knowledge, keys for tests

1. 3 months old infant operated on for esophageal atresia with distal tracheoesophageal fistula presented with stridor, neck hyperextention and apneic spells. What diagnostic modality is essential for establishing the diagnosis?
    A. Plain chest radiography
    B. Chest CT
    C. Tracheobronchscopy
    D. Esophagography
    E. Digital subtraction angiography
2. Parents of 6-months old infant note noisy breathing of the baby, neck hyperextention, dysphagia and recurrent bronchitis treated twice in the children hospital. What congenital malformation is the most likely to present with these symptoms?
    A. Double aortic arch
    B. Congenital isolated tracheoesophageal fistula
    C. Pulmonary agenesis
    D. Pulmonary sequestration
    E. Cyst of the lung
3. Parents of 6-months old infant note noisy breathing of the baby, neck hyperextention, dysphagia and recurrent bronchitis treated twice in the children hospital. What diagnostic study is the most simple and sensitive among listed below?
    A. Plain chest radiography
    B. Chest CT
    C. Tracheobronchscopy
    D. Esophagography
    E. Digital subtraction angiography
4. Newborn baby was diagnosed as having esophageal atresia with distal tracheoesophageal fistula at birth. There are no other malformations. What tactics will you concern?
    A. Observation
    B. Surgery including division of fistula and primary esophageal anastomosis
    C. Surgery including division of fistula and delayed esophageal anastomosis
    D. Surgery including gastrostomy and cervical esophagostomy
    E. Surgery including esophagocoloplasty

5. Newborn baby in 6 hours after birth developed respiratory disorders with dyspnea and cyanosis. Repeated physical examination revealed gradual shift of the heart to the right side. Left half of the chest is slightly enlarged, percussion shows tympanitis. Auscultation is suspicious for peristaltic sounds in the chest. Plain film showed mediastinal shift to the right and multiple air-filled cavities of different sizes within the left hemithorax. What is the preliminary diagnosis?
    A. Congenital left-sided emphysema
    B. Aplasia of the right lung
    C. Congenital left-sided diaphragmatic hernia
    D. Polycystic left lung
    E. Intralobar sequestration of the left lung
6. What is the operation of choice in patients with congenital left-sided emphysema involving apical segments of the lung?
    A. Left upper lobectomy
    B. Left lower lobectomy
    C. Left pulmonectomy
    D. Resection of lingual segments of the left lung
    E. Segmental resection of S1-S3 of the left lung
7. What congenital malformation should be included in differential diagnosis of right lung aplasia to prevent life-threatening surgery?
    A. Congenital left-sided emphysema
    B. Congenital diaphragmatic hernia
    C. Esophageal atresia
    D. Tension pneumothorax
    E. Bronchogenic cyst of mediastinum
8. You are caring for 4 months old infant presenting with respiratory disorders. Plain chest film showed shift of mediastinum to the right side, opacity in the right hemithorax and increased transparency in the left lung, which is hyperinflated and herniated into the right hemithorax. What study should you administer to differentiate between congenital emphysema and lung aplasia?
    A. Plain chest radiography
    B. Chest CT with i/v contrast enhacement
    C. Tracheobronchscopy
    D. Esophagography
    E. Echocardiograhy

Keys for tests

1. **C**
2. **A**
3. **D**
4. **B**
5. **C**
6. **E**
7. **A**
8. **B**

Tasks for final level of knowledge

1. Newborn baby in 6 hours after birth developed respiratory disorders with dyspnea and cyanosis. Repeated physical examination revealed gradual shift of the heart to the right side. Left half of the chest is slightly enlarged, percussion shows tympanitis. Auscultation is suspicious for peristaltic sounds in the chest. Abdomen is scaphoid. Plain film showed mediastinal shift to the right and multiple air-filled cavities of different sizes within the left hemithorax.
    1. What is the preliminary diagnosis?

2. What study should be done to differentiate this lesion from polycystic left lung?
3. What tactics of management should you propose?

Key Answer

1. Congenital left-sided diaphragmatic hernia.
2. Contrast GI series
3. Surgical correction after stabilization of the patient

2. 1 months old infant operated on for esophageal atresia with distal tracheoesophageal fistula is still unable to be extubated. However, spontaneous breathing is adequate through endotracheal tube. If extubated baby develops expiratory stridor, suprasternal, substernal and intercostal retractions, wheezing, cyanosis.
    1. What lesion is the most likely to appear with such picture?
    2. What diagnostic modality is essential for establishing the diagnosis?
    3. What is the operation of choice for this patient?

Key Answer

1. Primary tracheomalacia associated with esophageal atresia and tracheoesophageal fistula.
2. Tracheobronchoscopy on spontaneous breathing.
3. Anterior aortopexy

4. Parents of 6-months old infant note noisy breathing of the baby, dyspnea, chest retractions, neck hyperextention, dysphagia and recurrent bronchitis treated twice in the children hospital. The plain X-ray showed prominence of the mediastinum to the right side, bilateral mild emphysema. On lateral esophagography the posterior round-shaped indentation of esophagus at aortal level was revealed.
    1. What diagnosis is the most likely in this patient?
    2. What study is the most optimal to verify the diagnosis?
    3. What surgery is indicated in this patient?

Key Answer

1. Vascular ring, double aortic arch.
2. Digital subtraction angiography or contrast enhanced chest CT.
3. Division of vascular ring including division of ductal ligament and lesser aortic arch through left thoracotomy
4.

Materials for self-study of the students

Main tasks	**Notes (instruction)**
Repeat: Anatomy of the airway, lungs and pleura Physiology of respiratory system Pathogenesis of respiratory failure	To sketch out the anatomy of airway and lungs To represent the methods of diagnosis of congenital malformations of lungs and airway
Study: Pathogenesis of airway stenosis, lung aplasia, congenital emphysema, congenital diaphragmatic hernia The diagnostic possibilities of CT, angiography, MRI, ultrasonography, tracheobronchoscopy in infants and children	To make differential diagnosis of complete tracheal rings , compression and functional airway stenoses, lung aplasia and congenital emphysema, congenital and acquired lung lesions. To make the indications for surgical treatment in children with congenital malformations of respiratory system. To know the modern diagnostic modalities. To know the advantages and disadvantages of open and thoracoscopic procedures

THEME 9
CONGENITAL GASTROINTESTINAL ABNORMALITIES. ANORECTAL ABNORMALITIES.

Overview

Most of the congenital gastrointestinal (GI) anomalies result in some or the other type of intestinal obstruction, frequently manifesting with feeding difficulties, abdominal distention, and emesis at birth or within 1 or 2 days. Some congenital GI malformations, such as malrotation, have a very good outcome, whereas others, have a poor outcome.

A common type of anomaly is atresia, in which a segment of the GI tract fails to form or develop normally. The most common type is esophageal atresia, followed by atresia in the jejunoileal region and in the duodenum.

Immediate management includes bowel decompression (by continuous nasogastric suction to prevent emesis, which can lead to aspiration pneumonia or further abdominal distention with respiratory distress) and referral to a center for neonatal surgery. Also vital are maintenance of body temperature, prevention of hypoglycemia with intravenous 10% dextrose and electrolytes, and prevention or treatment of acidosis and infections so that the infant is in optimal condition for surgery.

Educational aims:

The aim of this chapter is to provide help in performing early diagnosis and management of children with congenital gastrointestinal abnormalities.

1. To analyze anatomic-morphological peculiarities of the abdominal cavity in children of different age groups.
2. To learn the relevant anatomy of the gastrointestinal tract and abdominal cavity in children.
3. To analyze peculiarities of the classic clinical manifestation of the congenital gastrointestinal abnormalities.
4. To learn features of clinical presentations of congenital gastrointestinal abnormalities in newborns.
5. To analyze lab studies and imaging studies in children with congenital gastrointestinal abnormalities.
6. To learn the differential diagnosis of different congenital gastrointestinal abnormalities.
7. To know the complications of congenital gastrointestinal abnormalities.
8. To know different surgical technique under congenital gastrointestinal abnormalities.
9. To master the preoperative management in children with congenital gastrointestinal abnormalities.
10. To Analyze the advantages and disadvantages of laparoscopic techniques under congenital gastrointestinal abnormalities.
11. To know the postoperative complications after surgery of congenital gastrointestinal abnormalities.
12. To master the postoperative management after congenital gastrointestinal abnormalities.
13. Look for the Anatomic location of anus in neonates.
14. Look for the evidence of pyloric stenosis on the abdominal wall.
15. Look for the signs of abdominal distension.
16. Must be able to define the dehydration stage in neonate and calculate the liquid and electrolyte volumes for rehydration.
17. Must be able to do the physical exam of children.
18. Must be able to evaluate the quantity and quality of meconium in neonates.
19. Must be able to insert nasogastric tube.
20. Must be able to interpret the results of abdominal x-ray and ultrasound diagnosis.

21. Must be able to perform the enema before the irrigography.

4. Basic skills and knowledge, necessary to know: (interdisciplinary integration)

Subjects	Skills obtained
1. Anatomy, topographic anatomy.	Palpation of different parts of intestines, knowledge of peculiarities of abdominal cavity in neonates.
2. Physiopathology.	Clinical interpretation of laboratory analyses.
3. Morbid anatomy.	To compare morphological changes, peculiar of Congenital gastrointestinal abnormalities. Anorectal abnormalities
4.Microbiology.	To master technique of collecting material for investigation. To interpret the results.
5. Pediatric diseases propaedeutics.	To collect complaints, medical history, to examine the child, to use additional methods of examination at abdominal cavity diseases.
6. Topographic anatomy and surgery.	To master methodic (schemes) of operative access and methods of operative intervention on abdominal cavity organs in neonates and children of different age groups.
7. General surgery.	To display methods of patient preparation for treatment, diagnostics and operation.
8. Hospital surgery.	To perform differential diagnostics, to ground methods of diagnostics and treatment of Congenital gastrointestinal abnormalities. Anorectal abnormalities in children.
9. Clinical pharmacology.	To classify medications necessary for treatment of acute surgical diseases, remember about pharma kinetics and methods of action of medications in neonates and children of different age groups.

4. Tasks for individual self-preparation for the lesson.

4.1. The list of key terms and characteristics a student must know:

Term	***Definition***
Hypertrophic pyloric stenosis	is narrowing of the opening from the stomach to the first part of the small intestine known as the duodenum, due to enlargement of the muscle surrounding this opening (the pylorus, meaning "gate"), which spasms when the stomach empties.
“Bull’s eye” sign	characteristic appearance of pyloric stenosis on ultrasound on cross section of the pyloric channel
Double-bubble sign	An x-ray gas shadows essentially pathognomonic for duodenal obstruction
Intestinal atresia	malformation where there is a narrowing or absence of a portion of the intestine
Hirschsprung’s disease	is a form of megacolon that occurs when part or all of the large intestine or antecedent parts of the

	gastrointestinal tract have no ganglion cells and therefore cannot function
Acetylcholinesterase (AChE) staining techniques	investigation of choice to evaluate suction biopsies to show an increased activity in the parasympathetic nerves of the affected zone as well as neurofibrils within the lamina propria and muscularis mucosa
VACTERL association	Association of vertebral and spinal cord, Anorectal, Cardiac, Tracheoesophageal, Renal and other urinary tract, Limb anomalies

CONTENT

Hypertrophic pyloric stenosis

Hypertrophic pyloric stenosis (HPS) is a common surgical cause of vomiting in infants. The reported incidence of HPS is 1–4 per 1,000 live births. There is a male-to-female ratio ranging from 2.5:1 to 5.5:1. Gastrojejunostomy was used to treat this disease until 1912, when extramucosal muscle-splitting pyloromyotomy was described by Ramstedt. This procedure has dramatically changed the outcome of infants with HPS

Etiology**.** Despite the frequency of pyloric stenosis, the etiology remains unclear. Genetic predisposition acting in conjunction with environmental factors is the most widely accepted explanation. The typical clinical picture develops at the age of 3-6 weeks. Cases at the age of 1 week to 3 months are described

Pathophysiology

Pyloric stenosis is characterized by hypertrophy of the pyloric musculature, leading to a mechanical obstruction of the gastric outlet in the affected infant. Thus, hypertrophied pyloric antral muscle fibers protrude distally into the duodenal lumen, producing a reflection of duodenal mucosa. Infants with a diagnosis of pyloric stenosis will show characteristically low chloride and hydrogen ions as measured in the serum leads to alkalosis.

Clinical manifestation

• Nonbilious vomiting is usually forceful and “projectile”.
• The infant is hungry after vomiting and eager to feed, only to vomit again.
• Weight loss occurs in severe cases.
• Signs of dehydration present in cases of repeated vomiting.
• Scaphoid abdomen especially noted after recent vomiting.
• Visible peristalsis may be observed in the upper abdomen, usually moving from the left hypochondrium towards the right side like “sand watch” symptom.
• A palpable mass is present in the right upper quadrant (90% in experienced hands); this is best appreciated while the infant is being fed with clear fluid.

Clinical Diagnosis

The clinical picture we can observe weight loss and dehydration coupled with an insatiable appetite lead to a characteristic face, with a furrowed brow, wrinkled appearance. In some infants, the distended stomach may be identifiable in the epigastrium, with active peristaltic visible through thin abdominal wall as sand watch symptom. An ovoid mass, commonly referred to as an “olive”, is palpable in the epigastrium or the right upper quadrant.

Ultrasound signs of congenital pylorostenosis: This would normally confirm the presence of a pyloric “mass”. The characteristic appearance of pyloric stenosis on ultrasound is that of a “doughnut” or “bull’s eye”.

- Pyloric muscle thickness over 3 mm
- Length of pyloric canal over 14 mm in children under 30 days.

Upper Gastrointestinal Contrast Study

In an occasional case where doubt still persists after ultrasound, an upper gastrointestinal (UGI) contrast X-ray may be done. The UGI series would show a narrow pyloric channel, the so-

called “string sign” and the “shoulder sign”, caused by the impression of the pylorus into the stomach.

Fibrogastroscopy can help in conformation of diagnosis.

Differential Diagnosis

The differential diagnosis of pyloric stenosis includes:

• gastro-esophageal reflux;
• viral enteritis;
• pylorospasm;
• duodenal stenosis/duodenal web;
• raised intracranial pressure.

Preoperative treatment usually lasts 24-48 hours, meticulous care and time should always be taken to correct fluid and electrolyte depletion before any surgical correction includes correction of water-electrolyte balance and metabolic disorders; aspiration of gastric contents for nasogastric decompression.

Surgical procedure: The standard operation is the Ramstedt pyloromyotomy. Classically, the operation has been approached through a right upper quadrant. The lower third of the stomach is then gently elevated using moist gauze to deliver the pyloric mass into the wound, A prolonged incision is made on the pyloric mass in the mid anterior surface through the serosa and superficial muscularis, beginning about 1–2 mm from the pyloroduodenal junction , underlying hypertropic muscle fibers are then divided using blunt dissection, Special care is taken to prevent mucosal perforation, Upward protrusion of the gastric mucosa indicates relief of the obstruction .

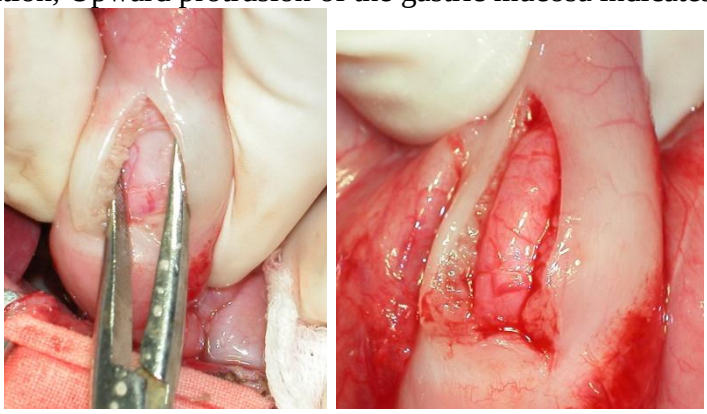


Fig 9.1. Pyloromyotomy.

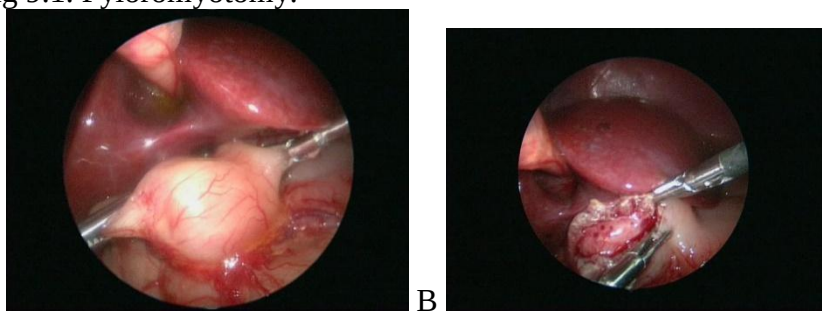
A  B

Fig 9.2. Laparoscopic pyloromyotomy. A – Enlarged pylorus, B – pyloromyotomy.

