SURGICAL DISEASES

in Children

ODESSA NATIONAL
MEDICAL UNIVERSITY

SURGICAL DISEASES IN CHILDREN

A manual

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Навчальний посібник містить сучасні дані щодо діагностики та лікування основних вад розвитку і хірургічних заворювань дитячого віку. Він призначений для підготовки майбутніх лікарів з надання невідкладної хірургічної допомоги й опанування методами клінічної та спеціалізованої діагностики найпоширеніших захворювань у дітей. Його складено відповідно до чинної програми та навчального плану з дисципліни «Дитяча хірургія» і рекомендовано для студентів 5–6 курсу міжнародного факультету.

Рекомендовано Міністерством охорони здоров'я України
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навчальної та навчально-методичної літератури
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INTRODUCTION

Pediatric surgery is one of the newest areas of medicine and at the same time one of the toughest sections of medical care for children, who represent a quarter of the population of Ukraine. It arose at the joint of two medical specialties — surgery and pediatrics, when it became clear that children and adults differ by anatomical and physiological characteristics that are more common at newborns and infants.

The possibility of early detection of many surgical diseases of children of different age groups, with the introduction of modern high-tech diagnostic procedures, as well as the current development of anesthesiology and resuscitation allowed to perform complex surgical procedures, even for newborns. Traditional operational interventions in pediatric surgery are now increasingly replaced by endoscopic surgery, which have a clear advantage because of the minimum trauma and the possibility of high-precision technology.

And we must distinguish that in spite of all the achievements of engineering and technology, the successful treatment of children with developmental and surgical diseases is based on the accuracy and timeliness of diagnosis by "a first contact" doctor — pediatricians, family doctors and "emergency" doctors. Often not only child's health but life also depends on their skills.

According to these ideas "Pediatric surgery" handbook was created. It contains current data for the diagnosis and treatment of the major drawbacks of development and surgical diseases of childhood. It is designed to train future doctors in emergency surgical care presentation and mastery of techniques and specialized diagnostics of the most common diseases of children. This knowledge is necessary for professionals of neighboring specialties — internists, pediatricians, surgeons, obstetricians and gynecologists. According to the principles of the Bologna Declaration, the handbook is structured on the section which provides optimal conditions for the students in the study of the discipline. Each section includes questions of test control and situational test.

This handbook forms the ability to use knowledge of pediatric surgery in the study of neighboring disciplines and in professional life.

The basis of the handbook is formed by many year experience of the team of pediatric surgery department of the Odessa National Medical University, and the achievements of pediatric surgeons in this country and abroad.

First department of Pediatric Surgery in Odessa was opened in 1938 and headed by its assistant professor Y. U. Kornman. In post-war times, the department was headed by outstanding pediatric surgeon professor M. L. Dmitriev. Over time, the department was in charge of M. L. Dmitriev's student — professor L. V. Prokopova and then professor N. G. Nikolaeva. Nowadays department is headed by professor O. O. Losev, who headed the work on the handbook.

For many years, the department staff is working on the problems of diagnosis and treatment of inflammatory diseases, neonatal surgery, orthopedics and trauma, trauma surgery, neurosurgery, urology, cardiovascular surgery.

Scientists of the department formed an original treatment of hematogenous osteomyelitis, bacterial destruction of the lungs, bone grafting, treatment of echinococcosis and malformations in newborns; 6 monographs, 7 manuals were published, more than 60 patents received. This material is also given in certain paragraphs.

Staff of the department hopes that this handbook will be useful in studying the subject of pediatric surgery and in further practice of pediatricians, pediatric surgeons, general surgeons, family doctors.

Section 1
URGENT PEDIATRIC SURGERY

1.1. BLEEDING FROM DIGESTIVE SYSTEM IN CHILDREN. PORTAL HYPERTENSION. BLEEDING FROM THE LOWER GASTROINTESTINAL TRACT SECTIONS

Specific objectives:

1. To master the list of diseases in which occures bleeding from the upper and lower parts of the digestive system in children.
2. Examine the main clinical symptoms of bleeding from the upper and lower parts of the digestive system in children.
3. Differentiate bleeding, depending on the cause.
4. Interpret auxiliary examination methods: ultrasound, X-ray, endoscopy, laboratory (P, AT, Hb, Ht, CBV).
5. To master gastric intubation, digital rectal examination, to characterize the composition of gastric contents and feces.
6. To substantiate and formulate a preliminary clinical diagnosis of the child with bleeding.
7. To master sequence of actions of a doctor in case of bleeding from the digestive system in children.
8. To master general principles of the treatment of digestive system illnesses in children, which are accompanied by bleeding and to determine the indications for surgical intervention.

BLEEDING FROM THE GASTROINTESTINAL TRACT. THE CONCEPT OF SHOCK INDEX

Gastrointestinal bleeding (GIB) occur in 5–8% of children, in 55% — they are caused by a stomach ulcer. Recognition of bleeding is a difficult diagnostic problem that requires professional skills and knowledge.

V. I. Struchkov classification is usually used.

Localization:

— from upper parts (esophagus, stomach, duodenum);
— from small intestine (ileum);
— from lower parts (thick).

2. According to the clinical course:
— active (ongoing);
— stopped.
3. By volume:
— massive (profuse);
— small (minimum).
4. According to the character:
— acute;
— chronic (hidden).
5. According to etiology:
— ulcerative;
— non-ulcerative.
6. According to severity of blood loss:
— mild;
— average;
— severe.
7. In terms of frequency:
— primary;
— recurrent.

Gastrointestinal bleeding may be caused by more than a hundred different diseases. ***The most common causes*** of bleeding are: duodenal ulcer, gastric ulcer, erosive gastritis, esophageal varices, Mallory-Weiss syndrome, erosive duodenitis, erosions and ulcers of the esophagus, vascular malformations, diseases of the small intestine.

Regardless of occurrence level the distinguish ulcerative and non-ulcerative bleeding. ***Non-ulcerative bleeding*** may be related to the localization in the digestive tract (tuberculosis, parasitic infestation, hemorrhoids, etc.) or due to processes outside of the intestines and stomach (thrombosis of the portal and splenic veins, blood disease, poisoning, uremia, vitamin deficiency). A non-ulcer one includes injury of the esophagus, stomach, liver (bruises, fractures, chemical and thermal burns), gastrointestinal foreign bodies, surgical complications and medical manipulations, prolonged unjustified use of anticoagulants.

The severity of gastrointestinal bleeding is determined by the volume of blood loss and the meaning of shock value index (SVI). They distinguish mild, moderate and severe bleeding degree.

Mild (I) degree of bleeding is characterized by a decrease in the number of red blood cells to $3.0 \cdot 10^{12}$, hematocrit (Ht) not less than 0.3 moderate pale skin. Heart rate (HR) exceeds the age limit by 10–15% at

normal blood pressure (BP), CBV is reduced on 15–20%. Shock index (SI) is 0.8–1.2.

Average (II) degree of blood loss is characterized by a decrease in red blood cells to $2.5 \cdot 10^{12}$, Ht to 0.2–0.25. Children are restless, sharp pale skin body is covered with a cold clammy sweat, heart rate above the age norm on 20-30%, blood pressure is reduced on 10–15%. SI — 1.3–2.

Severe (III) degree of blood loss: decrease in red blood cells $<2{,}5 \times 10^{12}$, Ht — <0,25. Children are slow, stupor is often observed, the body is covered with a cold clammy sweat, heart rate above the age norm on 50%, blood pressure is reduced on 30% or more. SI — > 2, CBV is reduced on ≥ 35%.

At the prehospital stage the doctor (family doctor, from hospitals, ambulances, admissions) can assess the severity of blood loss in terms of "**Allgöwer shock index**":

$$SI = \frac{\textit{heart rate in 1 minute}}{\textit{systolic blood pressure in mmHg}}$$

Normally, the shock index = 0.5 — 0.6. If Allgцwer index equals 0.7–0.8, the blood loss is 10% CBV (500 mL), 0.9–1.2 — 20% CBV (1000ml), 1.3–1.4 — 30% CBV (1500 ml) 1.5 or more — 40% CBV (2,000 ml).

The amount of blood loss (VC) be determined by using the hematocrit number.

$$VC = PCBV \cdot (PHt \cdot TNt)/PHt,$$

where PCBV — proper CBV in ml,
PHt — proper Ht in%,
TNt — true hematocrit in%.

Depending on the changes of blood loss coagulation of the child is also changing. At mild degree of blood loss fibrinogen concentration, platelet count, prothrombin index, fibrinolytic activity are increased or are in the normal range. The average degree of blood loss shown by decrease of the level of fibrinogen, platelet count, prothrombin index, and fibrinolytic activity is increased a little. Severe blood loss is accompanied by a significant decrease in the concentration of fibrinogen, thrombocytopenia, a decrease in thrombin time with an increase in fibrinolytic activity.

In severe blood loss degree changes in hemostasis, the redistribution of blood in the body, circulatory disorders quickly develop. This leads to the development of shock, acute renal or hepatic failure, myocardial and cerebral hypoxia, intoxication with hydrolysis products of blood proteins, which have effects on the bowel.

Acute bleedings are prolonged and intense. Chronic bleedings are not large in volume, but prolonged and tend to recur and are often also hidden. Small volume bleedings may not be accompanied by a visible reaction from the cardiovascular system and quickly offset by the redistribution of blood and vascular fluid. Massive bleeding (> 15–20% CBV) manifest by a clear clinical picture of hemorrhagic shock with multiple organ dysfunction.

GIB occur in children of all age groups: ulcerative bleeding predominate in children of school age (10–14 years), non-ulcer in the preschool years. GIB is more often in boys, regardless of age. Equally important among the causes of gastrointestinal bleeding has heredity (peptic ulcer disease occurs in 30–75% of cases).

The clinical picture of acute GIB depends primarily on the degree of blood loss degree, the nature of the underlying disease, age, compensatory capacity state of the organism. But GIB will be always accompanied by hematemesis, melena, hemodynamic disturbances, anemia, collapse.

The main symptom of bleeding from the upper parts of gastrointestinal tract is **vomiting with blood *(haemotemesis)***. It can be abundant in the form of "coffee grounds" and red blood with impure of food or without, single-, reusable, followed by loss of consciousness, preceded by melun or arise along with tarry feces.

At *profuse* bleeding vomiting occurs suddenly, although it may be preceded by increasing weakness, dizziness, nausea. Quickly appear pale skin, cold clammy sweat, tachycardia, and tachypnea. Reduced blood pressure, over the heart top is heard systolic murmur. This clinical picture is typical for patients with portal hypertension syndrome of Mallory–Weiss, gastric ulcer and duodenal when large arterial vessels are eroded. At considerable profuse bleeding it is marked "fountain" vomiting (portal hypertension).

The second symptom of gastrointestinal bleeding is black, tarry stool — ***melena***. The appearance of melena often indicates bleeding from the proximal gastrointestinal tract. However, melena is observed in many patients with pathology of the esophagus, portal hypertension. Slow blood flow to the lumen of the intestine causes dark stool. The gradual accumulation of blood in the colon leads to its decay: ferrous sulphate is formed, which gives the stool color from dark to black cherry. Necessary to eliminate intake of certain foods that contain a lot of blood (black pudding), as well as a symptom of ingested blood in children. Dark color of feces can also be marked with iron supplementation, bismuth, activated carbon, lots of cherries, blueberries, raspberries, red currants.

Development of acute bleeding leading to a mismatch of volume of vascular bed and BCC, which is accompanied by a decrease in blood pressure, accelerated heart rate, decreased cardiac output. In response to blood loss occurs defensive reaction in the form of spasm of blood vessels and reducing the volume of the vascular bed. Develops clinical hemorrhagic shock: growing weakness, dizziness, tinnitus, cold sweats, confusion, pallor, decreased blood pressure, thready pulse, transient syncope.

Thus, the direct clinical symptoms of GIB are:

1. Haematemesis — vomiting blood.
2. Haematochesia — discharge of unchanged or less changed blood from the rectum.
3. Melena — discharge of altered blood from the rectum in the form of tarry feces.

Indirect clinical symptoms of gastrointestinal bleeding are:

1. Paleness of skin.
2. Sedation.
3. Drowsiness.
4. Dizziness.
5. Cold extremities.

6. Increased and weakening pulse on peripheral vessels.

7. Reduction of blood pressure.

Hemorrhagic syndrome — is apparent bleeding or clinical signs of hidden blood loss (pallor, weakness, dizziness), the signs of internal bleeding (accumulation of fluid in one of the cavities) in conjunction with a reduction of age hemodynamic parameters or decrease of the amount of hemoglobin, erythrocytes, hematocrit compared to age norm.

Tasks of diagnostic algorithm of hemorrhagic syndrome at GIB:

1. Establishment of the existence of hemorrhagic syndrome
2. Level detection of bleeding source
3. Determination of the amount of bleeding
4. Determination of the causes of bleeding

Additional methods of research in the gastrointestinal bleedings:

1. Rhinopharyngolaringoscopy.
2. Fibrogastroscopy.
3. Fibrocolonoscopy.
4. Fluoroscopy of gastrointestinal tract.
5. Pneumocolography.
6. Hepatosplenoportography.
7. Angiography.
8. Ultrasound examination.
9. Radionuclide scintigraphy.
10. Laparoscopy.

The reaction of the organs and tissues to blood loss are represented in Table 1.

Stages of emergency measures in case of acute bleeding are represented in Table 2.

ULCER DISEASE OF STOMACH AND DUODENUM

Ulcer disease (UD) of the stomach and duodenum — chronic disease with polycyclic course, which is characterized by secretory, motor and trophic changes of the stomach or duodenum with formation of ulcers of the mucous membranes. Duodenal ulcer develops 10 times more often than the stomach one. The ratio of occurrence of duodenal ulcer in men and women ranges from 3:1 to 10:1.

Etiology and pathogenesis

The scheme of pathogenesis of ulcer disease is shown on Fig. 1.1.1

To etiological factors of peptic ulcer disease include following:

1) Genetic
2) Alimentary
3) Neuropsychic
4) Medicamentous
5) Infectious

Hereditary factor is important in the occurrence of peptic ulcer disease (30–38%). Is inherited reduced reactivity of the mucous membrane of the stomach and duodenum to the damaging effects of gastric acidity due to excess of glandular apparatus of its mucosa.

62% of children have level of basal gastric secretion at the rate of 10–15 mmol/l-g (at N<5 mmol/l-g).

Another role is played by nutritional factors (consumption of coarse and spicy foods, spices, smoked foods, excessive consumption of coffee and refined carbohydrates), which can cause mechanical injury to the mucosa or increased secretion and gastric motility. Secretion on an empty stomach may also cause acid-peptic damage to stomach and duodenum.

In children, a certain role belongs to bad habits — smoking, alcohol abuse. Nicotine, like alcohol, causes vasospasm of the stomach, especially in combination with the qualitative and quantitative eating disorders. Many researchers attach great importance to the neuro-mental factors (acute psychological trauma, mental stress), which can cause disorders of the nervous regulation of the gastro duodenal zone.

Ulceration is promoted by taking medications (salicylates, glucocorticoids), which cause a decrease in the secretion of gastric mucus and epithelial regeneration of the gastric mucosa, gastric vasospasm.

To explain the development of peptic ulcer they propose many theories. It was established that circulatory disturbances in the stomach wall can occur at violation of the arteriovenous shunts, which are located in the submucosal layer. Their opening leads to ischemia of mucosa, closing — to a permanent

Table 1

Reaction of the Organs and Tissues to Blood Loss

Organs and management	Early blood loss < 25% of volume	Pre-hypertensive 25% of volume	Hypertensive > 40% of volume
Heart	Weakness, weak pulse, increase in heart rate	Increase in heart rate	Hypotension, tachycardia, followed by bradycardia
CNS	Drowsiness, excitation, confusion, aggression	Mental confusion, lack of response to pain	comatose state
Skin	Cold, clammy	Cyanosis, decreased capillary refill, cold extremities	Pale, cold
Kidneys	Decreased diuresis	Increase in blood urea nitrogen	Anuria
Fluid therapy	20 ml/kg	20 ml/kg (×2) Er-weight 10 ml/kg (×1, ×2)	20 ml/kg (×2), Er-weight 10 ml/kg urgent transportation to the operating room

Table 2

Stages of Emergency Measures in Case of Acute Bleeding

Stage	Measures	Medicines
1.	Washing with ice-cold solution	Ice water, isotonic solution. sodium chloride, 2% solution of sodium bicarbonate, 5% solution, aminocaproic acid
2.	Hemostasis	A) Endoscopic Irrigation of source of bleeding with cooled solution, ACC, 10% solution of calcium chloride, 0,1% solution of noradrenaline Diathermocoagulation Photocoagulation Adhesive application Clipping of the vessel with metal clips B) Endovascular Introduction of 0.2-0.3 ml pituitrin for injection into the artery, 1 ml of 12.5% solution etamsylat, 5–10 ml, 5% solution of the ACC Embolization of the arteries C) Therapeutic Introduction of 5% solution of the ACC Introduction (intramuscularly) 1% solution vicasol, 12.5% solution of etamsylat, pituitrin Introduction (intramuscularly) 10% solution of calcium chloride, fibrinogen, 5% solution of ACC, pituitrin, omeprazole, sandostatin
3.	Infusion-transfusion replacement therapy	Transfusion of blood components, protein drugs, blood substitution fluids, the use of Blackmore probe
4.	With no effect — surgery	Indications for emergency surgery — Ongoing bleeding that is threatening, particularly in patients with hemorrhagic shock — Continued bleeding, when during massive blood loss all conservative measures, including therapeutic endoscopic, endovascular hemostasis are ineffective — Recurrent bleeding, which appears after a short break, especially in the duodenum ulcer, with “visible” vessel at the bottom of the ulcers

Pathogenesis of peptic ulcer disease

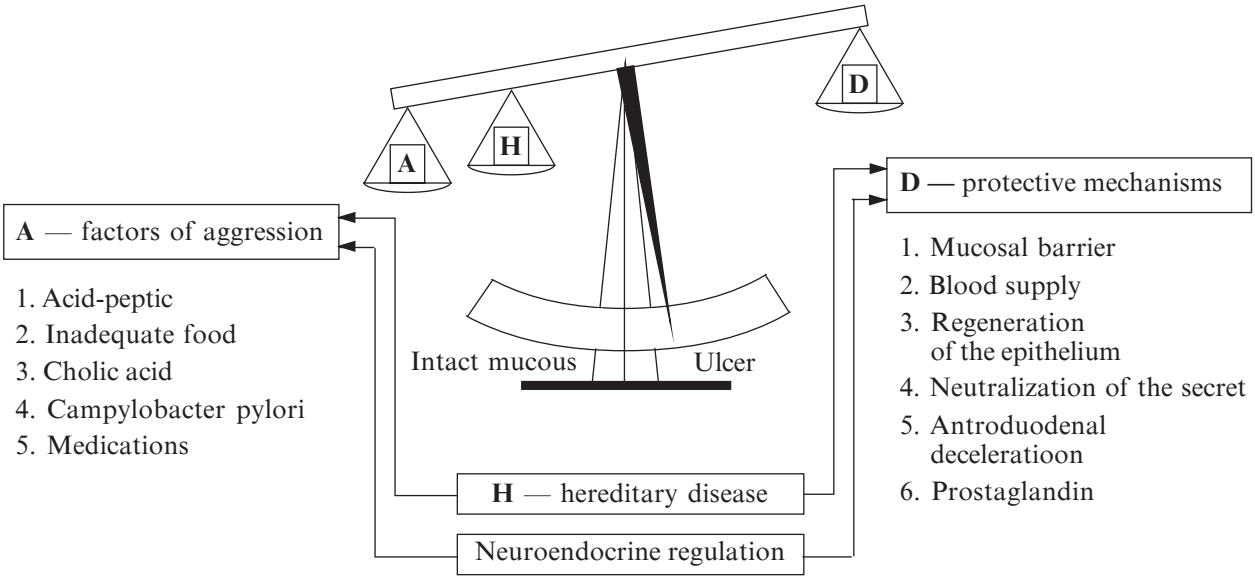


Fig. 1.1.1. The scheme of pathogenesis of ulcer disease

hyperemia. Such disorders may be caused by hypertonicity of vagus nerve, hyperreactivity of the adrenal glands cortex, increased tone and motility of the stomach. Hypoxia of mucous membrane, together with other factors contributes to the ulcer. Not lost a neurogenic theory of the pathogenesis of ulcer disease by Ressle, Bergman, A. D. Speranskiy, K. M. Bykov, I. T. Kurtsin is also important: violation of the central and autonomic nervous systems compared to other factors of ulcer formation. In Selye stress theory ulcer disease is seen as a manifestation of the adaptation syndrome in response to various stimuli. According to the author, any stressful stimulus results in an increased secretion of the hormone of the anterior pituitary and adrenal glands cortex, which increase gastric secretion. In accordance with the theory of mucosal barrier by Hollander, more importance is given to local factors: the violation of the protective gastric mucus formation, damage to the cells that form it, atrophy of the gastric mucosa. According to the theory of Dregsteda highlight in the occurrence of peptic ulcer is due to acid-peptic factor.

Considering the diversity of causes and pathogenic factors, features of clinical and morphological manifestations, peptic ulcer disease should now be considered a polyetiological and polypathogenetic disease.

The main factors of gastric ulcer are:

— Violation of nervous and humoral mechanisms that regulate the activity of gastroduodenal zone;

— A violation of local mechanisms of gastric secretion;

— Changes in the structure of the mucous membrane of the stomach and duodenum;

— Constitution and heredity;

— Environmental conditions.

Leading factors of duodenal ulcers are:

1. Violation of neurohormonal and local mechanisms of gastric secretion regulation, which predetermine the increased secretion of hydrochloric acid and pepsin;

2. Decrease in the resistance of duodenal mucosa.

Clinical symptoms. For patients with peptic ulcer disease is characteristic a typical ulcerative syndrome: pain, heartburn, belching, nausea, vomiting. The pain may be of a different nature: aching, burning, etc. The gradual development of pain, its progressive increase, which is associated with exacerbation of ulcerative process are typical. Pain in the epigastric region, along the midline of the abdomen, in the area of the xiphoid process, the right or the left upper quadrant is localized.

It may be early, late, hungry and night. Early pain appears within the first hour after a meal, later — after 1.5–4 hours after it. Early pain occurs in patients with gastric ulcer, late, night — with duodenal ulcer disease. Onset of pain may be due to physical stress, fatigue, anxiety. Some patients do not notice it, depending on the meal; others suffer from constant pain, which is frequently observed in chronic callous ulcer, presence of perigastritis is, periduodenitis.

Heartburn occurs in 50–80% of patients.

It appears at the beginning of the disease, often preceded by episodes of pain, occurs immediately or 2–3 hours after a meal and is due to decreased tone of the cardiac sphincter, gastro-esophageal reflux of acid with the development of esophagitis.

Vomiting occurs in half of patients, usually on the height of an attack of pain. Duodenal ulcer is caused by hypertonicity of the vagus nerve, which leads to disruption of the secretory and motor functions of the stomach, and gastric ulcer — a violation of the evacuation caused by swelling of the mucous membrane around the ulcer or spasm of the pylorus. Nausea is always preceded by vomiting. Belching at stomach ulcer can be acidic, with food or air, and is associated with impaired cardiac sphincter tone. Appetite at uncomplicated ulcer is not violated.

Intestinal symptoms especially with duodenal ulcer, include constipation, which may be accompanied by pain of spastic nature, caused by neuromuscular diskinesis of the colon and amplify at disease exacerbation.

Typical signs of a peptic ulcer are daily rhythm, frequency and seasonality of exacerbations. The occurrence of pain often occurs in the afternoon or at night, which is associated with mealtime and increased gastric secretion. Frequency — alternating periods of exacerbation with a period of remission — of different duration. Exacerbation of peptic ulcer disease occurs more frequently in spring and autumn, due to sharp fluctuations in meteorological conditions, unstable atmospheric pressure, and impaired vitamin balance.

The general condition of the patient is satisfactory. Belly of the usual form, palpation reveals mild tenderness in the epigastrium. Of great importance is the identification of areas of percussion pain (a symptom of Mendel) for duodenal ulcers — in the right half of the epigastrium with spread to the right upper quadrant, for ulcers of the lesser curvature — in the midline and somewhat to the left of it, with cardiac ulcer — at the xiphoid process. On examination reveals pain points: Boas (pain at pressing on transverse processes of the I–II of the lumbar vertebrae on the left), Openhovsky (pain with pressure on the spinous processes of the thoracic vertebrae X–XII.

In childhood, ulcers usually have 2 types of the course:

— With a severe pain, vomiting, and high acidity. In 85% of cases are located in the duodenum or pyloric area;

— "Silent" ulcers which are often diagnosed only in case of complications (stenosis, perforation, bleeding). Very early penetrate with a acute pain, and vomiting.

In patients with ulcers may develop the following complications: Penetration (9–13%), perforation (8 to 12%), stenosis (12.5%), bleeding (19–26%) and malignancy (0.2–0.5%).

Laboratory blood tests for the diagnosis of uncomplicated peptic ulcer disease have no practical importance, but play a role in assessing the severity of disease, dysfunction of other organs and systems. Leukocyte formula is normal; ESR is accelerated only during the acute illness.

Examination of feces reveals violations of digestion and absorption, hidden bleeding, inflammation, and various forms of diskinesis of the gastrointesti-

nal tract. If a peptic ulcer is accompanied by putrefaction and fermentation processes, the reaction of feces can be dramatically alkaline or acidic. At hidden bleeding in patients is revealed blood in the stool (sample-response of Gregersen). Scatological study reveals inflammation of the intestines and various forms of diskinesis.

Methods of special research. Special research techniques in patients with diseases of the stomach include: study of gastric secretion, motor function, and morphological changes of the mucous membrane, X-ray, endoscopic examination and tests on Helicobacter pylori.

Fluoroscopy and X-ray are among the most important methods of research of the stomach and duodenum. They allow determining their shape, size, location of ulcers, tumors, relief of mucous membrane of organs and their function. Abdominal X-ray reveals abdominal gas, radiopaque foreign bodies in the stomach.

At the appropriate clinical picture, presence of free gas beneath the dome of the diaphragm indicates perforated ulcer of the stomach or duodenum. X-ray with a contrast agents determines the shape, placement of organ, relief of mucous. After receiving a small amount of contrast agent, study the relief of the mucous membrane, at a tight filling of the stomach determine the shape, size, contours, mobility, evacuation, pain points, pathological changes (a symptom of "niche", filling defect, and other).

The main method of diagnosis is esophagogastroduodenoscopy (EFGDS) (Fig. 1.1.2).

Therapeutic management and choice of treatment. Therapeutic strategy depends on the duration of the disease, severity, presence of complications.

Conservative treatment of gastric ulcer and duodenum should be individual, etiological, pathogenetic, and complex, graded and include:

I. Diet therapy (diet number 1A, 1B by Pevsner).

II. Physiotherapy (ultrasound, peat-, mud-, ozokerite treatment).

III. Medical therapy:

1. Agents that inhibit helicobacter infection (denol, Trichopolum, oxacillin, ampiox etc.)

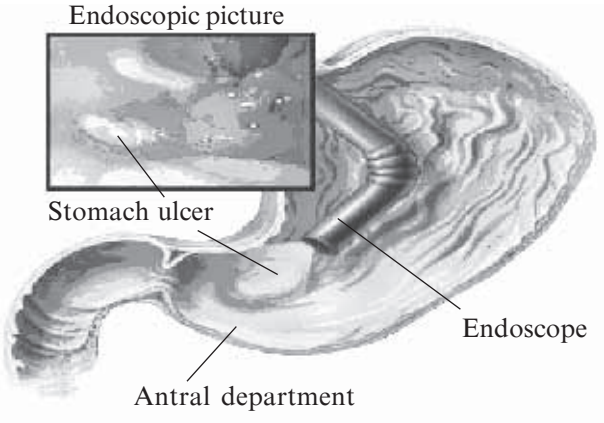


Fig. 1.1.2. Endoscopic picture at stomach ulcer depends on the localization process, the healing phase or exacerbation.

2. Antisecretory agents:

a) M-cholinolytics: nonselective (atropine, platifilin); selective (gastrotsepin).

b) H2 blockers of histamine receptors: cimetidine; ranitidine; famotidine; nizatidine; roxatidine.

c) blockers of H + K + ATPase: omeprazole.

d) Gastrin receptor antagonists (proglumide).

e) Antacids (Almagelum, phosphalugel, maalox, bismuth).

3. Gastrocytoprotectors:

a) Cytoprotective agent that stimulates the formation of mucus: carbenoxolone; synthetic prostaglandins — enprostil, cytotec.

b) Cytoprotectors that form protective foil: sucralfate; de-nol; smecta.

c) Enveloping and astringents.

4. Bismuth preparations — vikalin, vikair.

5. Means that normalize motor function of the stomach and duodenum (Cerucalum, Reglan, metoclopramide, Motilium), antispasmodics (no-spa, papaverine).

6. Reparants (solkoseril, sea buckthorn oil).

7. Tools of the central action (Dalargin, sedatives, tranquilizers).

IV. Local therapy (endoscopic): Adhesive CR-3; laser irradiation.

V. Hyperbaric oxygenation.

VI. Herbal medicine.

Surgical treatment

Absolute indications for surgery are severe complications of peptic ulcer disease:

1. Ulcer perforation.
2. Profuse bleeding or bleeding that does not stop by conservative way.
3. Scar-ulcerative pyloric stenosis and duodenum.
4. Malignization ulcers.

Absolute-conditional indications are the following complications:

1. Penetration and covered perforated ulcer.
2. Recurrent ulcer bleeding during treatment or recurrent bleeding in history.
3. Perforated ulcer history, the resumption of the clinical picture of peptic ulcer.
4. Recurrent ulcer after vagotomy and gastric resection.
5. Giant and callous ulcers and stomach ulcers that do not respond to conservative comprehensive treatment for 2–3 months of intensive therapy.
6. Postbulbar ulcer of duodenum.

Relative indications:

1. Uncomplicated gastric ulcer and duodenum with a severe pain and dyspeptic symptoms on condition of inefficiency of the whole complex of conservative treatment for 2-3 years.
2. Uncomplicated peptic ulcer and duodenum in combination with other diseases of the digestive tract, which require surgical treatment.

NON-ULCER BLEEDINGS: PORTAL HYPERTENSION

Most severe and difficult to predict complication of portal hypertension is bleeding from varices (BPB) of the esophagus and stomach, which in children is

the first manifestation of portal hypertension. The recurrence rate of hemorrhage makes 50–90%. (Nolte W., Hartman H. 1994).

It is most useful to divide (Whipple and Linton's classification) types of portal hypertension into:

1. Overhepatic;
2. Intrahepatic;
3. Underhepatic.
4. Mixed.

At extrahepatic form of portal hypertension (EHH) obstruction to blood flow is defined in the trunk region of the portal vein, a high level of pressure below the blockade and normal values in the blood vessels of the liver. That may be due to malformation of the trunk and branches of the portal vein, portal vein thrombosis in the defeat of its inflammatory process, compression of the vessel from outside scars, tumors of adjacent organs, and so on.

At extrahepatic form of HH in children bleedings arise solely from varices of the gastric cardia (85%). Clinical manifestations of extrahepatic portal hypertension are diverse. This explains the late diagnosis of extrahepatic form of portal hypertension in children. Only in 25–27% of cases portal hypertension is diagnosed before 3 years of age (A. G. Pugachev, A. V. Leontiev). In the neonatal period in 45–60% of patients was marked umbilical wound fester, after the falling away of the umbilical cord, or a statement of the umbilical catheter. During the first year, children are restless; they have an increase in the abdomen and loss of appetite. EHH is characterized by an increase in the spleen with a hypersplenism or without varicose veins of the esophagus and gastric cardia. Before bleeding the general condition of the child does not suffer. Often children can see an increase in the abdomen, bloating, increased pattern of veins of the anterior abdominal and thoracic walls. Children complain of a feeling of heaviness in the left upper quadrant, loss of appetite, unstable stool, nosebleeds, the appearance of "bruises" on the trunk and extremities (Fig. 1.1.3).

The liver can be increased in young children, as a consequence of hepatitis developed during phlebitis. Spleen is increased significantly, sedentary, and painful. Ascites is detected after previous bleeding, ascites quickly disappears with elimination of hypoproteinemia.

Often the first sign of the disease is suddenly developed stomach bleeding — in 60% of patients. Be-

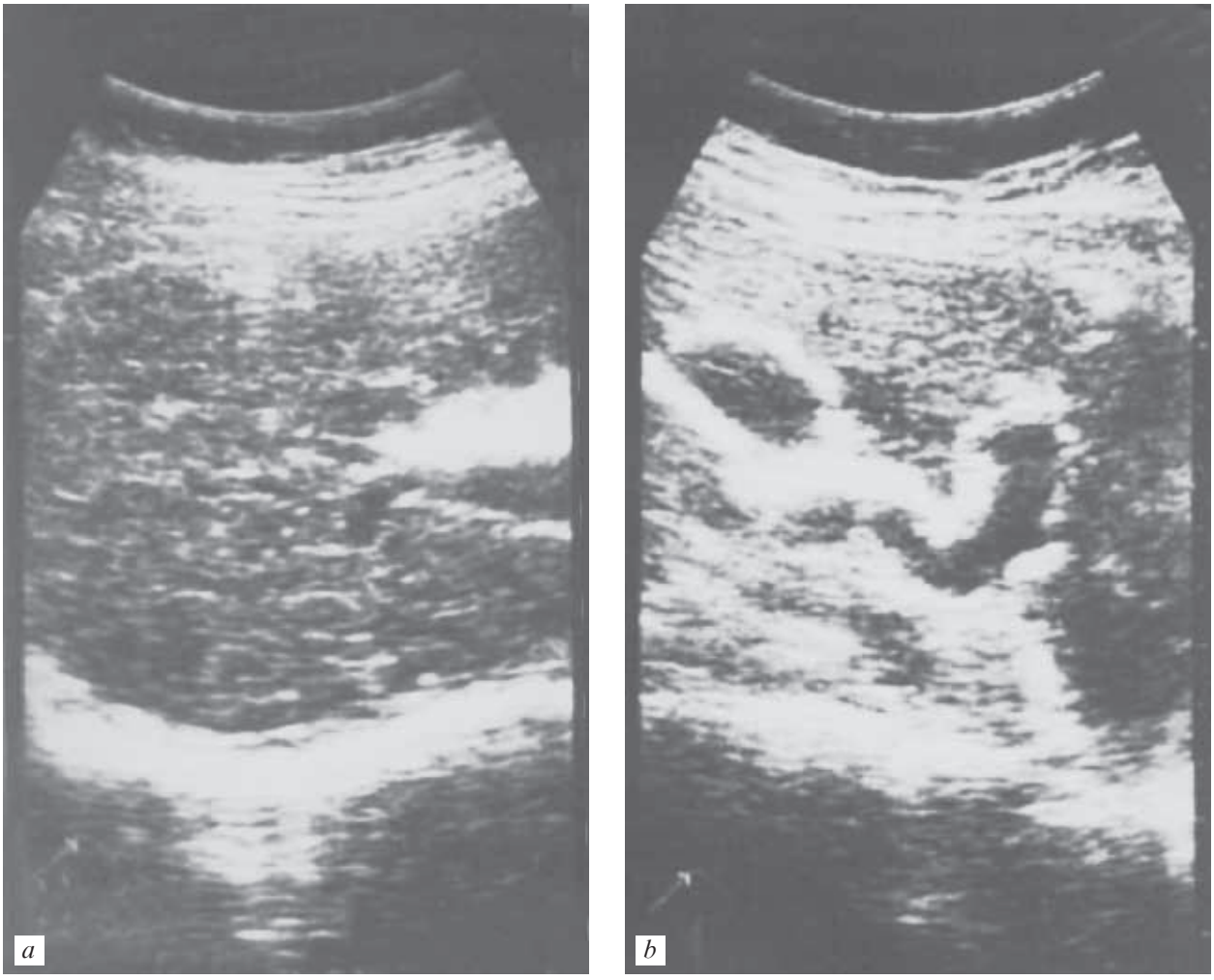


Fig. 1.1.3. Echogram of the patient with liver cirrhosis and portal hypertension: *a* — normal hepatic parenhyme; *b* — extended umbilical vein

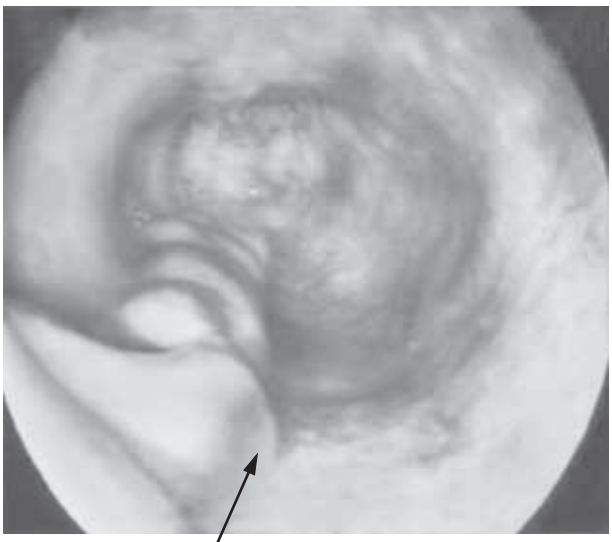


Fig. 1.1.4. Endoscopic picture at portal hypertension

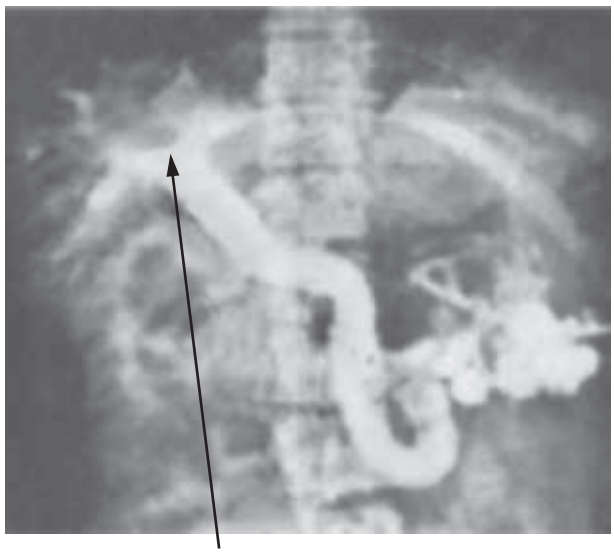


Fig. 1.1.5. Splenoportography at portal hypertension

fore the onset of bleeding children often complain of pain in the left shoulder, 1–2 days before or a few hours before the onset of bleeding may be the rise in temperature, sometimes to the febrile digits. At severe bleeding children become sluggish, appears weakness, paleness of nasolabial triangle, cold sweats and other symptoms of a cute blood loss. Soon there is vomiting with blood clots, usually plentiful.

Within hours, vomiting can be repeated until the onset of collaptoid state. Deaths after the first bleeding in children have not been observed (Fig. 1.1.4, 1.1.5).

Diagnosis of EHH is based on ultrasound, signs of which are:

1) Normal structure of the liver.

2) Lack of a well-formed trunk *v. porte* and its intrahepatic branches. Instead of trunk is determined the convoluted tangle of veins and fibrous tissue ("portal cavernoma").

3) Thickening of the lesser omentum to the size of the abdominal aorta.

4) Slow blood flow (at Doppler) in the veins of the mesentery and spleen, reverse blood flow in the vessels of the gland.

This method allows not only establishing the diagnosis of HH, but also options of anatomy of visceral veins, with which you can impose a decompressive anastomosis.

The next diagnostic test at EHH is esophagogastroscopy at which identify varicose veins in the esophagus and stomach. The likelihood of bleeding from these vessels can be predicted by the presence of congestion, "cherry spots", fibrinous deposits, dilatation and tortuosity of vessels of the esophageal mucosa.

Most complete picture of the anatomy of abdominal vessels gives visceral angiography.

Treatment begins with conservative attempts to stop the bleeding with a parallel examination of the child for a few hours (1–2 days).

To the stomach inject a probe with the purpose of draining, washing and stabilization. Sometimes use a probe of Blackmore for mechanical pressure on the bleeding vessels.

The patient is administered systemic infusion of vasopressin, which causes vasoconstriction of the arterial bed of the internal organs and thus reduces portal venous pressure. The initial dose is 0.2–0.4 /1.73 m^2/min, the drug is administered by continuous infusion. Volume of infusion therapy is reduced to 50% of daily needs, exclude drugs that improve blood rheology. With massive blood loss, designate red blood cell transfusions, fresh frozen plasma.

Currently, they use dopplerography for diagnosis, which allows not only to establish the size and constitution of the liver and spleen, but the caliber, patency, direction of blood flow in the portal system and the hepatic veins. The advantages of this method are in its non-invasive, possibility of using on the background of infusion therapy and resuscitation.

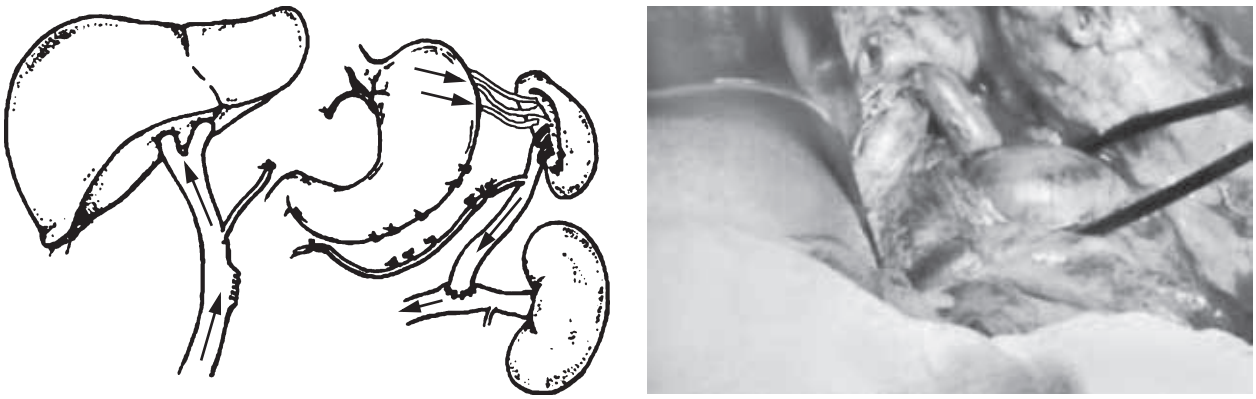
Esophageal endosclerosing is widespread due to the simplicity of implementation. At acute bleeding endosclerosing is carried out at intervals of 1.5 to 3 months until all varices are not obliterated.

Complications:

1. Ulceration of the esophagus;
2. Pleural effusion;
3. Atelectasis;
4. Fever;
5. Motility disorders of the esophagus;

The development of esophageal strictures; severe complications are rare:

1. Perforation of the esophagus;
2. Spinal paralysis;
3. Mesenteric venous thrombosis with myocardial ulcer;
4. Solitary brain abscesses;
5. The formation of tracheo-oesophageal fistula.



a *b*

Fig. 1.1.6. Distal splenorenal anastomosis in the treatment of portal hypertension. Complication of portal hypertension; *a* — scheme; *b* — stage of surgery

In order to avoid such complications, special ring or endoscopic bandaging of varices with thin ligatures is currently used.

Bypass surgeries, as indicated by Ashcraft, Holder (1997), can effectively reduce bleeding but almost not shown to children. According to authors, disadvantages of this method are:

1. In hepatic encephalopathy, as a result of retraction of portal blood to the systemic circulation;

2. In acceleration of liver injury due to a decrease or complete cessation of portal blood flow to the liver.

The incidence of encephalopathy in distant period ranges from 27 to 63%, particularly severe is the loss of: 1) visual memory; 2) spatial and temporal orientation, which can appear only in 15–20 years.

Even distal splenorenal anastomosis loses its theoretical advantage as high pressure in the "liver compartment" goes into low pressure in "splenic compartment", transforming itself in porto-systemic shunt.

Long existing splenomegaly may be accompanied by symptomatic hypersplenism. Treatment of this complication is solved by embolization of the splenic artery with surgical gel, which reaches 60–80% infarct of the splenic tissue. However, even after such operation the following complications are often observed: fever, aches, and sometimes gastrointestinal obstruction, atelectasis, pleural effusion. As for thrombocytopenia and leukopenia — they disappear, retaining immune functions (Fig. 1.1.6).

Mallory–Weiss syndrome (10% of bleeding from upper gastrointestinal tract) — the appearance of sharp linear ruptures of the gastric mucosa in the cardia, which are often accompanied by profuse bleeding (Fig. 1.1.7, 1.1.8).

The cause of this syndrome is often a paroxysmal cough, repeated vomiting, pancreatitis, acute cholecystitis, strenuous exercise, abdominal trauma. At the same time unexpectedly increased pressure in the stomach due to inconsistencies and closing of cardiac pyloric sphincter. Is possible falling of the mucous membrane of the stomach into the esophagus, and it's pinching in the cardiac sphincter. There are three stages of the syndrome:

— Damage only of the gastric mucosa;

— Rupture of the mucosa and submucosa;

— Rupture of all layers of the stomach and esophagus, development of peritonitis, mediastinitis, and pneumothorax.

Most often ruptures are localized by lesser curvature of the stomach, with observed single or mul-

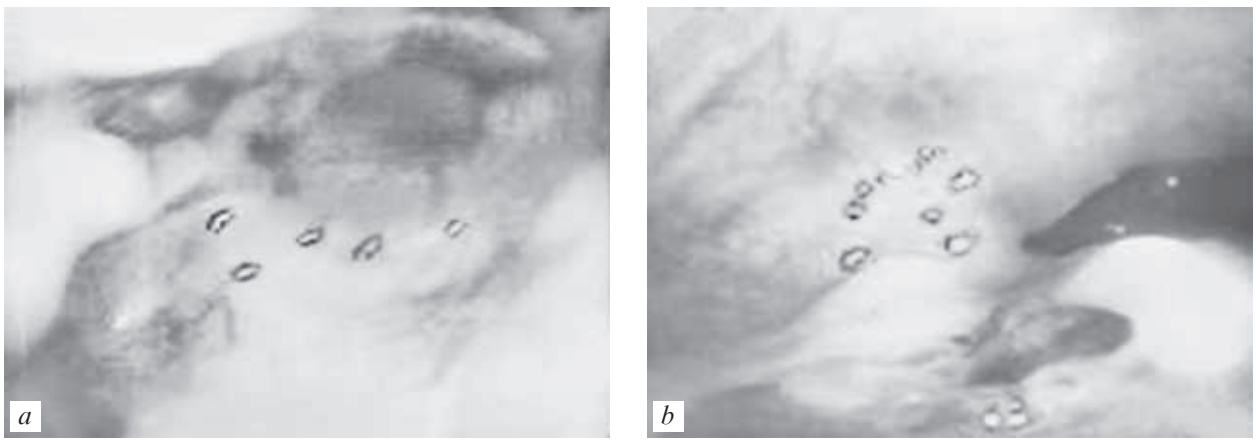


Fig. 1.1.7. Endoscopic picture at Mallory–Weiss syndrome *(a, b)*

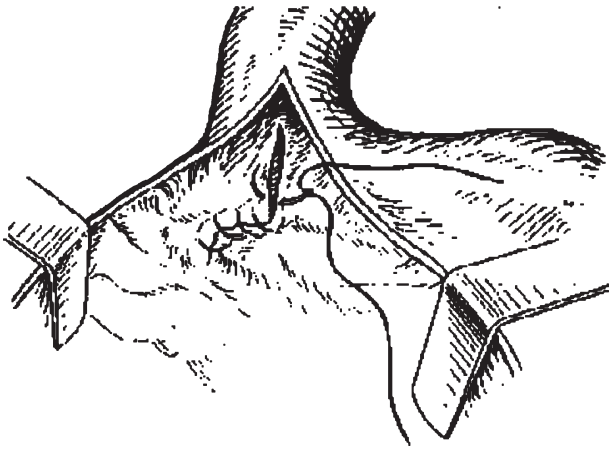


Fig. 1.1.8. Surgery at Mallory–Weiss syndrome

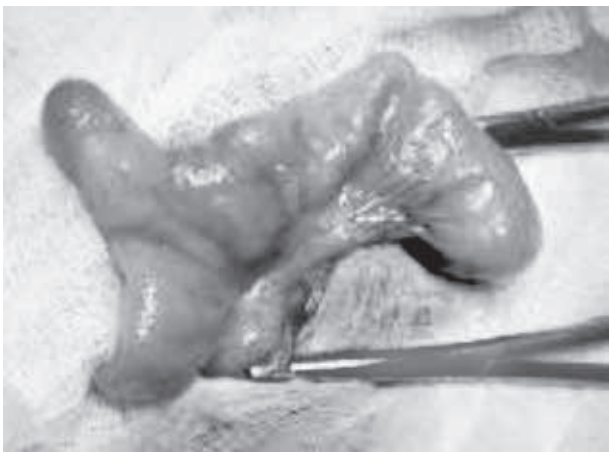


Fig. 1.1.9. Meckel's diverticulum

tiple diapedetic bleedings — bleeds entire surface of the gastric mucosa, or most of it.

Having discovered syndrome of Mallory–Weiss during endoscopy, V. D. Bratus and colleagues (1986) successfully produced diathermocoagulation in 189 patients and some of them have achieved final hemostasis without surgery. In some patients there was a need to make re-diathermy coagulation. The effectiveness of this method is confirmed by other authors. For the treatment of the syndrome Mallory — Veyss also use laser photocoagulation [Skobelkin O. K. and others, 1985; Marshall J., Vogelc K., 1985, and others] And the combination of cryotherapy with electrocoagulation [Pisanyj O. E., 1984]. In order to stop the bleeding can be used selective use of vasopressin and embolization of arteries [Classen M. et al., 1985].

BLEEDING FROM LOWER PARTS OF THE GASTROINTESTINAL TRACT. MECKEL'S DIVERTICULUM

First described by Johann Friedrich Meckel (1781 — 1833 years). Meckel's diverticulum (diverticulum of the ileum) is a congenital anomaly of the small intestine associated with the violation of inverse development of proximal part of the vitelline the flow (the flow between the navel and intestine), when it's proximal part is left not obliterated (uncovered) (Fig. 1.1.9).

During the first weeks of fetal development of human function embryonic flows — vitelline (ductus omphaloentericus) and urinary (urachus), which are parts of the umbilical cord. The first is used to supply fetal, connecting intestine with vitelline sac, the second carries out the flow of urine to amniotic waters. On 3rd–5th month of intrauterine life the opposite development of flows is marked: the vitelline is completely atrophied, turning in an average bundle, placed from the inner surface of the anterior abdominal wall.

Depending on the extent and the level of violation of vitelline flow obliteration, following variants are available:

1) Fistulas of navel: complete and incomplete;
2) Meckel's diverticulum;
3) Enterocystoma.

Meckel's diverticulum is the most common (70% of cases), in 50% of cases — in children up to 10 years, the remaining manifest before the age of 30 years. Frequency combination with other congenital malformations — up to 12%. Length of diverticulum averages 2–3 cm (1 to 26 cm), it may be as thick as the finger or narrow as the appendix, has a conical or cylindrical shape. Typically diverticulum is on the side of the ileum, opposite the mesentery (on free edge of intestine, anti-mesenteric), in average at a distance of 40–50 cm (3 to 150 cm) from baugin damper. It can be soldered by connective tissue (the remainder of vitelline flow) to the mesentery, the anterior abdominal wall or intestinal loops.

Meckel's diverticulum is considered a true diverticulum because histologically in its wall are all layers of the intestine, there may occur heterotopic located gastric mucosa (1882, Timmans) (able to produce hydrochloric acid), and pancreatic tissue (1861, Zenker). This is the reason for the erosion of its wall and intestinal bleeding.

Clinical picture

Uncomplicated diverticulum (95%) is asymptomatic. Diverticulum of the ileum is often discovered incidentally during laparotomy (operations on the organs of the abdominal cavity), undertaken on another occasion or in connection with the development of complications. Complications of Meckel's diverticulum are:

- Peptic ulcer with possible bleeding and perforation — 43%;
- Intestinal obstruction due to the strand, obstruction, volvulus and intussusception — 25.3%;
- Diverticulitis — 14%;
- Herniation (often little hernia — 11%);
- Umbilical fistula — 3.4%;
- Tumors — 3%.

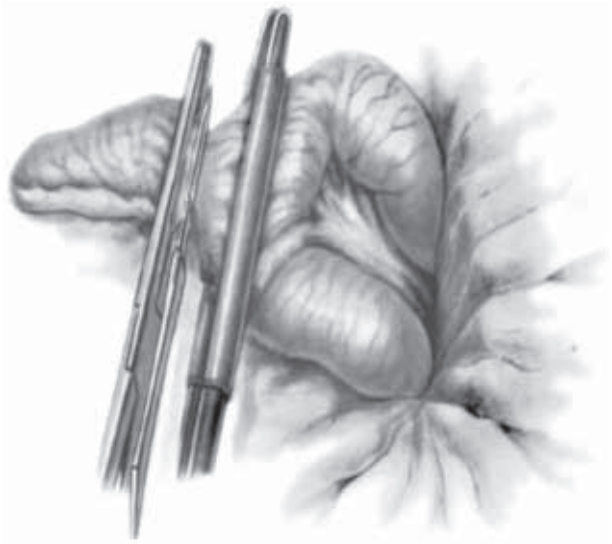
In children occurs peptic ulceration of islands of ectopic gastric mucosa, which is often the cause of massive gastrointestinal bleedings. Bleedings may

occur acutely and be abundant, but there is also observed chronic bleeding with small portions. These bleedings occur among full health, repeated at intervals of 3–4 months, which leads to anemia, pallor, tachycardia, collapse.

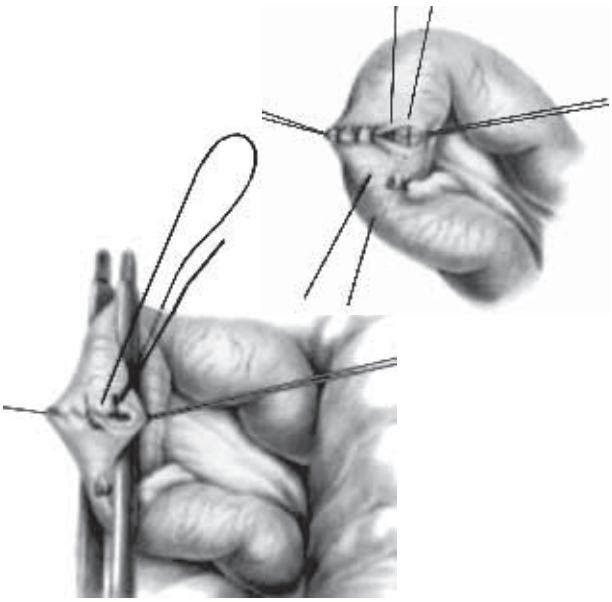
First bowel movements are usually dark, and dark (scarlet) blood without clots and mucus appears later. In contrast to gastrointestinal bleeding of another origin in Meckel's diverticulum there is no bloody vomiting. One of the major clinical manifestations of Meckel's diverticulum is recurrent abdominal pain, which is typical or other organic, functional and psychogenic disorders.

Treatment

Asymptomatic diverticula should not be removed. Meckel's diverticulum is to be removed at diverticulitis, ulcer diverticulum, intestinal obstruction caused by diverticulitis, fistulas navel, as well as at chance discovery of it during surgery. Is made diverticulum resection with suturing of the bowel wall (resectio diverticuli Meckelii) or wedge resection of the bowel (Fig. 1.1.10).



a



b

Fig. 1.1.10. Variants of operations at Meckel's diverticulum *(a, b)*

An important diagnostic feature of diverticulum is the presence of a pronounced supply vascular bundle or a small mesentery. The vessel is detected on one side of diverticulum and is located in its central part. In the presence of small mesentery, diverticulum may be disposed along the small intestine, closely adhering thereto. Near the diverticulum is sometimes found thin dense fibrous cord, extending from the mesentery to the parietal peritoneum of the umbilical ring.

Bleeding diverticula is quite a dense protrusion, sometimes whitish. Often diverticulum is located in the mesenteric edge. The small intestine may be deformed and tightened to the diverticulum. The degree of pathological changes is connected with the duration of ulcer bleeding of a diverticulum and its vastness.

Deep lesion of the intestinal wall with ulcerative process leads to severe perifocal reaction. To diverticulum is soldering oil induration and surrounding intestinal loops. All this leads to the formation of a conglomerate with coarse adnations, inside of which strongly deformed diverticulum with thinned walls is located.

POLYPOSIS OF THE INTESTINE

Colon polyps — a tumor formation, above the level of the mucous membrane in the form of spherical, mushroom or branching growths, sitting on a stem or wide basis. The basis of the polyp constitutes growths of glandular epithelium in the form of numerous glandular tubes or in the form of branched villus, covered with high columnar epithelium. Supporting tissue is represented by connective base containing muscle fibers. (S. A. Holdin) (Fig. 1.1.11, 1.1.12).

On the prevalence of polyps, V. D. Fedorov distinguishes solitary polyps, multiple (group, scattered) and diffuse (familial) polyposis of the colon.

There are various theories of the etiology and pathogenesis of polyps and polyposis of the gastrointestinal tract (inflammatory, embryonic, disregenerative, viral and others.). According to the theory of embryonic dystopia, the so-called true polyps are the result of misuse of the embryonic development of the gastric mucosa. This includes strabismus pancreatic tissues, brunner glands, which have high growth potential energy and are stored in the gastric mucosa from the embryonic period.

The clinical picture of colon polyps has not a sign that is characteristic only for polyposis. Some symptoms may appear depending on the number and size of polyps, their localization, histological structure and the presence or absence of malignancy. The clinical picture depends on the preceding or acceded to polyposis effects of inflammation of the intestinal mucosa, or other pathological process, on a background of which arose polyps.

Abnormal discharges (blood, mucus) during defecation — the most frequent clinical manifestation of colonic polyps in children. They occur in 55–89% of patients. At the beginning of disease, bleedings are rare and not abundant, but as the process evolves,

becomes permanent and considerable, and sometimes the blood stream is released. At the low location of polyps, blood drops have red color.

At high localization-blood color changes as it is mixed with feces. During the motion of hard feces in sigmoid colon at constipations may be damaged the surface of the polyp, which causes or increases the bleeding. Constant prolonged bleedings and joining of diarrhea cause general weakness, dizziness, headache, anemia and lead to depletion of the patient. Constipation and diarrhea are observed in 55.2% of patients. Persistent diarrhea dehydrates and depletes the body and aggravates the patient's general condition.

At multiple and diffuse polyposis severity of clinical manifestations is greatly increased by the addition of concomitant colitis. Diarrhea is accompanied by very painful tenesmus with the secretions of mucus and blood. Therefore the picture of disease is sometimes very similar to dysentery and patients are often wrongly admitted to the infectious department. In some cases, the disease is complicated by the phenomena of intestinal obstruction, intussusception, falling out of individual polyps.

Abdominal pain, pain in the rectum, itching and burning sensation in the anal passage is observed in 41.6–64.7% of patients. Pain in the abdomen and rectum are usually dull, nagging, radiating to the sacrum and lower back. They are more often localized in the left half of the abdomen and in the lower divisions.

Dyspeptic disorders — nausea, belching, heartburn, sometimes vomiting and loss of appetite — are observed in 8–10% of patients. General weakness, weight loss, anemia occur in 5–7% of patients. Asymptomatic disease is observed in 24% of patients with solitary polyps of the colon. At malignization of polyps and disseminated multiple polyposis manifestations of local and systemic clinical symptoms are more pronounced. To establish the nature of pathological changes in the colon also helps scatological study.

Signs of inflammation of the colon are the presence of mucus in the feces and positive samples for the level of soluble proteins.

Hereditary polyposis of the gastrointestinal tract (GIT), especially in children, is rare. There are several external symptoms (joint hypermobility, hamartomas, osteoma, dark spots), which under the law of linked genes with a high probability allow to speak in favor of systemic hereditary pathology. The most frequent is Peutz–Jeghers syndrome — intestinal polyposis with spotty pigmentation of skin, mucous membranes of the lips and cheeks. It is possible that it is a variant of juvenile polyposis of the intestine.

Peutz first described it in 1921, based on observations of a family of 5 children suffering from polyposis of the gastrointestinal tract in combination with pigmented spots on the face (lips, cheeks, around the mouth), and on the palms, and in 1949 Jeghers reported about 12 such patients, most of whom were members of the same family, with the characteristic triad: polyposis of gastrointestinal tract, the presence of pigment spots and hereditary of disease. Transmitted with complete penetrance but different expres-

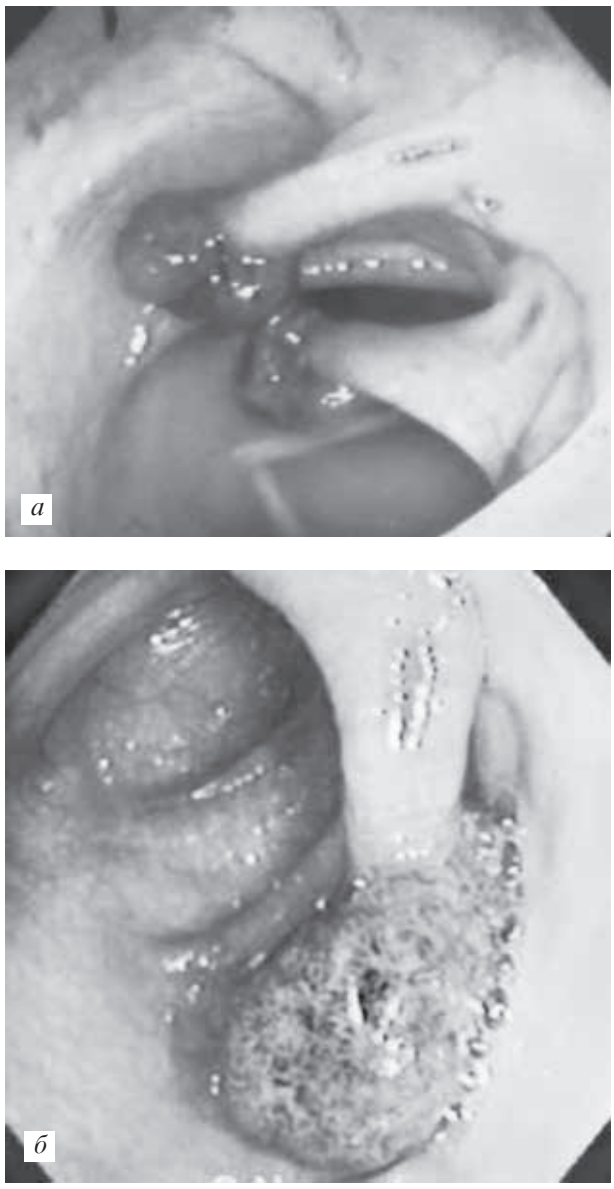


Fig. 1.1.11. Polyps on the stem *(a, b)*

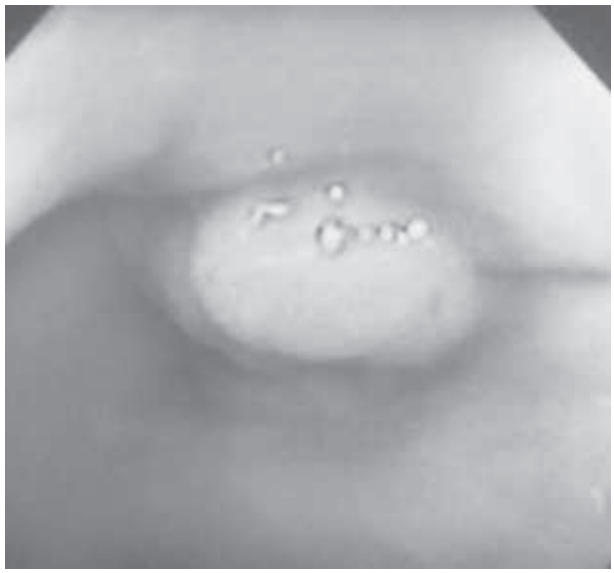


Fig. 1.1.12. Polyps on wide basis

sivity. In this case, on the face, especially around the mouth, in the mouth, more rare — on elbows, on the subungual lodges there do small dark spots of color «coffee with milk», resemble freckles. Spots appear in early childhood or have been since birth. In the small intestine, more rare — in the stomach and colon are detected multiple polyps with a relatively low probability of malignancy. In 50% of all polyps they are also found in close relatives. Malignancy of polyps is observed primarily during their manifestation already in early childhood. The probability of malignization increases after 35 years (Fig. 1.1.13).

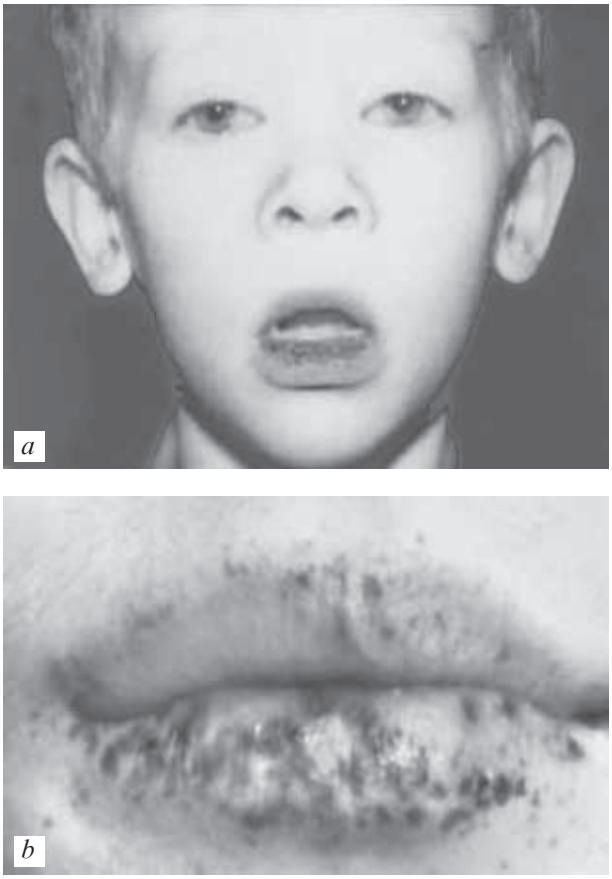


Fig. 1.1.13. Hyperpigmentation of lips' red border at Peutz—Jeghers syndrome *(a, b)*

Fig. 1.1.14. View of the bowel at Peyts—Jeghers syndrome

Clinical picture of Peutz–Jeghers syndrome is reduced to a non-specific abdominal pain, occult bleeding, iron deficiency anemia, although there may occur ileus, malabsorption syndrome with eating disorders. In 5% of cases, this type of bowel polyposis is combined with hormonally active ovarian tumors. There is a combination of Peutz–Jeghers syndrome with mixom syndrome (tumor of the heart) in the child.

Diagnosis of intestinal polyps is performed endoscopically. During a colonoscopy at detection of polyps, is certainly made their biopsy using biopsy forceps (Fig. 1.1.14).