Laparoscopic pyloromyotomy can be performed with similar efficiency and surgical outcome as traditional open pyloromyotomy. Improved cosmesis and avoidance of wound complications are major benefits of this procedure, and a tendency towards less postoperative emesis is a potential benefit that deserves further investigation.

Postoperative period:

- in uncomplicated cases: Several feeding schedules have been advocated after surgery. Traditional structured feeding regimens as opposed to more rapid initiation and advancing feeding schedules are probably unnecessary, feeding after 6 hours.

after the operation - breast milk or an adapted formula of 5-10 ml after 2 hours, gradually increasing the volume and intervals as per age;

- With complications (perforation of the mucosa) - nasogastric tube and starvation for 24 hours.

Normal feeding as per age can be achieved - in 5-6 days

Postoperative complications

- Perforation of the mucous membrane
- Bleeding.

Outcome

The majority of infants makes a full recovery postoperatively and do not require further medical care.

Duodenal atresia

Duodenal obstruction and jejunoileal atresia rank among the two most common causes of intestinal obstruction in newborns, duodenal obstruction is reported to be 1 in 5,000–10,000 births according to literature.

Congenital duodenal obstruction may be due to intrinsic or extrinsic lesions. Intrinsic duodenal obstruction may be caused by duodenal atresia, stenosis, and duodenal membrane with or without orifice. Extrinsic duodenal obstruction may be caused by malrotation with Ladd's bands or congenital vascular anomalies (aberrant artery and preduodenal portal vein) or annular pancreas.

Duodenal obstruction usually occurs at or below the ampulla of Vater in 85%.

Aetiology. It has been demonstrated that from gestational weeks 5 to 10, the duodenum is a solid chord. Intrinsic obstructions result from failure of vacuolization and recanalization. An annular pancreas results from fusion of the anterior and posterior anlage, forming a ring of pancreatic tissue that surrounds the second part of the duodenum. Extrinsic obstructions result from a variety of disorders of embryologic development specific to the pathology.

Clinical Presentation.

Prenatal. Duodenal obstruction can be diagnosed by prenatal ultrasound. Duodenal obstruction pres up to gestational week 20 with a double-bubble phenomenon due to the simultaneous distention of the stomach and the first part of the duodenum. a high index of suspicion must be maintained in cases of maternal polyhydramnios.

Postnatal Symptoms and Signs. The most common presenting features are bilious vomiting and feeding intolerance. Dehydration and electrolyte disbalance leads to worsen the condition of newborn. Repeated nonbilious vomiting is seen in cases of supra-ampullary obstruction (20%). Patients with a web or partial stenosis can show clinical symptoms later. Physical signs are nonspecific but can include upper-abdominal distention with scaphoid lower abdomen. Additionally, in the appropriate clinical context, observation of typical Down syndrome features should raise suspicion towards duodenal obstruction as the cause for neonatal intestinal obstruction.

Clinical diagnosis. In tertiary perinatal centers where a prenatal diagnosis has already been established, no further diagnostic work-up is typically necessary. In doubtful cases, a plain abdominal x-ray is the key method for diagnosis. An x-ray showing "double-bubble" symptom is essentially pathognomonic for duodenal obstruction (Figure 9.3). If no double bubble is seen, instillation of 10–15 ml of air immediately prior to a plain abdominal radiograph may help to demonstrate these findings. In cases of stenosis or perforated membranes, air may be seen in the distal gastro intestinal tract. Water-soluble contrast radiography is confirmatory, but it is generally needed only in cases of incomplete obstruction. Radiographic findings of annular pancreas are usually indistinguishable from other forms of duodenal obstruction.

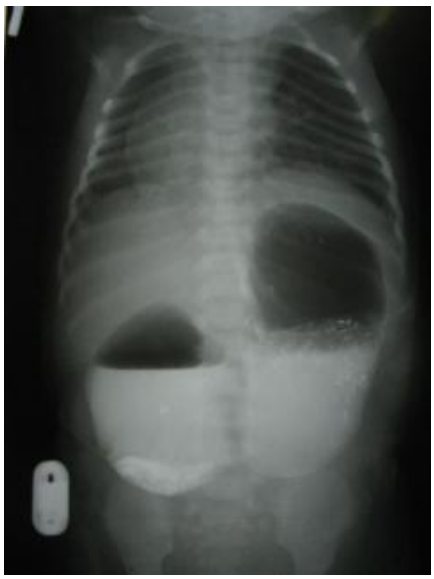


Fig.9.3: “Double-bubble” sign on x-ray with contrast. Note the lack of distal gas.

The most important differential diagnosis is duodenal obstruction due to malrotation, resulting in volvulus of the midgut loop or extrinsic compression related to Ladd´s bands across the duodenum. When no prenatal diagnosis is available, contrast radiography may be helpful to differentiate between these entities and can demonstrate the absence of the normal C-shaped curve of the duodenum or a classic “bird’s-beak” shape secondary to a volvulus. When the diagnosis still remains in doubt, prompt laparotomy is recommended because undiagnosed volvulus can result in gangrene of the entire midgut within hours.

If available, in cases of incomplete obstruction, esophago-gastro-duodeno-scopy can be done to prove the existence of an intrinsic obstructing membrane. An endoscopic approach to membrane resection can be utilized.

Management. Preoperative Care

Initial therapy consists of nasogastric decompression and appropriate replacement of fluid and electrolytes. Most of these newborn patients are premature, so special care must be taken to preserve body heat and to avoid hypoglycemia, especially in cases of very low birthweight, congenital heart disease, and respiratory distress syndrome.

General Intraoperative Considerations

General anesthesia with endotracheal intubation is required. The most approach is transverse, right upper quadrant incision. However, some centers are now employing minimal access laparoscopic methods for repair of duodenal obstruction.

A side-to-side duodenoduodenostomy is the standard repair for duodenal stenosis, atresia, or obstruction due to a preduodenal portal vein. In 1977, Kimura and colleagues described a modification of this procedure, known as the “diamond-shaped” duodenoduodenostomy. In this technique, a horizontal incision is made across the distal part of the proximal, dilated deodenum, and a length wise incision is made along the proximal aspect of the distal, of duodenum. This can achieve a greater diameter of the anastomosis for better emptying of the upper duodenum. In some cases, duodenojejunostomy can be an alternative and may afford an easier repair with minimal dissection. The choice of surgical procedure depends on the preference of the surgeon.

When an annular pancreas associated with duodenal obstruction is encountered, the treatment of choice is performance of a duodenoduodenostomy between the segments of duodenum above and below the area of the ring of pancreas. One should never consider division of the pancreatic ring because that could result in a pancreatic fistula while the underlying stenosis or atresia of the duodenum would remain unchanged.

In the case of an endoluminal membrane, duodenotomy and resection of the membrane can be done after localization of the ampulla of Vater. Alternatively, we can perform endoscopic resection of membrane or duodenoduodenostomy, if desired.

Postoperative Considerations and Complications

Intravenous infusions are continued in the postoperative period. Using a transanastomotic tube laying deep in the jejunum, feeding can be started as early as 48 hours postoperatively. Where available, parenteral nutrition via a central or peripherally inserted catheter can be very effective for longer-term nutritional support if transanastomotic enteral feeding is inadequate, not feasible, or not tolerated by the patient. All patients have a prolonged period of bile-stained gastric aspirate. This is mainly due to the ineffective peristalsis of the distended upper duodenum. The commencement of oral feeding is dependent upon a decrease in the volume of gastric aspirate and is often delayed for up to several weeks. Patients who have a severely prolonged return of duodenal function and have exceptionally marked dilatation of the proximal duodenum may benefit from reoperation and tapering of the proximal segment, although this is rare. Anastomotic leak, intraabdominal sepsis, and wound complications also are rare.

Prognosis. Although prognosis of intestinal atresia in general is good, an overall mortality of 7% for duodenal obstruction is seen. Associated congenital anomalies are identified as an independent risk factor for an impaired clinical course. Low birth weight and the problems of prematurity further increase mortality risk.

Anomalies of rotation (fixation) of the intestine - malrotation

Stages of normal intrauterine rotation of the intestine, rotation takes place around the axis of arteria mesenteric superior.

Stage I (7-12th week) rotation of the midgut (consists of small intestine and half of colon) outside the abdominal cavity by 90^0 around a. mesenteric superior and its transition from the sagittal to the frontal plane

Stage II: (11-16 weeks) moving the midgut into the abdominal cavity and turning it to 180^0 around a. mesenteric superior.

Stage III: (from 17 weeks to birth) lowering of the cecum into the right iliac region and intestinal fixation, ascending colon and descending colon fixes to the posterior abdominal wall.

Rotation did not take place (violation of embryogenesis at the **I stage**, nonrotation syndrome)

- Treitz ligament above and to the right of arteria mesenteric superior
- The small intestine occupies the right half of the abdomen, the large - the left
- Narrow root of the mesentery of the small intestine, which creates a risk of torsion

Incomplete rotation (violation of embryogenesis in **stage II**)

- Embryonic ligaments of the peritoneum fix the highly located cecum and compress the lower third of the duodenum
- Clinic of duodenal obstruction

Incomplete rotation (violation of embryogenesis in **stage III**)

- Embryonic ligaments of the peritoneum fix the highly located cecum and compress the lower third of the duodenum
- Clinic of duodenal obstruction
- Incomplete fixation (internal hernias)
- Potential hernia gate is formed when the mesentery of the colon and duodenum are not fixed retroperitoneally

Clinical manifestations of rotational anomalies occur:

- In 40% of patients during the first days of life
- 50% are diagnosed before 1 month
- In 75% of patients - up to 1 year
- In the remaining 25% - at any time of life, or is an accidental finding during laparotomy for another disease or section

Complications of rotational anomalies: Ice syndrome

1. Torsion of the midgut around the a. mesenteric sup.
2. High placement of the cecum and its fixation by embryonic ligaments

3. Compression of the duodenum with peritoneal ligaments

Clinic of acute (complete) torsion of the midgut

- Acute onset, usually in the first days of life
- Repeated bile vomiting
- Flatulence, sharp pain, muscle tension
- vomiting with blood, sometimes blood in feces
- Signs of dehydration and shock (hypotension, oliguria, progression of multiple organ failure)
- Skin discoloration

Clinic of chronic (incomplete) torsion of the midgut

- Recurrent or partial torsion causes impaired blood and lymph flow
- The main symptoms are intermittent abdominal pain and malabsorption

Physical examination does not reveal changes in the periods between attacks

During examination at the time of torsion - the clinical picture does not differ from acute torsion

Instrumental examinations

- X-ray in upright position
- Barium passage
- Irrigography
- Ultrasound
- Computer tomography
- Signs of normal rotation: the horseshoe of the duodenum crosses the midline, the duodenojejunal bend is located to the left of the spine at or above the pylorus
- Contrast breakage or spiral narrowing may be a sign of mid-torsion or other forms of high intestinal obstruction.
- Ultrasound signs of midgut torsion
- The superior mesenteric vein is spirally twisted around the superior mesenteric artery.

Intestinal atresia and stenosis

Atresia of the jejunum and ileum are common causes of bowel obstruction in the neonate, with a third of infants born prematurely or low weight for their gestational age.

Jejuno-ileal atresia has a prevalence rate of approximately 1:330–1:1,500 live births

The morphological classification into four types has both prognostic and therapeutic implications:

Stenosis are rarer generally occurs in 11%.

Type I atresia (23%) is a transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel. The bowel is usually of normal length (Fig 8.3).

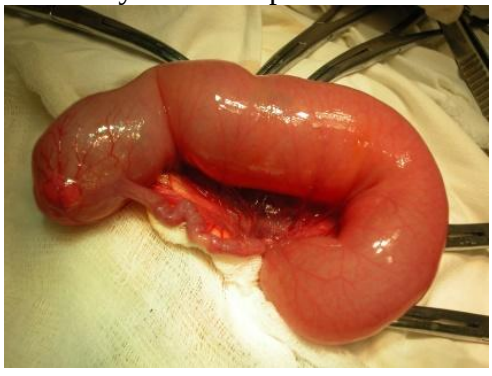


Fig 9.4. Type I intestinal atresia.

Type II atresia (10%) involves two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact.

Type IIIa atresia (15%) is similar to type II, but there is a mesenteric defect and the bowel length may be foreshortened (Fig. 8.4).

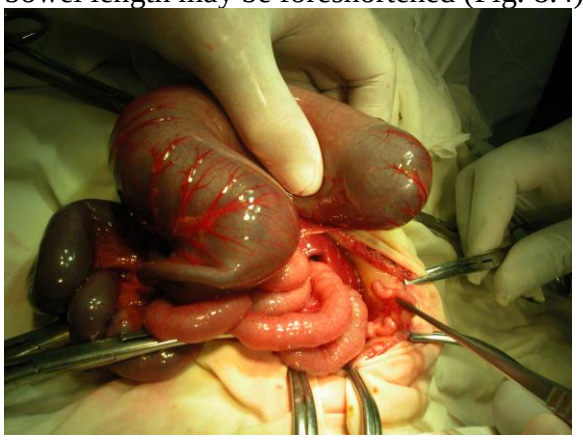


Fig. 9.5. Type IIIa intestinal atresia.

Type IIIb atresia (19%) ("apple peel" or "Christmas tree" deformity) consists of a proximal jejunal atresia, often with malrotation with absence of most of the mesentery and a varying length of ileum surviving on perfusion from retrograde flow along a single artery of supply (Fig. 9.5).

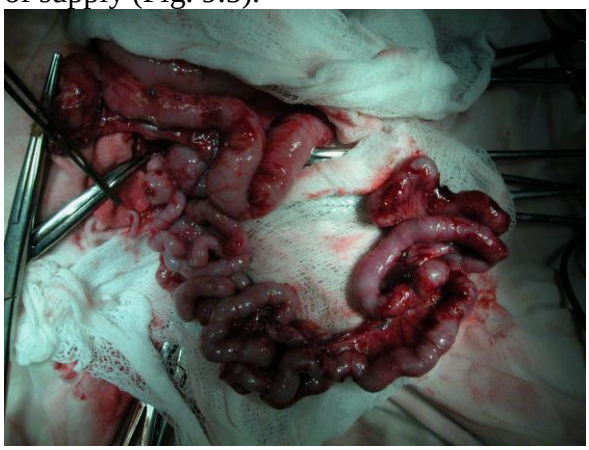


Fig. 9.6. Type IIIb intestinal atresia.

Type IV atresia is a multiple atresia of types I, II, and III, like a string of sausages. Bowel length is always reduced. The terminal ileum, as in type III, is usually spared.

Clinical Presentation

Atresia of jejunum is frequently associated with polyhydramnios. Many of these patients are born prematurely and often are small for their gestational age.

Antenatal ultrasound scanning may show dilated loops of bowel with vigorous peristalsis, which is diagnostic of obstruction. After birth most patients will have some degree of abdominal distention. The amount of distention will vary, depending upon the level of obstruction. Patients generally do not have abdominal tenderness or an abdominal mass. After aspiration of gastric contents, the abdomen will be less distended and visible peristalsis may be observed. There is usually a difficulty for passage of meconium, and typically small-volume gray mucoid stools are passed. Abdominal tenderness or peritonitis develops only with complications of ischemia or perforation. This commonly occurs with a delay in diagnosis and is due to increased intraluminal pressure from swallowed air and secondary volvulus of the bulbous blind-ending bowel at the level of the first obstruction.

In most patients, a simple abdominal x-ray with upright position and either left lateral projection are adequate to make the diagnosis which is based upon the presence of dilated, air-filled intestinal loops and air-fluid levels. In addition, plain abdominal x-rays will suggest the level

of obstruction based upon the number of dilated bowel loops. If there are two levels of fluid and gas it indicates higher intestinal obstruction. The presence of multiple dilated bowel loops without air-fluid levels suggests the possibility of lower intestinal obstruction, particularly if the intestinal content has a "ground glass" appearance. For the confirmation of the diagnosis of malrotation colonography is compulsory.

Preoperative management

All patients should receive fluid hydration prior to operative intervention. The nasogastric or orogastric tube should be passed to empty the stomach and decrease the risk of vomiting with aspiration.

In operation at the time of exploration, surgical treatment requires excision of the ends of the intestine involved in the atresia. It is also important to look for distal sites of obstruction, which can occur in up to 20% of patients and may not be immediately obvious due to lack of diameter change beyond the proximal atresia. These distal points of obstruction can be identified by flushing the distal intestinal lumen with saline to confirm intestinal continuity to the level of the rectum.