Radiology. X-ray at colitis is detected circular uniform narrowing of the intestine lumen. In narrowed place contrast mass is not delayed, while at inflating with air spastic colon department is not straightened. If polyps are accompanied by phenomena of colitis, in this case, are quite clearly visible rounded shades.

Clinical picture of colon polyps

Clinical picture of colon polyps depends on the location, the number of polyps and their histological structure.

Based on these features, 4 groups of clinical symptoms are:

1. Pain and discomfort (anal itching, heaviness, burning sensation in the anus and rectum, and pain in various parts of the abdomen).
2. Abnormal discharges from the rectum (mucous, bloody, mixed).
3. Violation of bowel function (constipation, diarrhea, constipation and diarrhea change, frequent urge to the bottom and difficulties of defecating)
4. The presence of tumor formation in the anus.

Bleeding from the rectum — the most frequent symptom at polyps is observed in 55% of children.

The diagnosis of colon polyps settle on the basis of history and physical examination of the patient, which includes an external examination of the anal region, digital examination of the rectum, examination by rectal mirrors, sigmoidoscopy, barium enema, colonoscopy, scopy with biopsy, laboratory methods and, in particular, scatological study. According to indications sometimes produce combined colon laparoscopy.

Examination of the anus. Attention should be paid to the coloring and condition of the skin, state of the anal mucosa ring. Detect the presence of cracks, hemorrhoids, varicose veins in the process of thrombosis and scarring. In cases when there is a falling of the polyp and it's infringement in anus, you can determine the nature of formation.

Manual research of the rectum — a widespread and mandatory method of study, which measures the state of the rectal mucosa at a distance of 10–11 cm; and at bimanual examination even of 12–13 cm from the anus. Manual research can be carried out with the patient on his knees with a slightly tilted forward trunk in the left lateral position, squatting and bimanual in patients' position lying on his back. At manual research you can reveal the presence of a polyp or other pathological process in the rectum, its size, shape, consistency, mobility, prevalence of the

process, the attitude to the underlying department of the intestine, the presence of abnormal discharge in the form of blood or pus, leaving a mark on the tip of the finger.

Treatment. Both conservative and various surgical methods of treatment are offered. Polyps of the colon are removed through a rectoscope.

HEMORRHOIDS — a disease, manifested in varicose veins of the lower rectum and anus. As the most frequent proctologic disease in adults, hemorrhoids as a disease in children is rare and mostly in older age. Normally, all people have, under the mucous membrane of the anal canal, hemorrhoidal venous plexus, in its structure similar to the erectile tissues of the genital organs. They are believed to play a significant role in keeping the intestinal contents, providing a complete closure of the anal canal at filling with blood. Among the main mechanisms of formation of hemorrhoids mark: varicose veins of the rectum, vascular hyperplasia and "shifting" of the rectal mucosa from its foundation.

Most of these violations occur in high-risk groups, which include children and adolescents with hereditary predisposition, suffering from constipation, operating hard physical work.

Hemorrhoids is traditionally characterized by two main symptoms — bleeding and falling of nodes from the anal canal. At this disease are also marked such symptoms as anal itching, discomfort in the anal canal, slime secretion. They distinguish acute (or thrombosis of hemorrhoids nodes, which manifests in increase and induration of hemorrhoids nodes, pain in the anus, inflammation of the mucous membrane of the rectum) and chronic hemorrhoids (the main manifestations are episodic bleeding and prolapsed hemorrhoid). In addition, inner, outer and combined hemorrhoids.

Cases of typical clinical picture of hemorrhoids for childhood are rare. In most cases, parents are turning to the surgeon with complaints that during defecation and straining appear knobby protrusions near the anus. Rather characteristic is the absence of subjective complaints on pain or itching and symptoms of bleeding or inflammation. In older children, first there is a feeling of a foreign body in the anal canal, followed by itching and burning. Soon adds pain, increased during defecation. One or more external hemorrhoids nodes swell, increase and inflame. Palpation of the area is sharply painful. The body temperature can rise to 38–38.5°C. Bleeding is usually absent.

Diagnosis of hemorrhoids is relatively simple. It is enough to have thorough history, external examination, digital examination and, in some cases, anoscopy (sigmoidoscopy) to diagnose hemorrhoids. Differential diagnosis should be made with rectal polyps, anal fissure, hemangioma, and paraproctitis (Fig. 1.1.15, 1.1.16).

Treatment of hemorrhoids. At initial manifestations of hemorrhoids they appoint conservative treatment (no surgery). To a complex of conservative treatment include measures to normalize the stool, preferably with the help of foods, containing plant fiber in large amounts (wheat bran, kelp, microcrystalline cellulose) or laxative preparations like forlaks, dufalac. Combined rectal suppositories: anuzol, ultraproct, doksiprokt, proctosedyl et al. are administered.

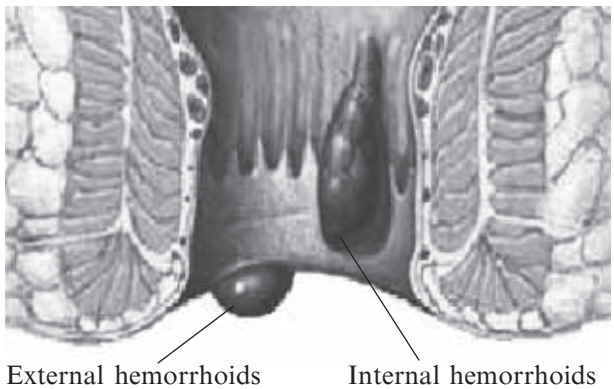


Fig. 1.1.15. Hemorrhoidal venous plexus

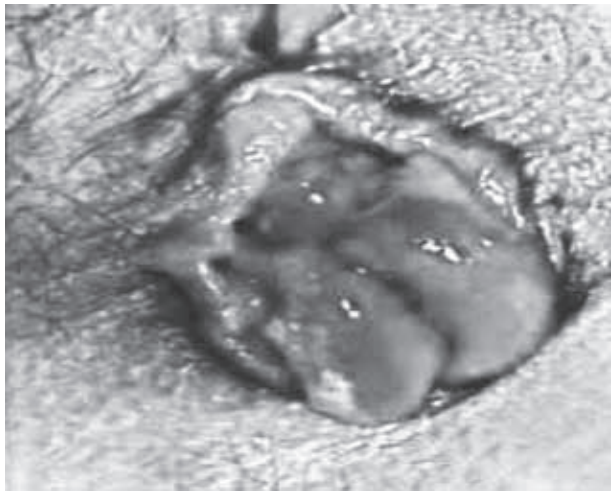


Fig. 1.1.16. Prolapsed hemorrhoids

In cases of acute hemorrhoids range of therapeutic interventions consists of assigning patients bed rest, sparing diet, medications for normalization of stool, cold lotions within the first few days. In rare cases, when conservative treatment is unsuccessful, resort to sclerotherapy, infrared coagulation, ligation of nodes with latex rings or their surgical removal (hemorrhoidectomy).

QUESTIONS FOR THE FINAL CONTROL

1. What indicators of clinical and laboratory tests (blood pressure, pulse rate, a general analysis and biochemical blood tests) show a hemorrhagic syndrome and bleeding in the abdominal cavity?
2. Name main clinical manifestations of bleeding from the upper and lower parts of the digestive system.

3. What are the main causes of bleeding from the digestive system.

4. Evaluate gastric contents and feces, depending on the height of the source of bleeding.

5. What are the general principles of treatment and bleeding arrest?

6. What are features of control over the clinical course of bleeding from the digestive tract? Determine the indications for surgical treatment.

7. Identify the indications for conservative and surgical intervention.

Practical Skills

1. Insertion of a nasogastric tube.

2. Temporary stop of venous bleeding in 1/3 of the right forearm.

3. Temporary stop of arterial bleeding from 1/3 of the hip, capillary bleeding of inner surface of the left forearm.

4. Setting the enema to a child of 7 years old, with evidences of gastrointestinal bleeding.

5. Determination of the shock index.

6. Determination of the extent of bleeding by objective data.

7. Conducting superficial and deep palpation of the abdomen.

1.2. ACUTE SURGICAL ABDOMINAL DISEASES IN CHILDREN

Specific objectives:

1. Learn the basic list of surgical inflammatory diseases of the abdominal cavity in children.

2. Learn the main clinical manifestations and local symptoms of inflammation of the abdominal cavity.

3. Differentiate symptoms of inflammatory diseases of the abdominal organs, which are need surgery.

4. Interpret additional research methods (X-ray, ultrasound, digital rectal examination), laboratory tests, medical and diagnostic laparoscopy.

5. Demonstrate features of examination of the child with surgical pathology of the abdominal cavity, to determine the local symptoms of inflammation of the abdominal cavity.

6. Analyze the causal relationships of inflammation of the abdominal cavity and their complications.

7. Learn the actions algorithm of the doctor in case of clinical inflammation of the abdominal cavity.

8. Identify specific actions of the doctor at a prehospital stage and the general principles of treatment of children with inflammation of the abdominal cavity which require surgical intervention.

ACUTE APPENDICITIS

1. Actuality of the problem

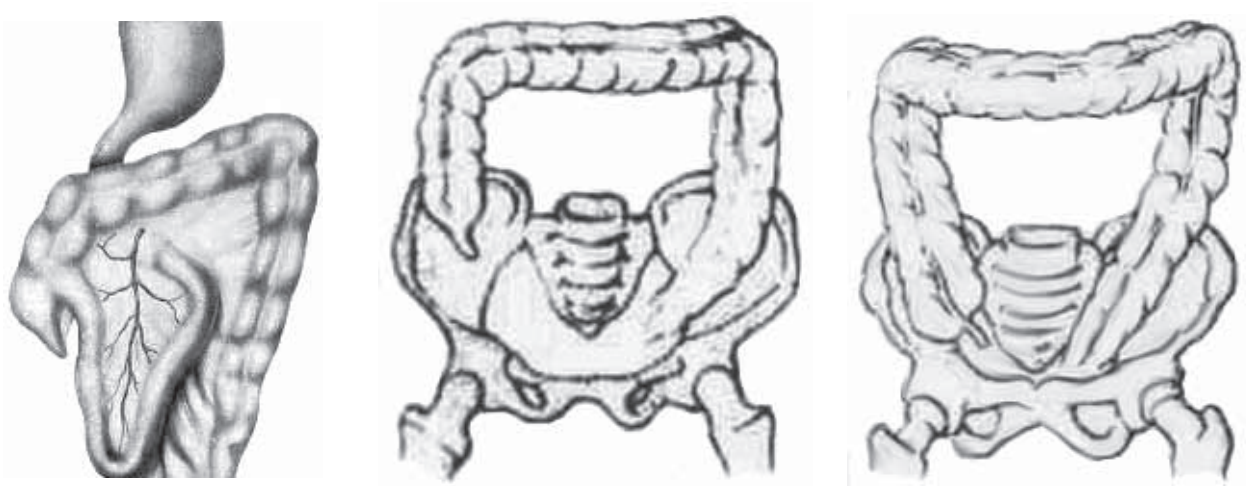
— The most frequent surgical abdominal pathology of children at which time determines the result.

— The number of diagnostic errors at a prehospital stage is over 50–70%.

2. Morphological and functional features of the abdominal cavity in children

— "Peak" of incidence occurs between the ages of 7 to 12 years (80%), and in children from 0 to 3 years the disease occurs in only 3% of cases. This is related to the funnel shape of the appendix (up to 2 years); a minor amount of lymphoid tissue in the wall of the appendix (up to 1 month there are no follicles, first 3 years — their number is small, only at 3 years appear reactive centers); V. Gerlachi is unexpressed or absent at this age, the wall of the appendix is thin, the crypt is not pronounced; immature nervous apparatus.

— Features of the ileocecal angle — only 60% of cases cecum is located in the iliac fossa, in the other 40% can be highly-situated, hypermobile, situated on the left (Fig. 1.2.1).



a *b* *c*

Fig. 1.2.1. Variants of appendix location *(a–c)*

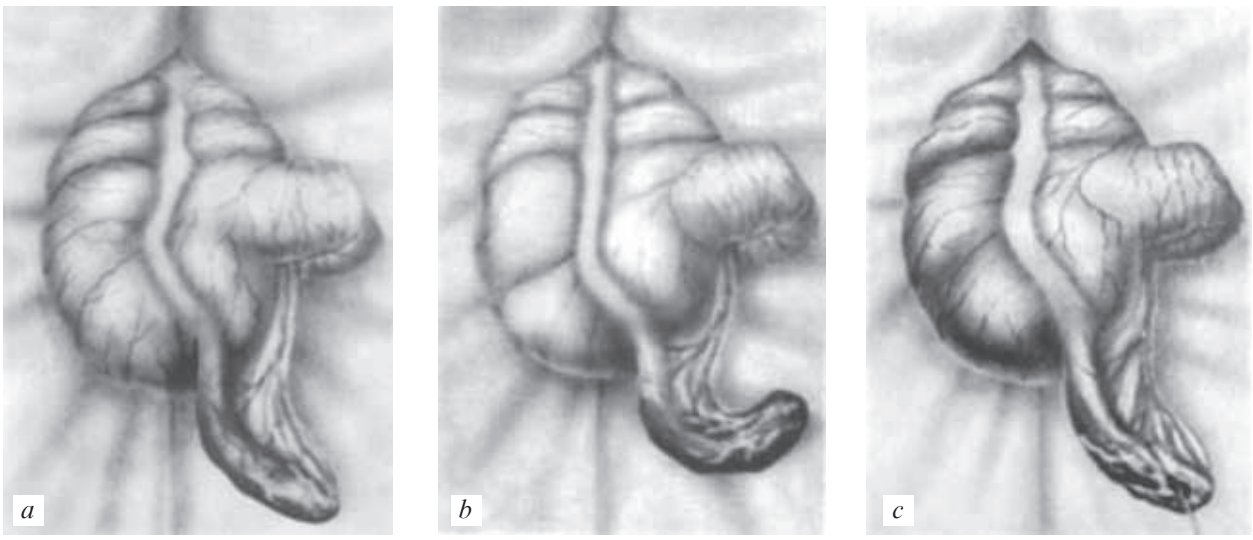


Fig. 1.2.2. Forms of acute appendicitis: catarrhal *(a)*, phlegmonous *(b)*, gangrenous *(c)*

— The abdomen has a relatively small size, the peritoneum is characterized by weak plastic qualities, omentum is short (only to 3 years reaches the level of the navel).

In addition, young children have the immaturity of the central nervous system, all of which leads to the rapid spread of purulent process in the abdominal cavity and the prevalence of the common symptoms over local in young children.

3. Clinical forms (Sprengel and A. Rusakov classification).

I) Simple — acute catarrhal appendicitis — appendix looks erythematous, edematous.

II) Destructive-phlegmonous appendicitis — appendage is covered with fibrinopurulent formation, there are purulent contents in abdominal cavity; gangrenous appendicitis — on the wall of the appendix there are areas of gangrene.

III) Perforated appendicitis — wall of appendix has perforated hole, content in the peritoneal cavity is purulent.

IV) Complicated: appendicular infiltrate, peritonitis.

Forms of acute appendicitis: catarrhal, phlegmonous, gangrenous see at Fig. 1.2.2.

4. Stages of emergency measures in acute appendicitis:

— Inspection on pre-hospital phase. Definition of local symptoms: Filatov, Sitkovskiy, Kocher, Shchotkin–Blumberg, Obraztsov, Moskalenko).

— Sending to surgical ward.

— Additional diagnostic methods. Digital rectal examination, thermometry, measurement of pulse, blood tests and urine tests. If necessary, ultrasound is used.

— General anesthesia. Laparoscopic or open appendectomy: examination of the abdominal cavity, exception of Meckel's diverticulum and pelvic pathology in girls.

Table 3

Diagnosis of Acute Appendicitis in Children of Different Age

Stages	Children of older age	Children of younger age
Complaints	Pain — appears in the epigastric region or navel and shifts to the right iliac region, has constant aching in nature, single vomiting, and refusal to eat.	Behavior changes, anxiety of a child, refusal to eat, repeated vomiting, loose stools.
Objective study	Subfebrile temperature, tachycardia, **local pain and muscle tension in the right iliac region**, positive symptom of Shchetkin–Blumberg.	Strengthening of screaming, pulling up of the right leg, pushing away the surgeon's hand on palpation of the right iliac region. **Inspection during sleep** — "pasty" muscle strain in the right iliac region. **Rectal examination** — pain, swelling, overhanging of the rectal wall, the presence of infiltration.
Laboratory study	Leukocytosis — 9–14 g/l, leukocyte formula is shifted to the left.	Leukocytosis — 12–25 g/l, leukocyte formula is shift to the left. Urinalysis — protein, leucocyturia, individual red blood cells. Coprogram — mucus, some leukocytes and erythrocytes.

Table 4

Diagnosis of Atypical Forms of Acute Appendicitis

Stages	Retrocecal location	Pelvic location	Subhepatic location	Left-hand location
Complains	Pain in the right flank, the right lumbar region, radiating to the genitals, dysuric phenomenon	Pain in the lower abdomen above the pubis, irradiation to genitals, dysuric phenomenon, frequent liquid stool	Pain in the right upper quadrant, repeated vomiting, Ortner symptoms	Pain in the left half of the abdomen, single vomiting
Objective study	Acute pain and muscle tension in the lumbar region on the right.	Rectal examination — the pain, the overhang, swelling of the bowel wall	Pain and muscle tension in the right upper quadrant, positive Shchetkin–Blumberg symptom	Pain and muscle tension in the left upper quadrant, positive Shchetkin–Blumberg symptom
Laboratory study	Urinalysis — leucocyturia	Urinalysis — white blood cells, red blood cells, squamous epithelium. Coprogram-mucus, red blood cells, white blood cells		

Diagnosis of acute appendicitis in children of different ages is represented in Table 3.

Diagnosis of atypical forms of acute appendicitis is represented in Table 4.

The most common surgical disease in children — **acute appendicitis**. Since it is encountered not only by pediatric surgeons, but also pediatricians, general surgeons, emergency physicians, and physicians in any other specialty. Diagnostic difficulties in acute appendicitis in children are associated with features of the reactivity of the child's body, the relative immaturity of the central nervous system, frequent atypical location of the appendix, and the features of its structure (Fig. 1.2.3).

Like adults, surgeons, pediatric surgeons use classification of Sprengel A. Rusakov, which is divided on pathological forms of the process and their clinical counterparts: catarrhal appendicitis (simple), abscess, gangrenous (destructive), ruptured (complicated). Children have same pathologic stages as adults, but their rates differ and the smaller the child, the greater the rate (Fig. 1.2.4).

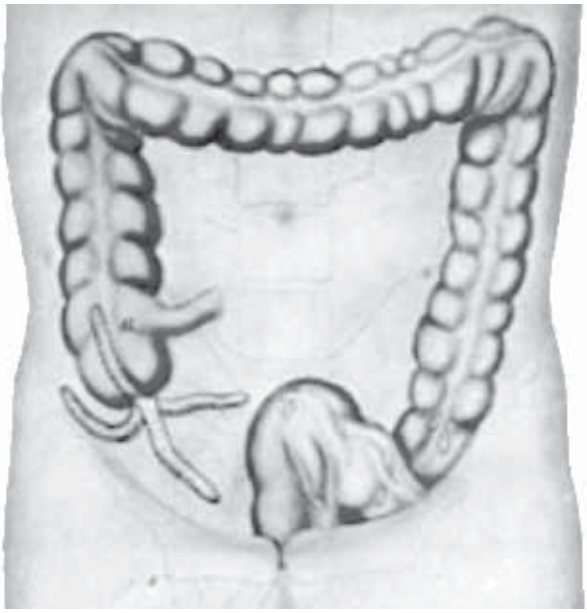


Fig. 1.2.3. Layout vermiform appendix

Clinical picture. In typical cases acute appendicitis begins gradually. Initially, there is pain, which during the first hours of the disease is localized in the epigastric region or around the navel, and then moves to the right iliac region — pain of final localization (G. P. Krasnobaev). Localization of pain depends on the location of the appendix: retrocecal — pain in the lumbar region, at subhepatic — in the liver. Due to the pain the older children often take forced position: on the right side, on the back, as this reduces the tension of the mesentery and peritoneal inflammation; in the left lateral position the pain increases (symptom of M. B. Sitkovskiy). Most often, the pain is constant, aching in nature. Pain — a subjective symptom and if older children indicate pain, the little kids act up (equivalent to pain — conduct disorder), restless, children refuse to eat, sleep disturbance. Describing the behavior of young children, T. P. Krasnobaev points out: "The child with acute appendicitis does not sleep himself and doesn't give sleeping to others" (Fig. 2.5).

Common symptom of acute appendicitis is vomiting in the early hours of the disease, which is reflexive in nature. Vomiting in older children, 1–2 multiple, observed in 80% of cases, in 20% of cases can only be nausea. If the appendix is located at the root of the mesentery, there is repeated vomiting. In young children, in 85% of cases single vomiting is repeated.

A typical picture of acute appendicitis is characterized by low-grade fever, but in young children and at complicated appendicitis — high. Symptom of the "scissors" (inconsistent heart rate and temperature) is characteristic only for older children.

At catarrhal appendicitis tongue is usually wet, but incrustated at the root, at phlegmonous — moist,

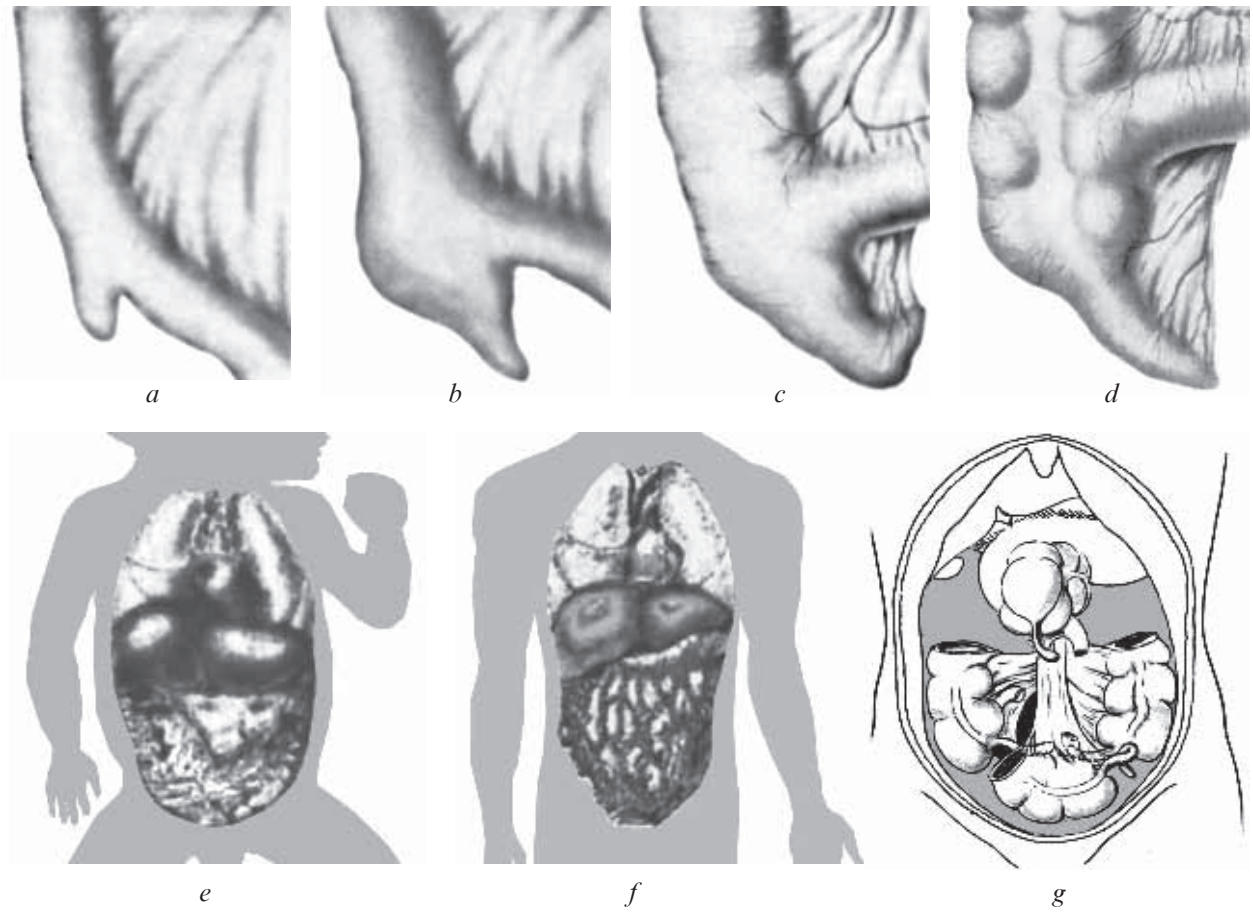


Fig. 1.2.4. A cone-form vermiform appendix *(a–d);* a short omentum in a 3-year old child *(e)*; omentum in an adult *(f); a* mobile blind gut (cecum) *(g)*

incrustated entirely; gangrenous changes are accompanied by dryness and incrustation of the tongue; at peritonitis it becomes massive.

In 35% of cases there is a delay of the stool, but in young children the stool is liquid, frequent, in connection with what Fevre introduces such a term as "diarrheal appendicitis". Frequent stools can sometimes be observed in older children with conditions of the medial location of the appendix. Urination, as a rule, is not broken, but the pelvic location of the appendix may be pollakiuria, which is associated with irritation of the bladder and may be accompanied by leukocyturia.

Given the above, abdominal pain — the main, leading symptom, but in young children predominate common manifestations (conduct disorder, anxiety,

Fig. 1.2.5. Superficial abdominal palpation contraclock wise *(a)*; superficial bimanual palpation by Krasnobayev *(b)*

high fever, repeated vomiting, frequent stools) that may properly orient the doctor into thinking about infectious diseases. As for the newborn, the difficulties in the diagnosis is so great, that appendicitis is diagnosed, as a rule, during surgery for peritonitis, but not in the preoperative period.

At objective research recognition of acute appendicitis in children is based on the identification of three main symptoms:

1. Provoked pain;
2. Defence musculaire;
3 Symptoms of peritoneal irritation.

At survey the shape of the abdomen is not changed, in the initial stages of the disease it is involved in the act of breathing. Superficial palpation of the abdomen begins with the left iliac region and is produced in a counterclockwise direction. Pain will grow at deep palpation (F. F. Filatov). At transition to the right half of the abdomen it is necessary to monitor the reaction of the child, expression and distract his attention to the conversation. In young patients about provoked pain indicate a symptom of "repulsion of hands" of the doctor and pulling the right foot.

At palpation of abdomen reveale muscle tension of the anterior abdominal wall in the right iliac fossa (passive muscle tension) — one of the leading objective symptoms of acute appendicitis. B. P. Voznesensky, S. D. Ternovsky, T. P. Krasnobayev, emphasizing its importance, write: "Where there is no defence musculaire, there is no acute appendicitis." In order to its better detection it is necessary simultaneously to pursue abdominal palpation on both sides — both hands of the doctor are placed parallel by plane palms on the anterior abdominal wall of the child (left — on the right half, right ~ on the left) and, like "playing scales" the doctor presses alternately right to left by determining the difference of muscle tone (Fig. 2.5). In children is rarely determined wooden belly, there is only a very mild but constant rigid tension of "dough" consistency.

Diagnostic value has a symptom of Shchetkin–Blumberg, which is traditionally identified or using gentle methods: local pain at percussion (symptom of Mendel); dosage percussion (at percussion from clearly intact areas in the direction to the center by A. R. Shurinku pain increases).

Most symptoms in pediatric practice are not used. **Mendel** wrote: "The symptoms of appendicitis should not be counted but weighed" and most important of them is the pain and defence. S. Y. Doletsky thinks that **"diagnostic key"** of acute appendicitis includes pain (independent and provoked) and defence. Symptoms of peritoneal irritation may be absent (retrocecal, retroperitoneal appendicitis, dense ambience of appendix by omentum).

S. Y. Doletsky introduced the concept of **"negative symptoms"** in the diagnostic phase: headache, muscle pain, rumbling in the stomach, liquid and offensive stools, hyperthermia, hyperleukocytosis. The first five symptoms testify against acute appendicitis but the last two — a complication of peritonitis.

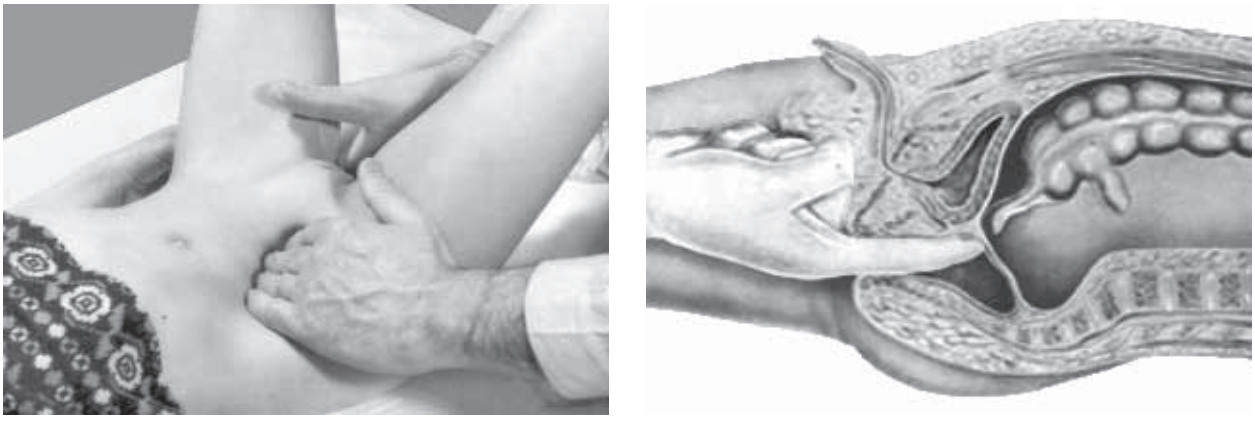
Taking into account the difficulty of collecting history, contact with a small child, the definition of "diagnostic key" in children is of prime importance and for this purpose use auxiliary survey methods:

1. Palpation by child's hand (the child will resist deep pressing).
2. Palpation in mother's hands (the doctoris behind baby's back, who sits on his mother's laps).
3. Examination during physiological sleep.
4. Inspection during medication sleep (enema with 3% chloral hydrate, anesthesia, in a dream is level ed active muscle tension caused by baby's distress, and provoked pain and passive defence musculaire remain).
5. Recto-abdominal bimanual examinations (verifies pelvic appendicitis and infiltration in the lower abdomen) (Fig. 2.6.).

Data of laboratory studies in acute appendicitis is not specific and can only indicate the presence of inflammation.

The clinical picture of atypical location of the appendix. Acute appendicitis can occur with moderate pain in the right upper quadrant, without vomiting and normal temperature or loose stools and dysuric phenomena without muscle strain. Atypical acute appendicitis depends primarily on the location of vermiform appendix in the abdomen.

At retrocecal intraperitoneal appendicitis muscle tension and pain at a palpation will be less than at a typical location. At the retroperitoneal location of



a *b*

Fig. 1.2.6. Recto-abdominal bimanual examinations *(a, b)*

appendix the abdomen may be soft all over, not very painful, symptom of Shchetkin–Blumberg is usually negative. In such cases, pain and muscle tension are identified in the right lumbar region, and pain radiates to the genitals or in the course of the ureter.

At pelvic location, pain is located in the abdomen, above the pubis, muscle tension is absent or weakly pronounced. Pain may radiate to the genitals, dysuric phenomenon is marked, loose stools with mucus. At involving bladder in inflammation urinalysis reveals white blood cells, red blood cells, squamous epithelium. At subhepatic location of appendix its inflammation begins with pain in the right upper quadrant, here is determined muscle tension and pain on palpation and effleurage on the right costal arch. In the reverse location of the internal organs, the mobile cecum or long appendix, all clinical manifestations of appendicitis will be localized in the left half of the abdomen. At gangrenous appendicitis clinical picture peculiarity is caused by lesion of the nervous apparatus of the appendix, which is manifested in subsided abdominal pain.

Stomach is involved in the act of breathing, soft, there is a slight tenderness at deep palpation. The relative well-being is marked before the development of peritonitis pattern. These children can be revealed tachycardia without the proper degree of hyperthermia, leukocytosis with a shift of leukocyte formula to the left.

Clinical picture in infants. Clinical picture of acute appendicitis in children under 3 years is characterized by rapid start. The child becomes restless, refuses to eat, there is repeated vomiting and rise in temperature to 38–40°C. Often there is diarrhea, frequent and painful urination. At palpation of the right iliac fossa a child resists examination, pushes the hand of surgeon, pulls up the right leg, a baby's cry is greatly enhanced. It is desirable to examine infants with suspected acute appendicitis during a medical or physiological sleep when active muscle tension disappears and passive tension, caused by inflammation, persists.

In children with suspected acute appendicitis they make digital examination of the rectum. Thus it is possible to identify very painful overhang and dope of rectal wall, at the late arrival of a child infiltrate is identified.

An objective assessment of muscle tension is allowed by electromyography study of the anterior abdominal wall. Laparoscopic studies in doubtful cases allow to unerringly visually and confirm or reject the destructive process in the appendix. In the absence of acute appendicitis laparoscopy allows in 1/3 of cases identify the true cause of abdominal pain syndrome.