After resection of the atretic segment, the surgeon faces with the difficult task of re-establishing continuity between intestinal segments with marked size discrepancies. Another consideration is the potential dysmotility of the proximal markedly dilated segment, which may result in delayed intestinal function and problems with bacterial overgrowth. Therefore, in patients with a relatively short segment of severely dilated proximal intestine, resection of the dilated segment with re-establishment of continuity by end-to-end anastomosis is a good option. However, in patients with long segments of proximal intestine that are significantly dilated, resection of the whole involved segment may result in inadequate remaining intestinal length to allow absorption of enteric nutrients (i.e., short bowel syndrome). Therefore, these patients frequently are treated by either imbrication or tapering enteroplasty of the proximal dilated segment. To date, no randomized studies have compared the outcomes for patients with intestinal atresia's with or without the addition of an enteroplasty or plication. In patients for whom the atresia is just distal to the duodenojejunal flexure, it may be advantageous to resect the dilated bowel, derotate, and taper the duodenum with primary anastomosis. This facilitates passage of a transanastomotic feeding tube and early restoration of foregut function. The total residual length of bowel should be measured with a tape and recorded, as this gives some guidance as to prognosis. Patients who have multiple atresia's (type IV) or an apple-peel deformity (type IIIb) present particularly challenging management problems. These patients may require multiple anastomoses and frequently will experience long-term delays in return of intestinal function. In addition, many of these patients will have short bowel syndrome due to inadequate residual intestinal length. In general, the formation of stomas is unnecessary and should be avoided because dilated bowel does not reduce in caliber, and fluid and electrolyte losses may be severe.

The most common postoperative complication is a functional obstruction at the site of anastomosis. Other less commonly observed complications include anastomotic leak and adhesive obstructions.

Most patients with intestinal atresia do not have associated life-threatening anomalies The judicious use of nasojejunal or gastrostomy transanastomotic feeding tubes for enteral feeding may be lifesaving.

Hirshprung's disease

Hirschsprung's disease (HD) is a functional intestinal obstruction resulting from the congenital absence of parasympathetic ganglion cells in the myenteric plexus of the distal bowel. The initial description of congenital agangliosis HD in 1886 was given by Harald Hirschsprung, who is rewarded as the Father of Danish Pediatrics. Successful treatment of the condition had to wait for 50 years until the pivotal role of the distal aganglionic segment and its vital role in the pathophysiology of Hirschsprung's disease was identified. The development of successful surgical management of this disease had changed historically the dominant approach in last decade has been the endorectal pull through and many prefer Duhamel and Soave Procedure ,one of the success stories of pediatric surgery in the modern era is TEPT and Laparoscopic assisted TEPT.

The HD incidence worldwide is approximately 1 in 5,000 live births with the male-to-female ratio is approximately 4:1. HD results in a functional obstruction of the bowel with dilatation of the proximal colon and hypertrophy of the muscles (i.e., megacolon). Macroscopically, HD features a narrow aganglionic segment and a transitional zone, and then the dilated proximal portion with a thickened bowel wall as a result of hypertrophy of the muscular wall of the intestine.

HD classically affects the rectum and sigmoid (70%), but can also involve a long portion of colon and can affect the entire colon. In 20% of cases, a long segment of colon is affected, with the total colon being aganglionic in 8–10%.

Extensive ultra-long aganglionosis with extensive small bowel involvement is uncommon, occurring in less than 2%. The functional abnormality always includes the internal anal sphincter and extends proximally for short or long distances, depending on the type.

The classic histological features of Hirschsprung's disease are the absence of ganglion cells in intramuscular myenteric (Auerbach's) plexus and the submucosal Meissner's plexus.

Acetylcholinesterase staining techniques is the investigation to evaluate suction biopsies to show an increased activity in the parasympathetic nerves of the affected zone as well as neurofibrils within the lamina propria and muscularis mucosa. This technique remains the gold standard on rectal biopsies.

The morphologic diagnosis of HD therefore rests on the following:

• Absence of ganglion cells in the submucosal layer (and/or intramyenteric (Auerbach's) plexus)

• Presence of the enlarged peripheral nerve trunks in the submucosa

• Increased AChE staining—proliferation of neurofibrils in the lamina propria and the muscularis mucosa (absent in normally innervated intestine).

Clinical Features. A delay in passage of meconium is the most important neonatal observation. The clinical presentation depends not only on the length of the aganglionosis. In the acute form of agangliosis there is a delay in the excretion of meconium in the first day or more, or a constant lack of self-defecation in the first days of life leads to an increase in abdominal volume due to flatulence. Vomiting on the first day after the birth of a child is replaced by profuse vomiting, on the second day in the gastric contents when vomiting there are impurities of intestinal contents. The general condition of the child is progressively deteriorating, increasing manifestations of exsiccosis and toxicosis. The picture of lower intestinal obstruction develops.

Acute HD in newborns is often complicated by enterocolitis. Instead of a temporary delay of a stool or diarrheal syndrome which is followed by increase in intoxication and dehydration of the child develops.

Diagnostic Investigations

Abdominal X-rays

The diagnostic accuracy of abdominal x-rays is 52%.

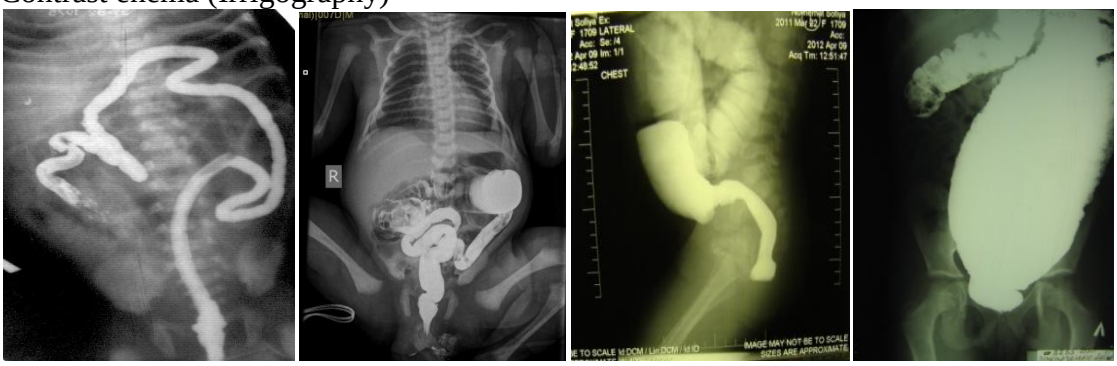
A. Contrast enema: • The narrow aganglionic segment is shown with a dilated proximal bowel segment, and a transitional zone is diagnostic. A lateral view may demonstrate the narrow rectum. Depending on the effected part of colon by this method four forms of HD are distinguished 1. totally effected colon, 2. subtotal form, 3. rectosigmoid form, 4. rectal form, the most common among them is rectosigmoid form.

Manometry

In healthy children, distention of the rectum results in the reflex relaxation of the internal sphincter is seen during manometry, which is absent in patients with Hirschsprung's disease.

Schematic presentation of HD

Contrast enema (Irrigography)



Intra operation

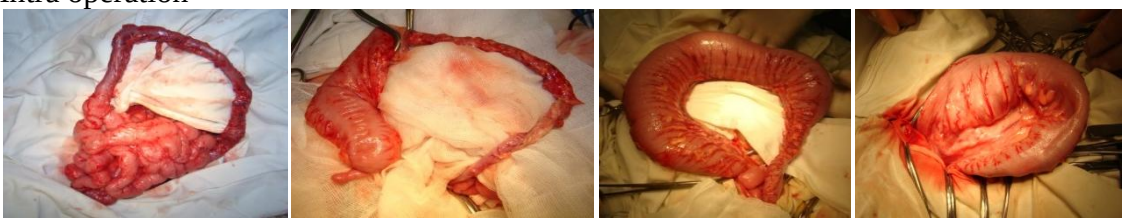


Fig. 9.7. Row 1. Schematic presentation of HD. A – Total form, B – subtotal form, C-rectosigmoid form, D- rectal

Row 2. Contrast enema. A – Total form, B – subtotal form, C-rectosigmoid form, D- rectal

Row 3. Intra operation. A – Total form, B – subtotal form, C-rectosigmoid form, D- rectal

Differential Diagnosis includes the following:

• Small left colon syndrome, particularly in diabetic mothers

• Meconium plug syndrome in neonates.

• Chronic idiopathic intestinal pseudo-obstruction (CIIP, or CHIPS)

• Other dysganglionosis of the gastrointestinal tract

• Acquired megacolon (generally at >1 year of age), resulting from anal fissure, anal or rectal stricture, anorectal malformations, tumor, or psychogenic reasons.

Principles of Surgical Management

In recent times the management of HD by colon irrigation techniques allows for one-stage surgery to be performed at a much earlier (mostly still within the neonatal period). Colostomy may still be necessary in resistant cases. Colostomy (which is located at site of normal ganglionated colon as determined by the presence of ganglion cells on frozen section) is the traditional way of relieving obstruction. This is then followed by a definitive pull-through procedure 3 to 9 months later.

In modern practice, temporary decompression is first attempted by means of irrigation with warm saline. If it's successful, it can be continued until definitive surgery.

Definitive pull-through surgical procedures have undergone numerous modifications since the original description by Swenson. Most of these modifications are based on the original concept described in 1948 by Swenson and Boley and adhere to the principle of removing the functionally

obstructive segment of aganglionic colon and pull through forming primary coloanal anastomosis. In recent years, the endorectal pull-through (ERPT) described by Soave has gained popularity.

Four surgical procedures have received reasonably wide acceptance.

Swenson Procedure

This was the original operation described by Swenson in 1948. It involves resection of the aganglionic segment deep into the pelvis and direct end to end anastomosis of the proximal colon to the anorectal canal.

Duhamel Procedure

In the Duhamel procedure (retro rectal pull-through), the lower part of aganglionic rectum is retained and the ganglionated colon is mobilized and pulled through posteriorly and anastomosed to the aganglionic remnant with a side-to side anastomosis.

Soave Procedure

The Soave procedure (extra mucosal endorectal pull-through), along with its variations, is the most frequently performed procedure in the world for short-segment Hirschsprung's disease. It has more recently been popularized as a laparoscopic-assisted or anal approach.

The procedure involves an extra mucosal resection of a retained aganglionic rectal segment. The rectal mucosa is removed and a muscular cuff retained. The ganglionated colon is pulled through this cuff and anastomosed to the dentate line in the rectum, thus forming a primary endorectal pull-through.

Trans anal Pull-Through

The trans anal pull-through approach is through the anus, thus avoiding abdominal scars. It is currently in vogue in many parts of the world, being mostly suited to a short aganglionic segment. It is similar to the Soave procedure, but is performed in reverse through the anus.

The technique involves the patient being placed in lithotomy and the rectum irrigated until clean. Retraction sutures are placed to expose the rectal mucosa and open the anus. Submucosal dissection is commenced 3–5 mm from the dentate line and the cut line controlled by multiple fine traction sutures. Following the completion of the submucosal dissection, the rectum is transected. The dissection is continued proximally until the peritoneal reflection where the sigmoid colon is mobilized and delivered. Following histological confirmation of ganglion cells in the proximal colon, the aganglionic segment is resected, and a sutured anastomosis is performed (Fig.9.8).

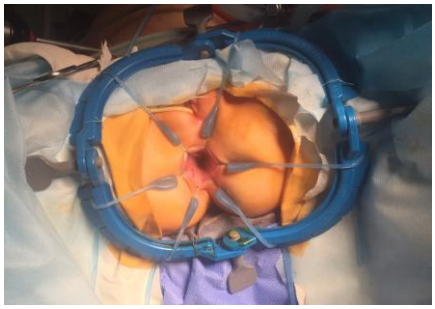
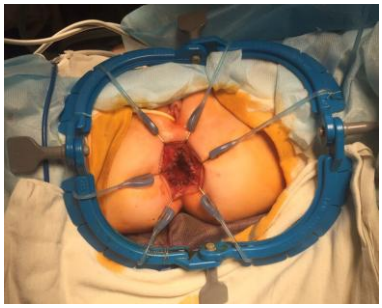
A.  B.  C. 

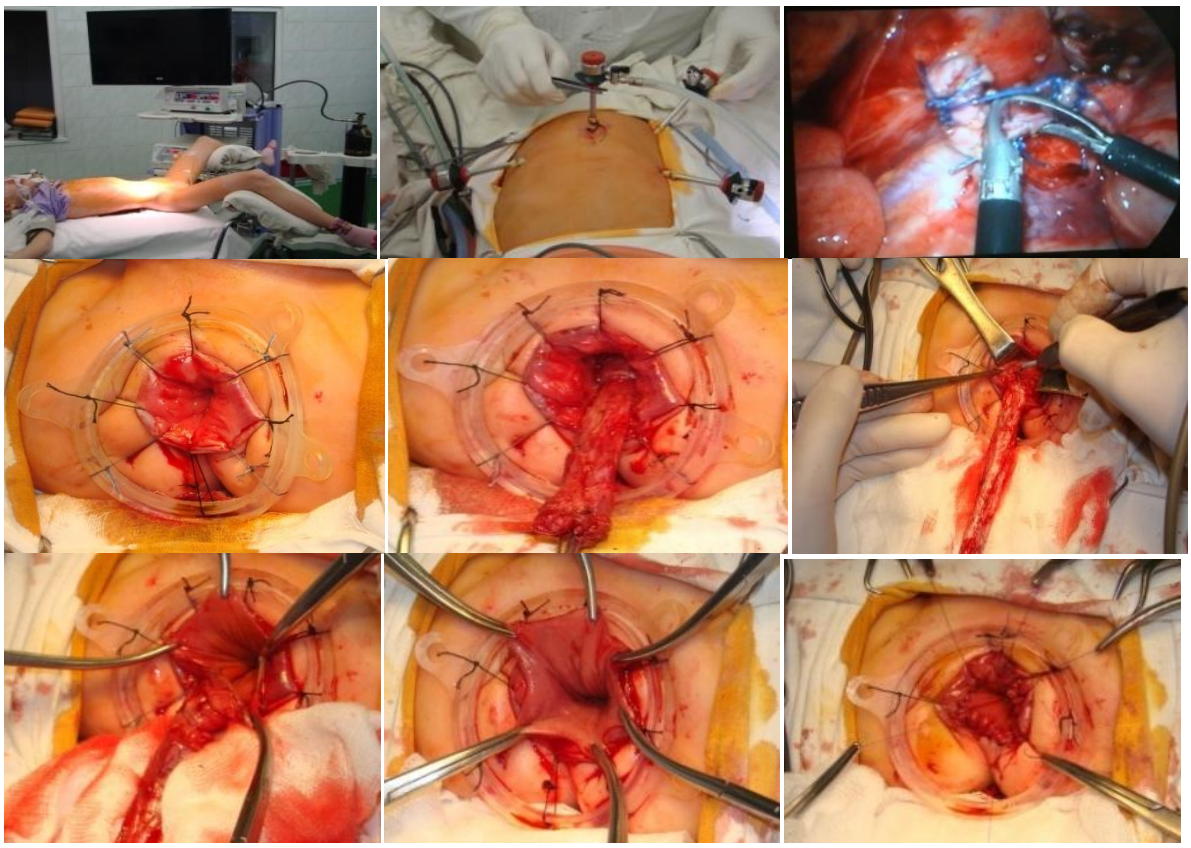
Fig.9.8. Trans anal Pull-Through

A. Retraction of anus. B. mobilization of aganlionic colon. C. colorectal anastomosis

Many recommend a laparoscopic first stage to mobilize the bowel and perform biopsies to facilitate the resection margins.

Many recommend a laparoscopic first stage to mobilize the bowel and perform biopsies to facilitate the resection margins.

Fig.9.9. Transanal Pull-Through TEPT with laparoscopic assistance



A. lithotomy position, B. installation if ports, C ligation of blood vessels with mobilization of colon, D. Retraction sutures, E, F, G Mobilization of aganglionic colon, H, I primary colorectal anastomosis.

Postoperative Complications of Hirschsprung's Surgery

Early postoperative complications include anastomotic leakage, prolonged ileus, adhesive obstruction, intestinal obstruction, and retraction of the neorectum. Wound sepsis, pelvic or presacral abscesses may be evident and enterocolitis associated with Hirschsprung's disease may be present.

Late Complications

It is generally recognized that the major long-term complications in the postoperative period following Hirschsprung's surgery are soiling, stenosis of colorectal anastomosis, constipation and Hirschsprung's-associated enterocolitis.

Sphincter achalasia may be difficult to treat, and previous attempts at repeated anal dilatation have had mixed success. We have found the topical application of a glyceryltrinitrate paste to be fairly effective, and it is a cheap alternative to the injection of botulin toxin advocated by some.

Prognosis and Outcome

HD is surgically correctable, and the majority of patients with the disease can live productive, satisfying lives. Most HD patients (93%) achieve acceptable anorectal function, given sufficient time to adjust. Long-term functional results are excellent in some, good in the majority, and poor in approximately 15–30%.Functional results depend on the length of aganglionosis, procedure performed, surgical complications, social circumstances, family support, and associated anomalies, among other factors. Psychological problems may be magnified in those with poor support systems.