Complications of acute appendicitis. With late diagnosis of acute appendicitis it is necessary to meet its complications: peritonitis and appendicular infiltrate. Perforation of the appendix manifests in increased abdominal pain and worsening of the patient's condition. There is repeated vomiting, fever up to 39–40°C. Skin becomes pale and dry, facial features are sharpened. Peritonitis has a typical symptom of difference in pulse rate and temperature. Stomach is not involved in the act of breathing; palpation is determined by a acute pain and muscle tension in all parts of the abdomen, more in the right iliac fossa. Symptom of Shchetkin–Blumberg is also positive in all departments. With increasing intoxication child becomes adynamic, sluggish, and lethargic, the intensity of pain in the stomach is reduced. Laboratory studies confirm the presence of severe inflammation.

Appendicular infiltrate may be formed in older children on 3rd–5th days from the beginning of the disease. In young children, the possibility of limiting the inflammatory process in the abdominal cavity is insufficient due to weak plastic properties of the peritoneum and omentum underdevelopment. In the formation of infiltration pain intensity decreases, but symptoms of intoxication, high temperature persist. In the right part of the stomach is determined a dense, very painful, tumor formation without clear boundaries. At an atypical location of the appendix infiltration can be detected in the pelvis, the left half of the abdomen, the right lumbar region. Are often marked dysuric phenomenon and loose stools. In blood leukocytosis, leukocyte formula shift to the left, increased erythrocyte sedimentation rate are revealed.

Taking into account the difficulty of diagnosis, and especially in children, one should follow the management enshrined by order of the Ministry of Health of Ukraine: "Children up to three years with vague abdominal pain should be admitted to a surgical hospital" where doctors — surgeons can correctly assess the whole set of clinical symptoms, discover the "diagnostic key" and act appropriately.

At late diagnosis of acute appendicitis develop complications caused by the destruction process and in the first turn — perforated peritonitis, which, depending on the process may be diffuse and local (unlimited and limited — infiltration, abscess). During peritonitis there are three phases (reactive, toxic, terminalon K. S. Simonyan), duration and severity of which depends primarily on the child's age and the prevalence of pathological process — and the smaller the child, the faster develops peritonitis, the more pronounced will be a violation of the general and the local state, the basis of which are progressive toxemia and dismetabolism on the background of morphological and functional immaturity of systems, organs and tissues. With the development of diffuse appendicular peritonitis child's condition is severe with significant fever, vomiting with multiple bile, pale skin, sunken eyes, the nose is pointed (Hippocrates face), dry tongue with a touch, tachypnea and tachycardia are pronounced, the stomach is not involved in respiration, painful and tense all over, pronounced signs of peritoneal irritation, marked oliguria, young children may have diarrhea. Local peritonitis is also characterized by high fever, intoxication, but course is milder, pain, defence musculaire and irritation of the peritoneum are limited, and at appendicular infiltrate it is defined as a painful tumor in the right iliac fossa.

Treatment. The basic principle of the treatment of acute appendicitis is early surgical intervention. Appendectomy is typically performed with immersion of the stump appendix under the purse-string suture, in

infants — without immersing of the stump (to prevent deformation of v. Bauhini and bowel perforation by seams). Children with pronounced symptoms of intoxication, metabolic disorders are in need of short-term intensive preoperative preparation. The only indication for conservative treatment is the presence of a dense still infiltrate. Treatment consists of antibiotics of broad-spectrum, introduction of antibiotics through the retroperitoneal microirrigator by the method of the department of physiotherapy treatment with anti-inflammatory action, active restorative therapy. At active treatment regression of the infiltrate takes place. A child who has had appendicular infiltrate, 1 month after completion of treatment should be hospitalized to perform an appendectomy. At suppuration of appendicular infiltrate it is necessary to make its opening. Postoperatively, is pursuing an active antibacterial, anti-inflammatory and detoxification therapy. After 2–3 months in the scheduled order appendectomy is performed.

In peritonitis conduct a 2–3-hour preoperative preparation, aimed at detoxification and correction of the disturbed functions, and then carry out a laparotomy and sanitation of the abdominal cavity (removal of the appendix, washing).

Differential diagnosis. Diseases, with which most often one have to differentiate acute appendicitis in children **up to 3 years**:

1. Intestinal colic, coprostasia.
2. ARVI, acute bronchitis, pneumonia, pleurisy.
3. Intestinal infection (salmonellosis, dysentery, yersiniosis, amoebiasis), dysbacteriosis, worm infestation.
4. Childhood diseases — measles, rubella, scarlet fever, chicken pox.

In older children:

1. Diseases of the biliary system — biliary dyskinesia, acute cholecystitis.
2. Diseases of the urinary system — pyelonephritis, glomerulonephritis, cystitis, urolithiasis.
3. Genital disease in girls — vulvovaginitis, adnexitis, apoplexy and ovarian torsion, hematocolpos.
4. Primary peritonitis.
5. Mesadenitis.
6. Crohn's disease.
7. Inflammation of Meckel's diverticulum.
8. Rheumatism, collagen.
9. Intestinal disease of Shenlejn — Henoch.
10. Malformations of the ileocecal angle — Jackson's membrane, Lane commissure, mobile cecum.
11. Psychogenic abdominal pain.

Meckel's diverticulum — a malformation of bile ductules, a cone-shaped protrusion located on the edge of mesenteric ileum in 20–100 cm from the ileocecal angle. Most often, a diverticulum is located freely in the abdominal cavity, in some cases the top of the diverticulum is connected by fibrous cord with parietal peritoneum in the navel. Inside the lumen the diverticulum is lined with a mucous membrane of enteric type, but sometimes is marked heterotopia of the gastric mucosa. Meckel's diverticulum is the result of incomplete regression of the intestinal part of the bile ductules. The presence of Meckel's diverticulum can lead to the following complications: intestinal obstruction, bleeding, inflammation.

Acute diverticulitis (catarrhal, phlegmonous, gangrene) begins with severe pain in the abdomen, which is soon localized in the navel or the right iliac fossa. Vomiting, fever, tachycardia are observed. The child refuses to eat, becomes sluggish. Palpation reveales pain in the navel or the right iliac fossa, muscle tension of the anterior abdominal wall, positive symptom of Shchetkin–Blumberg. The disease progresses rapidly, within 10–15 hours after the onset occurs diverticulum perforation and peritonitis develops. Differential diagnosis of acute appendicitis and diverticulitis is extremely complex, but it has no practical value. Both diseases require urgent surgical treatment. Used in acute appendicitis access by Volkovich–Diakonoff is convenient for resection of Meckel's diverticulum.

Mesadenitis — inflammation of the lymph nodes of the mesentery of the intestine — most often caused by adenovirus infection, so the frequency increases during periods of acute respiratory viral infection. The disease is manifested by acute pains in the abdomen, nausea, vomiting, and fever up to 38–39°C. Moderate muscle tension of the anterior abdominal wall and spilled soreness. Sternberg's symptom is pain at deep palpation in the course of attachment the root of the intestinal mesentery, which is typical for mesadenitis; in the left lateral position the pain shifts to the left — Klein's symptom.

Sometimes it is possible to probe bags of enlarged lymph nodes. At pronounced pain syndrome they use laparoscopy, which allows not only to inspect the abdominal cavity, but also do a lymph node biopsy. If mesadenitis is found during surgery, it is also necessary to make a biopsy of a lymph node of the intestinal mesentery. Treatment of mesadenitis consists in the appointment of broad spectrum antibiotics, desensitization drugs, multivitamins. Mesadenitis may have Yersinia, tuberculosis, lambliasis.

Among **gynecological diseases** abdominal pain syndrome can occur in inflammatory diseases of the uterus, ovarian apoplexy, torsion of an ovarian cyst. In acute salpingitis or oophoritis is marked pain in the lower abdomen, more on the right, sometimes indistinct signs of peritoneal irritation, low-grade fever, and leukocytosis. Rectal examination reveales tenderness of appendages. The disease occurs mainly in girls of 10–14 years old, mostly on the right. Violation of the integrity of the ovary with hemorrhage into the abdominal cavity is most often cited in the period of ovulation, when due to increased levels of ovarian hormones flushing, vasodilation and increased permeability of the vascular wall of the ovary take place.

Disease begins acutely, among full well-being from strong cramping pain in the abdomen, accompanied by dizziness, nausea, vomiting. Typically, into the abdominal cavity is poured 20–100 ml of blood, which is not accompanied by hemodynamic disturbances. At a significant blood loss is marked increased heart rate, decreased blood pressure, a fall in hemoglobin. At rectal examination is revealed a painful ovary during displacement of the uterus. Surgical treatment is indicated in the bleeding intensity

and consists in suturing of rupture and aspiration of blood from the abdominal cavity. If the bleeding is minor and stopped on its own, a conservative treatment is conducted: bed rest, cold on the lower abdomen, hemostatic drugs.

Primary peritonitis most commonly occurs in girls aged 5 to 10 years and is associated with the penetration of infection into the abdominal cavity through the vagina. Clinically distinguish two forms of the disease — simple and toxic. Toxic form begins acutely with severe pain in the abdomen, repeated vomiting, fever up to 39°C. Frequently they observe mucopurulent vaginal discharge, high leukocytosis. A few hours after the onset of the disease, the general condition of a child is estimated as severe.

On examination of abdomen reveal muscle tension and soreness in all departments, but more at the bottom right, the positive symptoms of peritoneal irritation. More common is a simple form of primary peritonitis in which intoxication is not expressed, subfebrile temperature, is marked moderate pain in the lower abdomen or only in the right iliac region. The use of laparoscopy in emergency pediatric surgery with high accuracy determines the cause of abdominal pain syndrome. In acute salpingitis fallopian tubes are hyperemic and edematous, mesenteric vascular tubes are injected, the ovaries are swollen and have a dull color, sometimes is marked hyperemia and hyperemia of vessels of pelvic peritoneum. Patients with inflammatory changes of the uterus should receive a course of antibiotic therapy, and then course of anti commissural therapy. Laparoscopic signs of pelvic peritonitis are detection of a large number of clear or turbid effusions in the pelvic cavity, sticky exudate, so reaching for the manipulator, is marked redness and swelling of the uterus and its appendages. The highest inflammatory changes are found in the ampullar part of the fallopian tubes. At toxic form of pelvic peritonitis effusion becomes purulent. Treatment of pelvic peritonitis consists in aspiration of effusion from the peritoneal cavity, introduction of antiseptic solution. Postoperatively antibacterial and detoxification therapy is conducted.

Acute cholecystitis is a rare disease in childhood. Infection can penetrate into the gallbladder by hematogenous, lymphatic or ascending enterogenous way. Inflammation is promoted by stagnated bile, which in children is most often caused by abnormalities of the gallbladder or cystic artery. Calculous cholecystitis in children is rare. Typically, acute cholecystitis begins with severe pain in the right upper quadrant or epigastrium. Pain is often paroxysmal in nature and lasts from a few minutes to several hours. When there is an obstacle for bile flow, which is often a mucopurulent clot, pain becomes very intense and causes the child to rush in bed or take forced position in which the child remains for a long time, afraid to move. Attacks are usually accompanied by repeated vomiting, not bringing relief, rapid breathing and heart rate, raising the temperature to 38–39°C, bloating, and constipation. Pale skin, covered with sticky sweat, approximately in half of cases there is jaundice.

At palpation of abdomen reveal acute pain and muscle tension in the right upper quadrant, positive symptoms of Ortner, Murphy, Zakharyin. The liver is enlarged and painful. Treatment of acute cholecystitis begins with conservative measures: bed rest, antibacterial and detoxification therapy, are appointed analgesic, antispasmodic and antifermental drugs. Absence of effect from conservative treatment is an indication for emergency surgery. If phlegmon or gangrene of the gallbladder are detected, they use cholecystectomy. If conduction of bladder removal is not possible because of the severe condition of the patient or the presence of significant infiltrate, it is necessary to perform a cholecystostomy. Surgery for acute cholecystitis ends with drainage of obstructive area.

Much more frequently than inflammation, there are biliary dyskinesia and cholecystopathy, related to violation of the diet, neuroses, helminthic invasion, dysbacteriosis, gastritis, duodenitis, and developmental abnormalities of the biliary tract.

Acute peritonitis — a disease characterized by swelling, inflammation, and necrosis of the pancreas, in children is rare. Its reasons can be duodenally-pancreatic reflux, dyskinesia of duodenum, injury, including operating, stress situations, infectious diseases (measles, mumps, dysentery and other) proceeding with a toxic-allergic component. The pathogenesis of pancreatitis is autolysis of pancreatic tissues by the ejection to the blood a large amounts of trypsin, elastase and other proteolytic enzymes. The first stage of acute pancreatitis — swelling of the pancreas begins with general non-specific symptoms: weakness, refusal to eat, uncertain abdominal complaints. After a few hours pain localizes in the epigastrium, has surrounding nature, radiating to shoulder girdle and are accompanied by repeated vomiting. The temperature may be normal or subfebrile, pulse is fast, and blood pressure is normal or slightly reduced. The child can take forced position, usually on the left side. Belly is not swollen, is involved in the act of breathing, muscle tension is absent. Such discrepancy between pronounced complaints of pain in the abdomen and the absence of objective evidence of inflammation in the abdominal cavity is typical for swelling of the pancreas. The most informative laboratory parameter is to define a higher level of amylase in the blood and urine. Ultrasound is useful for diagnosis. With the progression of the disease fatty and hemorrhagic pancreatic necrosis develop. Younger children are initially restless, crying out in pain, take forced position, then motor unrest is replaced by adynamia. Older children indicate severe pain in the upper abdomen, which often has belting nature. Despite the thirst, caused by repeated vomiting, the child refuses to drink, as each sip of water causes repeated vomiting. Pale skin, with cyanotic shade, dry tongue. Pulse is fast, blood pressure is reduced. The temperature is usually subfebrile; its raise up to 38–39°C indicates the development of fat necrosis. The child's condition is progressively deteriorated, exsicosis, toxicosis develop quickly. Examination reveals swollen belly, muscle tension, pronounced anterior abdominal wall tension, positive symptoms of peritoneal irritation. The blood anal-

ysis determines high leukocytosis, electrolyte disorders. Elevated levels of amylase in the blood and urine. Its sharp decline at the deterioration of the patient is a sign of total necrosis. The most informative method of diagnosis of acute pancreatitis is laparoscopic study, during which foci of fat necrosis in the form of whitish-yellow spots on the parietal and visceral peritoneum, in omentum, hemorrhagic effusion, hemorrhagic imbibition of omentum and retroperitoneal space are detected.

The abdominal cavity exudate contains increased levels of amylase. Difficulties at differential diagnosis are primarily connected with the fact that acute pancreatitis is a rare disease in children, and the doctor first thinks of a typical children disease. Very acute pain in the upper abdomen, frequent vomiting and rapid deterioration of the child testify agains acute appendicitis. The most typical symptoms of acute appendicitis — local pain and muscle tension of the anterior abdominal wall in the right iliac region is absent. Laboratory studies allow us to define the diagnosis. Principles of conservative treatment of acute pancreatitis are: to provide the rest of the pancreas, for which for 3–4 days enteral nutrition is forbidden; the struggle with pain syndrome, the most effective method is prolonged epidural anesthesia; antifermental therapy; antibiotic therapy; elimination of water and electrolyte balance disorders; detoxification therapy. Surgical treatment is used in the absence of conservative therapy effect at the progression of the disease. In the postoperative period using active drug therapy.

Crohn's disease — a nonspecific granulomatous inflammation that can affect any part of the digestive tract. Most often, about three-quarters of cases, the disease affects the terminal part of ileum. The exact etiology of Crohn's disease is unknown, although currently most authors attribute this disease to tissue diseases. Chronic granulomatous inflammation affects all layers of the intestinal wall with the formation of narrow, deep longitudinal ulcers in the mucosa, granulomas, increased mesenteric lymph nodes appear. Clinically, the disease is characterized by paroxysmal abdominal pain, repeated vomiting, fever up to 38°C. Sometimes the pain is so pronounced that a child is operated, suspecting acute appendicitis. During the operation, detect hyperemia, infiltration and thickening of the wall of the terminal ileum, and sometimes are affected blind and initial division of the colon. Regional lymph nodes are significantly increased in size. Sometimes, the disease takes a slow character and manifests in periodic complaints on abdominal pain, weakness, loss of appetite, dyspeptic disorders, and weight loss. In children rarely develop complications of Crohn's disease: perforation, intestinal stenosis, bleeding, and fistula. In the diagnosis help X-ray and endoscopic study of the colon. Treatment of Crohn's disease in children is conservative and the appointment of a diet with low fat level and high in protein and carbohydrates, are used vitamins, antihistamines, sedatives. They use antibacterial therapy. Surgical treatment is indicated at development of complications.

Nonspecific ulcerative colitis is an inflammation of all or a segment of the colon, which is based on autoimmune disorders. On the mucous membrane of the large intestine are formed erosion and ulcers, on the spots of which subsequently grows granulation tissue and occurs scarring. The disease usually begins with mild pain in the left half of the abdomen, blood in the stool. Stool, depending on the severity of the disease, appears from 3 to 10 times a day, there is a temperature rise, rapidly develops anemia, dysproteinemia, dysbacteriosis.

At endoscopic research of the colon are identified redness, swelling and bleeding of the mucous membrane on the surface of which are formed erosion and ulcers. Unlike Crohn's disease, ulcers have a transverse direction, are not spread to deeper mucosal layers, frequently damage rectum. Treatment is mainly conservative: nutritional therapy (a large number of high-grade, easily digestible proteins), preparations of Salazopyrin (treatment course is 4–8 months), in severe cases — detoxification and steroid therapy. Surgical treatment is indicated at development of complications — perforation of ulcers, profuse bleeding, toxic dilatation of the colon, fulminant disease course.

QUESTIONS FOR THE FINAL CONTROL

1. Give the main list of surgical diseases in children that cause inflammation of the abdominal cavity.

2. What are the main clinical manifestations and local symptoms of inflammation of the abdominal cavity?

3. What are the main clinical manifestations and local symptoms typical for inflammation of the abdominal cavity in newborns?

4. Name the features of clinical picture of acute appendicitis in young children of age less than 3 years old.

5. What are the indications for conservative treatment and the nature of peritonitis in the newborns?

6. Name the features of preoperative preparation at inflammation of the abdominal cavity.

7. What are the clinical signs and auxiliary diagnostic methods specific for destructive forms of cholecystitis?

8. Give the general principles of treatment of cholecystitis and define the indications for surgical treatment.

9. What are the main causes of acute pancreatitis?

10. State the indications for conservative and surgical treatment of acute pancreatitis, its volume and methods.

Practical skills:

1) Implementation of the superficial and deep palpation of the abdomen.

2) Determination of the symptoms of an acute abdomen in a child of 9 years old (muscle tension, irritation of the peritoneum).

3) Interpretation of liver ultrasound.

4) Diagnosis of abdominal cavity infiltrate.

1.3. ACQUIRED INTESTINAL OBSTRUCTION

Specific objectives:

1. To master a list of diseases that cause acquired obstruction.

2. Identify the main clinical symptoms of all types of acquired obstruction of the digestive system.

3. Identify the main reasons that cause intussusception, early and late adhesive obstruction, obstructive and dynamic obstruction.

4. Conduct differential diagnosis of acquired intestinal obstruction with other somatic and surgical diseases that cause abdominal syndrome.

5. Interpret supporting research techniques: ultrasound, sensing of the stomach, rectal and X-ray studies, posing a cleansing enema, using of the vapor tube, conducting pneumocolonography, laboratory and biochemical analyzes.

6. Demonstrate techniques of gastric intubation, digital rectal examination, conducting pneumocolonography, examination of the patient with acquired intestinal obstruction.

7. To analyze the causal relationships of acquired intestinal obstruction occurrence in some patients, justify and formulate a preliminary clinical diagnosis.

8. Learn the algorithm of actions of the doctor at intussusception, mechanical and dynamic acquired intestinal obstruction.

9. Justify basic principles of the treatment of diseases that are accompanied by acquired intestinal obstruction and define the indications for conservative and surgical treatments.

ACQUIRED INTESTINAL OBSTRUCTION

In children and in adults, acquired intestinal obstruction is divided into two main types — mechanical and dynamic. In children, in a group of mechanical obstruction mark obstructive, strangulated and intussusception. In turn, the cause of obstructive obstruction is often coprostasia at a congenital stenosis of the colon, Hirschsprung's disease, megacolon or fistulous form of rectal atresia. Strangulation obstruction is sometimes caused by a violation of the inverse development of yolk flow, or a consequence of other malformations. Nevertheless, pediatric surgery often faces the dynamic intestinal obstruction, with adhesive intestinal obstruction and intussusception.

DYNAMIC INTESTINAL OBSTRUCTION

Dynamic obstruction — one of the most common forms of intestinal obstruction in childhood, among all types of intestinal obstruction makes 8–10%.

Distinguish paralytic and spastic forms. In newborns and infants dynamic obstruction occurs as a result of functional disability of the digestive system against the background of a generic craniocerebral trauma, pneumonia, intestinal infections and sepsis, as well as after operations on abdominal organs and thoracic cavities. In older children, the dynamic obstruction occurs more frequently in the postoperative period. Phenomena of paralytic obstruction is supported by hypokalemia, due to the large loss of salts and fluids with vomitus, and insufficient intake of potassium in parenteral nutrition.

Clinical picture and diagnosis. Dynamic obstruction is characterized by repeated vomiting with greenery, growing bloating, constipation and gas and severe intoxication. As a result of the high standing of the diaphragm occurs difficulty in breathing. The abdomen is soft, peristalsis is not heard.

X-ray examination reveals multiple Kloiber cups, but their diameter is small and expanding is uniformly, while in mechanical obstruction are especially enlarged intestinal loops over the obstacle (Fig. 1.3.1).

Treatment. At dynamic obstruction we must first establish its cause. At the same time carry out the

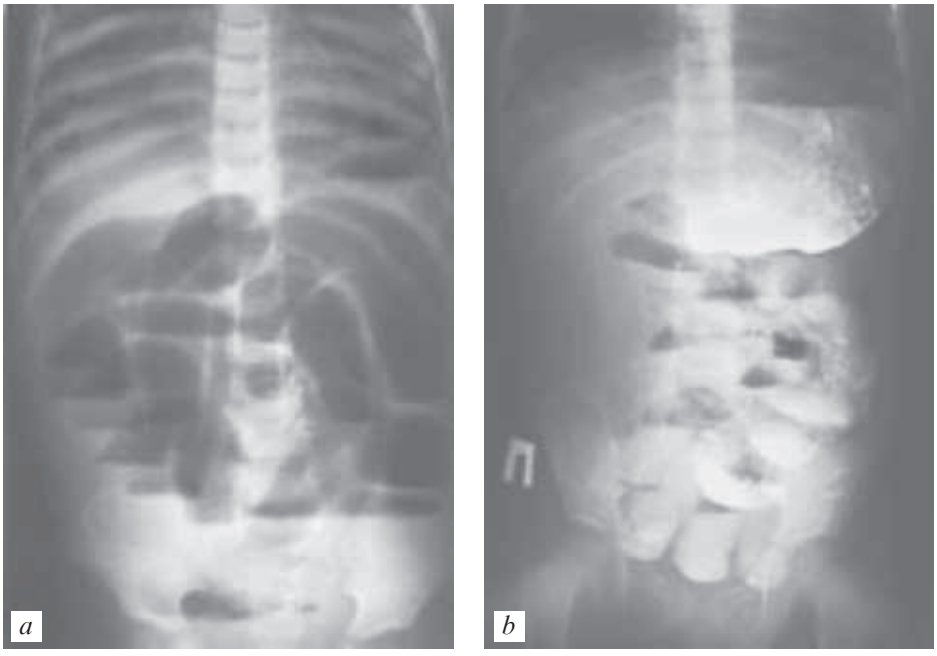


Fig. 1.3.1. Dynamic obstruction: multiple horizontal levels throughout the intestine *(a, b)*

struggle against intestinal paresis. Treatment regimen of enteroparesis includes:

1) Direct stimulation of the contractile activity of muscles of the gastrointestinal tract (cleaning, siphons and hypertonic enemas, intravenous drip of potassium and sodium chloride solutions with ECG monitoring, electric stimulation);

2) The blockade of reflexes arc that determine inhibition of motor activity of the intestine (the use of neostigmine, perirenal novocaine blockade);

3) discharge of the gastrointestinal tract (constant gavages, intestinal intubation).

Stages of treatment of dynamic obstruction are presented as follows:

1. Conservative therapy of the underlying disease.
2. Correction of homeostasis infringements.
3. Drainage and gastric lavage.
4. Subcutaneous or intramuscular administration of drugs that stimulate the motility of the intestines (Neostigmine, Cerucalum, etc.).
5. Regional sympathetic blockade by epidural anesthesia.
6. Intravenous 0.25% solution of novocaine, Cerucalum 0.5 mg/kg.
7. Physiotherapy (solljuks, UHF, microclyster with hypertonic sodium chloride solution).
8. Venting tube every 2 hours for a period of 10–15 min.

Conservative treatment is effective if it is carried out in this sequence for 12–24 hours.

ADHESIVE INTESTINAL OBSTRUCTION

This type of obstruction is in second place after the intussusception. Acute **adhesive intestinal obstruction** in children — one of the most severe and widespread diseases in abdominal surgery. It is always important to remember that if a child appeared with abdominal pain and anamnesis had any surgery on the abdominal organs, it is necessary first of all to think of acute adhesive intestinal obstruction. Most often adhesive intestinal obstruction occurs after surgery for acute appendicitis (80%), much less — after laparotomy at malformations of the intestine, intussusception and traumatic injuries of the abdominal cavity.

To conventional classification of acute adhesive intestinal obstruction (early and late with the delimitation of both on acute and subacute forms) it is advisable in late adhesive obstruction allocate hyperacute form of the disease.

Division of intestinal obstruction on severity of clinical manifestations largely determines the diagnostic and therapeutic management. Indications for surgical intervention are determined not so much by disease stage (early, late) as by its sharpness.

Clinical picture and diagnosis. Hyperacute form of adhesive intestinal obstruction is manifested by clinical picture which is similar to a state of shock. In the early period are marked toxemia, a rapid increase in exsicosis phenomena, there is a sharp, cramping abdominal pain, during which the patient sometimes falls apart, there are uncontrollable vomiting, pronounced increased peristalsis. In case of late arrival there is pronounced intoxication, is marked plentiful stagnant vomiting (fecal vomiting), "peritoneal" belly; intestinal peristalsis is sharply reduced or nonexistent. Such a pattern is most common in strangulation obstruction.

X-ray are determined clear horizontal levels (Kloiber cups), "arch" in a sharply stretched loops of the small intestine.

In acute and subacute forms of the disease symptoms are less pronounced, but the children also complain of paroxysmal pain in the abdomen; vomiting occurs, increases peristalsis. Clinical manifestations depend on the duration of the disease. In later stages the clinical picture is characterized by phenomena of exsicosis, repeated vomiting of stagnant character, moderate swelling and asymmetry of the abdomen, rarer, but reinforced peristaltic contractions. X-ray are identified multiple horizontal levels and gas bubbles in a moderately dilated loops of bowel. Traditional X-ray method for the diagnosis of acute adhesive intestinal obstruction requires an average of at least 8–9 hours and allows only confirming or excluding the fact of mechanical bowel obstruction. Diagnostic errors in these cases lead to untimely or futile surgery. Perspective and highly informative diagnostic method is laparoscopy.

Treatment. Patients with hyperacute form of the disease operate on an emergency basis after a short pre-operative preparation. Subacute or acute forms of treatment should start with a complex of conservative measures, including:

1) Gastric emptying (constant probe) with its periodic washing after 2–3 hours;

2) Ganglionic blockade;

3) Intravenous stimulation of intestine: 10% sodium chloride in 2 ml at 1 year of life; 0.05% solution of neostigmine 0.1 ml at 1 year of age;

4) A siphon enema after 30–40 min after stimulation.

At the same time X-ray control the passage of barium sulfate suspension of the intestines. These activities are carried out on the background of correction of homeostasis disorders, hemodynamic stabilization, and restoration of microcirculation. The use of this management in subacute and acute forms can cut adhesive intestinal obstruction by conservative measures in more than 50% of patients.

Surgery at the failure of conservative measures is to eliminate obstacles (colliotomy). In total adhesive process even in the acute stage may perform the full viscerosis and horizontal intestinoplications (Noble operation) with the help of medical adhesive without overlapping seams.

Causes of **obstructive ileus** in children are most often coprostasia, rarer — a tumor, ascarids.

COPROSTASIA — the bowel obstruction with dense feces. Occurs in children at any age. The reason can be in sluggish bowel function in frail children, and malformation of the anterior abdominal wall muscles, accompanied by atony of the abdominal organs. Often contribute to the development of Coprostasia anomalies and malformations of the co-

lon (megadolichocolon, Hirschsprung's disease, a congenital and cicatricial narrowing of rectum).

Clinical picture and diagnosis. In anamnesis there are always references to early constipation. Stool, as a rule, can be obtained only after the enema. Wrong diet and lack of child care lead to fecal debris, formation of fecal stones, some of which were thought to be an abdominal swelling.

At complete obstruction of the intestinal lumen the child's condition worsens, increases bloating, vomiting appears, symptoms of intoxication develop.

To conduct *differential diagnosis* between tumor and coprostasia of the bowel helps consistency of tumor formation, which has dough character at coprostasia. There is a positive symptom of "pits", which remains after pressure. In doubtful cases use a contrast X-ray examination, during which the contrast mixture flows around fecal stone from all sides and its shadow is clearly contoured.

Treatment. It is necessary to use repeated enemas with 1% sodium chloride solution at room temperature. If ordinary enemas do not help, do re-siphon enema to a fully diluted feces and restoration of patency of the intestine. Improper technique of siphon enemas can lead to severe complications, since during blurring of compacted fecal masses with warm isotonic sodium chloride solution begins fluid absorption and develops severe fecal intoxication, accompanied by a sharp deterioration in the state up to cerebral edema. In addition to siphon enemas, they appoint a diet rich in fiber, vegetable or mineral oil inside, light laxative means, course of electrical stimulation of the bowel with "Amplipuls" apparatus, total 15–20 sessions.

After the liquidation of Coprostasia make radiopaque study of intestinal tract. Helminthic obstruction in recent years practically does not occur. The reason of blockage in the described observations was a tangle of ascarids staying at the ileocecal valve. If conservative measures (siphon enema) fail to eliminate obstruction, surgery is performed. After surgery they prescribe deworming.

INTUSSUSCEPTION IN CHILDREN

Actuality of the problem

A. The most common in children (70%) is intussusception of intestines of all kinds of acquired intestinal obstruction.

B. Early diagnosis (up to 24 hours) of intussusception allows greater use of conservative treatment, which is much easier and less traumatic.

Intussusception — introduction of one part of the intestine into the lumen of another — the most common form of acquired intestinal obstruction. This option of intestinal obstruction occurs predominantly in infants (85–90%), most frequently in the period from 4 to 9 months. Boys become ill nearly 2 times more often than girls. In children older than 1 year old intussusception is rare and in most cases can be linked to the organic nature (diverticulum of ileac intestine, lymphoid tissue hyperplasia, polyp, malignant neoplasm, and others.).

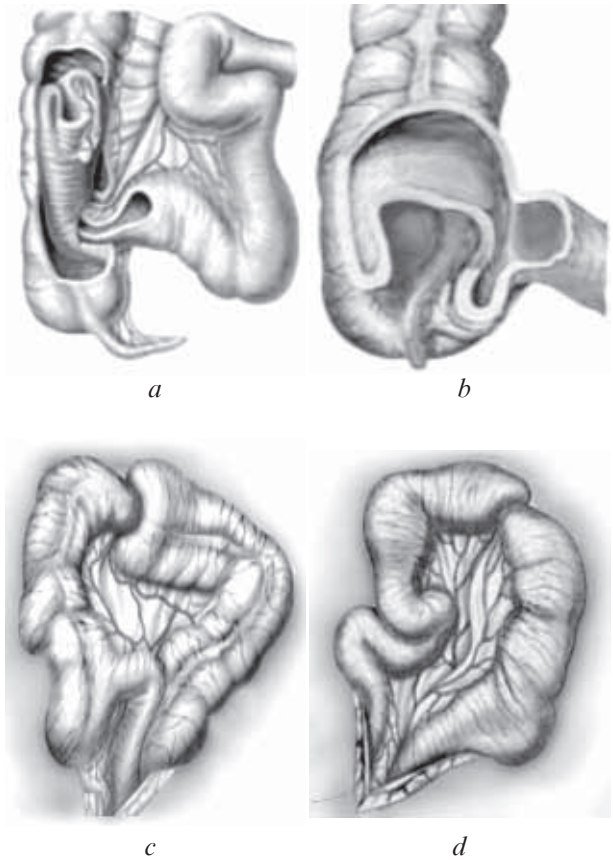
Matters correct rhythm of peristalsis disorder, consisting in violation of coordination of longitudinal and circular muscles reduction with a predominance of the contractile ability of the latter. To uncoordinated contraction of the muscular layers may lead changes in diet, using of complementary foods, inflammatory bowel disease, including enterovirus infection.

Intussusception refers to a mixed or combined type of mechanical obstruction, as it combines elements of strangulation (pinching of mesentery embedded intestine) and obturation (closing of intestinal lumen with invaginates. Depending on the location they distinguish *ileocecal* (over 95%), *small bowel* and *colonic* intussusception.

The term "ileocecal intussusception" is collective and refers to all types of intussusception in ileocecal angle. Of all the forms of intussusception in the area most often occurs *iliac colon*, when small intestine is implemented through ileocecal valve into ascending colon (Fig. 1.3.2).