Anorectal malformations (ARM)

Anorectal anomaly is a congenital absence of the anal canal and rectum and may have connection with fistula to the genitourinary system or perineum. The term "imperforate anus" has traditionally been used to describe all anorectal abnormalities in females and males. The frequency of atresia ranges from 1 in 4000-5000 newborns. It is well known that anorectal defects are more

common in boys than in girls. The most common variant of the defect in boys is atresia of the anus with rectourethral fistula, and in girls - recto vestibular fistula.

The diagnosis of ARM should be made during the first physical examination of newborn in the delivery room (Fig.9.10).

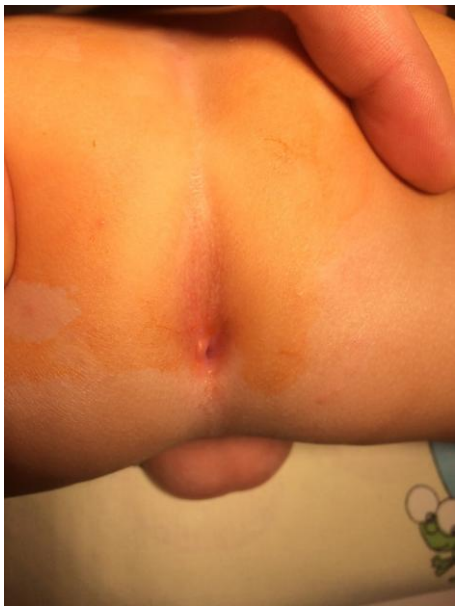


Fig. 9.10. Anorectal malformations – fcence of the anus in typical place. Perineal fistula (arrow).

In the prenatal period ARM can be diagnosed with ultrasound examination which allows you to visualize dilated intestinal loops in the fetus, which gives reason to suspect congenital intestinal obstruction.

After the birth of a child, a mandatory general examination, including perineal examination, in most newborns gives grounds to establish a preliminary diagnosis.

Newborns with anorectal atresia and the clinical picture of acute intestinal obstruction require immediate surgical intervention after additional examination and clarification of the form of atresia. Surgery is performed after preoperative preparation.

Posterior Sagittal Anorectoplasty (PSARP), popularized by Alberto Peña in the 1980s, is currently the most commonly utilized procedure for repairing ARMs.

Diagnosis

Some children are not born in health care facilities, however, and the absence of an anus (Fig. 9.11) sometimes is not noted by the family until hours or days after birth, when it is noted that the child's abdomen is distending and the infant has not passed meconium. When consulted for a newborn with ARM, surgeon should initially note the child's gender and examine the perineum carefully to see whether there is any evidence of meconium from a perineal fistula or along a midline raphe or whether the child has a true bucket-handle deformity. If the child is male and there is no initial evidence of visible meconium, the child should be observed for 12 hours or more to see whether meconium appears in the perineum. During this time, gauze is placed over the penis so that the urine can be examined for evidence of meconium. If there is meconium in the urine, the diagnosis of a recto urinary tract fistula is made, and no further diagnostic procedures are indicated.

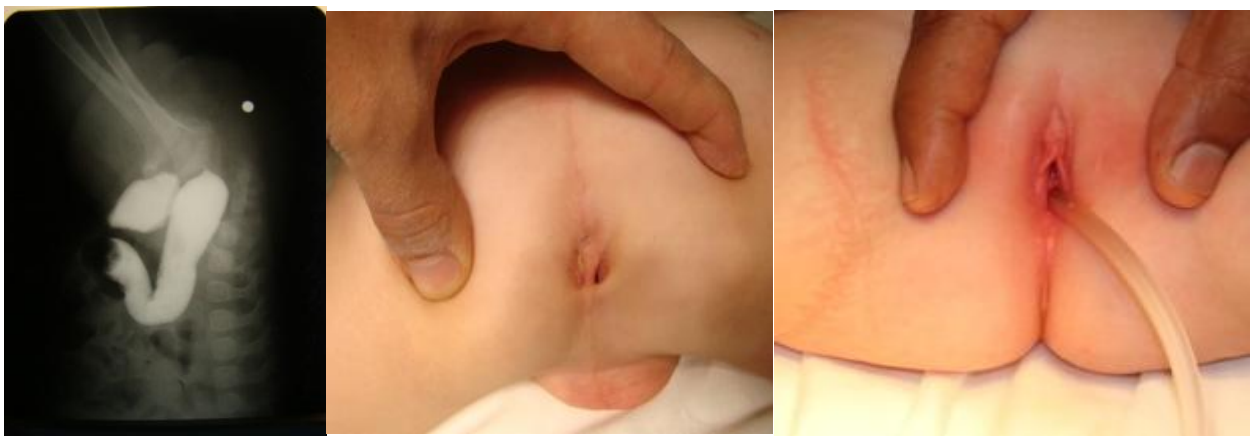


Fig. 9.11. Anorectal malformations
A. Invertogram B. perineal fistula in male, C recto vestibular fistula in female

If, however, there is no meconium identifiable in the urine or on the perineum, an invertogram is performed because a very small percentage of children (more common in children with Down syndrome) will have a blind pouch without a fistula.

The child is placed in a prone jackknife position with the buttocks higher than the rest of the body for at least 30 minutes. After that time, a radio-opaque object (drop of barium or coin) is placed on the anal dimple, a cross-table lateral x-ray is performed, and the distance between the rectal air bubble and the perineal skin is measured. If the distance is less than 1 cm, the lesion can be treated as a low lesion, but any distance greater than 1 cm should be managed as a high lesion. In females, the labia should be grasped for traction and the posterior portion of the vestibule (just external to the hymen) examined for a recto vestibular fistula. In persistent cloaca, there is only one perineal opening, and therefore the separate openings for urethra, vagina, and rectum are not visible.

(Table 9.1)Classification of ARMS in accordance to anorectal sphincter

TYPE	MALE			FEMALE	
High (Supra-Levator)	Ractal atresia				
	Anorectal agenesis	Without fistula		Without fistula	
		With fistula	Rectovesical Rectourethral	With fistula	Rectovesical Rectocloacal Rectovaginal
Intermediate	Anal agenesis	Without fistula		Without fistula	
		With fistula	Rectobulbar	With fistula	Rectovaginal - low
					Rectovestibular
Low (Trans-Levator)	At normal site	Covered anus – complete Anal stenosis			
	At perineal site	Anterior perineal anus Anocutaneous fistula – covered anus (incomplete)			
	At vulvar site			Vulvular anus Anovulvular fistula Anovestibular fistula	

When the diagnosis of ARM is made, the child should be examined for other components of the VACTERL (Vertebral and spinal cord, Anorectal, Cardiac, Tracheoesophageal, Renal and other urinary tract, Limb) complex of anomalies. Diagnostic modalities must be appropriate for the particular locale. An orogastric tube is used to test for patency of the esophagus. An ultrasound, if available, is the best way to initially assess the urinary tract for abnormalities.

Classification of arms

*Males***:** Rectoperineal fistula, rectourethral fistula, bulbar, prostatic, recto bladder neck fistula, imperforated anus without fistula, rectal atresia or stenosis

Females: rectoperineal fistula, rectovestibular fistula, persistent cloaca, imperforated anus without fistula, rectal atresia/stenosis. (Table 9.1)

Treatment

After the establishment of diagnosis, we can select the surgical approach, in many cases colostomy is recommended as the initial treatment for high lesions, whereas low lesions can be treated primarily with an anoplasty. Low lesions that can be treated without a colostomy include those with evidence of meconium in the perineum or a bucket-handle lesion and those with a blind pouch less than 1 cm from the anal dimple, as demonstrated on invertogram. The most common lesions, including rectourinary tract fistulas in males and rectovestibular fistulas in females, should be treated as high lesions with an initial colostomy.

Alberto Peña has repeatedly emphasized the importance of a double-barrel colostomy to achieve total diversion of faeces. This is particularly important in males because there is usually a fistula between the distal colon and the urinary tract, and undiverted stool in the distal colon may cause repeated urinary tract infections (UTIs). It is best to place the colostomy in the distal descending or proximal sigmoid colon to make the distal limb shorter than it would be in a transverse colostomy.

Many male children with ARMs have urine flow from the urethra into the distal colon and then come out from distal stoma, and if urine stays in the colon for long periods of time, it can cause a significant metabolic acidosis.

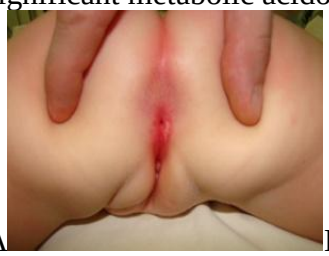
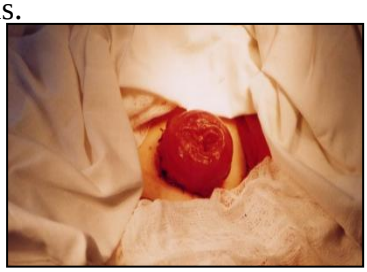
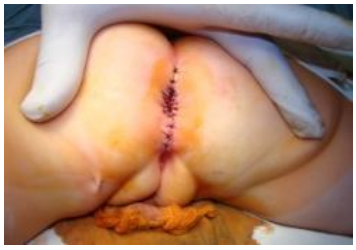
A  B  C 

Fig. 9.12. Anorectal malformations stages of operation

A. diagnosis of ARM, B. colostomy, C Posterior Sagittal Anorectoplasty

When the child reaches approximately 2–3 months of age, and if the child appears in very good nutritional status as evidenced by a weight of 8–10 kg, a definitive posterior sagittal anorectoplasty (PSARP) can be performed. In males, a distal colostography should be performed prior to operation to determine the site of the fistula.

Posterior Sagittal Anorectoplasty Technique (PSARP)

PSARP is performed with the child in a prone, jackknife position. Then urinary catheter is inserted, Muscle stimulator is used for mapping the external sphincter muscles and can mark the place for neo rectum.

After the revision of the tissue's rectum is identified and separated from the fistula then mobilized, after which the rectum is pulled through the center of the sphincter muscles and neo anus is made.

Bowel Management Programs

Even under the best of circumstances, some children particularly those with high ARMs suffer from constipation or fecal incontinence caused by encopresis (escape of liquid stool around a large, hard fecal impaction). The overriding principle behind bowel management is that the child learns to evacuate in a socially acceptable place (home) at a socially acceptable time (before or after school). The primary technique used to achieve continence is an enema regimen.

The child is given enemas each night or early in the morning at home to completely evacuate the colon. The child can then go to school with confidence, knowing that there will be no stool accidents.

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Tests for initial level of knowledge

1. What metabolic changes develop during hypertrophic pyloric stenosis?
   A. Hypochloraemic hypokalaemic respiratory alkalosis
   B. Hyperchloraemic hyperkalaemic metabolic acidosis
   C. Hypochloraemic hypokalaemic metabolic alkalosis
   D. Hypochloraemic hypokalaemic metabolic acidosis
2. Which of the following is the most common clinical sign of hypertrophic pyloric stenosis?
   A. Constipation
   B. Dehydration
   C. Abdominal pain
   D. Vomiting
3. At what age usually appear hypertrophic pyloric stenosis?
   A. 3-6 days
   B. 3-6 weeks
   C. 3-6 month
   D. 3-6 years
4. Which x-ray sign is pathognomic for duodenal atresia?
   A. Apple peel sign
   B. Double-bubble sign
   C. "Doughnut" sign
   D. "Bull's eye" sign

5. What is characterized for Type I intestinal atresia
    A. Proximal jejunal atresia, often with malrotation with absence of most of the mesentery and a varying length of ileum
    B. Transluminal septum with proximal dilated bowel in continuity with collapsed distal bowel
    C. Two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesentery intact
    D. Two blind-ending atretic ends separated by a fibrous cord along the edge of the mesentery with mesenteric defect
6. What is the etiology of Hirschsprung's disease?
    A. Failed migration of nervous ganglion
    B. Bowel obstruction
    C. Constipations
    D. Failed vascularization of intestinal wall
7. What method of diagnostics confirm the diagnosis of Hirschsprung's disease?
    A. Ultrasound
    B. X-ray of abdomen
    C. Ct of abdominal cavity
    D. Acetylcholinesterase staining techniques
8. What operation is most commonly used for anorectal malformations repair?
    A. Ramstedt pyloromyotomy
    B. Transanal endorectal pull through
    C. Pena operation
    D. Kimura operation
9. What is the most common complication after anorectal malformations?
    A. Constipation
    B. Fecal incontinence
    C. Vomiting
    D. Abdominal disproportion

Keys for tests:

1. C
2. D
3. B
4. B
5. B
6. A
7. B
8. D
9. C
10. A, B.

Tests for final level of knowledge

1. You see a 4 weeks old infant which has nonbilious forceful vomiting, hungry after vomiting with signs of dehydration, scaphoid abdomen and palpable mass in the right upper quadrant. You consider that child has hypertrophic pyloric stenosis. Which of the following investigations will confirm the diagnosis?
    A. Ultrasound
    B. X-ray
    C. CT
    D. MRI
2. You see a thin malnourished 5-year-old girl who has the history of constipations from the neonate period, with defecation once a week. She presents abdominal distention. Rectal

findings show beyond the aganglionic zone an explosive evacuation of soft stool. Which methods of investigation should be performed in order to confirm the diagnosis?

A. Contrast enema
B. Manometry
C. Ultrasound
D. Acetylcholinesterase staining techniques

3. You are caring a neonate on a ward who has developed bilious vomiting and abdominal distension. On abdominal x-ray you see the "double-bubble" sign and parents' constant absence of meconium evacuation. What type of operation is indicated in this case?

A. Diamond-shaped duodeno-duodenostomy
B. Ramstedt pyloromyotomy
C. Posterior sagittal anoproctoplasty
D. Transanal endorectal pull through

4. You see a neonate with Intestinal obstruction presents with bile-stained vomiting, abdominal distention, delayed passage of meconium. On contrast enema you recognize narrowed rectum and dilatated sigmoid bowel. What type of operation is the most common in this case?

A. Swenson Procedure
B. Duhamel Procedure
C. Soave Procedure
D. Neonatal pull-through

5. You see 2-day child with unperforated anus, with vomiting, distended abdomen, signs of dehydration. Which will be first step of surgical treatment?

A. Pena procedure
B. Posterior sagittal anoproctoplasty
C. Ramstedt procedure
D. Colostomy

Keys for tests:

1. A
2. A, D
3. B
4. D
5. D

Tasks for final level of knowledge

Task 1. You are called to the neonate's department to see a 1-month boy. He presented with nonbilious vomiting, weight loss, signs of dehydration present in cases of repeated vomiting and palpable mass in the right upper. On ultrasound the sign of "doughnut" is present. Your working diagnosis is hypertrophic pyloric stenosis.

1. What is your management of this patient?
2. Which type of operation is preferable?

Key Answer

1. Nasogastric decompression, correction of electrolytes, operation
2. Ramstedt pyloromyotomy/ laparoscopic pyloromyotomy.

Task 2. 9-year girl presents with sever constipations from the neonatal period, abdominal pain, abdominal distention and malnutrition.

1. What pathology you consider?
2. What methods of diagnosis you will perform to confirm the diagnosis?
3. Which surgical treatment is method of choice?

Key Answer

1. Hirschsprung's disease
2. Contrast enema, manometry, acetylcholinesterase test.
3. Transanal endorectal pull through

Task 3. You see a neonate with Intestinal obstruction presents with bile-stained vomiting, abdominal distention, delayed passage of meconium. On contrast enema you recognize narrowed rectum and dilatated sigmoid bowel. With what you will make a differential diagnosis?

Key Answer

1. Meconium plug syndrome in neonates.
2. Chronic idiopathic intestinal pseudo-obstruction
3. Other dysganglionosis of the gastrointestinal tract

Materials for self-study of the students

Main tasks	Notes (instruction)
Repeat: Anatomy of Congenital gastrointestinal abnormalities. Anorectal abnormalities	To sketch out the anatomy of peritoneal cavity Top represent the methods of diagnosis of Congenital gastrointestinal abnormalities. Anorectal abnormalities in children
Study: Pathogenesis of Congenital gastrointestinal abnormalities. Anorectal abnormalities Features of Congenital gastrointestinal abnormalities. Anorectal abnormalities in children The diagnostic possibilities of ultrasound examination, CT, MRI in children	To make differential diagnosis of acute appendicitis and nonsurgical diseases in children To make the indications to surgical treatment in children with abdominal pain To know modern diagnostically methods To know the advantages and disadvantages of classic and laparoscopic approaches

Theme 10

ABDOMINAL WALL DEFECTS. GASTROSCHISIS, OMPHALOCELE. RARE ABDOMINAL WALL DEFECTS

Overview:

The prevalence of malformations of the anterior abdominal wall is 1: 2000 newborns, with plum belly syndrome 1: 40,000 newborns. Despite the fact that the success of surgeons and anesthesiologists can reduce mortality and the number of complications, they remain high in this group of patients. This is largely due to the presence in this group of newborns of comorbidities, severe background diseases and conditions, the components of which worsen the course of the disease and the postoperative period, worsening the prognosis of surgical disease.