The reasons for such topical and age frequencies are a number of background factors:

1. Immaturity of nervous system of the intestine (generating longitudinal and circular musculature discoordination of peristalsis by dysfunction).
2. Immaturity v. Bauhini.
3. Long mesentery, mobile cecum.



a *b* *c* *d*

Fig. 1.3.2. Types of intestinal intussusception: *a* — iliocolonic; *b* — blind colon; *c* — colonic; *d* — ileocolic

4. Significant difference in diameter of thick and thin intestine.

Less common is blind colon intussusception, in which the bottom of the cecum invaginates into the ascending part of the colon together with the appendix. Isolated introduction of the small intestine in a thin (small bowel intussusception) and thick into a thick (colonic intussusception) in total marks not more than in 2–3% of all patients with intussusception.

At intussusception they distinguish the outer tube (vagina) and internal (invaginate). The initial division of the intruded intestine is called the head of invaginate.

Direct factors (initiating) are following:

A. Functional:

1. Alimentary (improper introduction of complementary foods, violation of nutrition regimen);

2. Inflammatory diseases (enterocolitis, dysentery),

B. Organic:

1. Tumors of intestine;

2. Malformations of intestine (diverticulum, doubling).

Functional causes (95%) are triggering factors mainly at a critical age (in infants), organic (5%) — in children after one year of age.

Clinical picture and diagnosis. Clinical manifestations of intussusception depend on its type and duration. A result of introduction the bowel wall occurs infringement; during peristalsis increases length of the invaginate body, head remains unchanged. Promotion of invaginate due to peristalsis increasingly stretches and squeezes the blood vessels and nerves of the mesentery. Venous outflow is disturbed; there is stasis, edema, followed by diapedetic bleeding, swelling of the intestinal wall, inflammatory changes in it, and fibrin deposition between the inner and outer cylinders, their bonding. Invaginate migrates along the intestine, so that may fall out through the rectum — prolapsus invaginati. Circulatory disorders lead to necrosis of the intestine (especially in the head of invaginate — the area of largest infringement), which can lead to peritonitis. Thus, **Mondor** wrote, "... the disease is galloping and we, clinicians, should not trail behind it like a snail."

Typical symptoms are:

— Paroxysmal anxiety (the equivalent of abdominal pain);

— Once or twice vomiting

— Constipation and gases

— Dark bloody discharges from the rectum,

— Palpable "tumor" in the stomach.

In most cases the disease begins abruptly, among overall health and occurs usually in well-fed children.

Suddenly, the child begins to dramatically worry, yells, strains. "...A child is in a huge horror, his scream spreads all over the house. This is similar to the cries of parturient, but the woman in this case blushes and the child turns pale "(Narris). Mondor writes that "pinching — intraperitoneal drama, to which the child responds with such force that leads parents to horror". Older children are trying to take the knee-elbow position, which is a pathognomonic sign.

Anxiety attack ends as suddenly as it starts, but after a short period of time begins again. Typically, these vivid clinical manifestations are observed in children suffering from iliac colon implementation.

Attacks of pain in the beginning of the disease are frequent with small intervals of silence (3–5 minutes). This is due to the waves and progress of intestinal peristalsis of invaginates inside the intestine. In the light period, the child usually settles down for 5–10 minutes and then there is a new twinge. Shortly after the outbreak of the disease appears vomiting, which is reflexive in nature and related to the infringement of the mesentery, invaginated part of the colon. In the later stages of development of intussusception, occurrence of vomiting is caused by complete intestinal obstruction.

Temperature mostly remains normal. Only in the neglected forms of intussusception temperature increases. Stool can be normal within few hours due to emptying of distal part of intestine. After some time blood mixed with mucus discharges from the rectum instead of feces. This is due to deranged blood circulation in the invaginated section of intestine; often a symptom appears in no less than in 5–6 hours after the onset of the first attack of pain in the stomach.

In some cases, there is no discharge of blood throughout the entire period of the disease and is mainly observed in the form of blind-colon intussusception. This is due to the fact that these patients have almost no strangulation and obturation conditions prevail. So, the clinical manifestations of blind-colon and colonic intussusception forms are less pronounced: no child's anxiety, seizures of abdominal pain are less intense. With these forms of intussusception in the initial stages of the disease vomiting is observed in only 20–25% of patients.

Examination of the abdomen for suspected intussusception should be performed between bouts of pain. Unlike all other forms of intestinal obstruction abdominal distention is not observed at intussusception, especially within first 8–12 hours of the disease. This is due to the fact that intestinal gases for a while are penetrating into the lumen of invaginate. During this period, the stomach can be mild, affordable for deep palpation in all departments. In the right of the navel, often to the right hypochondrium, tumor formation of mild elastic consistency, not very painful at palpation, can be found. In 60% of cases find a "roller" (moderately agile and painful) along the large intestine, usually in the right upper quadrant. A **Dance** symptom is detected (desolation in the right iliac fossa). As blood is "a sign of salvation," appears within the first 3–6 hours only at 40% of children, according to Mondor "... no need to wait for it, you need to meet it." For this aim rectal examination or l enema (isotonic solution, 0,5 L) is conducted.

Terms of manifestation of these symptoms depend on the level of obstruction, duration of the disease (the lower the obstruction, the later the vomiting becomes multiple and the earlier broken stool

and gases are violated; over time flatulence appears).

Ombredane writes: "Recognition can be done with a precision of an algebraic equation: signs of obstruction + blood from the anus (the intestinal epistaxis) = infringement of the intestines. This is the basic equation. "The clinical picture of invagination is so typical that Mondor says that "...the diagnosis can be made by phone," i.e. the diagnosis can be made even from anamnesis.

The amount of blood is small, it may only be in the diapers. As a rule, the blood is mixed with mucus, has the character of a "currant jelly" (Mondor). Discharges does not contain bile, pus. Mondor wrote: "Blood on the diapers shows the doctor the diagnosis and therapy," and that is the importance of this feature, the same author points out: "The intestinal bleeding at intussusception — the main symptom. This is an ominous sign, threatening symptom, but it is informative, the most valuable, most significant, saving symptom."

Great importance for the early diagnosis of intussusception has X-ray examination. within the first 12 hours of the disease on survey X-ray can be seen a slight decrease of pneumatization of the intestine, at a later time — the signs of mechanical obstruction (variegated Kloiber cups) (Fig. 1.3.3, 1.3.4).

Contrast study — pneumoirrigography — is carried out in the early stages as follows. Into the rectum, under radiologic control with Richardson balloon, gently blow the air (40 mm Hg) and monitor its gradual spread in the colon to detect head of invaginate — find a block in the passage of air and shadow of invaginate in the form of "cockades", "sickle", "trident" etc.

Invaginate is clearly visible on the background of gas in the form of spherical shade with clear contours, often in the angle of hepatic colon.

Differential diagnosis. Intussusception is often mistaken for dysentery. However, dysentery disease is preceded by prodrome (weakness, loss of appetite, sometimes increasing the overall body temperature), in the period of developed clinical picture has a 2–3 day fever, rumbling in the stomach, stool in the form of "rectal spittle" (contains feces, pus, mucus) "raspberry jelly" (mucus with blood of light-red color because of haemorrhagia per diabrosin), whereas at invagination the disease is developed at the top of full health, the temperature within the first 12 hours increases and the stool is dark-red ("currant jelly" due haemorrhagia per diabrosin), doesn't contain feces and pus.

Treatment. Intussusception can be eliminated by both conservative and surgical treatment. Conservative smoothing is indicated at early admission of the child to the clinic (during first 12 hours of onset). During the diagnostic X-ray studies continue pumping air for unfolding of invaginate — pneumodesinvagination (under pressure of 120 mm Hg). Upon completion of the study introduce into the rectum gas outlet tube for removing excess gas from the colon. After straightening of invaginate the child usually calms down and falls asleep.

Clinical efficiency criteria of desinvagination are:

1. Disappearance of Dance symptoms.
2. The phenomenon of "cotton".

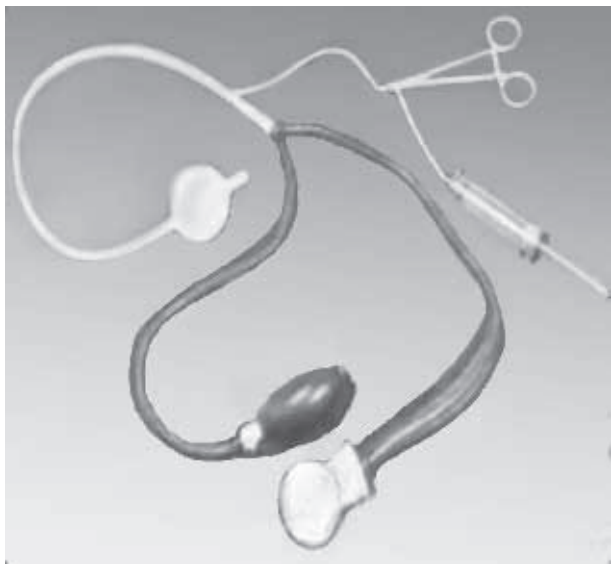


Fig. 1.3.3. Apparatus for pneumodesinvagination

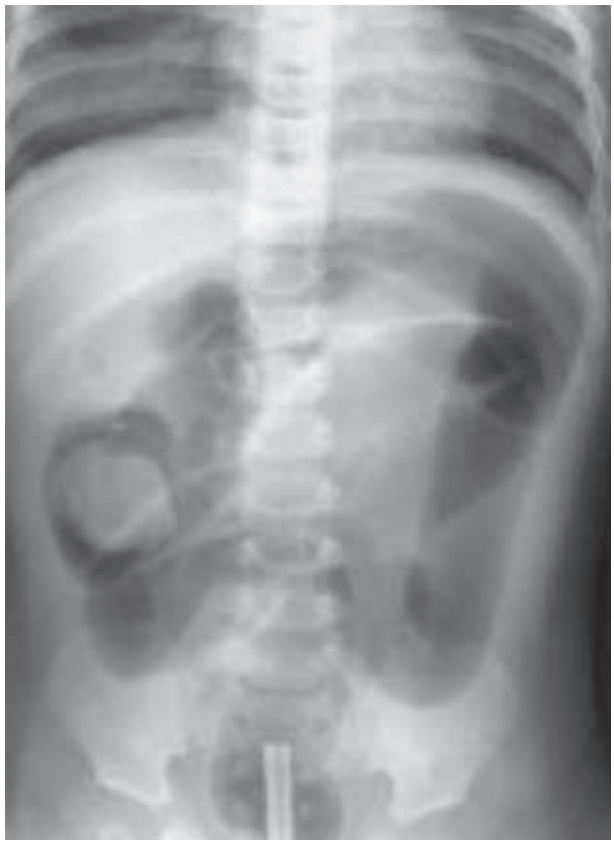


Fig. 1.3.4. "Block" symptom of air passage at pneumodesinvagination

3. Pressure drop on the blood pressure monitor.
4. Passing flatus.
5. Regurgitation with air or outlet through gavages. X-ray efficiency criterion of pneumodesinvagination — symptom of "honeycomb" ("small bubbles"), which is caused by the passage of air into the small intestine.

To be finally ensured in the full unfolding of invaginate, the child is required to be admitted to hospital for observation and study of the gastrointestinal tract dynamics with barium, which is given in the jelly and is monitored while it moves through the intestines.

Usually in the absence of small bowel intussusception contrast agent after 3–4 hours is found in the initial parts of the colon, and after a while barium suspension appears with stool. Method of conservative unfolding of intussusception is effective in an average of 65%.

Indications for conservative treatment are:
1. Blind-colon intussusception.
2. Colonic intussusception.
3. The first 24 hours of disease.
4. Age of children up to 1 year.

Contraindications to conservative treatment and at the same time the indications for surgery are:
1. Small bowel intussusception.
2. Colon-iliac intussusception (in these forms it is impossible to create a therapeutic pressure).
3. More than 24 hours of disease.
4. Age of children is over 1 year (are likely organic causes).
5. Recurrence of intussusception (according to the above reason).
6. Failure of conservative treatment.

In the case of admission later than 12 hours from onset of the disease increases dramatically the probability of circulatory disorders of the restrained intestine. Increased intraintestinal pressure in this case is dangerous, and at the dilatation of invaginate it is impossible to assess the viability of the affected parts of the colon. In such cases, as well as at the failure of conservative unfolding give indications for surgical treatment.

Surgical treatment consists of laparotomy and manual desinvagination which produce not by pulling embedded intestine, but by a cautious "milking" of invaginate captured by the whole hand or two fingers (Fig. 1.3.6).

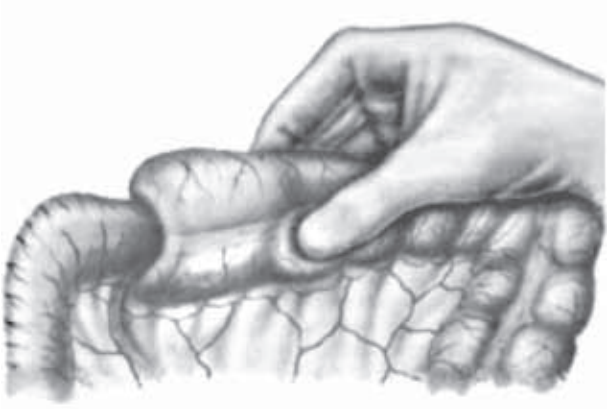


Fig. 1.3.6. Procedure of intraoperative milking back of invaginate

If you are unable to carry out desinvagination or necrosis of the bowel was found, produce resection within healthy tissue with anastomosis placement. Such management is logical and justified, but not perfect. Often pronounced infringement and necrosis of invaginate develop in a few hours from onset of the disease, and in a period exceeding 12 hours, desinvagination during surgery does not cause difficulties, gut is minimally changed.

Inclusion of laparoscopy to the complex of diagnostic and treatment measures at intussusception can significantly increase the percentage of patients cured conservatively. The aim of laparoscopy — visual control over the unfolding of invaginates and assesses the viability of intestine. The indications for this method are:

1) Failure of conservative treatment in the early stages of disease.

2) An attempt of conservative unfolding of invaginate at late arrival (excluding the complicated forms of the disease).

3) Elucidation of the causes of intussusception in children older than 1 year.

Laparoscopy visually determines the site of introduction of the ileum into the large intestine. The cecum and vermiform appendix are often also involved in invaginate. Instrumental palpation determines expressed induration of the colon at the site of introduction. After detection of invaginate conduct its desinvagination by introducing air into the colon through the anus under a pressure of 100–120 mm Hg. Desinvagination is considered effective at detecting of dome unfolding of the cecum and terminal ileum, filled with air. In the absence of rapid circulatory changes and space-occupying lesions (major cause of intussusception in children older than 1 year) is completed laparoscopy. Such management can significantly reduce the number of laparotomies at intussusception.

The prognosis depends on the timing of surgical hospitalization. At early detection and timely produced surgery deaths from intussusception are usually not observed.

Procedure of Pneumoirrigography

Under anesthesia into the rectum to a depth of 15–20 cm they inject rubber catheter № 9–10, coupled with Richardson balloon and mono vacuum meter; air is pumped to 40 mm Hg. They carry out an X-ray revealing colonic intussusception, continue pumping air to 100 mm Hg into the intestine, making pause, then again increase the pressure, so repeating until the complete unfolding which is controlled by X-ray. Signs of unfolding will be: filling with air of the entire colon, lack of invaginate, the appearance of air in the small intestine — a "honeycomb" symptom. After conservative unfolding of intussusception, patients start to drink after an hour, if there is no vomiting they appoint the usual age diet.

1.4. CLOSED TRAUMA OF CHEST ORGANS IN CHILDREN

CLOSED TRAUMA OF ABDOMINAL CAVITY ORGANS AND RETROPERITONEAL SPACE IN CHILDREN

A. CHEST AND THORACIC ORGANS TRAUMA

Specific objectives:

1. To master a list of the most common mechanisms of thoracic cavity organs injury during trauma.
2. To recognize major clinical displays of chest organs damage.
3. To differentiate the damage to chest organs.
4. To interpret subsidiary research methods (ultrasound, X-ray method, CT).
5. To demonstrate the examination of the chest: inspection, percussion, palpation, auscultation.
6. To identify the characteristics of the course of various injuries of the chest cavity.
7. To analyze the cause and effect ligaments of thoracic cavity organs injuries in different patients, substantiate and formulate preliminary clinical diagnosis.
8. To propose an algorithm for the doctor's actions during trauma of thoracic cavity organs injuries and management of patient's conduction.
9. To interpret general principles of treatment of thoracic cavity organs injuries, to determine the indications for surgical treatment.
10. Ti define of etiologic and pathogenetic factors, features of clinical motion, diagnosis and treatment of chest injuries.

Relevance of the problem

A. Damages of the thorax are fairly common; they make up to 3.4% of all childhood injuries.

B. Recognition of chest organs injuries — responsible and difficult diagnostic problem during determining the indications for urgent surgery.

Trauma is the leading cause of infant mortality at the age over 1 year. Among all injuries the most difficult are caused by road traffic accidents and falls from a great height. As a rule such severe damages are accompanied with trauma of several organs of different body systems (combined injuries). Often, on the background of the skeletal and cranial trauma, associated injuries of internal organs of thorax and abdomen appear later than during their isolated injury, or even are undetectable, which significantly increases mortality in this group of patients.

In Ukraine every year due to various injuries die about 2,000 children, and in each fourth case the cause of death are chest organs injuries, in every fifth case — the trauma of the abdomen. The leading place among causes of mortality takes craniocerebral injury. Damages of abdomen and thorax, as well as damage to other organs and systems are more common in boys aged 7–14 years, due to their active and often uncontrollable behavior.

Damage to internal organs is divided into closed and open. First group occurs much more frequently (in 85% of cases).

Trauma of thorax during severe injuries (falling from a great height, road accidents) is often combined. Therefore, in this group of patients respiratory symptoms may be both a display of craniocerebral injury, as well as lung damage.

Special features of chest injuries in children include a low incidence of fractures of ribs and sternum, due to elasticity of thorax; tensed pneumothorax in children is more dangerous because of the non-fixed mediastinum; the most frequent type of injury — a lung bruise; and once again it should be recalled that in 50% thorax trauma cases are combined. The most severe injuries of thorax that threaten the patient's life include open and tension pneumothorax, large hemothorax, cardiac tamponade, running thorax. These injuries require adequate help at a prehospital stage and within the first few minutes of being in the hospital, as even artificial lung ventilation in the presence of these injuries can be ineffective for serving respiration and blood oxygen saturation.

Chest trauma is divided into open and closed, with or without damage to internal organs. The most common trauma of thorax is bruised lung, in which there is bleeding into the lung tissue with the formation of intrapulmonary hematomas and areas of atelectasis. Patients complain of pain during breathing, shortness of breath, and cough with bloody sputum. During objective examination are determined the lag of one half of thorax in breathing, areas of deadened sound and reduced breath, the area of which depends on the amount of damaged lung parenchyma. Survey X-ray identifyes areas of atelectasis and speckled shades of pulmonary hemorrhages. As a rule, lung contusion is complicated by the development of traumatic pneumonia and requires appropriate treatment: antibacterial, mucolytic, hemostatic therapies, adequate pain relief, elevated position, oxygen therapy, breathing exercises, physical therapy (Fig. 1.4.1).

More severe injury — lungs ruptures with formation of hemothorax, pneumothorax, or hemopneumothorax. It should be noted that the cause of bleeding in the pleural cavity, besides of lung rupture, can be damage to intercostal and other vessels, which is usually a consequence of ribs fractures. Formation of hemothorax or pneumothorax (especially valvular) is accompanied by a very severe condition of the patient. Dyspnea, cyanosis, and accessory muscles participation in breathing, swelling of the nose wings, weakened shallow breathing, and severe pain take place. Pneumothorax is often accompanied by the formation of mediastinal and subcutaneous emphysema.

During valvular (stressed) pneumothorax, syndrome of intrapleural tension causes displacement

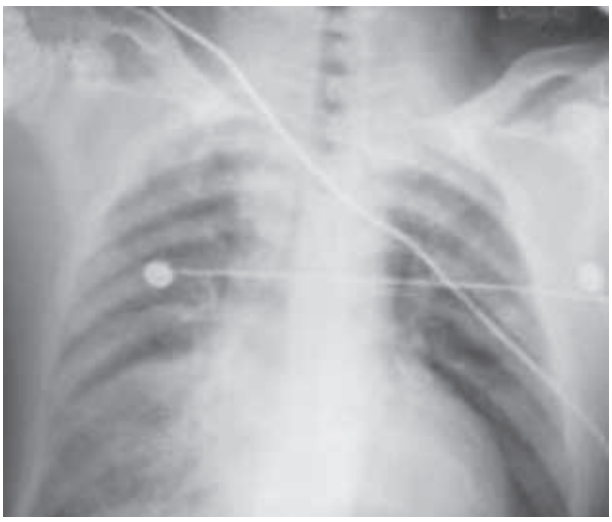


Fig. 1.4.1. X-ray of lung injury

and compression of the mediastinal organs, impaired return of blood to the right heart sections, which is an immediate threat to the life of a patient and requires immediate assistance (puncture of the pleural cavity). X-ray during break of lung determines its collapse, the presence of free air in the pleural cavity during pneumothorax or its extensive blackout during hemothorax. The leading sign of intrapleural tension is the displacement of mediastinum in a healthy side (Fig. 1.4.2–1.4.5).

Treatment management during lung rupture is determined by the nature of injury, the amount of bleeding, condition of the patient, the presence of associated injuries. If the patient is stable, the local treatment may be limited by a puncture or drainage of the pleural cavity. If punctured blood from the pleural cavity doesn't clot — the bleeding stopped, if clots — bleeding continues (Gregoire sample). Puncture of the pleural cavity during hemothorax is made in VI–

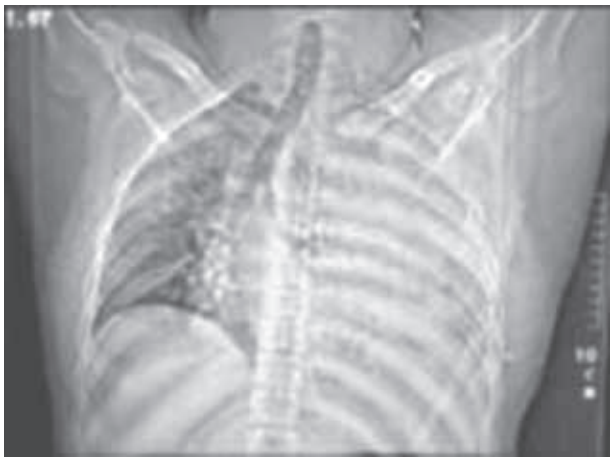


Fig. 1.4.2. X-ray of hemothorax

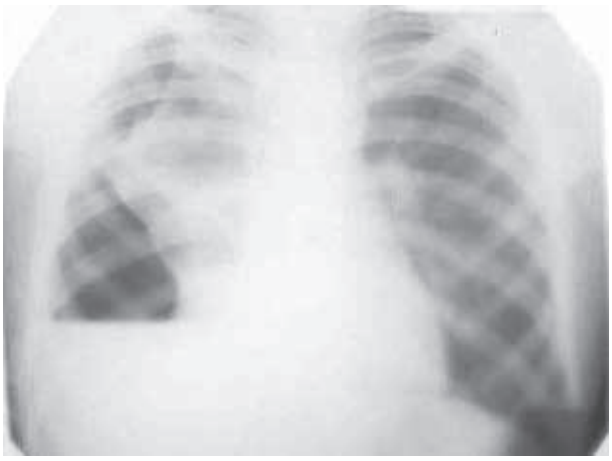


Fig. 1.4.4. X-ray of hemopneumothorax

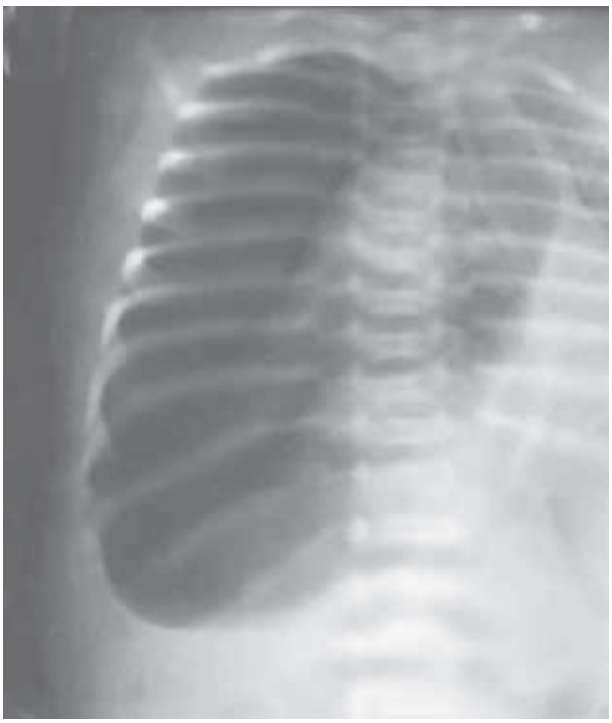


Fig. 1.4.3. X-ray of pneumothorax

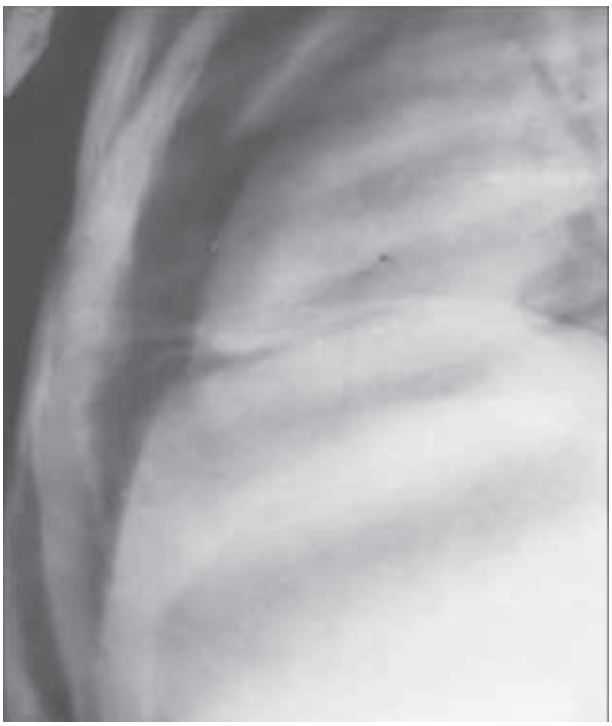


Fig. 1.4.5. X-ray of pneumomediastinum

VII intercostal space at the middle or posterior axillary line on the upper edge of the ribs, during pneumothorax — in II–III intercostal space at the middle clavicular line. If necessary, drainage of the pleural cavity (large hemothorax, the presence of bronchial fistula) drainage pipe is installed through the same points of the chest.

Intensive intrapleural bleeding, signs of damage to major vessels and large bronchi are indications for urgent surgery. The volume of surgery can range from wound closure of the lung to forehead — or pneumoectomy, but you should always strive to fulfillment of conservative surgery. An important point of surgery during trauma of the chest is the revision of other organs (heart, diaphragm, large vessels) that often accompany lung injury or are themselves the cause of hemothorax. Surgical treatment is carried out for 10–20% of patients with closed trauma of lungs. Much more frequently indications for surgical treatment occur in the presence of an open chest injury (stab and gunshot wounds, falling on sharp objects). Feature of operations in such cases is a revision of chest organs, which is not feasible in choosing access through the wound channel. Optimal access is lateral thoracotomy in the fourth–sixth intercostal space.

Severe consequence of closed chest trauma is damage to heart and large vessels. During aorta rupture (most often — in the region of the neck distal to the left subclavian artery), 80–90% of patients die before hospitalization. Blunt trauma of the heart leads to its injury or rupture. The most susceptible to rupture the right atrium and ventricle. Damage to the heart leads to the development of complications such as cardiac tamponade (accumulation of blood between the sheets of pericardium). Leading symptoms of cardiac tamponade are hypotension, which cannot be explained by the presence of other injuries, deafness of heart sounds, jugular venous distention. *Urgent care* consists in puncture of pericardium and blood aspiration to restore adequate filling of the cavities of the heart with blood. Then, an urgent surgery is performed, the purpose of which is the closure of the damaged areas of the heart (access — median sternotomy or left thoracotomy, depending on the presence of associated injuries).

Dull chest trauma can lead to heart injury, which is accompanied by reduced blood flow through the heart muscle and its ischemia. Corresponding changes are detected at electrocardiography, may be determined arrhythmia. Patients with cardiac injury need constant ECG monitoring to restore normal mobility of the heart muscle (Fig. 1.4.6, 1.4.7).

Isolated fractures of the ribs without damage to intercostal vessels and lung tissue do not cause significant respiratory disorders, are diagnosed on examination, palpation of the chest and on the basis of X-ray examination. More severe rib injury is balloting chest, at which there are at least three fractured ribs on two lines. Such compound fracture is possible under the influence of a considerable force, so this injury is often accompanied by damage to the lungs and heart. The main *clinical manifestation* of balloting chest is paradoxical movement of the damaged area of the chest during breathing. During large area of damage its retraction on inspiration can significantly impede breathing. X-ray shows the number and location of fractures. *Treatment consists* of adequate pain relief, detection and treatment of lungs lesions, heart and other injuries. If adequate breathing is not restored, conduct the pulmonary ventilatioon. Surgical treatment of balloting chest is indicated at significant deformation of the chest and in cases when patient will have thoracotomy for another injury.

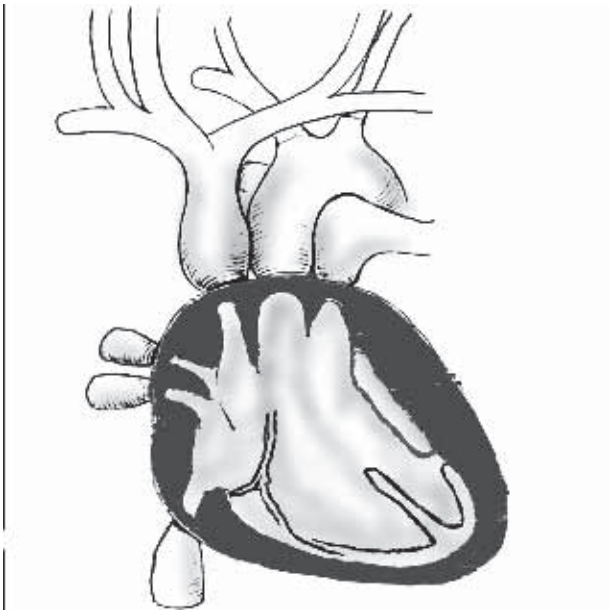


Fig. 1.4.6. Cardiac tamponade

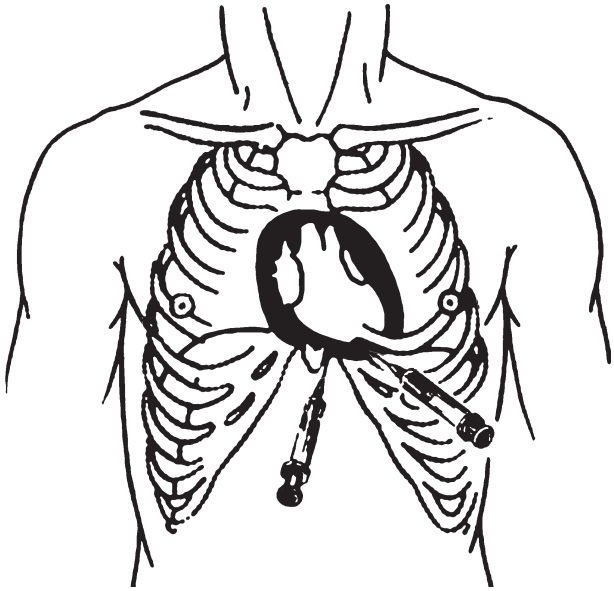


Fig. 1.4.7. Puncture at tamponade

DAMAGE TO ESOPHAGUS

Determination of etiologic and pathogenetic factors, clinical features, diagnosis and treatment.

Relevance of the problem.

A. in children damage to esophagus is relatively rare. But the severe course and complexity of treatment of esophagus injuries form a problem that needs urgent resolution because of the threat of severe complications.

B. Recognition of esophageal injury — responsible and difficult diagnostic problem that manifests in progressive subcutaneous emphysema and inflammation of the mediastinum.

Esophageal damage in only 1% of cases are the consequence of a closed chest injury. Much more often they are due to open or iatrogenic injury (bougienage of esophagus, esophagogastroscopy, removal of foreign bodies of the esophagus).

The diagnosis of esophagus rupture is established on the basis of *complaints* of a strong chest pain that is worse when swallowing, fever, general condition of the patient deteriorates, occurs dyspnea and cough, purulent mediastinitis is rapidly developing. *On examination* is often detected subcutaneous emphysema. The diagnosis is confirmed during the X-ray. The presence of mediastinal emphysema and contrast flow beyond the esophagus. Esophagoscopy should not be conducted because of the additional trauma. The best results of rupture treatment are achieved during surgical *treatment* within the first 24 hours after perforation to the development of purulent mediastinitis. The operation involves suturing of perforation hole and drainage of the mediastinum (upper mediastinum is drained by Razumovsky through neck mediastinotomy, mid and low back sections — by Nasilov through outside pleural access). If necessary gastrostomy is superimposed.

B. CLOSED ABDOMINAL INJURY

Specific objectives:

1. To master a list of the most common mechanisms of abdominal organs injury during trauma.
2. To recognize the major clinical manifestations of hollow and parenchymal organs injuries.
3. To differentiate damage to hollow and parenchymal organs.
4. To interpret support research methods (ultrasound, X-ray examination, CT, paracentesis, laparoscopy, radionuclide scintigraphy.
5. To demonstrate the examination of the abdomen: inspection, percussion, palpation, auscultation, digital rectal examination.
6. To identify the features of the course of damage to various organs of the abdominal cavity.
7. To analyze the cause and effect ligaments of abdominal organs injury in different patients, justify and formulate preliminary clinical diagnosis.
8. To propose an algorithm for the doctor's actions during intraperitoneal bleeding and management of the patient's treatment.
9. To interpret the general principles of treatment of the abdomen injuries, to determine the indications for surgical treatment.