Educational aims:

After completing this part of module students should be able to:

1. Recognise the main features of gastroschisis and omphalocele in newborn children.
2. Counsel parents about the likely diagnoses and management.
3. Know what the possible complications are.
4. Know the indications for urgent treatment or referral to a specialist center.
5. Know the outcome of abdominal wall defect.
6. Recognise of complications.
7. Discuss the overall prognosis and management of gastroschisis and omphalocele.

A student must know:

1. Embryology of the abdominal wall.
2. Embryogenesis of the abdominal wall defects.
3. Antenatal considerations of the abdominal wall defects.
4. Incidence and associated conditions.
5. Pathogenesis of omphalocele and gastroschisis.
6. Prenatal diagnostic procedures.
7. Clinical features of omphalocele and gastroschisis.
8. Preoperative treatment.
9. Modern approach to abdominal wall closure.
10. Complications of abdominal wall defects.

A student must be able to:

1. Describe the clinical features of abdominal wall defects.
2. Evaluate the prenatal ultrasound and MRI investigations.
3. Evaluate the prenatal screening laboratory tests.
4. Take care for children with silo sac.
5. Analyze the blood tests to evaluate the type of coagulopathy.
6. Appoint additional investigations in children with abdominal wall defects.

Terminology

Term	Definition
Omphalocele (exomphalos)	herniation of intraabdominal contents through a full-thickness umbilical defect which is covered by a sac composed of peritoneum internally and umbilical cord amnion externally.
Gastroschisis	defect in the abdominal wall usually just to the right of the umbilicus. The herniated viscera are not covered by a sac
Beckwith-Wiedemann syndrome	exomphalos-macroglossia-gigantism syndrome (EMG syndrome)

Pentalogy of Cantrell	anterior diaphragm and pericardial defects, a short bifid sternum, and cardiac defects associated with an omphalocele sac or, at least, hypotrophic epigastric skin
Bladder exstrophy	Abnormal development of the lower body wall results from defective enfolding of the caudal pole of the embryo and deficient incorporation of the yolk sac and allantois; this is associated with malformation of external genitalia
Prune belly (Eagle Barrett) syndrome	Rare congenital abnormalities, is a result of increased apoptotic cell death in the body wall placode, which leads to insufficient mesodermal cell deposition. Muscle fibers are absent and replaced by a thick collagenous aponeurosis.

CONTENT

Omphalocele

Omphalocele (Embryonic hernia) is a severe malformation, a defect of the anterior abdominal wall, which is accompanied by underdevelopment of the abdominal organs. There is an inversely proportional relationship: the larger the hernia, the more underdeveloped the abdominal cavity.

Up to 54% of patients with omphalocele have been reported to have a cardiac abnormality (septal defects, tetralogy of Fallot, ectopia cordis). Chromosomal abnormalities can be found in up to 20%, and an association with Down syndrome has also been reported. Patients with omphalocele are more likely to be large for gestational age (macrosomia or more then 4 kg in birth weight).

Physical examinations***:***

The abdominal wall defect can range in size from 2–10 cm. Virtually all intraabdominal structures can be eviscerated including the liver. The omphalocele is covered by an outer layer of amnion, a middle layer of Wharton's jelly, and an inner layer of peritoneum. This robust covering protects the gastrointestinal tract during intrauterine life and bowel function is usually normal at birth.

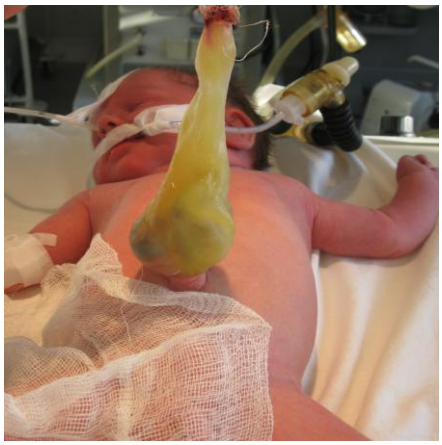
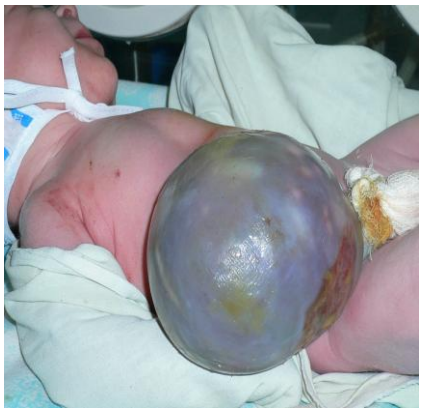
A.  B. 

Fig. 10.1. Newborn with omphalocele. A – small omphalocele, B – large omphalocele

Treatment of umbilical cord hernias begins immediately after hospitalization. A nasogastrc tube should be placed early to decompress the intestines. A rectal examination aids this with evacuation of meconium. Maintenance of body temperature is important. Ventilator support and supplemental oxygen should be supplied as needed.

Because of the fraquency of associated heart defects, cardiology evaluation and echocardiography are in order. Intravenous fluids are provided at maintenance rates, 1 mg of vitamin K should be administered, as well as prophylactic antibiotics.

There are two methods of treatment - conservative and operative. The first positive result of conservative treatment was described in 1751 (Storch), and the first surgical intervention for umbilical cord hernia was performed in 1836 (Berard). Only since 1889. surgical treatment of this defect is becoming more common.

Although the bowel in omphalocele is protected by the sac, operaration is still urgently needed to increase the chance for primary closure.

Operative closure. In the operating room the sac and abdomen are prepared with an antiseptic and excess cord is removed. The operative strategy for the repair of omphalocele depends on the size of the defect, the extent of visceroabdominal disproportion (VAD), and the severity of the associated anomalies.

Primary closure of the omphalocele can be accomplished in small defects (generally 2–5 cm in diameter). In large omphaloceles with the liver herniated, care must be taken in the reduction of the liver to avoid torsion of the hepatic veins, obstruction of portal vein inflow, obstruction of the inferior vena cava, or injury to the hepatic capsule. When there is significant VAD and loss of abdominal domain the reduction of the hernia can be approached via a staged silo technique (Fig. 10.2).

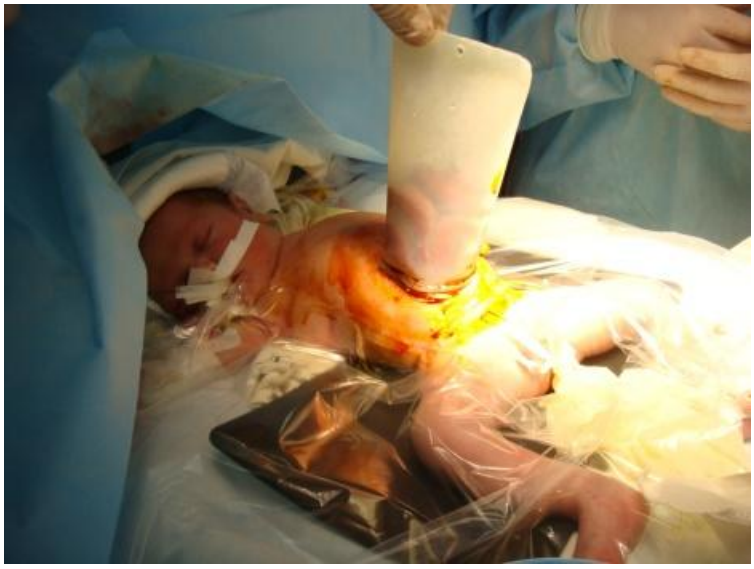


Fig. 10.2. Silo plasty of large omphalocele with handmade silo sack.

Alternatively, the surgeon can create an intentional ventral hernia by elevating skin flaps or inserting a prosthetic mesh. But this method has poor long-term results (Fig. 12.3).

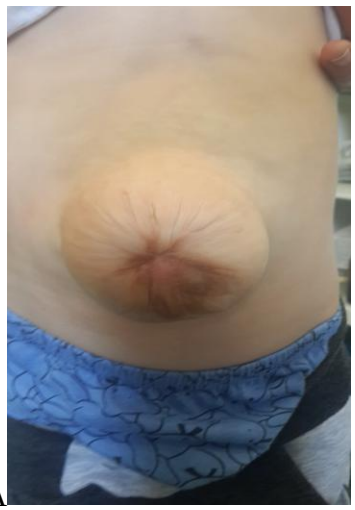
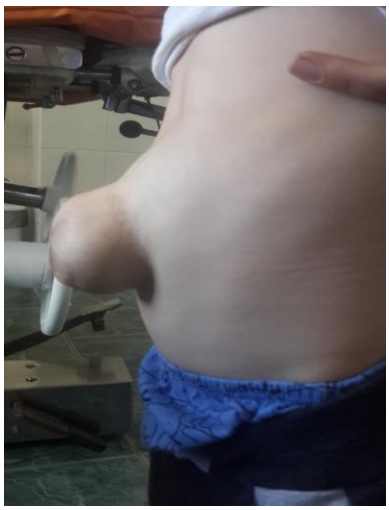
A  B 

Fig. 10.3. Ventral hernia after skin flaps plasty of omphalocele

Gastroschisis (from the Greek - divergence of the abdomen) - is an eventration that occurs in utero. The umbilical cord attaches normally, so the pathology is an extraumbilical malformation of the anterior abdominal wall. It occurs with a frequency of 1: 6000 newborns, 40% of which are premature. The defect of the anterior abdominal wall is localized to the right of the umbilical cord. Its occurrence is associated with circulatory disorders in the distal part of the right omphalomesenteric artery, the development of necrosis of the anterior abdominal wall to the right of the umbilical cord and the formation of a defect of the abdominal wall. Due to the defect falls a short section of the midgut three times larger in diameter than the defect. The loops of the intestines are soldered together, opaque, swollen, covered by the fibrinous peel, and impaired outflow of venous blood and lymph. Fibrinous layers on the intestinal loops are associated with changes in the composition of amniotic fluid after the onset of renal function in the fetus. This period is characterized by hypoperistalsis and malabsorption of the intestines, their lumen is filled with thick and viscous meconium.

Today, the diagnosis is usually made antenatally by ultrasond (US). Omphalocele can be distinguished from gastroschisis by the presence of a sac and presence the liver in the defect. Antenatal US also detects associated anomalies. In gastroschisis these anomalies are usually intestinal atresias.

Physical examinations

The abdominal wall defect is fairly uniform in size and location: a 3-5cm vertical opening immediately to the right and inferior of the umbilical cord. All patients with gastroschisis have intestinal nonrotation. The stomach, small and large intestines and occasionally the gonads can be found outside of the abdominal wall. The liver is almost never eviscerated. There is no peritoneal sac over the herniated contents.

Inflamed intestine is thick and edematous, the loops of bowel are matted together, and the mesentery is congested and foreshortened (Fig. 10.4).

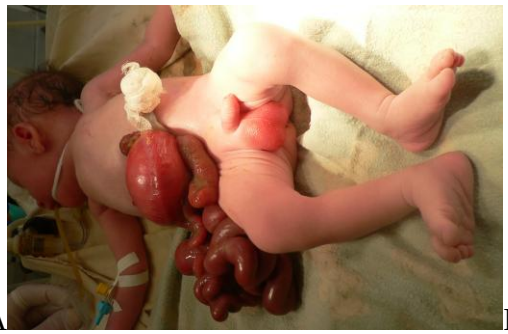
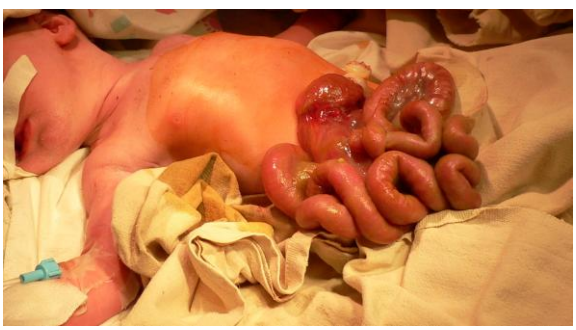
A  B 

Fig. 10.4. Newborn with gastroschisis

Treatment

Most patients with gastroschisis are premature, and close attention must be paid to heart preservation, and respiratory support. The large surface area of exposed intestine leads to fluid and heat loss. The best way to control this problem is to place most of the infant immediately in a plastic drawstring bowel bag to control evaporative heat and fluid loss. The patient also needs increased intravenous fluid volume. After removal of the newborn from perinatal shock, the defect is closed. The surgeon holds the umbilical cord up, and loop by loop, immerses the intestine into the abdominal cavity. In most cases, by time the patient is ready for ckin preparation, the bowel has been totaly reduced. If the primary cloasuse of abdominal wall is impossible, other surgical maneures can be used, it may be Silo plasty.

In the postoperative period, intestinal passage is slowed due to trophic disorders and the development of peritonitis. Prolonged parenteral nutrition and intensive care are provided.

Rare abdominal wall defects

One unusual form of omphalocele is the **cephalic fold defect, or pentalogy of Cantrell**, in which the abdominal wall defect is supraumbilical and the hart is in the sac through a defect in the pericardium and the central tendon of the diaphragm. the other elements of pentalogy are a heart defect and a sternal cleft. Some patients have the ectopia cordis thoracis. It is the malformation when the heart is outside the thoracic cavity, inside the omphalocele sac, and not covered by the pericardium.

Caudal fold defect, or cloacal extrophy, is anouthe unusual form of omphalocele. Abdominal wall defect is infraumbilical and accompanied by bladder extrophy, epispadias, diastasis of the pubic rami, and imperforate anus.

Prune belly (Eagle Barrett) syndrome occurs as a result of increased apoptotic cell death in the body wall placode, which leads to insufficient mesodermal cell deposition.

Underdevelopment of the muscles of the anterior abdominal wall can be of varying degrees. All muscles of the anterior abdominal wall may be absent if the pathology occurs in the early stages of fetal development, when muscle development is just beginning. More often there is a partial absence of transverse and oblique muscles, because the violation of embryogenesis occurs in the late stages of fetal development. The defect is more common in boys Clinic. This pathology is characterized by a triad of symptoms: bilateral cryptorchidism, muscle aplasia, dysfunction of the urinary system - Parker's syndrome. A characteristic feature is a flaccid and thin anterior abdominal wall, through which the contours of the abdominal cavity are contoured, noticeable intestinal motility. The abdomen is spread out like a frog's belly. Palpation in the protrusion reveals the organs of the abdominal cavity. The diaphragm in such children is high, resulting in shortness of breath, stagnant wheezing in the lungs. A wide rib angle is detected.

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Tests for initial level of knowledge

1. The earliest way to diagnose an anterior abdominal wall defect is:
   A. by physical exam
   B. by history
   C. by fetal ultrasound
   D. by fetal CT scan.

2. The following are correct regarding omphaloceles except:
   A. is usually covered by a translucent membrane
   B. is frequently associated with other congenital malformations
   C. is lateral to the umbilical stump
   D. is within the umbilical ring.

3. The following are true about gastroschisis:
   A. occurs lateral to the umbilical stump
   B. can be diagnosed antenatally
   C. at birth often have edematous matted intestinal loops
   D. all of the above.

4. Treatment of abdominal wall defects includes:
   A. immediate surgical repair
   B. pushing the intestines back into the abdominal cavity while still in the delivery room
   C. provide immediate optimal resuscitation and stabilization first, and then surgery
   D. always do primary closure in both lesions.

5. Gastroschisis is a malformation of the abdominal wall that
   A. is commonly associated many different genetic disorders
   B. usually occurs to the left of the umbilicus but the umbilicus is intact.
   C. only involves a few layers of the abdominal wall.
   D. has no membranous covering.
   E. allows abdominal contents to herniate through the base of the umbilicus.

6. The incidence of gastroschisis worldwide is about
   A. 1 per 10,000 live births.
   B. 2 per 10,000 live births.
   C. 3 per 10,000 live births.
   D. 4 per 10,000 live births.
   E. 5 per 10,000 live births.

7. Omphalocele (Exomphalos) is a midline defect
   A. where the abdominal contents herniate to the right side of the umbilicus.
   B. that rarely contains the liver, bowel, and stomach.
   C. that allows abdominal contents to herniate through the base of the umbilicus.
   D. that has no membrane covering.
   E. where the abdominal contents herniate to the left side of the umbilicus.