DAMAGE TO PARENCHYMAL ORGANS OF ABDOMINAL CAVITY

Relevance of the problem

A) Closed injuries of parenchymal organs of the abdomen, according to different authors, are observed in 10–16% of all injuries.

B) Among traumatic injuries of abdominal organs in half of the cases spleen is injured.

C) Recognition of injuries of abdominal cavity organs — responsible and difficult diagnostic problem.

Trauma of the abdominal cavity organs, as well as chest trauma, is divided into **open and closed.**

Closed injury of abdominal organs to two clinical syndromes: abdominal bleeding (in case of parenchymal organs damage) and peritonitis (at rupture of hollow organs).

Among abdominal cavity organs in children, **splenic traumas** take place most frequently (25–27% of all injuries). They distinguish the following types of splenic injuries:

— Superficial anguishes of capsules
— Subcapsular hematomas
— Capsule and parenchyma ruptures
— Crushing of the spleen, splenic avulsion from the vascular pedicle

The main and permanent sign of spleen injury is constant nagging pain in the left upper quadrant and epigastric region. Younger children can not accurately localize the pain and often complain of spilled pain. Older children are beginning to complain of spilled pain some time after the injury, which is associated with the spread of blood through the abdominal cavity. However, the greatest intensity of pain remains in the projection of the spleen. The pain usually increases at deep breathing and irradiates in left shoulder and shoulder blade. Vomiting and rapid breathing are rare. Rib fractures at splenic injuries in children almost never occur. The most typical display of internal bleeding in children are pallor of the skin and mucous membranes. Thus heart rate and blood pressure can maintain within the age norm for a few hours after injury. Sometimes children occupy forced position: lying on the left side with knees pressed against stomach. Attempt to change the situation leads to increased pain that forces the child to return to his former position — a symptom of "Roly-Poly". On abdomen examination is marked lagging behind of the left half during breathing process, are sometimes visible abrasions in the projection of the spleen. Soreness and muscle tension are localized in the left upper quadrant, rarely spread throughout the abdomen. In some cases there are inconsistencies of severe pain in the abdomen and a slight muscle strain — Kulenkampf symptom. In the abdominal cavity in the near future after the injury free fluid appears, which is displayed by toning in sloping areas of the abdomen. During rectal examination can determine the accumulation of blood in

the pelvis. Due to the fact that into the abdominal cavity is mainly poured blood which has been deposited in the spleen, during the first hours after the trauma red blood parameters (erythrocytes, hemoglobin) vary slightly. More typical for splenic lesions is growth of the number of white blood cells, especially within the first hours after injury.

In the *diagnosis* of splenic lesions help radiology, ultrasound, computed tomography, laparocentesis or micro laparotomy with using the method of "groping catheter", laparoscopy. During stable condition of a child ultrasound and CT can detect location, size and depth of a ruptured spleen, the presence of subcapsular hematoma or blood in the abdominal cavity. Such techniques as angiography and radioisotope study can evaluate the nature of splentic injury, but they are not widespread in medical practice. In cases when the child's condition does not allow computed tomography and ultrasound, which is most often associated with the presence of severe concomitant injury, unconsciousness, to confirm the diagnosis of a ruptured spleen use a technique of "groping catheter." This research involves the input of catheter into the abdominal cavity through a small midline cut below the navel, with help of which is determined the presence of blood in the peritoneal cavity (Fig. 1.4.8).

Currently up to 80–90% of children with splenic injuries are treated conservatively in a way that consists in strict bed rest for 10–14 days, use of cold on injured area, and the appointment of infusion and hemostatic therapy. Such management is possible due to the structural features of the spleen in children. In most cases bleeding from parenchyma of this organ stops spontaneously. Conducting conservative treatment is allowed only in specialized medical institutions, where it is possible to perform surgery at any time of the observation.

Surgery is indicated in cases when, despite the ongoing infusion and hemostatic therapy, the condition of the affected remains unstable (growing tachycardia — more than 130 beats per minute, hypotension — systolic blood pressure less than 80 mm Hg, decreased diuresis, hematocrit, anemia increases). One of the indicators of unstable hemodynamic and continuous bleeding is the need for blood components transfusion in the amount of 30–40 ml/kg. Surgical management is determined by the degree of damage to the spleen, but, if possible, should be performed organ preserving operations, remembering of the high risk of more severe course of any infections and septic diseases in children after splenectomy.

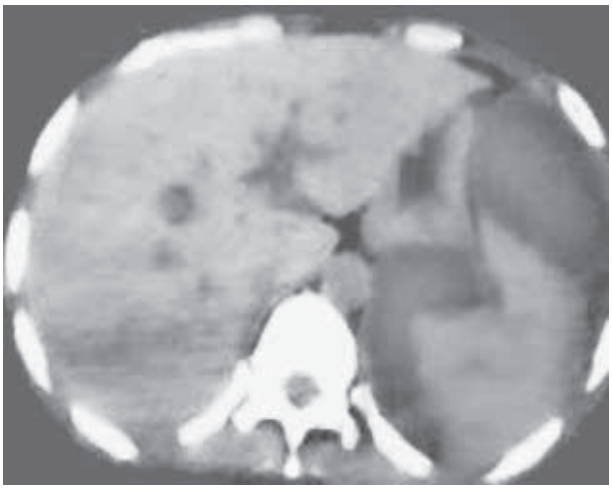


Fig. 1.4.8. Damage to the spleen (CT)

Two moment splenic rupture occurs during damaged parenchyma without damaging the capsule, resulting in a subcapsular hematoma, which, during continued bleeding, gradually increases in size, and in a few hours or days after the injury can spontaneously break the capsule of the spleen. In this case, the child's condition worsens and develops a picture of bleeding to the abdominal cavity. During identifying the subcapsular hematoma of the spleen they indicate hospitalization, conservative treatment and are indicated.

Liver damage takes the second place among injuries of the abdomen (15–17%), but these injuries are the cause of half deaths, caused by the trauma of the abdomen. More than half of the cases of liver injury are accompanied by damage to the spleen, injury of other organs and body systems.

Liver damage can be divided into three groups:

— Subcapsular hematomas,

— Liver ruptures with damage to the capsule,

— Central liver ruptures (central hematomas) when a cavity forms inside of parenchyma, filled with blood and bile.

In most cases, the child's *condition* after the injury is regarded as severe because of the development of shock. The main symptom of liver damage is a constant pain, usually in the right upper quadrant or right half of the abdomen. Localization of damage also affects the distribution of pain. During damage to the liver dome pain is localized in the lower right part of the chest, during damage to the back surface of the liver and its separation from coronary ligaments pain extends to the lumbar region, during damage to the lower surface of the liver pain is determined closer to the navel.

The pain may irradiate to the right shoulder girdle and the shoulder blade, during propagation of blood on the right side channel, pain will be determined in the right iliac region, above the vagina. Vomiting reflex occurs often after an injury. Sometimes there is vomiting of coffee ground color — a consequence of hemobilia (breakthrough of central hematoma in the bile ducts). Pallor, tachypnea, tachycardia, decreased blood pressure take place. Tension of the muscles of abdominal wall, the positive Shchetkin–Blumberg symptom, free fluid in the abdominal cavity, later appearing blunting in sloping areas of the abdomen, flatulence are observed in most patients. Symptoms of peritoneal irritation are particularly pronounced and crisp in case of damage to exterior bile ducts and the development of bile peritonitis.

"The navel" symptom — an acute pain during pressing on the stomach that occurs as a result of tension of the liver round ligament. Quite quickly Anemia develops. Liver damage is diagnosed like spleen injury: computed tomography, ultrasound, technique of "groping catheter", angiography. Ra-

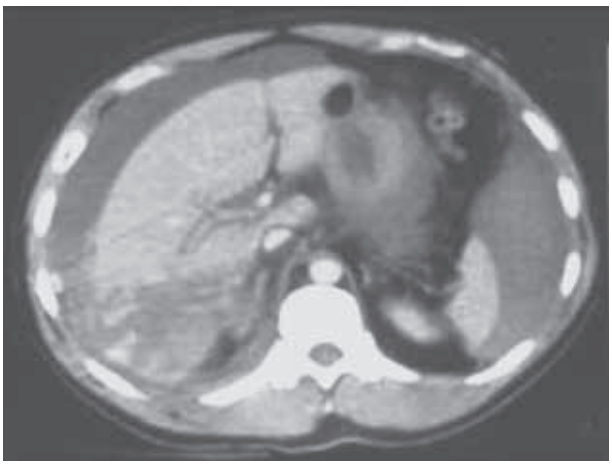


Fig. 1.4.9. Liver damage (CT)

dioisotope study allows revealing trauma of the bile ducts. A CT scan should be carried out with the use of a contrast agent that allows identifying areas of blood accumulation, the presence of active intra-abdominal bleeding (Fig. 1.4.9).

The presence of active intra-abdominal bleeding and hemodynamic instability *are indications for surgical treatment*, which is carried out in 20–30% of all liver injuries in children. In most cases, as during spleen injury, bleeding stops spontaneously and hemodynamic parameters are stabilized, allowing conservative treatment. One of the methods of arrest of arterial bleeding from the liver parenchyma, which is actively being developed at present, is angiographic embolization of damaged artery.

Damage to pancreas at children are rare. Most often they are marked with concomitant injury of the abdominal cavity and retroperitoneal space. Damages of pancreas are the result of forced hits in the epigastric region (for children is most typical handlebar hit from falling), or as a result of prolonged compression of the upper part of the abdomen. Immediately after the injury children *suffer from* severe pain in the epigastric region, collapse and shock develop. Pain may irradiate to the lumbar region, in the left cost vertebral angle, at significant damage the pain has encircling character.

Vomiting occurs, which during the development of traumatic pancreatitis becomes uncontrollable. Pulse is frequent of weak filling; blood pressure in most cases does not change. On examination of abdomen one can detect traces of trauma in the epigastric region (abrasions, bruising), the stomach lags behind in the act of breathing; there is tenderness over the navel and in the left, moderate tension of the abdominal muscles, especially marked in the epigastria. Shchetkin–Blumberg symptom is weakly positive. In the dynamic observation symptoms of peritoneal irritation increase. During two moment damage to pancreas in first hours after the trauma the child is relatively stable, moderate epigastric pain, single vomiting. Then, suddenly, the child's condition worsens, strong abdominal pain, intractable vomiting, clinical shock develops.

Great importance in the diagnosis of pancreatic injuries have repeated determination of amylase level in the blood and urine within the first hours after injury lowering amylase level may occur, which is associated with vascular spasm. Then, the more time passes since the injury, the more pronounced is increase in the activity of the enzyme. Lipase activity in the blood increases only on 2–3 days after injury. Among additional research methods for identifying nature of the damage to the pancreas computed tomography and ultrasonography can be used.

Treatment: at suspecting pancreatic injury a child gets a parenteral nutrition, is assigned strict bed rest and therapy course, aimed to prevent the development of pancreatitis, every 4–6 hours the level of amylase in the blood and urine is determined. In the absence of effect from conservative therapy during the day, at revealing of extensive damage to the gland, during break of excretory duct cancer they indicate surgery. Technique of surgery is defined by character of the identified lesions and can range from a cavity drainage of omental bags to resection of the distal part of the gland or creating pancreatic jejuno anastomosis. Postoperative complications (fistula, pseudocyst of the pancreas, abdominal abscess) are observed in 10–25% of cases.

DAMAGE TO HOLLOW ORGANS OF THE ABDOMINAL CAVITY

The urgency of the problem

A) Most injures, because of anatomical and physiological features, small intestine, and namely its proximal parts near Treyts ligament and the distal ileum.

B) Isolated penetrating fractures of the stomach and colon are very rare.

C) The most common mechanisms of injury:

1) Crushing (between the anterior abdominal wall and the spine) — crash-injury;

2) Rupture of the mesentery or mesenteric edge of ulcer;

3) Rupture of overstretched intestinal loop.

D) Closed injuries can be penetrating and non-penetrating.

E) During penetrating ruptures develops clinical peritonitis.

Damage to hollow organs of the abdomen (stomach and intestines) is most often caused by open penetrating wounds but at 2–6% of patients with blunt abdominal trauma occur these particular organs. *The clinical picture* of stomach rupture is characterized by severe shock and the rapid development of peritonitis. There are acute pains, especially pronounced in the epigastric region, frequent urge to vomit, vomit masses are meager, may contain an admixture of blood. The abdomen is not involved in the act of breathing, is determined muscle tension and positive Shchetkin–Blumberg symptom, during percussion is often determined the disappearance of liver dullness borders. X-ray reveals free gas (Fig. 1.4.10).

Dull abdominal trauma may be accompanied by the formation of hematoma in the wall of the stom-

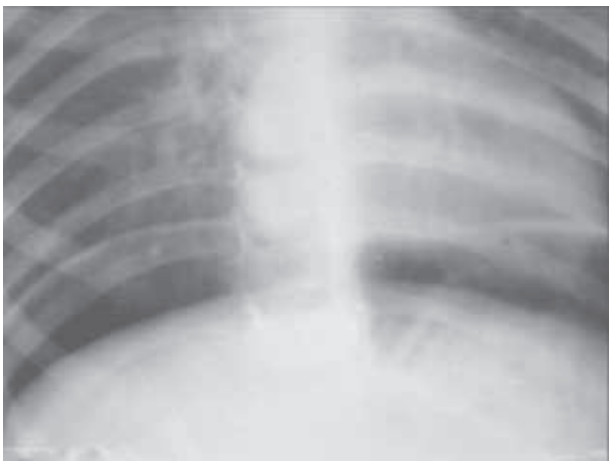


Fig. 1.4.10. Pneumoperitoneum

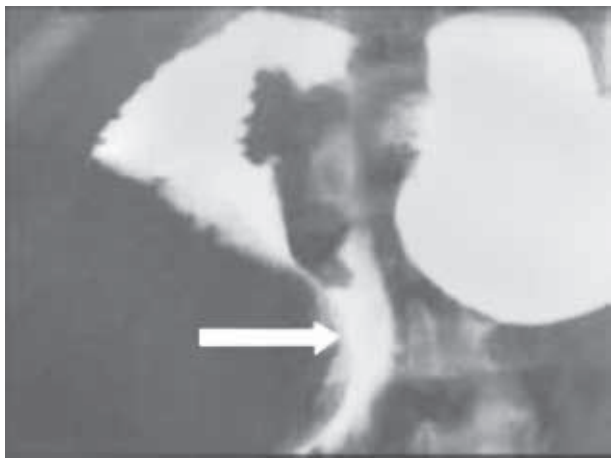


Fig. 1.4.11. Damage to the duodenal ulcer

ach, tears of mucous or serous-muscle membrane of the stomach. Condition of the child after the injury may be severe, but the antishock therapy quickly leads to improved condition of the patient, the clinical picture of peritonitis develops. In case of serous-muscular layer damage may develop signs of intra-abdominal bleeding.

During stomach ruptures they indicate *urgent laparotomy*, found gastric rupture or tear of serous-muscular layer is sutured with double-row seam.

Intestinal damages are divided into bruises, accompanied by the formation of hematoma in the wall of the colon or wall ruptures; complete ruptures of the bowel wall with the arrival of its contents into the abdominal cavity. A rare type of bowel injury is its separation from the mesentery. As during closed and in open abdominal injuries complete rupture of the intestine is accompanied by development of the *clinical picture* of peritonitis. The child complains on pain in the abdomen, which are almost always accompanied by vomiting. The general condition of the child is severe. He is pale, non dynamic, facial features are sharpened, the pulse is frequent, weak filling, and body temperature is increased. During abdominal palpation acute pain over the site of damage is defined, with the development of peritonitis, pain spreads throughout the abdomen. Spilled muscle tension and positive Shchetkin–Blumberg symptom are identified. On the survey X-ray of the abdomen, which must be mounted in a vertical position of the patient, or in later position in the supine position, free gas in the abdominal cavity is defined. However, the lack of this feature does not exclude hollow organ injury.

In some cases, perforation of the intestine may be covert or small sized, which leads to slower development of peritonitis. In case of damage to the intestine within the first hours there is a moderate leukocytosis with a shift of leukocyte formula to the left. With the development of peritonitis, leukocytosis and a shift to the left are growing.

Injuries of the intestinal wall can be asymptomatic, but the presence of a hematoma in the intestinal wall can lead to necrosis of the wall, its perforation and development of peritonitis.

Features of clinical course are seen during **damages to the retroperitonial part of the duodenum.** Within the first hours after the injury only manifestations of the damage can be complaints on mild epigastric pain associated with the formation of a retroperitoneal hematoma. But over time, the intensity of the pain increases, they are located slightly above and to the right of the navel. Deterioration of the child, the occurrence of vomiting, fever up to 38–39C°, the growth of intoxication is associated with the formation of a retroperitoneal phlegmon. In the epigastric region is defined muscle tension and a acute pain during palpation. Pain can be defined and in the lumbar region on the right as well. At suspicion of injury of retroperitoneal part of the duodenum they indicate X-ray with water-soluble contrast agent — in penetrating ruptures contrast material goes beyond the duodenum (Fig. 1.4.11).

Causes of rupture of the rectum in children may include: falling with crotch area on a sharp object, fractures of the pelvis, damage at a rectal temperature measurement, or entering of the vapor tube. Intraperitoneal rupture of the rectum is accompanied by severe pain in the abdomen and perineum, vomiting, fever, muscle tension and positive symptoms of peritoneal irritation in the lower abdomen. Bleeding from the rectum can be marked.

The established diagnosis of bowel injury is an indication for urgent surgical treatment after a brief anti-shock therapy.

C. TRAUMATIC INJURIES OF THE URINARY SYSTEM

Specific objectives:

1. Identify anatomical structures, the damage to which is most probable.
2. Highlight and group clinical symptoms that characterize soft tissue injury, symptoms that indicate bleeding and features which characterize the gap of the urinary system organs.
3. Illustrate clinical features on an example of the patient with traumatic injury and formulate preliminary diagnosis, determine the condition of the patient, presence of shock.

4. Differentiate damages, depending on the anatomical structures of the urinary and reproductive systems and type of injury: bruise, rupture, concomitant injury.

5. Make a plan of inspection and interpret supporting research methods (ultrasound, X-ray method, urography, urethrography, CT, etc.), laboratory and biochemical analyzes, hemodynamic parameters (heart rate, blood pressure, Hb, Ht, BCC, CVP).

6. Demonstrate the possibility of bladder catheterization and suprapubic bladder puncture.

7. Identify the characteristics of the course of various injuries of sexual and urinary systems.

8. Analyze the cause and effect of damage and highlight the main complications: bleeding, uroplania.

9. Propose an algorithm for the doctor's actions during traumatic injuries and determine the management for the individual patient.

10. Render an urgent help at the major injuries of the urinary system in children and perform the necessary medical procedures: measurement of blood pressure, blood type and Rh-factor, bladder catheterization or suprapubic puncture, procaine blockade.

11. Define the indications for surgical intervention, depending on the type of injury: suturing of rupture, cystostomy, nephrostomy, drainage of around cystic and perinephric space, para uretheral hematoma, hemi- or nephrectomy, perineal and scrototomy.

Kidney trauma occurs in children as often as splenic injury (25–27% of all injuries that occur as a result of closed injuries of the abdomen and retroperitoneum). Children, due to the nature of the structure and location of the kidneys are injured more frequently than adults. The presence of renal anomalies in children also predisposes to injury. The most important is hydronephrosis. Kidney trauma can be divided into two groups: mild (kidney trauma, subcapsular hematoma, rupture of the renal parenchyma without damaging the renal pelvis system) and severe (deep ruptures with damage to the excretory system, damages of the pedicle, crush injury of kidney).

The main clinical feature of kidney injury is hematuria, which is observed in 95% of patients. This symptom can be absent in the case of damage to kidney pedicle or pyelitis urethral segment. It should be noted that such symptom as microscopic hematuria is very common in trauma of the abdomen, the abdominal wall injury. For severe damage to kidney is more typical macrohaematuria, the intensity of which increases, whereas with light injuries hematuria quickly disappears. Also, almost all of the patients have pain in the abdomen or lumbar region; however, we must remember that children poorly localize pain that may be a cause of diagnostic error. The appearance of swelling in the lumbar region indicates the formation of retroperitoneal hematoma or urohematoma. Severe kidney injuries and the presence of concomitant trauma (damage to the liver or spleen) may result in symptoms of shock (Fig. 1.4.12).

In all cases of suspected kidney injury (increasing hematuria, the presence of urohematoma, severe trauma of the stomach or the lower parts of the chest, fractures of the lower ribs) it is necessary to use additional methods. The most objective are contrast-enhanced computed tomography and excretory urography. The timely release of contrast by the kidneys and the lack of access of contrast outside the kidney allow to exclude a severe injury and to appoint conservative therapy.

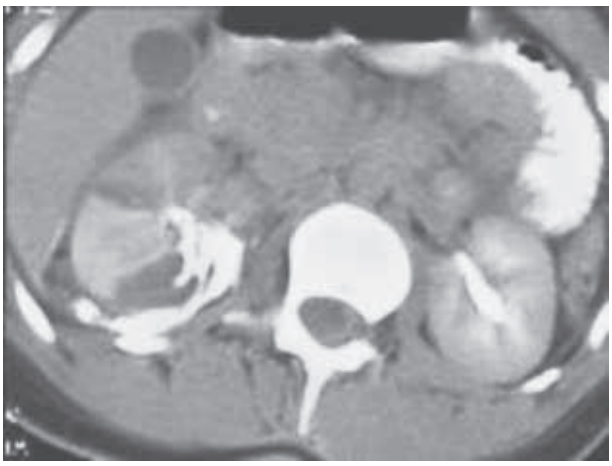


Fig. 1.4.12. Damage to the kidney (CT)

When indicating surgical treatment one consider how stable the patient's condition is and the possibility of associated trauma. The instability of vital signs and growth of urohematoma are the indications for surgery. An important advantage of the excretory urography is an opportunity to evaluate the presence and function of the second kidney, so, cases of removal of a single kidney at its injury are described. An effective method of determining the extent of damage to the kidney and the indications for surgical treatment is angiography. Special value this method has with suspected kidney vascular injury. Thrombosis of the renal artery, the active bleeding, kidney devascularization of segments are also indications for surgery. Large renal vessels injury causes the development of 90% of long-term complications — arteriovenous fistula, encapsulated hematoma, hypertension.

An important feature of the surgical treatment of renal damage is an effective vascular control that allows you to stop bleeding during surgery. Ligatures under vessels of a kidney must be supplied outside the renal fascia, near the site of their connection to the aorta and inferior vena cava. This manipulation allows you to reduce the number of nephrectomy three times during surgical treatment of kidney trauma.

Trauma of the bladder are caused by the child falling on his stomach at filled bladder or at fracture of the pelvis. Depending on the location of damage, complete ruptures of the bladder wall are divided into intra- and extraperitoneal. Extraperitoneal ruptures are identified 3–4 times more often. One of the types of injuries are only tears of the bladder mucosa, which appear in hematuria and pain in the suprapubic region.

Because the rupture of the bladder is often associated with fractures of the pelvis and damage to other organs, *the condition of the patients* is usually very severe. Injury of the bladder is characterized by such symptoms as pain in the lower abdomen, muscle ten-

sion of abdominal wall and symptoms of peritoneal irritation. The most important symptom of bladder wall damage is hematuria, and in 95% — macrohaematuria.

To confirm the diagnosis of injury of the bladder it is necessary to make a retrograde cystography or computer tomography with advanced bladder filling with contrast agent. Both of these studies allow us to accurately determine the nature and location of injury of the bladder toward streaks of contrast outside the bladder.

If there is no streaks of contrast, after research they remove contrast from the bladder through a catheter and take another shot. If this image determines accumulation of contrast in the bladder, with high probability, we can assume partial rupture of the bladder wall. The advantages of computed tomography are the possibility of a simultaneous examination of the kidneys and the abdominal cavity, which is of great importance, as in half of the cases bladder injury is combined.

The only contraindication to the installation of a urinary catheter and inserting of contrast into the bladder is **urethra trauma**. Its symptoms include discharge of blood from the meatus drop by drop (urethremorrhagia), lacunar and perineal hematoma, swelling of the scrotum, the inability to urinate. To confirm the diagnosis of urethral injury they indicate conducting an ascending urethrography (Fig. 1.4.13).

The presence of intraperitoneal rupture of the bladder is *an indication for surgical treatment*. After aspiration of urine and blood from the abdomen wall of the bladder is sutured by double row seam without locking mucosa into seams (the presence of ligatures in bladder lumen leads to the formation of concretions). The operation is completed by bladder drainage (through urinary catheter or with suprapubic cystostomy.) Boys should be avoided prolonged catheterization, as it is often complicated by the formation of strictures of the urethra. Extraperitoneal rupture of the bladder needs both methods: conservative treatment (installation of a urinary catheter at small gaps, which close by itself) and surgery (suturing of bladder wall with its catheter cystostomy + drainage around cystic space by Buyalsky).

At ruptures of urethra the best long-term results (no strictures of the channel) are achieved by applying the primary seam of urethra within the first 24 hours after injury. Previously they form suprapubic cystostomy which remains after surgery for adequate drainage of the bladder. If necessary, the operation is finished by draining the lacunar space and the pelvic fiber on Buyalsky. If it is impossible to impose a primary seam of urethra, rupture edges are sewed to the skin, cystostomy is formed. Later a secondary urethra plastic is made.

If there is a severe injury, regardless of the location and number of injuries, crucial importance in saving the life of the patient has adequate first aid.

Its basic principles:

— Condition stabilization of patient by the ABC system;

— Detection of life-threatening injuries and providing the necessary help;

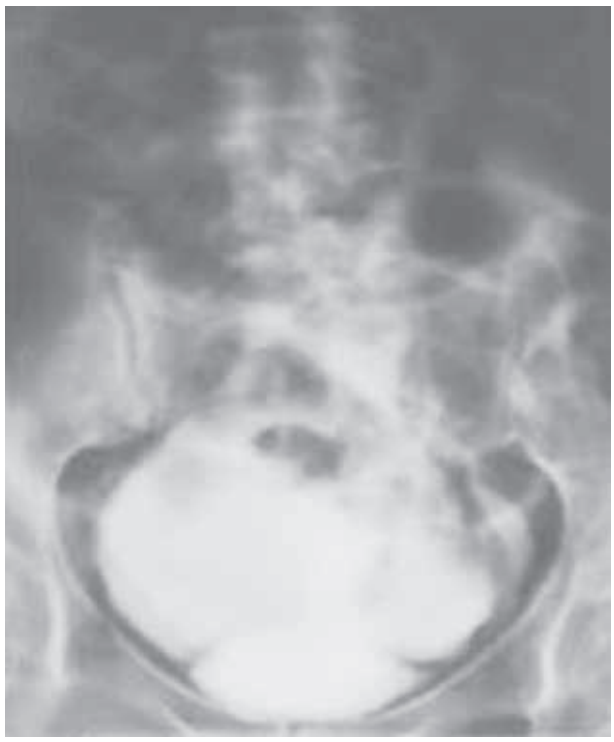


Fig. 1.4.13. Damage to the bladder

— Urgent hospitalization (within 1 hour) to a specialized hospital;

— Parallel examination and treatment in a hospital.

The first priority **A (airway)** — airway patency. The reason for their obstruction may be direct trauma to the face and neck with the damage to respiratory tract, their swelling, presence of foreign bodies. Control of airway patency is achieved by their readjustment, if necessary intubation is performed, tracheotomy.

After the restoration of airway patency it is necessary to assess the adequacy of breathing — **B (breathing)**. This requires auscultation of the lungs (breathing should be done on both sides), calculate respiratory rate, and in the hospital X-ray of the chest. At this stage it it is necessary to identify the damage, which prevents adequate breathing (open or tension pneumothorax, large hemothorax, cardiac tamponade) and, if necessary, conduct puncture of pleural or pericardial cavity, close the wound of the chest at the open pneumothorax. Adequate breathing while ballotating thorax is achieved by conducting a ventilator.

Restoration and maintenance of blood circulation (C) is achieved by external bleeding arrest, conducting fluid therapy with crystalloid solutions (the first stage — the infusion of Ringer's solution in a volume of 20 ml/kg body weight), chest compressions is carried out in the absence of cardiac activity.

Assessing the level of consciousness of the patient is performed by Glasgow (Glasgow coma scale). Great importance in providing first aid and transporting of the patient is given to preventing of hypothermia.

Table 5

Normal Vital Signs in Children of Different Age

Age	Pulse (beats/min)	Systolic blood pressure (mm Hg)	Respiratory rate (breaths/min)
Newborns	120–140	60–90	40–60
Nursling	110–140	75–95	30–60
Younger children	100–120	80–100	25–40
Preschool children	80–110	80–110	20–35
School children	70–100	85–120	18–25
Teenagers	60–90	100–120	12–16

Transportation must be carried out on rigid stretcher, at suspicion on fractures of limbs — with their compulsory immobilization, on patient's neck put cervical collar.

Upon arrival at the hospital diagnostic and therapeutic measures are carried out in parallel. In all cases cardiac activity is monitored (continuous ECG-monitoring), during mechanical ventilation — continuous monitoring of parameters, pulse oximetry, the installation of a nasogastric tube, bladder catheterization (the only contraindication — urethral injury). The stability of the patient's condition is assessed by vital signs (pulse, blood pressure, and respiratory rate), level of consciousness, urine output (minimum 1ml/kg/hour), the level of hemoglobin and hematocrit.

Remembering the high frequency of associated injuries in severe trauma (car accident, a fall from a height), it is necessary to make a survey of all body systems. At revealing associated trauma treatment plan and sequence of surgical interventions may vary considerably depending on the nature and number of injuries, the patient's condition (Table 5).

QUESTIONS FOR THE FINAL CONTROL

1. Basic mechanisms of abdominal cavity injuries.

2. Classification of injuries at abdominal trauma.

3. Main symptoms of parenchymal organs injuries.

4. Main symptoms of hollow organs damage.

5. What additional research methods are most informative at parenchymal injuries and hollow organs of the abdominal cavity?

6. What are the indications for emergency surgery at continuous bleeding that arose as a result of parenchymal injuries and hollow organs of the abdominal cavity?

7. What indicators of clinical and laboratory researches (blood pressure, pulse, general analysis and biochemical studies of blood) show hemorrhagic syndrome and bleeding in the abdominal cavity?

8. What clinical symptoms are marked at esophageal perforation?

9. Which X-ray method is the most informative for the diagnosis of esophageal injury?

10. Which survey methods are used at trauma of the chest cavity?

11. What is the indication for surgery at hemothorax?

12. What methods are used for diagnosis of injuries of the esophagus?

13. What is important to identify at the patient with traumatic urinary system?

14. What changes can be detected by palpation of the lumbar region of a patient with a kidney injury?

15. What is the basis justification of preliminary diagnosis of traumatic kidney damage?

16. What methods of diagnosis will help in identifying kidney injury?

17. What are the main features of children examination with an injury of bladder and urethra?

18. What is necessary to identify at a patient with a traumatic injury of the bladder?

19. On the basis of what principles is formed a list of injuries for the differential diagnosis at traumatic injuries of pelvic bones and pelvic organs?

20. What are the most reliable diagnostic methods that will help in the identification of urethral trauma?

21. Features of physical examination of children with scrotal trauma.

22. What is most important to determine at formation of treatment management in patients with traumatic damage to urinary system?

1.5. FEATURES OF CHILDHOOD TRAUMA. FRACTURES OF EXTREMITIES, BONES, PELVIS, SPINE IN CHILDREN

The rate of childhood injuries has increased in recent years, a growing number of complications in the treatment of common fractures of limbs, spine, pelvis, which often leads to disability among children. Hence the urgency of the problem of child injury, the importance of the role of the physician in providing first aid at the prehospital and hospital stages.

Specific objectives:

1. Familiarize with classification of fractures in children.
2. Master main clinical symptoms of fractures.
3. Master technique of examination of the patient with a fractured limbs, spine, pelvis.
4. Master types of transportation, the amount of first aid, medical care.
5. Familiarize with the basic principles and methods of fractures treatment in children.
6. Know the terms of consolidation of fractures.
7. Know the complications in the treatment of fractures and their prevention.
8. Familiarize with methods of rehabilitation therapy.
9. Interpret additional research methods for fractures; evaluate the data of X-ray and ultrasound examinations.
10. Identify the indications for conservative or surgical treatment methods depending on the type of fracture and the child's age.

FEATURES OF FRACTURES IN CHILDREN (EXTREMITIES, SPINE, PELVIS)

Types and features of child injuries

Injury means the sudden effect of environmental factor on the body that violates the anatomical integrity of tissues and physiological processes. Damage in children is quite frequent and makes up 30% of all surgical pathology of childhood.

Childhood traumatology has its own characteristics that significantly distinguish it from adults' trauma. First of all, these are features of child injuries structure, i. e. circumstances and factors of injury. They distinguish the following types of child injuries: school (identical to industrial injuries in adult), sports, outdoors, home, generic.

In children 75% of the damage accounts for household injuries. In adults there is no such type of damage as school trauma but there is industrial traumatism. Generic injuries occur during birth. In the structure of generic child injuries the highest rate concerns traumatic cerebral trauma, the second place is occupied by broken limbs, the third — damage to the peripheral nervous system (paresis, paralysis), the fourth — spinal injury, and the fifth — damage to internal organs.