8. Gastroschisis is a defect resulting from a vascular compromise of
   A. the left umbilical vein
   B. the superior mesenteric artery
   C. the inferior mesenteric artery

D. the splenic artery
E. the omphalomesenteric artery

9. The most common associated anomaly(s) seen with gastroschisis is (are)
   A. prune belly syndrome
   B. genitourinary tract abnormalities
   C. congenital heart defects
   D. intestinal tract complications
   E. hydrocephalus
10. Associated anomalies seen with gastroschisis include all of the following except
   A. intestinal tract complications such as malrotation, atresia, and stenosis.
   B. ischemia of bowel sections due to kinking, and obstruction.
   C. congenital heart defects
   D. genitourinary tract abnormalities

Keys for tests

A. C
B. C
C. D
D. C
E. D
F. A
G. C
H. E

Tests for final level of knowledge

1. Omphalocele can be seen with numerous different chromosome abnormalities including triploidy, which is a fetus with
    A. 45 chromosomes missing an X
    B. 47 chromosomes with an extra X
    C. 47 chromosomes with an extra Y
    D. 47 chromosomes with an extra number 13
    E. 69 chromosomes
2. During the development of the embryo, if the caudal fold tissue fails to fuse, it results in
    A. Cloacal extrophy of the bladder
    B. Omphalocele
    C. Gastroschisis
    D. Pentalogy of cantrell
    E. Ectopia cordis
3. During the development of the embryo, if the lateral tissue folds fail to fuse, it results in
    A. Cloacal extrophy of the bladder
    B. Omphalocele
    C. Gastroschisis
    D. Pentalogy of cantrell
    E. Ectopia cordis
4. Beckwith-Wiedemann syndrome may be associated with all of the following except
    A. An enlarged liver
    B. Enlarged kidneys
    C. Growth restriction
    D. Omphalocele
    E. Macroglossia
5. The ultrasound findings for gastroschisis may include all of the following except
    A. Multiple echogenic free floating bowel loops near the anterior abdominal wall
    B. No evidence of a membranous covering or sac
    C. Polyhydramnios
    D. Oligohydramnios
    E. An umbilical cord that inserts into the mass
6. Factors that can sometimes make it difficult to identify gastroschisis include all of the following except
    A. the size of the defect
    B. the fetal size
    C. the gestational age
    D. too much amniotic fluid (polyhydramnios)
    E. the fetal position
7. It is important not to make an incorrect diagnosis of a fetal anomaly between the 8th and 12th week of gestation because
    A. The fetal liver and spleen are not completely formed yet.
    B. The embryonic bowel normally protrudes into the base of the umbilical cord during this time period and could mimic an anomaly.
    C. The amount of amniotic fluid is too low at this point in gestation to make an accurate assessment.
    D. The fetal stomach bubble is not visible enough to confirm the presence of bowel.

E. The umbilical cord at this gestational age looks like bowel on ultrasound imaging.

8. The ultrasound findings for omphalocele include
   A. A solid appearing, round, echogenic mass adjacent to the anterior abdominal wall
   B. An umbilical cord that inserts to the right of the mass
   C. An umbilical cord that inserts to the left of the mass
   D. No evidence of a membranous covering or sac
   E. A herniation of abdominal contents to the right of the umbilicus
9. With a diagnosis of gastroschisis,
   A. the fetus should have follow-up ultrasounds to watch for macrosomia.
   B. the fetus should have follow-up ultrasounds to watch for oligohydramnios.
   C. karyotyping (genetic amniocentesis) should always be recommended because most cases of gastroschisis are associated with chromosome abnormalities.
   D. the patient should deliver by cesarean section
   E. if the case is an isolated gastroschisis the overall prognosis is very poor with a survival rate of less than 10%.
10. Regarding a potential diagnosis of gastroschisis, one potential concern that may result in some prenatal testing centers offering further genetic testing, such as amniocentesis, is
    A. the issue that an omphalocele may have ruptured through its membrane covering, mimicking the appearance of a gastroschisis.
    B. the presence of bowel ischemia due to kinking.
    C. the presence of oligohydramnios.
    D. the presence of an elevated maternal serum alpha-fetoprotein.
    E. the presence of bowel thickening due to prolonged exposure to amniotic fluid.

Tasks for final level of knowledge

Task 1. A 25 year second gravida female was sent for routine antenatal ultrasound at 27 weeks of gestation. Her last pregnancy was normal full term hospital delivery 4 years back with delivery of normal healthy child. She didn't have any major disease and there was no family history of delivery of abnormal child. Maternal serum AFP was not tested. Her calculated gestational age by ultrasonography was approximately 26 weeks and 2 days. On scanning of abdomen, there was large anterior abdominal wall defect in the midline with herniation of liver and bowel loops through it. The contents were covered with a membrane. Umbilical cord was inserted in the center of the covering membrane.

1. What is the diagnosis?
2. What additional investigations are needed?
3. What is the best place for delivery?
4. What is the prognosis?

Answer Key.

1. This is a typical prenatal ultrasound features of an omphalocele.
2. Maternal serum AFP, karyotype analysis by amniocentesis or chorionic villous sampling.
3. Infants with an abdominal wall defect should be delivered at a perinatal center, where immediate neonatal and surgical expertise are available.
4. Prognosis depends of additional anomalies.

Task 2. A 31-year-old women was referred for evaluation due to the finding of an omphalocele by routine second trimester screening ultrasound. The pregnancy course had been uncomplicated up to this point, including normal prenatal labs. Serum screening had been declined by the patient. Her obstetric history is significant for three full-term vaginal deliveries without

complication. The remainder of her history and the family history is unremarkable. No environmental exposures were elicited.

1. What is the plan of next evaluation?
2. Is a karyotype indicated in this situation?

Answer Key

1. Comprehensive evaluation performed at 25-weeks gestation included detailed ultrasound, echocardiography.
2. Karyotype is necessary if omphalocele is diagnosed.

Task 3. Comprehensive evaluation of 38 year old pregnant women performed at 25-weeks gestation included detailed ultrasound, echocardiography and ultra rapid fetal MRI. Biometrics were appropriate for gestational age. Ah abdominal wall defect containing liver, gallbladder and bowel was seen. Gender could not be determined as the finding in the ultrasound image distorted the visualization between the fetal legs. The pubic bones were splayed and no bladder could be identified. Further imaging of the fetal spine was seen in the MRI demonstrating a neural tube defect. The left kidney could not be visualized and amniotic fluid was normal. A two-vessel umbilical cord was noted. Fetal echocardiogram was normal.

1. Based on the constellation of findings, what is the likely diagnosis?
2. What is the significance of the ultrasound finding?
3. What are associated anomalies seen with this disorder?

Answer Key

1. Large omphalocele, exstrophy of the cloaca, imperforate anus and spinal defects.
2. The finding is the classic appearance of the “elephant’s trunk” in which the terminal ileum prolapses through the cecum and is often seen between the fetal legs. This can also further obscure gender determination.
3. Single umbilical artery, limb defects (arthrogryposis, club feet, syndactyly), cardiac, gastrointestinal (atresia, malrotation, bowel duplication), neurologic (hydrocephalus) and urogenital (renal agenesis, ectopic kidney, duplicated collecting system, ambiguous genitalia, abnormal scrotum, uterine abnormalities) anomalies have been described.

THEME 11

CONGENITAL ABNORMALITIES OF THE LIVER AND BILIARY TRACT: BILIARY ATRESIA (BA) AND CHOLEDOCHAL CYST (CC).

Overview

Jaundice in newborn is one of the most common reasons when a child is brought to medical attention. Late diagnosis of biliary atresia leads to development of liver fibrosis, cirrhosis, portal hypertension, and following end stage liver disease. Early diagnosis is essentialy important for successful surgical treatment.

Choledochal cyst is a congenital anomaly involving cystic dilation of various ducts of the biliary tree. A monograph published by Vater in 1723 presented a series of studies describing normal and abnormal anatomy of the biliary tree, including the first description of a fusiform dilatation of the common bile duct. In 1959 Alonso-Lej published the first series of patients with choledochal cysts. This landmark paper was the first to describe a classification and led to a better understanding of the pathophysiology of this anomaly, which in turn has led to new approaches in treatment. There is a well-documented female dominance (4 to5:1). There are some theories of etiology of choledochal cyst: congenital weakness of the common bile duct, improper formation of the bile ducts during the primitive stages of the development, an anomalous arrangement of pancreaticobiliary ductal system, abnormal insertion of the duct into the duodenum and neuromuscular incoordination of the sphincter.

Educational aims:

The aim of this part of module is to provide help in identifying children with biliary atresia who need surgical intervention, and to provide guidance on the initial stabilization and management of a variety of common surgical problems.

A student must know:

1. Relevant anatomy of biliary tree and abdominal cavity in children.
2. Classification of biliary atresia.
3. Classification of choledochal cyst by Alonso-Lej, Todani modification.
4. Etiology, pathophysiology, clinical presentation and management obstructive cholangiopathy.
5. The classic clinical manifestation of biliary atresia.
6. Features of clinical presentations of biliary atresia in newborn.
7. Lab studies and imaging studies in children with biliary atresia.
8. Differential diagnosis of biliary atresia and other diseases manifesting jaundice.
9. Complications of biliary atresia. Clinical presentations of the liver cirrhosis.
10. Preoperative management of children with biliary atresia.
11. Postoperative complications and management after Kasai’s operation.
12. The classic clinical manifestation of the choledochal cyst.
13. Features of clinical presentations of choledochal cyst in newborns and children.
14. Lab studies and imaging studies in children with choledochal cyst.
15. Differential diagnosis of choledochal cyst, other cysts of abdominal cavity, and infectious hepatitis.
16. Complications of choledochal cyst. Clinical presentations of the cholangitis.
17. Treatment of choledochal cyst.
18. Advantages and disadvantages of laparoscopic cystectomy.
19. Postoperative complications after cystectomy (open and laparoscopic).
20. Liver transplantation in children.

A student must be able to:

1. Identify BA:
   - Color of stool in newborn.
   - Enlargement if liver.
   - Other causes of jaundice in newborn.
2. Color of skin, hemorrhages on the skin.
3. Make the physical exam of children**.**
4. Look for evidence of peritoneal irritation
5. Use lab tests to support the diagnosis
   - CBC to look for an elevated leukocyte count and formula's shift to the left.
   - Urinalysis
   - Liver function tests (direct and indirect bilirubin, alaninaminotransferase (ALT), aspartataminotransferase (AST), gammaglutamilaminotransferase (GGT), alkalin phosphatase (AP).
6. Identify CC:
   - Point of pain in the right subcostal area.
   - Evidence palpable tumor in right subcostal area.
   - Jaundice, acholic stool.
   - Vomiting.
7. Palpation of anterior abdominal wall.
8. Make the physical exam of children with choledochal cyst.
9. Look for an evidence or absence of peritoneal irritation
10. Use lab tests to support the diagnosis:
    - Liver function tests (direct and indirect bilirubin, alaninaminotransferase (ALT), aspartataminotransferase (AST), gammaglutamilaminotransferase (GGT), alkalin phosphatase (AP), serum amylase levels.
    - Blood tests for hepatitis profile.
    - Urinalysis.
11. Be able to interpret ultrasound data, CT, MRI cholangiography in the pediatric population.

TERMINOLOGY

Term	***Definition***
Biliary atresia	Obstructive progressive cholangiopathy
Obstructive jaundice	Hyperbilirubinemia due to mechanical reasons
Acholic stool	Noncoloration of stool due to pigments absence.
"Fibrous remnants"	Biliary atresia represents a necroinflammatory destructive cholangitis, resulting in progressive destruction and obliterating fibrosis of the ducts.
Cirrhosis	Diffuse liver process characterized by fibrosis and the conversion of normal liver architecture into structurally abnormal nodules.
Cholangitis	Infection of biliary system.
Portal hypertension	Portal hypertension is defined as an elevation of portal blood pressure above 5 mm Hg.
Kasai operation	Hepatoportojejunostomy with Roux-en-Y jejunojejunostomy.
Choledochal cyst	Choledochal cysts are considered to be congenital anomalies of the biliary tract characterized by varying

	degrees of cystic dilatation at various segments of the biliary tract (extrahepatic or intrahepatic.
Triad of Villard	The classic triad of intermittent abdominal pain, jaundice, and right epigastric mass.
Jaundice	Jaundice (conjugated hyperbilirubinemia) is the most common manifestation of CC.
Abdominal pain	May be a presenting symptom, often with elevated serum amylase levels.
Ultrasonography	Should be a the initial procedure in the evaluation of suspected choledochal cyst.
Spontaneous perforation .	Spontaneous perforation of CC in infancy is not a rare event.
Cholangitis	Chronic or acute inflammation caused by bacteria entering the bile ducts due to reflux of the intestinal content to biliary tract.
Pancreatitis	Persistent or recurrent infection of pancreas , may occur secondary to proximal pancreatic duct or sphincter stenosis or stones.

CONTENT

Biliary atresia (BA)

Biliary atresia is a disorder of the newborn, occurring in 1 in 15000 live birth worldwide. It is obstructive obliterative cholangiopathy characterized by inflammation of the bile ducts leading to progressive fibroprofilative obliteration of the extrahepatic biliary tree and extent to the intrahepatic bile ducts. The progressive destruction of the bile ducts leads to cholestasis, liver fibrosis, and cirrhosis. Without definitive surgical therapy, the natural history invariably includes progressive liver fibrosis, cirrhosis, end stage liver disease, portal hypertension, and death, usually within 2 years.

Fig. 11.1 Patient with biliary atresia

Diagnosis

The typical patient with biliary atresia is a full-term, healthy appearing infant with normal birth weight, who develops jaundice that persists at 4-6 weeks and is associated with acholic stools, dark urine, and hepatomegaly (Fig 1.). There is often a delay in diagnosis because nearly two thirds of all newborns develop jaundice, overwhelming majority of which are due to physiologic jaundice (usually lasts only 2-3 days) or breast milk jaundice (can last up to 4 weeks). Only a small part of these cases are due to neonatal cholestasis (1/500 cases of jaundice in infants between 2-4 weeks old). Nevertheless, biliary atresia should be suspected in any infant who remains jaundiced for

more than 2 weeks (3 weeks if breast-fed). From the major reports in the literature, it has been found that both the jaundice clearance rate and the long-term outcome are related to the age at surgery. It is therefore mandatory to establish an early diagnosis so timely surgery can be performed.

An infant with evidence of cholestasis (elevated total bilirubin with greater than 20% conjugated) must undergo a complete workup to rule out both extrahepatic and intrahepatic causes. The evaluation of extrahepatic causes of neonatal jaundice starts with laboratory tests demonstrating increasing serum alanine aminotransferase (ALT), aspartate aminotransferase (AST), alkaline phosphatase, and especially gamma-glutamiltranspeptidase (GGT), which is usually disproportionally elevated in infants with biliary atresia. Because the cause of pathologic jaundice in an infant is diverse, biochemical evaluation must include hepatitis serologies, TORCH screening (toxoplasmosis, rubella, cytomegalovirus, and herpes simplex), α_1-antitrypsin level, serum ferritin, and sweat chloride test or direct genetic analysis to rule out cystic fibrosis.

A fasting abdominal ultrasound will demonstrate either a contracted or absent gallbladder (Fig.11.2). Other findings that have been reported with a focused and detailed ultrasound include abnormal gallbladder configuration, shape or contractility, nonvisualisation of the common bile duct, enlarged hepatic artery diameter, and a focal area of increased echodenesity anterior to the bifurcation of the portal vein (*the triangular cord sign*), which represents the fibrotic remnant of the extrahepatic biliary tree. Ultrasound does not allow us to confirm the diagnosis of biliary atresia but it can support the diagnosis, and is useful to exclude other extrahepatic causes of neonatal cholestasis, such as a choledochal cyst. Percutaneous liver biopsy is often the next step in the diagnostic workup. In infants with biliary atresia, there will be expanded fibrous portal tracts with edema, fibrosis, and inflammation, bile ductile proliferation, and canalicular and bile duct plugs. These histologic findings are diagnostic of neonatal cholestasis, they are nonspecific. Uncorrected biliary atresia results in histologic evidence of cirrhosis as early as 3 to 4 months of age.

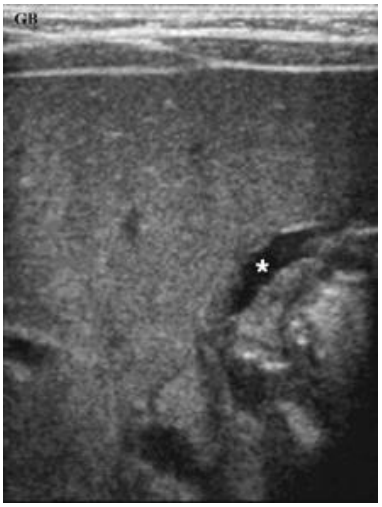


Fig. 11.2 Abdominal ultrasound demonstrates a contracted gallbladder (white star - GB)

Endoscopic Retrograde Cholangiopancreatography (ERCPG) and Magnetic Resonance Cholangiopancreatography (MRCP) are not routinely used because of limited availability and experience with mentioned procedures in neonates. The final step and still the gold standard for the diagnosis of biliary atresia is to demonstrate an atretic biliary tree by abdominal exploration and intraoperative cholangiogram.

Treatment.