The second feature of childhood trauma is the correlation of cause and effect relations of damage to the child's age. In newborns there are such injuries, which does not happen in older children, for example, as a result of the passage through birth canal arise cephalohematoma, generic tumor. In children less than 3 years old prevails household trauma, most often — burns, which in this age make up 30%. At preschool children also prevails domestic trauma, but it is mostly due to falling.

The third feature of childhood trauma is the correlation between the nature of damage to the morphological and functional features. For example, fractures on the growth zone (epiphysiolysis, apophysiolysis, meta epiphysiolysis) are observed only in childhood, because growth zones are and function only in children, fractures of the type of "green branches", "wicker", "vine", subperiosteal injuries.

The fourth feature — correlation of symptoms and course of lesions with morphological and functional characteristics of the child. This feature is common to all types of injuries in children due to abundant vascularization of damage; is always accompanied by considerable swelling and hematoma, due to the immaturity of the central nervous system and receptor apparatus in children is often observed traumatic shock; during significant burns phase — burn shock is reduced.

The fifth feature of childhood traumatology is the priority of sparing approaches in diagnosis and treatment. A child is never deliberately revealed such symptoms of fractures as crepitus and abnormal mobility; doctors prefer conservative treatment, and if they resort to operation, they use less traumatic methods.

The sixth feature of the pediatric trauma — the occurance of acquired defects due to damage to the immature structures. Damage to areas of growth in children can lead to further dysfunction of these areas, occurance of strains and shortening of the limbs and disability.

BONE FRACTURES

Features of bone injuries in children are caused by morphological and functional features of the locomotor apparatus.

In children there is a high proportion of the organic component of bone, and the younger the child, the less minerals his bone tissue is full of, i. e. the more pronounced the elastic features of the skeleton. A young child's epiphysis is mostly a cartilage tissue, which performs the function of a damper. The periosteum is thick, elastic; its blood supply is plentiful.

This leads, on the one hand, to the occurrence of subperiosteal fractures, on the other one — their rapid healing. Good vascularization of the skeleton leads to significant swelling and bruising, and at the same time to rapid consolidation. Due to the elasticity of ligamentous apparatus in children rarely occur dislocations and injuries, and the presence of growth zones explains the occurrence of fractures that run through phisar plates.

Features of fractures in children:

1. Fractures occur rarely, if comparing to rate of traumatic factors and injuries occurrence.
2. Rarely there are traumatic dislocations.
3. Rarely there are fragment fractures.
4. Fractures are often going through the zones of growth: epiphysiolysis (gap between the epiphysis and the metaphysis), epiphysiolysis or osteoepiphysiolisis (gap of epiphysis with metaphyseal part), apophysiolysis (gap of apophysis) (Fig. 1.5.1.–1.5.3).
5. Subperiosteal fractures are common.
6. Fractures are often of the "green branches" type (Fig. 1.5.4, 1.5.5).

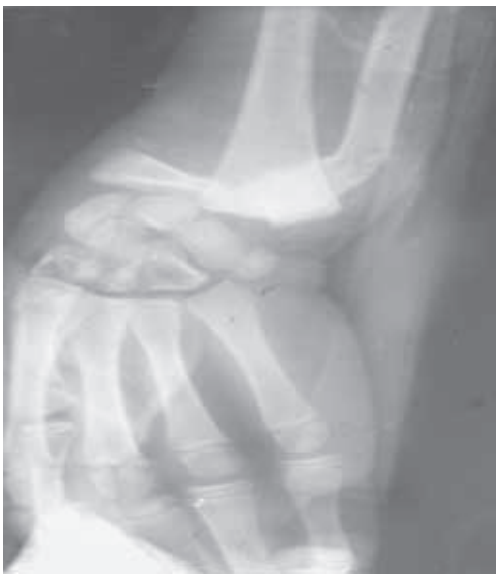


Fig. 1.5.1. Radial bone epiphysiolysis

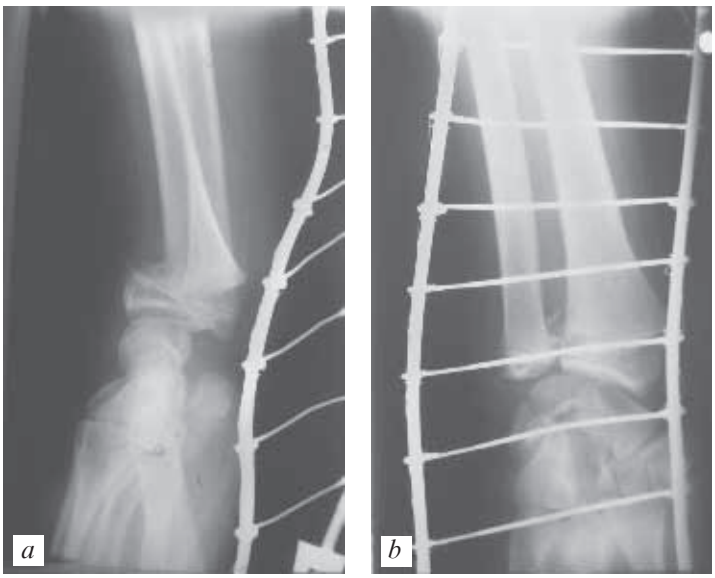


Fig. 1.5.2. Radial bone epiphysiolysis *(a, b)*

7. Fractures are accompanied by significant swelling and hematoma (Fig. 1.5.6).

Clinical manifestations of fractures are divided into probable and reliable, the probable clinical symptoms include deformation, local pain and dysfunction. Intensity of deformation depends on the presence or absence of displacement of fragments, the thickness of soft-tissue membrane around the damaged segment, the size of the accompanying swelling and bleeding in the tissues. Local swelling and hematoma may be insignificant during compression, fractures with slight displacement of bone fragments. The nature of displacement of fragments (width, length, angle, rotational) is determined by a direction of the traumatic factor, the level of the fracture tension of the muscles. The latter causes several typical displacements. For example, at fracture of hip total bone damage in the upper third is accompanied by abduction, flexion and external rotation of the proximal fragment as a result of thrust of sciatic and iliopsoas muscles, in the bottom third — shift of the distal fragment in the dorsal side due to reduction of three-headed calf muscle (Fig. 1.5.7).

The leading symptoms are pain and dysfunction, pain can be independent and provoked. Provoked pain is detected by percussion, palpation and by using special methods. For example, Werneul's positive symptom (compression of pelvis by the iliac crest bones entails pain) is a sign of a fracture with violation of integrity of the pelvic ring. A dysfunction is indicated by forced position of limbs, restriction of movements, inability to rely on the limb. Limitation of movement can be determined by means of special signs. For example, at epiphysis of front ilium the patient cannot move forward (a "reversing" symptom), at fracture of pubic bone one cannot take his heel off the bed, lying on it (a symptom of "stuck heel"). These clinical signs are constant, observed at all kinds of fractures.

The reliable signs are abnormal mobility and crepitus of fragments, but in children deliberately they are not defined, it additionally traumatizes the

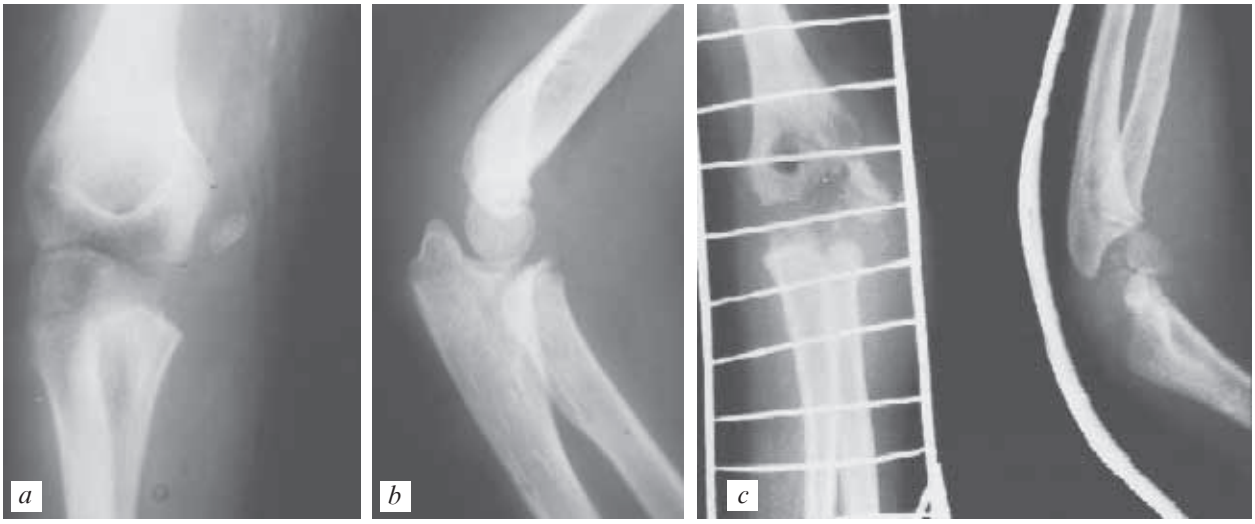


Fig. 1.5.3. Apophysiolysis *(a–c)*

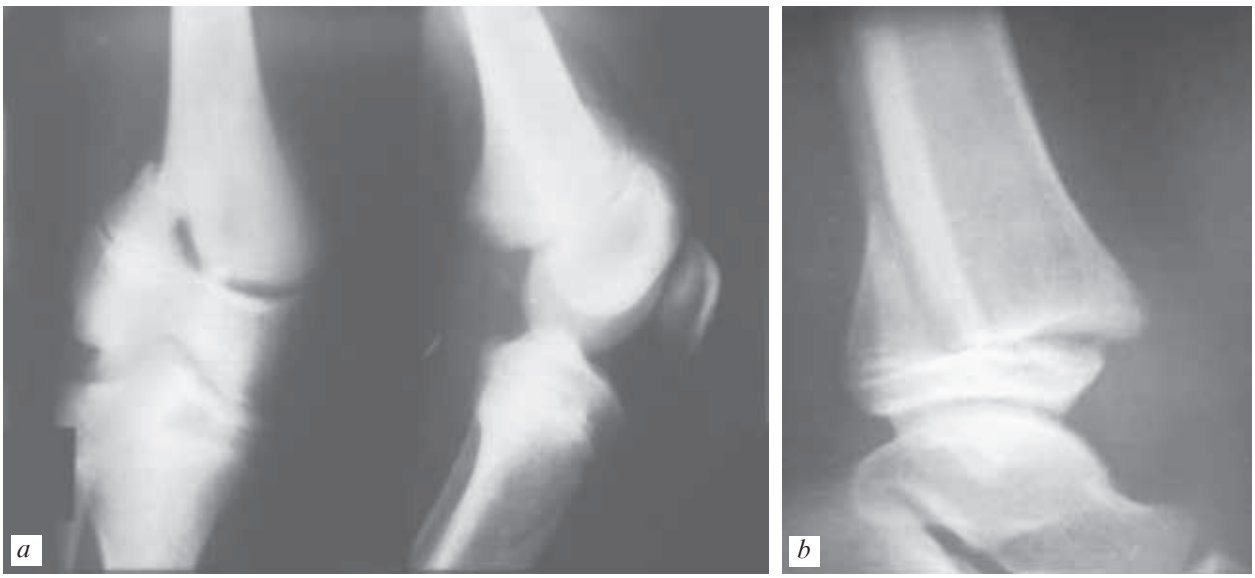


Fig. 1.5.4. Osteoepiphysiolisis *(a, b)*

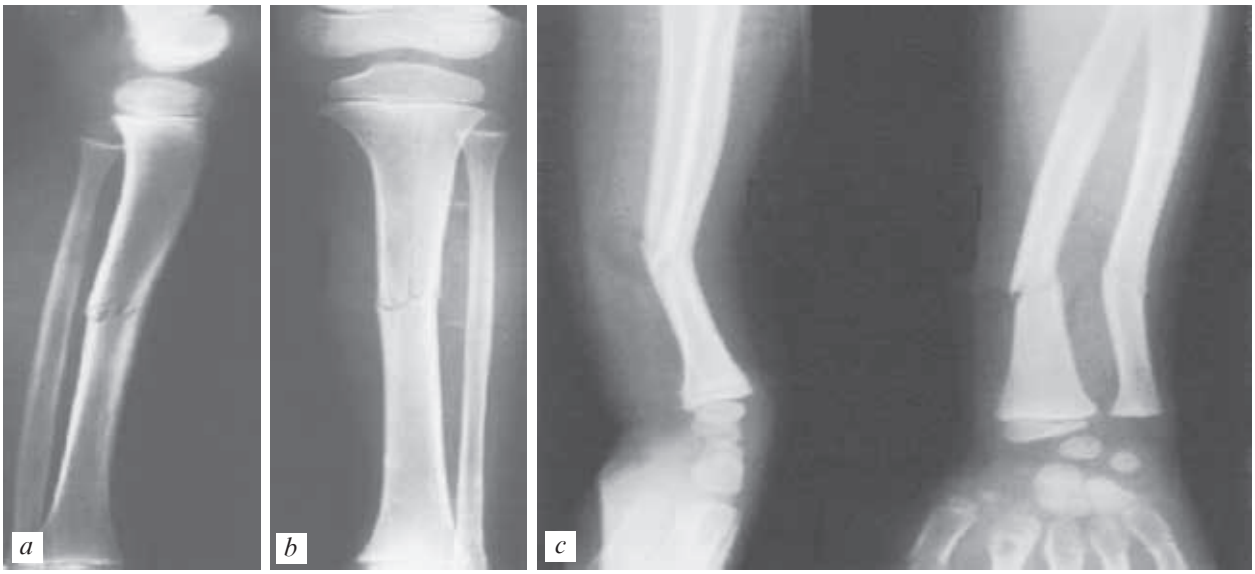


Fig. 1.5.5. Subperiosteal fracture of the "green branches" type *(a–c)*

child and worsens the offset. These symptoms may be determined spontaneously during transport immobilization during moving the patient.

The main mean of supporting diagnosis is X-ray exam, which allows not only to detect the fracture, but also to determine its nature, the type of displacement. X-ray is made in two projections. At difficulties of exam result interpretation (fractures without displacement, damage to areas of growth in young children) the contralateral limb X-ray is conducted, which helps to diagnose the pathology by comparison.

The main thing in treatment of fractures is to achieve consolidation and complete recovery of function. Fracture healing processes take place in stages and under favorable conditions the distinguish the following stages.

Stage I — fibrous callus — is characterized by organization of blood that has accumulated in the area of the fracture. Blood on 4–5th day of injury trans-

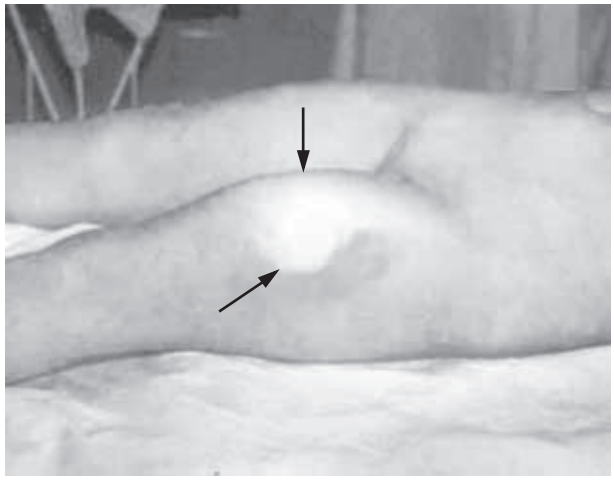


Fig. 1.5.6. Fractures in children are accompanied by a large swelling and hematoma

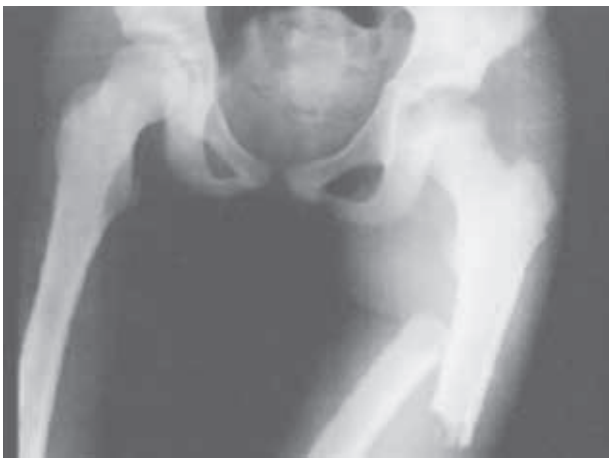


Fig. 1.5.7. Displacement of bone fragments at hip fracture

forms into blood clot, fibroblasts form and within the first two weeks fibroreticular tissue that surrounds the fragments in the form of coupling in the fracture zone appears. Fibroblasts are osteogenic progenitor cells. Their sources are perimyelis, periosteum, and vascular endothelium. Fibrous callus is deprived of mineral components, has no mechanical strength and X-ray is not contrasted.

Stage 2 — soft bone callus — develops, passing cartilaginous, at undisturbed osteogenesis during adaptation of fragments and a secure fit. Fibroblasts, which belong to the melanocytes, proliferate, produce collagen, alkaline phosphatase activity increases and calcium formation takes place, i. e. parallel-fibrous tissue and mineralization appears. Primary bone structures form. This process proceeds starting with the 7th day after the injury and lasts an average for 1–1.5 months. Soft bone callus is by X-ray contrasted, has a mechanical strength sufficient to hold fragments, but deforms easily under pressure.

Stage 3 — maturation of bone structures — lasts for 2.5–4 months and is characterized by the formation of lamellar bone tissue, regenerate restructuring and structural adaptation.

The smaller the child is, the shorter these stages are and more intense is the course of a reparative process. Taking into account dynamics of normal repair process, it should be provided by timely reposition, reliable fixation by sparing methods. Neglecting these principles may result in a breach of physiological healing, delayed consolidation, deformation of regeneration, formation of distortions and false joints.

General principles of fractures treatment in children (M. V. Volkov)

— gentle attitude;
— individual approach;
— timely repositioning of bone fragments;
— reliable fixation;
— early functional treatment.

Methods of treatment can be conservative and operative, based on the principle of sparing treatment, the advantage should be given to conservative methods. By means of reduction and fixation, they are divided on ***immobilization*** (complete immobility of the injured segment during the period of fixation), ***functional*** (the union of the principle of rest and motion — the immobility of damaged segment, while maintaining joint function) and ***combined*** (combining functional and immobilization methods).

The choice of treatment method depends on the patient's age, type of fracture and displacement of fragments, localization of damage, injury term and condition of the child. Thus, in infants they use only conservative methods for fractures of femur diaphysis — functional and combined, at chronic fractures, which are not properly fused, — operational ones.

In children, the priority is given to conservative treatment, which provides a one-step closed reposition (if necessary) and fixation of the injured segment with plaster cast. Reposition of fragments is not conducted when there is fracture without displacement; a slight shift (subperiosteal fracture); displacement is allowable.

The latter includes such a displacement of the fragments, which can be neglected, because during growth the bones lengthen and thicken, thus providing the opportunity for spontaneous correction. The smaller the child, the more opportunities for correcting deformation. In this regard ***admissible displacement of fragments*** at fractures of diaphysis in infants is considered a total displacement across the width; displacement along the length no more than 2 cm; displacement at an angle — no more than 30°. In children of the first year of life allowable total displacement in width; in length — not more than 1 cm; angular displacement is unacceptable. In older children at fractures of the metaphyseal and diaphyseal in the absence of angular and rotational displacement is allowed almost complete shift in width, and the distance of fragments is evaluated as follows: excellent — the width displacement by 1/3; good — the width displacement on 1/2; satisfying — the displacement by width on 2/3.

In all these cases reposition is not carried out, and only fixation and rehabilitation therapy are carried out.

Reposition is indicated in case of inadmissible displacement of the fragments, as angular and rotational displacement, fractures in the area of growth even with slight disblacement in children of all age groups in the future may lead to deformity and shortening.

Simultaneous closed reposition is used in cases when it may terminate in effective immobilization; it is used mainly at epiphyses, transverse meta- and diaphyseal fractures of the long bones, at para- and intra-articular fractures and lesions of small bones. Reposition is preferably carried out manually under general anesthesia. At simultaneous closed manual reposition in case of damage to large segments with significant displacement along the length sometimes traction devices are used. Mostly these are devices for shoulders and hips; they are used mainly for older children.

At closed reposition they walk after three rules:

1) A comparison of distal fragment by the central;

2) At traction should be contra-traction;

3) The force should be applied along the axis of the segment.

As immobilizing agents tires and plaster bandages are mainly used. Means of immobilization provide secure fixation in those cases when the motion state remains in traumatized segment and in two adjacent joints. It should be remembered that in children because of the morphological and functional characteristics, injuries caurse significant soft tissue swelling, in connection with which the plaster immobilization should be only in the form of tires that do not cover more than 2/3 of the diameter of the limb or in the form of gypsum dressings slotted all through, which will prevent compression of the soft tissues.

Immobilization method of fixing has some *advantages*: a relative simplicity of the method, the mobility of the patient; possibility of treatment on an outpatient basis. Its *disadvantages* are contractures of adjacent joints, the possibility of secondary displacement of fragments (in this regard, after the edema dissipates it is necessary to carry out X-ray control and strengthen the bandage), the difficulty of monitoring the wound in open fractures, concomitant injuries.

In newborns at birth trauma immobilization using a plaster is not used, because their skin is easily damaged, its diaper rash, maceration occur, inflammation may assist. These children are used fixing soft bandage (Dezo bandage — at damaged upper limbs, bandaged legs in fully extended position by Crede–Keffer at hip fractures), or cardboard splints.

Functional treatment method consists of functional reposition of fragments ("reposition in time") and fixation of the injured segment by constant stretching. Traction may be skeletal, soft (sticky plaster) for plaster "boot". Gentle stretching is carried out on soft tissues, skeletal — directly on the bone: the power of the first one — no more than 3 kg, the second one is intended for severe loads. Therefore, they use a soft traction in children aged 2–3 years old, when there is no need to put considerable efforts, skeletal — in children after 3 years in oblique and transverse fractures of the hip, shoulder, shin bones (Fig. 1.5.8).

At functional treatment they walk by the following rules:

1) The limb should be in the mid physiological position

2) The need in providing limb's rest

3) Fragments should be compared

4) The load should be gradual

5) There should be contratraction (most frequently by the body of the patient).

At proper treatment strategy within the first 3 days (reposition stage) self reposition of bone fragments happens; at unsatisfactory standing in the same period they a one-step closed manually reposition in the conditions of traction is performed. The next stage (retention stage) — constantly adapted traction for up to 3 weeks is provided. During this period callus is formed, and then (reparation stage) maturation of bone structures occurs.

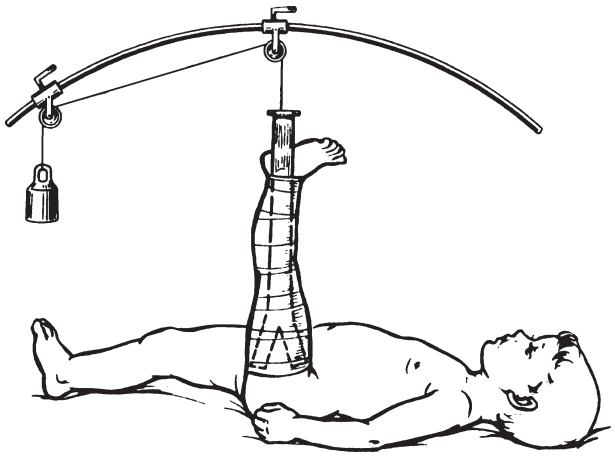


Fig. 1.5.8. Adhesive plaster traction by Schede at femur fracture in a child up to 3 years

The advantages of functional method are undeniable: high reliability (there is no secondary displacements), absence of joint contractures; correction capability of standing of the fragments and treatment of common soft tissue injuries. But there are *disadvantages:* the patient is prescribed bed rest, the hospital term of the patient in the hospital increases; you need a qualified daily observation. Furthermore, in the supine position with upturned feet blood circulation worsens, there is a risk of stagnation phenomena in the lungs, intracranial pressure increases. In this regard, at hip fractures, shoulders, shin bones, they usually use a combination of treatment that start with stretching (Fig. 1.5.9, 1.5.10) After the formation of callus (an average of 3 weeks) the patient is applied a plaster cast to until the end of the total period of fixation and he is discharged from the hospital.

The patients continue therapy at the outpatient department. General terms of fixation of the injured segment depend on the age of the patient and localization of damage. In newborns consolidation of the fracture of the clavicle takes 1 week, shoulder — 10 days, hip — 2 weeks. At school children medium terms of fixation at fractures are: clavicle — 3 weeks, upper third of the shoulder — 4 weeks, middle — 5 weeks, bottom — 3 weeks ; bones of the forearm, wrist — 4 weeks; upper third of thigh — 4 weeks, medium — 6 weeks, bottom — 5–6 weeks, shin bones, foot — 5–6 weeks, in the younger age group these terms are by 1–2 weeks less. Rehabilitation measures are beginning to take place immediately after the end of immobilization (physical therapy, massage, physiotherapy, spa treatment)

Surgical treatment is used rarely — in 3–4% of cases. It is carried out at an inefficiency of conservative methods (unsatisfactory state of fragments), long-standing and wrongly consolidated fractures.

Open reposition is carried out taking into account the morphological and functional characteristics of

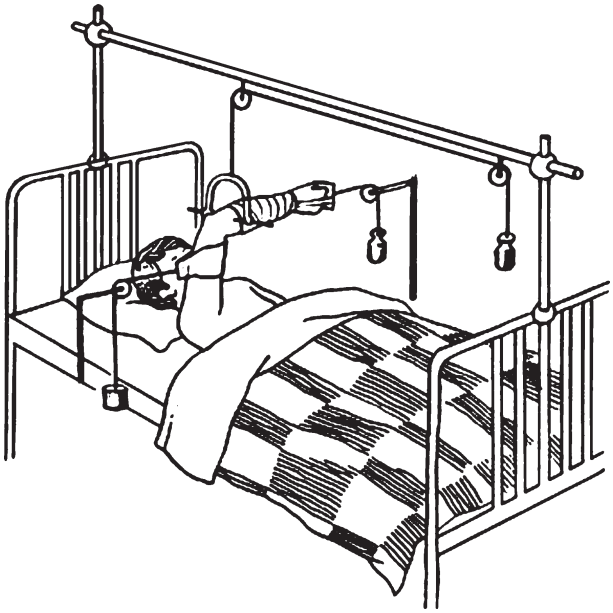


Fig. 1.5.9. Skeletal traction at humerus fracture

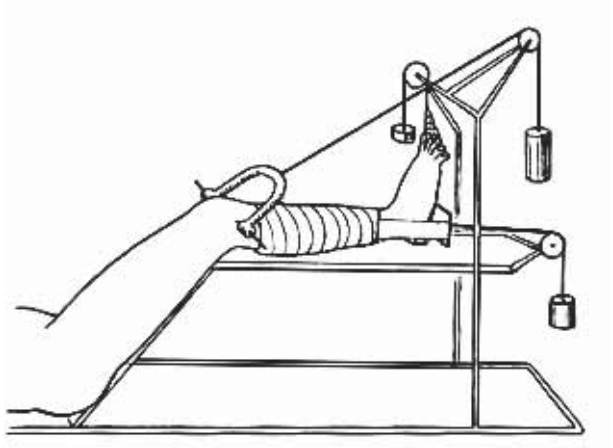


Fig. 1.5.10. Skeletal traction at femur fraction

children's age, using sparing methods; as fixing means Kirshner needles are mostly used, at hip fractures — Bogdanov rods; plate osteosynthesis metal plates are not used. At combined injuries (fractures combination with advanced soft tissue injuries), preference is given to external fixation by compression-distraction spoke apparatus by Ilizarov, by which allows not only to reposition, to reliably fix traumatized segment, but also to provide optimal conditions for the treatment of wounds and wound infection in a patient's mobility conditions. The disadvantages include the possibility of surgical treatment of delayed consolidation, the formation of a false joint, and suppuration around the spokes, clamps and the development of osteomyelitis.

VERTEBRAL FRACTURES

Vertebral fractures are divided into uncomplicated and complicated. Uncomplicated are considered spinal injuries, in which the pathological process does not involve spinal cord and its roots. Complicated forms of vertebral fractures are characterized by the development of neurological symptoms.

In children compression fractures of bodies, fractures of transverse and spinous processes arches are often observed.

Fracture of the spinous processes of vertebrae

Fracture of the spinous processes of vertebrae in children occurs at direct mechanism of injury (a hit in process region). Child is bothered by severe pain in the area of damaged process that increases with flexion and extension of the spine. In the process projection find swelling, acute pain on palpation. X-ray reveals fracture line in the lateral projection.

Treatment

The area of fracture is injected with 3–5 ml of 1–2% novocaine solution. The patient is placed on a bed with a shield. Physiotherapy is conducted.

Fractures of transverse processes

Fractures of transverse processes occur mainly in the lumbar vertebral region as a result of direct or indirect mechanism of injury — at sharp bending the trunk. Unexpected sharp decline in the quadratus lumborum, which is attached to the crest, ilium, five transverse processes of lumbar vertebrae and ribs XII, may cause a fracture of one or more transverse processes.

Clinical picture and diagnosis

Clinically, fracture of the spinous process is characterized by pain that is localized apart from the spinous processes, and increases with body bending to the opposite side (Payr symptom). Hyperextension in the hip joint as a result of tension of large buttocks muscle also increases pain. The presence of abrasions, bruising, and hematoma in the lumbar region in the projection of the fracture requires the exclusion of kidney damage.

X-ray study clarifies and defines the location of the fracture. It should be remembered that transverse processes have a nucleus of ossification, which can be taken for bone fragments. Errors in radiodiagnosis are possible by superimposing the shadows of gas in the intestine, psoas muscle shadow and the shadow of transverse process.

Treatment

At fracture of one or two transverse processes the patient is placed in bed with a shield for 2–3 weeks, pain disappears, they start physiotherapy for back muscles. At fracture of three or more transverse processes, which indicates a pronounced tension of lumbar muscles, from the lower part of the pelvis to the nipples on 4–6 week a plaster corset is imposed. After a few days they start dosage activity. After removal of corset conduct a course of gymnastics with a gradual increase in its intensity. Sport is permitted in 3–4 months.

Compression fractures of vertebral bodies

Compression fractures of vertebral bodies occur mostly at falling on legs, buttocks and forced flexion of the trunk. Mechanism of injury can be represented as follows: at falling there is a strong bending of the spine, compression of the vertebral bodies and intervertebral discs. If the compressive force exceeds the elastic limit of vertebrae and cartilage, compression occurs, induration of cancellous agent, bone beams are converging, and vertebra takes the form of a wedge. At falling headfirst cervical and upper thoracic vertebrae suffer, at falling on legs and buttocks are damaged mainly bodies of lumbar and lower thoracic vertebrae.

Clinical picture and diagnosis

Among clinical signs most characteristic are constant mild pain in the area of damage with limited mobility in the spine, protective back muscle strain at the site of injury, difficulty in breathing (posttraumatic apnea). In case of damage to the lower thoracic and lumbar spine there can be pains in the abdomen with diffuse nature with tension in anterior abdominal wall, which disappear quite quickly. The exact pain localization over the area of spinous process of vertebra compression is managed at palpation and percussion. Functional dosage load at a moderate flexion of the head, trunk, rising of straight lower extremity causes pain in the injured vertebrae.

Diagnosis is based on clinical and X-ray examinations. Picture of vertebra compression fracture on spondylogram is characterized by the following features: a wedge-shaped deformation of different degree, slide of the locking plates with the formation of wedge-shaped ledge, stairs form deformation of the front surface of the vertebral body, the violation of the topography of intersegmental furrow, intervertebral space, formation of kyphotic curvatures of the spine as a result of a shift back of the vertebral body into the spinal canal, a subluxation in the intervertebral joints (Fig. 1.5.11).

Treatment

Main task in the treatment of compression fractures is early and complete unloading of the front spine unit. At the same time they achieve a correction of sphenoid deformation of compressed vertebra and prevent its subsequent deformation, and spinal cord is protected from compression. Such discharge is easily achieved by traction.

The patient is placed on his back on a hard bed (under the mattress is placed a wooden shield), head side of the bed is lifted on 25–30 cm using coasters. On both armpits they use cotton-gauze webbings, to which loads fix.

At damages of cervical or upper thoracic vertebras (to IV thoracic vertebra) traction is performed with the help of Gleason loops. Along with the traction they conduct reclination by placing a bag with sand under prominent spinous processes. Bag pressure on a corner kyphosis promotes preservation of lordosis with a fan-shaped position of the vertebral bodies.

Patients from the first day are prescribed physical therapy on four periods to build a strong muscular corset and good posture. At compression fractures of two or three vertebras, as well as at compression of vertebral body in the anterior region more than on a half (unstable fracture) in the period of rehabilitation is prescribed wearing stabilizing corset within 6–12 months. it is necessary medical check-up during 1.5–2 years.