Hitherto Kasai's portoenterostomy procedure, when performed early in life before 8-10 weeks of age, gives best results for biliary drainage, and widely accepted as the gold standard

surgery for biliary atresia. The goal of the hepatoportojejunostomy (Kasai operation) is to establish the bile flow from still patent bile ductuli below the fibrous plate of the liver capsule (Fig.3).

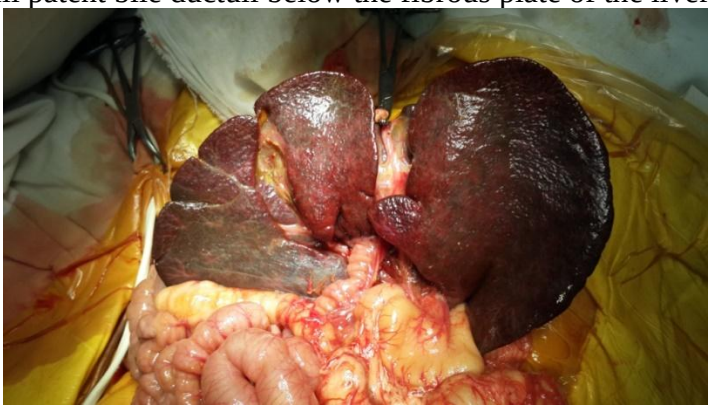


Fig.11.3 Kasai procedure – hepatoportojejunostomy

Anesthetic management

General endotracheal anesthesia is administered by standard technique. An urinary catheter and nasogastric tube are placed. A weight-based dose of prophylactic parenteral antibiotic covering gram-negative enteric bacteria is administered 30 minutes before skin incision.

The operation is divided into two stages: diagnostic confirmation and Kasai portoenterostomy.

Operative procedure

Step 1: Diagnostic confirmation.

The liver in biliary atresia is characteristically firm, dark, and cholestatic in color, typically has many subcapsular telangiectatic vessels on its surface, representing early cirrhosis and portal hypertension changes (Fig.4).

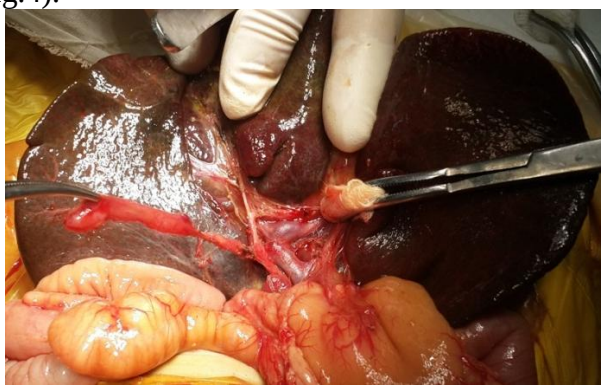


Fig.11.4 The liver in biliary atresia: view after hilum dissection (BA type IIb)

Intraoperative cholangiography is performed by placing a 20-gauge intravenous catheter into the gallbladder remnant under direct vision and securing it with a purse-string suture. No passage of contrast into the liver confirms the diagnosis of biliary atresia. Intrahepatic flow into anatomically correct ducts, even when they are small, is inconsistent with biliary atresia, and Kasai portoenterostomy is not indicated in such scenario.

Step 2: Kasai Portoenterostomy.

The initial dissection of the fibrous remnant begins just below the level of the cystic duct junction with the fibrous mass. Superior dissection of the fibrous tissue proceeds just anterior to the portal vein and hepatic artery. Dissection should proceed until the fibrous remnant has come into approximation with the capsular surface of the liver within the bifurcation of the portal vein. Division of the fibrous mass is then performed. The plane of division should be at the level of the surface of the liver capsule. There is no advantage of deep dissection (“taking a core”) at the location of the fibrous mass. A smooth, glistering cut surface should be a result. Hemostasis is achieved by placing an epinephrine-soaked gauze and gentle pressure (*never use electrocautery!*). The specimen should be oriented and sent to pathology.

While awaiting hemostasis at the site of the divided fibrous mass it is time for creation a Roux-en-Y limb. The proximal jejunum is divided 20-30 cm distal to the ligament of Treitz and a

20-30 cm Roux-en-Y limb is created by performing an end- to- side jejuno-jejunostomy with a single layer of interrupted absorbable sutures. The mesenteric defect is closed and the Roux-en-Y limb is passed through a defect created in the transverse mesocolon so as to lie in a retrocolic position. End of the limb is sutured to the porta hepatis using interrupted fine absorbable monofilament sutures. The posterior rim of the anastomosis, between the portal vein and the fibrous plate, is performed with horizontal mattress sutures add the knots tied internally. The anterior half of the anastomosis is then completed with the knots tied externally.

To reduce the risk of postoperative cholangitis it is necessary to create an antireflux valve within the jejunal loop, going from porta hepatis. Drainage of the anastomotic area is optional. The viscera are returned to the abdominal cavity, hemostasis is confirmed, and the incision is closed in layers with absorbable sutures (Fig.5).

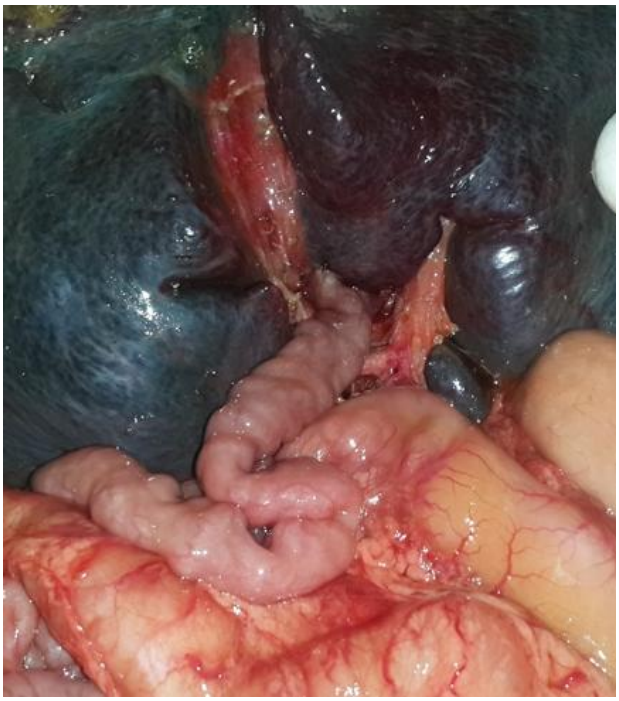


Fig.11.5- final view of Kasai procedure

Postoperative care.

Patients are usually maintained on bowel rest, with a nasogastric tube. The clinical hallmark of a functioning portoenterostomy is the production of pigmented stools. Stools should be green, tawny or brown. Persistently acholic stools, even when intermittent, are concerning for inadequate bile drainage. Prophylactic antibiotics should be continued throughout the peri-operative period.

Corticosteroids are thought to stimulate bile flow by inducing canalicular electrolyte transport. Some believe they might also limit the progression of bile duct injury and fibrosis and prevent the closure of microscopic bile ducts through their anti-inflammatory and immunosuppressive effects. Reports from Japan suggests significant benefit with an aggressive regimen that includes prednisolone 10 mg intravenous twice a day for 7 days, then 20 mg orally every day for 4 days, gradual tapering of subsequent doses, and stopping when the total bilirubin level will be normalized. Ursodeoxycholic acid is another therapeutic option used frequently to encourage bile flow and decrease the toxicity of circulated bile acids.

Most patients with biliary atresia have ongoing issues with malnutrition, mainly due to increased caloric needs in the setting of fat malabsorption and fat-soluble vitamin deficiencies. They require life-long nutritional support with oral fat-soluble vitamin supplementation and a high-calorie, high-protein diet with at least 125% of recommended caloric intake. Although the

exact numbers may vary, the overall survival rate of children after surgical treatment for biliary atresia is approximately 60% at 5 years and 35% at 10 years.

Liver transplantation.

Liver transplantation is the only treatment option providing long-term survival in patients who fail to establish active bile drainage following Kasai portoenterostomy. Even in patients who underwent successful Kasai procedure, progression of hepatic fibrosis and cirrhosis anyway occurs. Such patients manifest a sequel of portal hypertension, including ascites, hypersplenism, and variceal hemorrhage. Patients with worsening liver function (e.g., hyperbilirubinemia, coagulopathy, hypoalbuminemia) are considered for liver transplantation. Outcome after liver transplantation is excellent, with 1-,5-, and 10-year patient survival of 95, 90, and 88% correspondingly, and graft survival rates of 87,82, and 81%.

Choledochal cyst (CC).

Choledochal cysts (CCs) are one of the more common bile duct anomalies in children and occur in one in 10,000 live births. The malformation more common in girls.

Three patterns of presentation have emerged before in 1959 Alonso-Lej published their landmark paper describing their classification of ductal dilation. The first is a cystic mass in abdomen identified prenatally, the second is jaundice presenting in infancy, and the third is ascending cholangitis, obstructive jaundice, or pancreatitis presenting in later in childhood. The anatomic configuration of CCs were first classified by Alonso-Lej and subsequently modified by Todani into five subtypes (Fig.11.6)

A Type 1

This is by far the most common form of cyst. It involves most of the common bile duct, cystic duct, gall bladder and common hepatic duct. They frequently communicate with the duodenum trough a lumen so small that it can barely be received at surgery.

- For a type 1 CC, the dilation is confined to the common bile duct. Isolated choledochal dilatation in Type 1 is additionally divided into three types: fusiform type A, tubular type B, and cystic type C.
- The dilation in patients with a type 4 CC either extends in continuity into the intrahepatic biliary tree or there may be a short portion of normal-caliber common hepatic duct with isolated or multifocal intrahepatic duct dilatation.

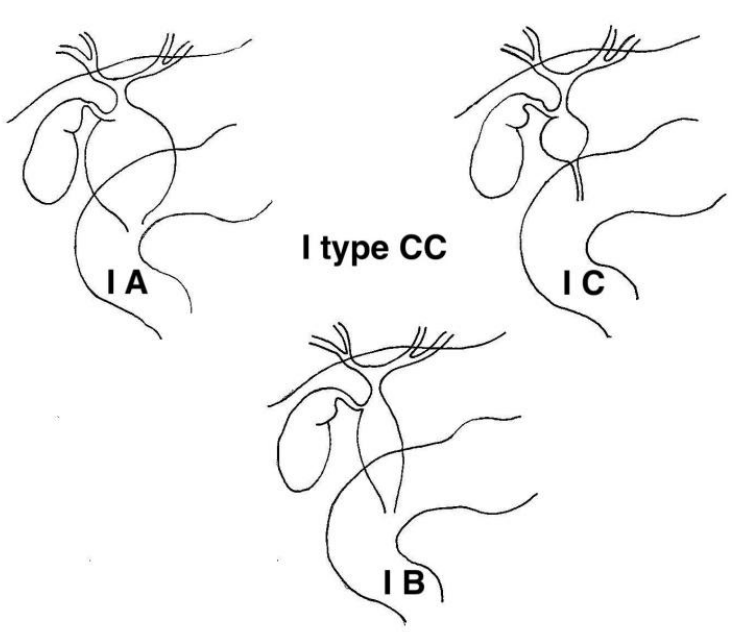


Fig.11.6 Anatomic classification by Alonso-Lej, subsequently modified by Todani, type I

A Type 2

It is diverticulum arising from the wall of the bile duct. This abnormality is thought to occur because of a localized weakness in a segment of the common bile duct. This type of cyst is an outpouching or diverticulum of the common bile duct.

A Type 3

Also known as a choledochocele, Type 3 CC consists of a dilatation at the distal end of the common bile duct that protrudes into the lumen of the duodenum. Endoscopically, its appearance is similar to that of a ureterocele found within the bladder. This type is located at the distal end of the common duct. Often it is completely contained within the duodenal wall. It causes obstruction through compression of the normal duct and is sometimes considered a duplication cyst.

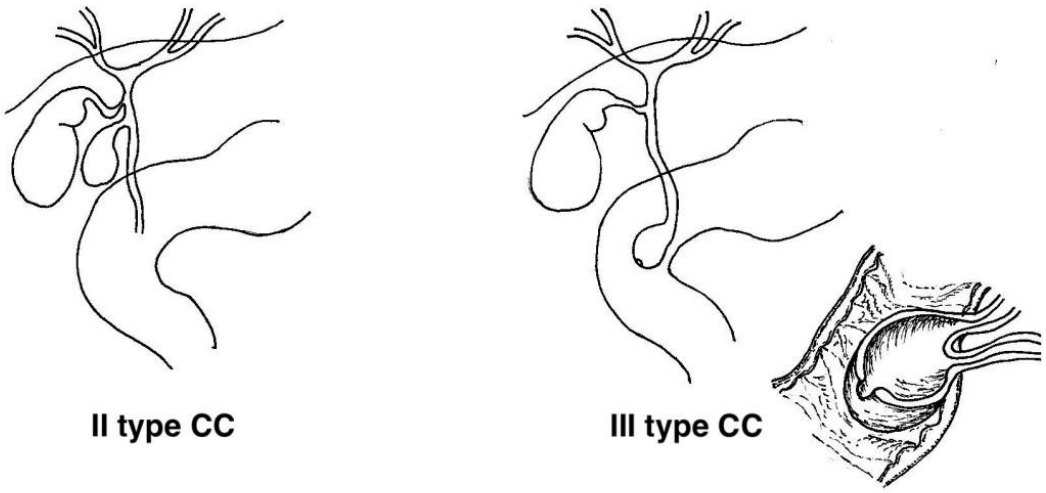


Fig.11.7. Anatomic classification by Alonso-Lej, subsequently modified by Todani, types II, III

A Type 4 (a, b)

This cystic lesion involves both the intrahepatic and extrahepatic biliary system.

A Type 5

This type are single or multiple intrahepatic cysts. There is no extrahepatic component in this type. The intrahepatic biliary cysts may be localized to one lobe or segment, but is it usually a bilateral diffuse process. This lesion often called Caroli's disease. Caroli's disease often occurs in association with hepatic fibrosis and polycystic kidney disease.

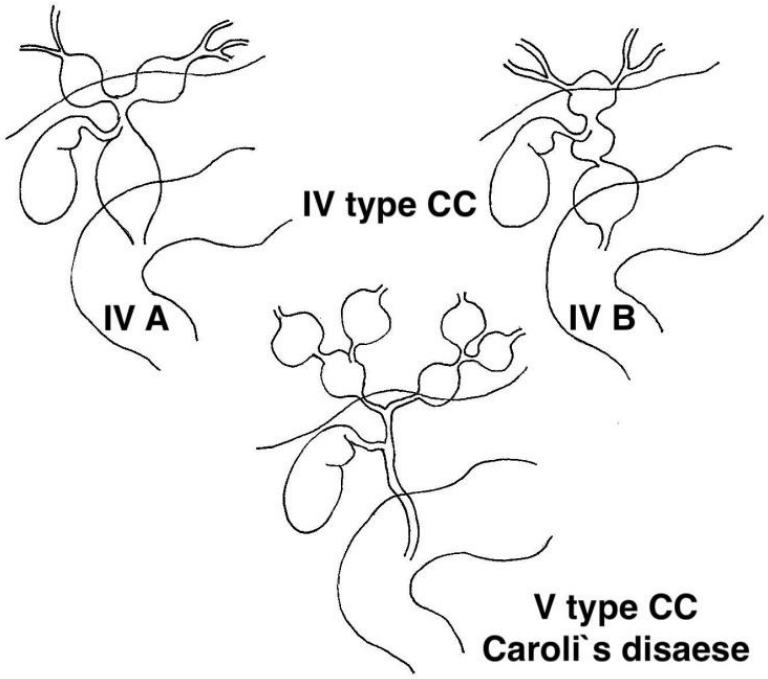


Fig.11.8. Anatomic classification by Alonso-Lej, subsequently modified by Todani, types IV, V

Presentation

Classically a patient presents abdominal pain, jaundice, and a right upper quadrant mass; however, rarely are all three all three symptoms are present at diagnosis. Some patients are diagnosed with cholangitis when they present with Charcot triad: jaundice, fever, and right upper quadrant pain. Others have recurrent episodes of pancreatitis as a result of sludge within a CC, which causes intermittent obstruction of the pancreatic duct. A giant CC can spontaneously perforate. Because bile causes chemical peritonitis, these patients require urgent care. A cystic anomaly of the biliary tract can be detected prenatally. The differential diagnosis is a CC or cystic variant of biliary atresia. If the biliary tract is obstructed, biliary atresia is likely. If the biliary tract is patent, a CC is likely.

Diagnosis

If choledochal cyst is suspected, the diagnosis is confirmed by blood chemistry studies and abdominal ultrasonography. All liver function tests are elevated during period of acute pain: direct hyperbilirubinemia, elevated GGT (gammaglutamyltransferase), and modest elevation of transaminase values. Serum amylase and lipase can be elevated if there is an accompanying pancreatic inflammation.

Abdominal ultrasound is the imaging test of choise for making the diagnosis of CC. A cystic dilatation of the common bile duct and gallbladder is the most common finding. The dilatation can extend into the common hepatic duct, but typically does not extend into the hepatic ducts.

Cross-sectional imaging of the abdomen is necessary to define the anatomic configuration of a CC. Computerized tomography or magnetic resonance imaging should be used to define the anatomic extent of disease (Fig.11.7). Hepatobiliary scintigraphy (HIDA) scan is helpful to determine patency of the biliary tract. On occasion, endoscopic retrograde cholangiography (ERCPG) or, more recently, magnetic resonance cholangiopancreatography may be helpful to clearly define the anatomy, especially in cases when the cyst size is small.

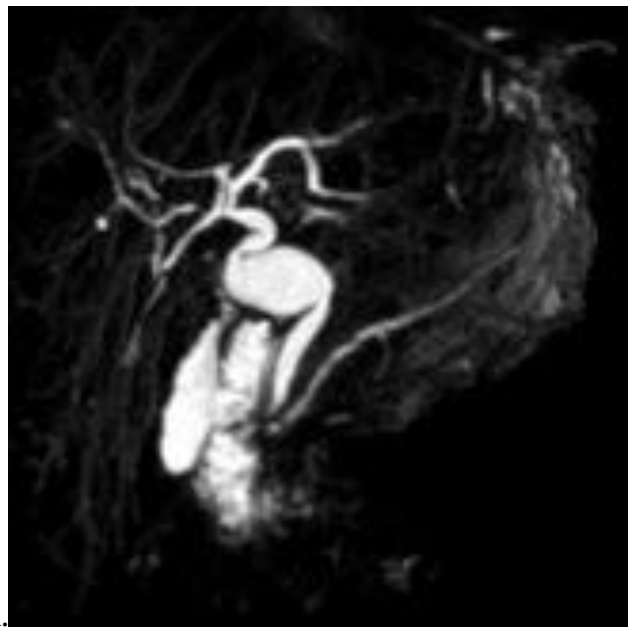
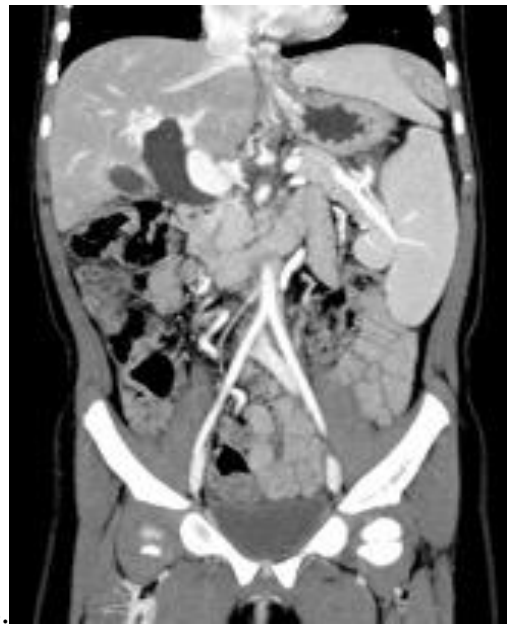
A.  B. 

Fig.11.9 Image of CC magnetic resonance cholangiopancreatography (A) and CT scan (B)

Pathology

Examination of the cyst wall demonstrates chronic inflammatory changes and often complete denaturation of epithelium. In undiagnosed or incorrect treated cysts, chronic epithelial inflammation rarely leads to malignant degeneration. Cholangiocarcinoma has been reported in choledochal cysts first diagnosed in adults and in cyst remnants not resected after childhood diagnosis.

Treatment

Treatment consists of cyst excision and biliary reconstruction with a Roux-en-Y choledochojejunotomy for all type 1 and for type 4 cysts. In the past, anastomoses of a loop of bowel to either the gall bladder or cyst wall to re-establish bile drainage (operations of internal drainage) was considered adequate treatment. The realization that malignancies can develop in cyst remnants therefore the cyst is to be resected radically.

For type 2 cyst resection should be made with primary repair of the common duct.

Type 3 cysts are resected via a transduodenal approach.

Type 5 (and some type 4) lesions pose a more difficult treatment problem. For intrahepatic cysts localized to one lobe or segment, hepatic resection is only rarely beneficial. Many of these patients may eventually require liver transplantation.

Prenatal diagnosis of choledochal cysts is more common recently. Cysts rarely cause problems in the newborn period, and therefore it is almost never necessary to operate right after birth. A period of 4-6 weeks for observation is usually a good idea. A waiting period allows the baby to grow and may lower anesthetic and surgical risks.

Preoperative preparation**.**

Before induction of general anesthesia. A saline enema is given on the evening before surgery to ensure cleansing of the large bowel.

Operative treatment.

Types 1 and 4 Choledochal cysts.

A right subcostal incision is used to enter the peritoneal cavity. An intraoperative cholangiogram performed through the gallbladder is helpful to determine where the proper hepatic duct appears normal and where the pancreatic duct joins the biliary tract. The common duct is circumferentially mobilized, which allows the application of the traction to the CC, during which identification of the hepatic artery and portal vein can be performed. Dissection should proceed distally until the duct narrows to normal caliber where the cyst can be divided. Biliary drainage is achieved by an end-to-end hepaticojejunostomy into a 40-50 –cm limb of a Roux-en-Y brought to

the hilum in a retrocolic position. A mucosa-to- mucosa anastomosis is important because it limits the likelihood of a stricture in postoperative period. It is used a single-layer interrupted anastomosis using absorbable suture to complete the hepaticojejunostomy. Sutures are placed the way all the knots lie in an extraluminal position, thereby reducing the likelihood of choledocholithiasis. For reducing the risk of postoperative cholangitis is need creating of antireflux valve on the jejuna loop, going from porta hepatis.

Type 2 Choledochal cyst.

For patients with a type 2 CC, excision of the diverticulum and the extrahepatic biliary tract with a Roux-en-Y hepaticojejunostomy to re-establish drainage of the proximal bile ducts is the procedure of choice. Although simple excision of the diverticulum with ligation at its base can be performed, excising of the extrahepatic biliary tract is generally recommended because of the risk of the development of a cholangiocarcinoma in the remaining extrahepatic biliary tract.

Type 3 Choledochal cyst.

For patients with a type 3 CC, the recommendation for treatment vary according to the type of epithelium found within the CC. An ERCP should be performed before surgical intervention. At the time of ERCP, biopsy of the mucosa lining the cyst should be performed. If the biopsy reveals mucosa of duodenal origin, the lesion can be treated by a sphincterotomy or meatotomy to enlarge the opening and to relieve the obstruction. This can be done endoscopically or with open surgery.

Postoperative Care.

Postoperative antibiotics.

Suppressive antibiotic started and maintained for at least 5 months. If no episodes of cholangitis occur during the 5 months period, antibiotics are discontinued.

The use of Ursodeoxycholic acid may be beneficial in the postoperative period, but if no complications arise, it can be discontinued.

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Tests for initial level of knowledge

1. What is the most common symptoms of the biliary atresia?
    A. Jaundice with colored stool
    B. Hemolytic anemia
    C. Conjugated hyperbilirubinemia from first living day, acholic stool, absence of gallbladder at the ultrasound.
    D. Jaundice with unconjugated hyperbilirubinemia.
    E. Congenital syphilis.

2. Which of the following is the complication of biliary atresia?
    A. Cholangitis.
    B. Encephalopathy.
    C. Fibrosis with following cirrhosis.
    D. Ileus.
    E. Pneumonia.
3. All mentioned are common complications of biliary atresia, except?
    A. Biliary cirrhosis.
    B. Malnutrition.
    C. Encephalopathy.
    D. Hemorrhages.
    E. Umbilical hernia.

4. All mentioned features of biliary atresia, except?
    A. Jaundice from first days of life.
    B. Uncolored stool.
    C. Direct hyperbilirubinemia.
    D. Absence of normal gallbladder in ultrasound.
    E. Jaundice with pigmented stool.

5. What is the critical term of surgical treatment of biliary atresia?
    A. Before 1 year.
    B. Before 6 months.
    C. Before 2 years.
    D. Before 2 months.
    E. At 2 weeks of life.
6. Which of the following diseases present with obstructive jaundice?
    A. Neonatal hepatitis.
    B. Hemolytic disease of newborn.
    C. Infectious hepatitis.
    D. Biliary atresia.
    E. Hepatitis B.
7. What is biliary atresia?
    A. Progressive obstructive cholangiopathy.
    B. Congenital absence of gallbladder.
    C. Alagille syndrome.
    D. Byler disease.
    E. Cholestasis.
8. What is the best initial investigation in patients with suspected biliary atresia?
    A. Ultrasonography.
    B. Plain X-ray in vertical position.
    C. Liver biopsy.

D. Duodenal intubatioin.
E. Cholangiography.

9. Who is most likely to develop biliary atresia?
    A. Newborn
    B. Children between 3 and 6 years
    C. Children younger than 3 years
    D. Toddlers
    E. Teenagers
10. What is the most common indication for liver transplantation in children?
    A. Allagille syndrome.
    B. Byler disease.
    C. Biliary atresia
    D. Neonatal hepatitis.
    E. Methabolic diseases of liver.

Keys for tests:

1. C
2. C
3. C
4. E
5. D
6. D
7. A
8. A
9. A
10. C

Tests for final level of knowledge for BA

Task 1. You see a 1 months girl with jaundice. She has had jaundice from the first days of life. The stool has acholic character. The jaundice was referred by nurse as physiologic. Liver enlarged to 2 cm below rib arch. You perform a blood test that is normal. What should you do next?

A. Arrange to admit her to hospital.
B. Reassure her mother that it is most likely a biliary atresia.
C. Perform ultrasound, liver tests, MRI cholangiography.
D. Start preoperative treatment.

Task 2. Newborn has a jaundice from first days of life. In blood analysis indirect hyperbilirubinemia, ALT and AST elevated. Stool is periodically acholic. What diagnosis is suspected? What should you do next?

Answer Key: neonatal hepatitis is suspected. You should: perform abdominal ultrasound after hunger pause for detecting of contraction of gallbladder. Urgently determine type of blood group, partial and total bilirubin, reticulocyte cell count and peripheral smear. MRI cholangiography. Presence of hepatomegaly. Exclude perinatal infections (TORCH titers, hepatitis profile).

Task 3 .Baby of 4 months admitted in pediatric surgery unit with jaundice which begun after birth. Child have got conservative treatment with diagnosis of neonatal hepatitis which was noneffective. Liver decreased in size, firm in palpation. Diagnosis of biliary atresia was established.What treatment is possible?

Answer Key: Kasai's procedure not indicated due to age of child. Liver transplantation is the treatment of choice in this case, because hepatoportojejunostomy is effective only before 60 days of child's life.

Task 4. Child was operated 5 years ago with diagnosis of biliary atresia. Treatment was effective, jaundice disappeared. Unfortunately, last time jaundice appeared again, progressive liver dysfunction ensues, liver function tests demonstrate a cholestatic picture with a modest but persistent abnormality in serum transaminase values. What treatment is indicated?

Answer Key: When bile drainage after successful portoenterostomy stops and liver cirrhosis is developed liver transplantation is indicated. Liver transplantation has become the treatment of choice in children with failed Kasai operation.

Tests for initial level of knowledge for CC

1. Which of these symptoms can help to suspect choledochal cyst?
   A. Intrahepatic cyst on ultrasound.
   B. Intermittent jaundice, palpable tumor in right subcostal area, abdominal pain.
   C. Parenchimous jaundice.
   D. Obstructive jaundice.
   E. Calculous cholecystitis.

2. Which of mentioned is a procedure of choice in choledochal cyst?
   A. Cystoduodenostomy.
   B. Cystojejunostomy.
   C. Cholecystostomy.
   D. Cholecystcystectomy.
   E. Cystectomy with hepaticojejunostomy, Roux-en-Y jejunojejunostomy.
3. The most informative noninvasive diagnostic method for choledochal cystis?
   A. Duodenography.
   B. Plain X-ray in up-right position.
   C. Liver tests and common blood analysis.
   D. Liver biopsy.
   E. MRI cholangiography
4. All mentioned are complications of choledochal cysts, except?
   A. Cholangitis.
   B. Pancreatitis.
   C. Stone formation.
   D. Biliary cirrhosis.
   E. Ileus.
5. How to establish the diagnosis of choledochal cyst by imaging studies?
   A. Abdominal ultrasound.
   B. Technetium-99 HIDA scan.
   C. Abdominal computed tomography.
   D. Magnetic resonance cholangiopancreatography.
   E. All above.
6. What procedure should be performed in 3-type of choledochal cyst?
   A. Cystoduodenostomy.
   B. Cystojejunostomy.
   C. Duodenotomy, papillosphincterotomy, marsupialisation of the cyst.
   D. Cholecystectomy.
   E. Cystectomia with hepaticojejunostomy, Roux-en-Y jejunojejunostomy

7. What laboratory abnormalities are associated with choledochal cyst?
   A. Conjugated hyperbilirubinemia increased alkaline phosphatase, and other serum indicators of obstructive jaundice.
   B. Hyperleucocitosis with left shift of formula.
   C. Dysproteinemia.
   D. Unconjugated hyperbilirubinemia.
   E. Trombocytopenia.

8. Should cholecystectomy be included as part of the treatment for choledochal cyst?
    A. Never.
    B. Always should be performed.
    C. Sometimes.
    D. Over 1 year.
    E. If cholecystitis is developed.

9. What type of choledochal cyst is most often revealed?
    A. 1 type (fusiform)
    B. 2 type (diverticulum)
    C. 3 type (choledochocele)
    D. 4 type (intra- and extrahepatic dilation)
    E. 5 type (Caroli's disease).

10. What treatment of choice of complicated Caroli's disease ?
    A. Cystoduodenostomy.
    B. Cystojejunostomy.
    C. Liver transplantation.
    D. Cholecystectomy.
    E. Cystectomia with hepaticojejunostomy, Roux-en-Y jejunojejunostomy

Keys for tests:

1. **B**
2. **E**
3. **E**
4. **E**
5. **E**
6. **C**
7. **A**
8. **B**
9. **A**
10. **C**

Tasks for final level of knowledge

Task 1. 5-year-old girl admitted with jaundice, acholic stool, pain in right subcostal area, there is palpable "tumor" in this area. In ultrasound was revealed choledochal cyst of 1 type by Alonso-Lej classification

1) What is your management for this patient?
2) What is the best time of operation in child with choledochal cyst?
3) Is it possible to use internal draining procedure in treatment of choledochal cyst?

Key Answer: The operation of choice in choledochal cyst is tjtal excision of the cyst – coledochocystectomy, cholecystectomy with hepaticojejunostomy and Roux-en-Y jejunojejunostomy. Procedure of internal draining (cystoduodenal or cystojejunal) are uncorrect: cyst wall never decreased in size due to absence of muscle layer that leads to cholestasis, calculousis, cholangitis and malignancy in adults.

Task 2. Girl 6 years old admitted in infectious department with hepatitis. Jaundice disappeared over 1 week. It was second episode of jaundice over 6 months after the first one.

1) What disease you should suspect?
2) What instrumental methods you should use?

Key Answer:

1) Recurrence jaundice is characterized to coledochal cyst.
2) Abdominal ultrasound, MRI cholangiography, liver tests.

Task 3. 10-year-old child with confirmed diagnosis of Caroli disease. Periodically manifests attacks of cholangitis that required intensive therapy with massive antibiotic therapy.

What treatment is indicated?

Key Answer: Patients with Caroli's disease are difficult to manage because they tend to develop severe recurrent bouts of cholangitis, subsequent biliary cirrhosis, and progressive segmental ductal ectasia. A variety of technics may be needed for management of intrahepatic cysts. Partial hepatic lobectomy may be done when the disease is localized and amenable to resection. Liver transplantation is indicated also.

Task 4. Diagnosis of choledochal cyst was established prenatally, the newborn does not manifest jaundice. What appropriated timing of surgical correction when a choledochal cyst has been noted in utero?

Key Answer: Delaying of operation may lead to hepatic fibrosis, sometimes even noted at birth, which is known to be reversible with surgical repair, as well as the potential complications of cyst enlargement and inflammation, formation of proximal stenosis, hepatic disfunction, and perforation. Therefore, is indicated early correction within 2-4 weeks of life.

Materials for self-study of the students

Main tasks	Notes (instruction)
Repeat: Anatomy of liver, extrahepatic bile ducts Physiology of liver.	To sketch out the anatomy of liver Top represent the methods of diagnosis of biliary atresia.
Study: Pathogenesis of biliary atresia. Features of biliary atresia.	To make differential diagnosis of biliary atresia. and nonsurgical jaundices.

The diagnostic possibilities of ultrasound examination, CT, MRI in children Kasai's operation.	To make the indications to surgical treatment in children with biliary atresia. To know modern diagnostically methods.