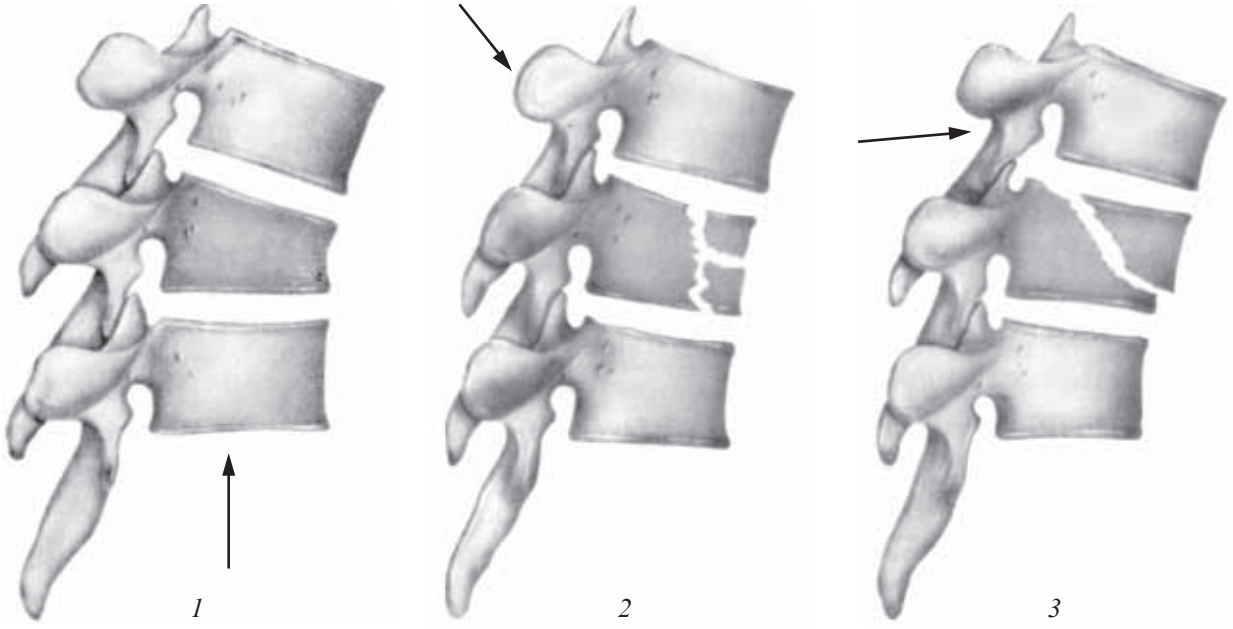


Fig. 1.5.11. Variants of vertebral fractures: *1* — wedge-shaped fracture as a result of vertical compression; *2* — comminuted fracture as a result of a sharp corner bending; *3* — fracture-dislocation as a result of bending under the action of strength in the transverse direction.

FRACTURES OF THE PELVIS

Fractures of the pelvis in children in most cases are related to street (traffic) trauma or a fall from a height. 62.3% of child patients arrive at the hospital in a severe and very severe condition.

Characterizing damage to the pelvic bones and its organs in the child a certain value belong to anatomical and physiological features of the pelvis, weakness of ligamentous apparatus in pubic and sacroiliac joints; presence of cartilage layers, which divide all three pelvic bones (iliac, ischial and pubic); elasticity of pelvic ring, associated with the flexibility of bones. All this contributes to the fact that in children, for example, instead of the typical two-way vertical fracture of the iliac bones which is observed in adults, there is a rupture and separation of the iliac-sacral joint, mostly on one side.

At damages of the pelvic bones they distinguish following types of damage:

— Isolated fractures of individual bones without damaging the integrity of pelvic ring;

— Fractures with violation of the integrity of pelvic ring:

— Fractures of the anterior part of the pelvic ring with injury of sciatic and pubic bones on one or both sides, a break of symphysis pubis or a combination of these injuries;

— Fracture of posterior pelvic ring, which includes fractures of the sacrum, ilium, and the gap of the sacroiliac joint;

— Malgaigne fractures;

— Fractures of the acetabulum;

— Fracture — dislocation — a fracture of the pelvis with a dislocation of the sacroiliac or of pubic joint.

Type of fracture is characterized by the clinical picture and determines the term and amount of therapeutic measures.

The most severe are fractures, which are accompanied by a significant retroperitoneal bleeding, damage to the pelvic organs. Urethra or bladder is damaged, rarer — the rectum, vagina. Damage to the urethra occurs mainly in boys, the gap is localized in the membranous unit of the urethra where it passes under symphysis pubis, near the bladder neck. Bladder rupture occurs due to an increase of hydrostatic pressure on wall, damage occurs at the apex of the latter, at the junction of parietal peritoneum on the bottom of the bladder. Rupture of the pelvic diaphragm in children at blunt abdominal trauma, which is combined with fractures of the pelvis is a very rare phenomenon.

Clinical picture and diagnosis. Symptomatology of damage is determined by the localization of the pelvic bones, the nature of the fracture and associated injuries. At monofocal and especially polyfocal fractures pelvis position of the patient is often forced — on the back with stretched, diluted and rotated outward lower limbs. At fractures of anterior part of the pelvic ring legs bent at the hip and knee joints and dilute on "frog paws" type by Volkovich. At fracture of pubic joint legs more often bent at the hip and knee joints, passive dilution causes acute pain.

Typical and most frequent symptom at pelvic fractures — the pain of a permanent nature, corresponding to the region of injury, which is enhanced at low active movements and attempts to passive movements with feet. By gentle and consistent palpation of pelvis is localized pain, moderate at bruises and sharp at fractures, swelling, and sometimes deformation, indicating on the localization of the fracture. At rupture of pubic symphysis palpatory detect painful groove between pubic bones. Symptoms which are observed at fractures of the pelvis, — increased pain at compression of iliac crest (Verneuil symptom), while stretching the pelvis on the iliac crest (Larrey symptom), onset of pain with gentle rhythmic pressing on rump, with your fingertips under hands (symptom of "balloting sacrum" of Drachuk). Regardless of age a characteristic symptom of "adhering heel" described by V. V. Gorinevska: supine lying patient is unable to raise the extended leg because of the emerging pain that is worse from pressure of iliopsoas muscle on a broken bone, but the patient is not taking the heel off the bed, relatively easy drags his leg to the body, bending at the hip and knee joints.

Very rarely they observe asymmetry of the pelvis. Pelvic fragment offset in the proximal direction and its rotation outward give the impression of shortening of lower limb.

Fractures of the pelvis in children are always accompanied by the appearance after 2 hrs after injury bruising and swelling, which indicates the location of the fracture. Open fractures of the pelvis are rare. At fractures of the pelvis with a shift of fragments, as well as the violation of the integrity of pelvic ring with local changes there are significant signs of traumatic shock. Child is pale, skin is covered with a cold clammy sweat, and he wants water and weakly moaning. Blood pressure decreases, pulse is frequent of weak filling and tension.

The diagnosis is established after the X-ray of the pelvis and hip joints. Radiology of pelvic fractures in children is not difficult and is based on X-ray symptoms: the fracture line, change of cartilage zones width in comparing symmetrical formations, deformation of pelvic contours, seals of bone structures, their shift.

Treatment. Prescribe bed rest in the "position of the frog" on a hard bed for a period of 3–5 weeks, depending on the severity of the damage. In this case, legs are slightly bent at the hip and knee joints, they are placed on a soft cushion. Exercise therapy is performed on 2nd–3rd day; physiotherapy is prescribed only at related injuries, nervous disorders and prolonged immobilization. By the end of the third week in uncomplicated fractures there is a consolidation of the fracture, which is controlled clinically and X-ray.

In severe cases, besides the usual antishock measures (transfusion of blood and blood products, glucose-novocaine mixture, narcotic analgesics, heart drugs, hormones, vitamin therapy, hyperbaric oxygen therapy, and others.) is carried out intrapelvic anesthesia by Shkolnikov–Selivanov. Pain relief on Shkolnikov–Selivanov — a highly antishock and anesthetic event. It is performed as follows: put the patient on his back, with a fine needle produce anesthe-

sia of the skin on 1–2 cm inside from the front upper spine. Then long needle, connected to a syringe containing 0.25% novocaine solution is introduced to a depth of 10–12 cm, so that its cutting edge slides on the inner wall of the ilium. At needle moving inject 0.25% solution of novocaine (children 3–6 years — 30–40 ml; 6–9 years — 40–60 ml; 9–12 years — 60–70 ml; 12 to 15 years — 70–100 ml), which provides the effect of intrapelvic anesthesia on 18–24 hours (Bairov G. A., Ulrich E. V., 1976). At unilateral pelvic fracture novocaine solution is injected on the side of the fracture at the indicated dose, at bilateral — at half the dose from each side.

By reducing painful irritation of pelvic bone fractures, intrapelvic anesthesia promotes the disappearance of false symptoms that simulate damage to the abdominal organs.

At pelvic bone fractures of different localization main method of treatment is conservative. To eliminate the vertical displacement of one half of pelvic skeletal traction for the distal metaphysis of femur is used with a gradual increase in load up to 7–8 kg, until reduction and consolidation will not be achieved (for 4–6 weeks) Differences of pubic symphysis at its ruptures are eliminated with using "hammock" or tightening belt. Surgical intervention may be indicated for ruptures of the symphysis, fracture — dislocations, polyfocal fractures, with a large divergence of fragments (if conservative treatment would be ineffective), at concomitant damage to urethra, bladder. For stable osteosynthesis use metal plates, rods, screws, spokes-rod and rod apparatus.

QUESTIONS FOR THE FINAL CONTROL

1. Features of childhood trauma.
2. Main clinical symptoms of fractures, diagnosis, allowable bone fragments displacement in children.
3. Immobilization treatment of fractures in children. Advantages and disadvantages of the method.
4. Functional treatment methods of fractures in children. Advantages and disadvantages of the method.
5. Surgical treatment of fractures in children.
6. Limb fractures in children. Classification, clinical features, diagnosis, treatment. The value of early rehabilitation.
7. Spine fractures in children. Classification, clinical features, diagnosis and treatment.
8. Fractures of pelvis in children. Classification, clinical features, diagnosis and treatment.

Student must have the following knowlege

1. To issue the case record.
2. To carry out clinical researches.
3. To take the anamnesis.
4. To interpret the X-ray film.
5. To make immobilization of extremity.
6. To make a plan of treatment.

1.6. PURULENT SURGICAL INFECTION IN CHILDREN

SURGICAL SEPSIS

Observed in recent years growth in the share of septic diseases in pediatric pathology and severity of nosocomial infections can be explained by several factors. First of all, it changes the microbial landscape: gram-positive flora, marked in monoculture of 30–60's, was replaced by microbial associations of gram-positive and gram-negative aerobic and anaerobic bacteria. Furthermore, improper use of antibiotics inevitably leads to the emergence of acquired resistance, which in turn is accompanied by a lack of response from the standard antibiotic therapy.

So, in 50–60 years worldwide there was a wave of hardest staph infection (staphylococcus pneumonic plague), struck mostly young children and was accompanied by a high mortality rate of up to 70% in infants. Note that the geography of this wave followed exactly haphazard and unnecessarily wide use of penicillin.

In recent years, the study of bacterial infections trigger mechanisms at the molecular level has supported depth understanding of sepsis etiopathogenesis. This disease is fundamentally different from other infections with the fact that develops with the participation of opportunistic pathogens, so in order to achieve recovery there is no need in their complete elimination from the host body.

Sepsis — a generalized inflammation that arises in response to an excess of microorganisms and/or their fragments.

You can also define sepsis **as life-threatening condition associated with the rapid multiplication of micro-organisms and is characterized by multiple organ dysfunction syndrome of varying severity; leading, as a rule, is a violation of the respiratory function.**

The most common general symptoms of sepsis in children are: fever (sometimes with hectic temperature curve), tachycardia, and tachypnea, progressive intoxication, skin icteritiousness, early accession of septic pneumonia, the emergence of other secondary sites of infection.

Systemic inflammatory response syndrome (SIRS) is one of the common symptoms of sepsis.

Criteria or SIRS in children include:

1) Body temperature: rectal above 38°C (oral is above 37.8°C) axillary above 37.2°C, or rectal below 36°C (oral below 35.8°C) axillary below 35.2°C;

2) Tachycardia: increased HR at or above upper limit of the age norm (Table 6).

3) Tachypnea: increased BH to or above the upper limit of the age norm or hyperventilation (PCO_2 higher than 32 mm Hg) (Table 7).

4) The number of leukocytes in peripheral blood: more than 12×10^9/l (leukocytosis) or less than 4×10^9/l

(leukopenia) or there is not less than 10% of leukocytes immature forms (sum of metamyelocytes, myelocytes and stab).

In order to confirm the child's SIRS there is a need to install not less than two of the above symptoms. Having SIRS criteria only points on system nature of the organism response to some pathological processes, but is still not a cause of diagnosis. For example, SIRS develops not only in sepsis, but also as a result of trauma, burns, hyperthermia (overheating) in the absence of any other signs of infection.

For diagnosis of childhood sepsis must be available signs SIRS, developing on the background of the infection.

Theoretically, there are three possible combinations:

1) SIRS and source of infection: the child with purulent-inflammatory lesions at any localization has severe disease, joins SIRS, which determines the threat of generalization — the diagnosis is "sepsis" and is initialized intensified treatment;

2) SIRS and bacteremia: a child that has no obvious sites of infection, showing signs of SIRS, at the same time is found a positive blood culture. Diagnosed with "sepsis", which is an indication for antibiotic therapy;

3) SIRS and clinical symptoms of infection: at the child that has no obvious sites of infection, appears SIRS, blood culture is negative, but there are clinical signs of infection. This is the most difficult situation to make "sepsis" diagnosis. It is placed for health reasons, as the delay in the appointment of antibiotic therapy can lead to the development of septic shock with fatal outcome when objective indicators of sepsis will be obtained only at autopsy.

Table 6

Heart rate with sepsis in children

Age	Heart rate (beats/min)
Newborn1	60 and more
3 years	140 and more
4–5 years	130 and more
7 years	120 and more
8–10 years	110 and more
12–14 years	90 and more

Table 7

Breathing rate with sepsis in children

Age	Breathing rate
Newborn	40–60
1 month	48
2 month	43
3 month	41
4–6 month	40
7 month	37
8–10 month	36
11–12 month	35
2 years	31
3 years	28
4 years	26
5 years	24
6 years	26
7 years	23
8 years	22
9 years	21
10 years	20
12–13 years	19
14–15 years	16–18

To date, it is advisable to consider two sepsis variants.

First, often encountered option — **sepsis as a complication of surgical infection**, when the "worse local state is(purulent foci), the worse is patient's general condition". **In this situation, sepsis substantially reflects achievement of certain extent severity of the patient's condition**. In such cases sepsis in the formulation of diagnosis should take the appropriate place: for example, acute hematogenous osteomyelitis, sepsis. This procedure determines diagnostic and treatment management — a priority not of immunomodulation and extracorporeal detoxification attemptsbut of adequate drainage of the purulent source.

The second option — sepsis as a rare disease — pyosepticemia when determining criterion is the occurrence of metastatic (pyemic) purulent foci. Then, in the formulation of the diagnosis after the word "sepsis" should follow the designation of a primary source of infection, followed by an enumeration of locations pyemic (secondary) purulent foci.

Clinical features and diagnosis of surgical sepsis. The primary focus of sepsis can be not only inflammatory processes in organs and tissues, but also infected foreign bodies, implanted prosthetic devices (endoplastitis), and vascular catheters (catheter sepsis). In addition, the source of sepsis in severe illness may be translocation of intestinal bacteria.

Symptoms of sepsis depending on the timing of their appearance can be divided into early, that help with diagnosis in the initial phase of sepsis, and late that characterize the next phase of sepsis and its complications. Early symptoms may include primary (100% of cases) and (or) metastatic suppurative foci, accompanied by manifestations of inherent toxic-infectious syndrome, and bacteremia (78.6%).

Late sepsis symptoms include warning signs of decompensation or development of the sub-functions of the various organs and systems of the patient due to intoxication or septic metastases and bacteremia or a combination. The most frequent late symptoms of sepsis — cardiovascular, respiratory, renal failures, central nervous system lesions (impaired consciousness), multiple organ failure.

Thus, the clinical picture of sepsis is the basis for its diagnosis.

Great importance for clinical course and outcome of sepsis have its complications, which include manifestations of insufficiency of vital organs and systems functions of the patient, septic shock, exhaustion, bleeding, thrombosis, etc.

Features of patient's examination with sepsis include daily physical examination, designed to assess the general condition, the nature of change (location, lesion volume) at the site of infection and the search of possible purulent (metastatic) lesions. For the same reason it is advisable to use the entire available arsenal of modern diagnostic techniques (X-ray, ultrasound, computed tomography, magnetic resonance imaging, angiography, etc.).

Laboratory signs of sepsis. Despite attempts to identify the specific laboratory criteria for sepsis, currently there are no pathognomonic tests for sepsis. Nevertheless, laboratory data undoubtedly is a valuable complement of clinical picture.

It is known that sepsis is characterized by neutrophilic leukocytosis with a left shift, and in some cases there may be a leukemoid reaction with the number of white blood cells up to 50–100 th/ml and higher. Bacteremia, especially in children, the elderly, patients with alcoholism, can cause the development of neutropenia. An early manifestation of latent infection may be thrombocytopenia. It should be noted that, according to the literature, the incidence of disseminated intravascular coagulation syndrome at bacteremia is quite low and is only 11%, whereas thrombocytopenia at sepsis may occur in 56% of patients. Morphological changes of neutrophils at sepsis include toxic granularity, occurrence of Dole cells and vacuolation. Production of red blood cells at sepsis is reduced. According to M. I. Kuzin and L. L. Shimkevich, anemia at sepsis is observed in all cases, and in 45% of patients hemoglobin concentration is below 80 g/l.

Routine determination of the concentration of serum electrolytes, urea level, creatinine, hepatic indicators helps to identify the source of infection.

The most important method of confirming the diagnosis at sepsis is the microbiological examination (microscopy and culture) of blood, urine, cerebrospinal fluid, sputum, secreted from wounds or fistulas, as well as tissue of purulent focus. It is important not only to identify the detected microorganisms, but also quantitative assessment (degree of contamination).

For detecting bacteremia inoculation of blood is preferably carried out either as soon as possible after the beginning of rising temperature or chill, or 1 h before the expected temperature rise, preferably before the antibiotic. It is advisable to produce 2 to 4 blood samplings at intervals of not less than 20 minutes, as the increase in the frequency of samplings increases the likelihood of abjection. Blood sampling is carried out from a peripheral vein (not from the subclavian catheter). Blood is recommended to be distributed in two vials for aerobic and anaerobic incubation for 7 days.

General principles of intensive therapy for sepsis in children:

1. Intensive therapy should begin from the moment of admission to the hospital with sepsis and implemented in full. This principle is often violated because the doctor on duty, as a rule, limits with minimum assignments.

2. Intensive therapy should be carried out in parallel with urgent examination of the child, turning, with the evolution of diagnosis, from symptomatic to pathogenic and then into etiotropic.

3. Children with sepsis should be admitted to departments of intensive care unit (ICU).

4. In compiling the program of intensive therapy should be involved three specialists: anesthesiologist, a pediatrician and pediatric surgeon (in his absence general profile surgeon).

5. Intensive care must be carried out simultaneously in three areas: the impact on the source of infection, the impact on the patient's organism, the impact on the microflora.

Impact on the source of infection requires its early and complete sanitation, methods of determining the nature and characteristics of the main process.

Impact on the child's organism must come from the pathogenesis of multiple organ failure: it is necessary to provide detoxification, supplementation of BCC, respiratory support, normalization of the cardiovascular system, syndrome therapy.

A sustained infusion therapy necessitates catheterization of main veins. Catheterization of the great veins can only be carried out by specially trained doctors. Catheterization and catheter care should be carried out under strict aseptic and antiseptic trained personnel.

Antibiotic therapy may be **empirical or directed (causal).** *Empirical* antibiotic therapy means prescription of antibiotics directly at child's admission to the hospital before the results of microbiological studies. Unfortunately, empirical antibiotic therapy is often administered blindly. Such management is unacceptable, especially in sepsis, since the adequacy of empiric therapy may be fatal for the child. Thus, empirical therapy should be oriented.

Primary guidelines for empirical therapy can be bacterioscopy of Gram-stained smear of clinical material, obtained from the lesion, and information about possible pathogens specific to a particular state. The method allows determining the presence and ratio in material Gram-positive bacteria quickly, stained in dark-purple color and Gram-negative microorganisms, stained in dark-red color.

Starting empirical antibiotic therapy

Neonatal sepsis: penicillins with extended spectrum and β-lactamase inhibitors, cephalosporins of II–III generation in combination with aminoglycosides. In severe cases — carbapenems.

Urosepsis: III generation cephalosporins + aminoglycosides, imipenem, meropenem

Intra-abdominal sepsis, diffuse purulent peritonitis: III generation cephalosporins + aminoglycosides + metronidazole, imipenem, meropenem

Post catheterization sepsis: vancomycin, oxacillin or cephalosporins I–II generation + aminoglycosides, imipenem, meropenem

Bacterial endocarditis: III generation cephalosporins + aminoglycosides; vancomycin; imipenem, meropenem

Bacterial destruction of lungs, nosocomial pneumonia with severe course: cephalosporins of II–III generation + aminoglycosides; imipenem, meropenem.

In our opinion, in large multi-profile children's hospitals on the basis of bacteriological laboratories should be organized service of clinical microbiology, providing constant informing of clinicians about the state of resistance of hospital flora and promoting adequate choice of antibacterial drugs.

BACTERIAL LUNG DESTRUCTION

Concrete aims:

1. To master the list of diseases which cause intrathoracic tension.
2. To recognize the basic clinical symptoms of intrathoracic tension.
3. To differentiate intrathoracic tension depending on the origin.
4. To interpret the auxiliary methods of examination: ultrasound, roentgenologic, laboratory and biochemical analyses, indexes of hemodynamics (BP, P, t°, Hb, Ht).
5. To show the technique of pleural puncture, the puncture of intrapulmonary formations.
6. To explain the technique of thoracocentesis, drainage of pleural cavity, imposition of the system of passive or active aspiration.
7. To identify the features of course of separate diseases of lungs and pleura, which are accompanied by intrathoracic tension.
8. To analyse thecausal nexus of origin of intrathoracic tension at separate patients, base and formulate a previous clinical diagnosis.
9. To offer the algorithm of action of doctor at the syndrome of intrathoracic tension and patient management.
10. To interprete general principles of medical treatment of diseases, that is accompanied by intrathoracic tension and to define a indications to surgical treatment.

Actuality. Destructive pneumonia course (bacterial lung destruction (BLD) has a high rate of progression of the disease with pronounced changes in lungs and pleura, which is associated with high virulence of microorganisms, the presence of microbial associations and decrease in sensitivity of microorganisms to many antibiotics.

Definition. BLD is a necrotic process, affecting segment, part or the entire body with a wide variety and dynamic of structural and clinical manifestations of the disease. Limited or massive necrosis of lung tissue under the influence of pyogenic microorganisms is accompanied by formation of cavities decay.

According to the classification of Yu. F. Isakov et al. there are ***intrapulmonary*** and ***pleural*** forms of BLD. Intrapulmonary form is divided into infiltrative forms, lung abscess bullous form, lobitis. Pleural is divided into pyothorax, pneumothorax pyopneumothorax. It is assumed that one form of destructive lung disease may progress to another.

Classification of BLD:

Primary (aerogenic, bronchogenic)

Secondary (hematogenous, lymphogenous)

By the pathogen:

1. Gram-positive *(Staphylococcus)*
2. Gram-negative *(Escherichia coli, Pseudomonas aeruginosa, klebsiela)*
3. Mixed

By complication type:

1. Pulmonary complications (lung abscess, bulla, lobitis)
2. Pleural (pyothorax, pyopneumothorax, pneumothorax)
3. Complications of other organs (progressive mediastinal emphysema).

Etiology and pathogenesis of destructive pneumonia in children. While pathogenic staphylococcus remains to be leading in the etiology, it should be noted that BLD at the present stage is a disease caused by mixed infection. To implement the inflammatory process it is necessary that the antigenic irritation in the body exceeds the protective capacity of the immune system. There are no specific pathogens of purulent infection. Most of them can be distinguished from healthy individuals or from daily environment. Purulent pathogens of surgical infection (with decreasing frequency) include *Staph. aureus* and *Streptococcus* from gram-positive bacteria group, as well as a large group of gram-negative bacteria: *Pseudomonas aeruginosa, proteus, E. coli, klebsiella,* non-clostridial anaerobic flora. 60% of cases have mixed flora.

It should be noted that each of these microbes exist in large number in more or less virulent strains. To virulence factors include aggression enzymes (hyaluronidase, fibrinolysin, coagulase), endotoxin (lipopolysaccharide) and exotoxins (exfoliative toxin eritrotoxin, enterotoxins, and others) Depending on external conditions bacteria can change their virulence.

For example, when the temperature rises above 37°C streptococci increase the synthesis of exotoxins several times, and iron deficiency states contribute to the growth of Escherichia coli. Microorganisms may also have a capsule or special proteins of the cell wall, preventing phagocytosis or intracellular lysis.

Bacteria causing suppurative inflammatory diseases are relatively few antigenic compared with specific pathogens. This leads to a tendency to chronization and long-term persistence of virulent strains. For example, Streptococcus pneumoniae (*Strep. Pneumoniae*) is covered with polysaccharide capsule, which can stimulate the production of only IgM. Therefore, immunologic memory on pneumococcus is quite short. For gram-positive bacteria are most common skin lesions, subcutaneous fat, bones, lungs, gastrointestinal tract; for gram-negative pathogens groups — intestinal damage, joints, urinary tract. Gram-negative microflora plays a leading role in the development of postoperative complications and pathogenesis of postresuscitative disease. It is especially dangerous for infants, deprived of natural factors of protection against it. Sometimes a microorganism can be judged by the appearance of pus. Staphylococcus — the most pus organism while the pus is usually yellow and thick; only at pemphigus of neonatal exudate is serous. Pyogenic streptococci also gives or watery yellowish exudate is serous. For pneu-

mococcus is characteristic dense, viscous pus. At outbreaks with mixed flora pus is usually brownish. Gram-negative organisms produce sparse whitish or grayish pus. Some anaerobes (groups of *Bacteroides melanogenicus*) may be painted in black exudate is serous.

Clinical course. Infiltrative pulmonary form is before destructive stage of acute lung bacterial destruction development, according to the clinical picture is not much different from other etiologies of acute pneumonia. Rapid onset of the disease: in 60% of cases the beginning of lung bacterial destruction is preceded by acute respiratory infection, which blocks the ciliated epithelium, thereby creating the conditions for the penetration of S. aureus in the alveoli. On the 2nd–5th day course of ARVI is marked by a sharp rise in temperature up to febrile digits. From the first days of the disease physical changes in the lungs are clearly revealed: strengthened voice trembling, in differend degree pronounced shortening of percussion sounds, at auscultation — crepitation and microvesicular rhonchuses in significant numbers. In infants, on the foreground are general toxic symptoms: loss of appetite, vomiting, enteric syndrome, progressive exsicosis.

Infiltrative stage can be resolved as follows:

1. Is subjected to reverse development, leaving no significant changes in the structure of lung tissue.

2. End with the formation of bullae. In this case, there is a danger of the valve mechanism in draining bronchi; such bulls can increase dramatically in size and even lead to the development of the "syndrome of intrathoracic voltage" rupture of bullae and the development of pneumothorax.

3. Abscess formation infiltration — the formation of abscesses. The child's condition is deteriorating: Hyperthermia increases to 39–40°C with a pronounced hectic curve, daily temperature fluctuations in the range of 1.5–3°C, purulent intoxication progresses, with a pronounced neurotoxicosis, exsicosis. These physical methods are directly dependent on the size of the abscess and the degree of emptying, so at not draining abscesses they will match those at infiltrative stage, at emptying along with plenty moist rales can be detected abdominal symptoms.

Pulmonary-pleural forms of BLD can be primary, ie pleura leaves are involved in the process almost simultaneously with the lung parenchyma, and secondary that develop as a result of drainage of the abscess into the pleural cavity.

Pyothorax is clinically manifested in progressive deterioration of the patient with the growing signs of purulent intoxication. Depending on the amount of pus in the pleural cavity physical symptomatology will vary within very wide limits, up to dumb lung.

Pyopneumothorax — the most severe form of chronic destructive pulmonary pleural process. Develops at the break of subcortical abscess to the bronchus and pleural cavity and can manifest the picture of pleuro pulmonary shock: state of extreme severity, acute tachypnea with indrawing compliant places of chest, pulse is frequent, weak filling, pale skin with an earthy tint, paroxysmal painful cough, chest pain, mainly on the affected side. The child is restless, rushing, trying to find the most advantageous position. On the affected side is marked flattening of intercostal spaces, the lag of the chest half at breathing. Percussionally is determined tympanitis and mediastinal shift to a healthy way. By auscultation breath is sharply weakened.

Diagnosis. The study begins with the multi-chest X-ray, which task is to determine the phase of the disease, localization, character, contour, intensity and homogeneity of the liquid, determine the state of lungs, mediastinal shift.

At the first stage by a clinical laboratory and X-ray investigations in all patients unable to put topical diagnosis. X-ray analysis of the data and operational comparisons showed that the diagnosis can be formulated as the final on the basis of a conventional X-ray examination.

To determine the nature and volume of the liquid should be carried out of pleural cavities. To survey are subjected all patients with pleurisy. Analysis of the ultrasound results allows determining the structure of the liquid, the presence of adhesions, encysted cavities. It should be noted that the resulting characteristics of the object advantageously complement radiographic criteria, increasing informative survey in general.

Puncture taking of exudate for cytological study helps to clarify the morphological diagnosis and the phase of the disease.

Thoracoscopy is performed to update the status of the pleural cavity, the presence of bronchial fistulas the volume and nature of exudate.

X-ray diagnosis. The purpose of diagnosis is to identify the bacterial destruction of the lungs in uncomplicated and complicated states for the adequate therapy. In most cases, patients with BLD come to the surgical ward at the stage of complications due to the similarity of the clinical picture with lobar pneumonia, in connection with which patients are treated in a pediatric hospital and in some cases even receive therapy on an outpatient basis. The absence in the initial period of the characteristic clinical signs of the disease leads to the fact that the surgical hospital patients arrive already at development of pulmonary and pleural pulmonary complications.

Important questions are dynamic observation of the process in the pleural cavity of the bacterial destruction of the lungs, given the rapid changes in local data, their variety, and often severe, and even rapid process course.

The data obtained in the course of radiation research in the vast majority of cases appears crucial in establishing pyonecrotic lung disease, its location and extent. The main method of diagnosis and follow-up of the state of the pleural cavity is still the X-ray. However, the total number of X-ray, produced in a child with bacterial degradation of lungs, sometimes varies from 8 to 25, which is highly undesirable in children due to high radiation load. All the patients are performed X-ray in frontal and lateral projections.

Early X-ray signs of BLD are rapidly merging into segmental and polysegmentary infiltrates focal shadows, which are located in one or more portions

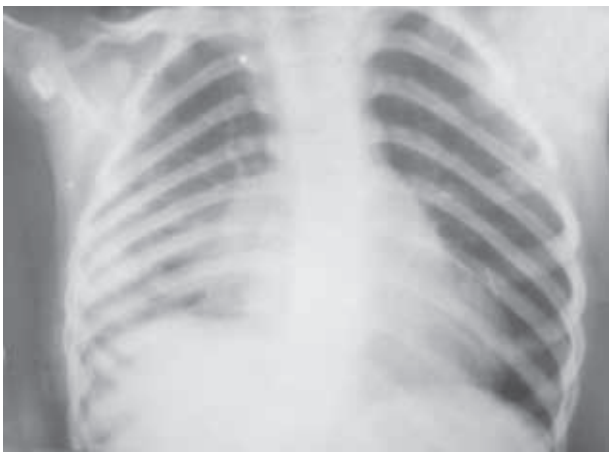


Fig. 1.6.1. Infiltrative pneumonia, direct projection

of the affected lung. At the height of the disease appear signs of reactive pleurisy as a narrow strip of compacted paracostal and interlobar pleura. Affected segments in the initial phase of inflammation are increased in volume and deform the course of interlobar gaps, often collaborate adjacent intact segments, vascular pattern of which is thickened (Fig. 1.6.1).

Ultrasound diagnosis. At decrease of lung tissue airiness due to pneumonic infiltration or atelectasis, it becomes echografically visible, has decreased echogenicity and homogeneous echo structure. Contour of pneumonic outbreak contour at onset is even, form may be different. In the case of lobar pneumonia outbreak contour follows the shape of the share; at segmental may have a pyramidal shape with the base facing the pleura, at focal — wrong, close to round. In airless pneumonic foci echogenic, branched, discontinuous strips are visible, which are air-filled bronchi, as well as tubular anechoic structures — containers or fluid-filled bronchi and rarer — thin linear echogenic strips — intersegmental connective tissue septums. Pneumonic areas have lower or middle echogenicity. At resorption of infiltrate multiple confluent hyperechoic areas of pneumatization appear in them. Ultrasound allows the of predestructive BLD phase course control and early enough to record the occurrence of pleural complications.

A possibility of daily monitoring of pathological process in the lungs and pleura was very important.

At complicated pneumonia course airless areas increase in size, a few small merge into larger ones (Fig. 1.6.2). Focal-drain form may be accompanied by suppuration. In these cases, in airless parts of the lung small areas of increased echogenicity appear in the center of which then arise anechogenic inclusion with a vague outline, which may be surrounded by echo-positive rim. Such sites are often multiple and are emerging centers of destruction.

Additional information gives computed tomography. On axial images is more clearly revealed typical subpleural localization of infiltrate, which is adjacent to costal or interlobar pleura to broad base. The structure of the inflammatory infiltrate in the center of which one or more segments of uniform density is determined, deprived of bronchial lumen, is also typical.

With the development of process already on the 2nd–5th day since disease has started, against infiltration rounded air cavitary formations by X-ray defines "dry bulls" of various sizes that arise due to necrosis of the lung parenchyma, their number varies from 2 to 5, single bull are rare. Around these cav-

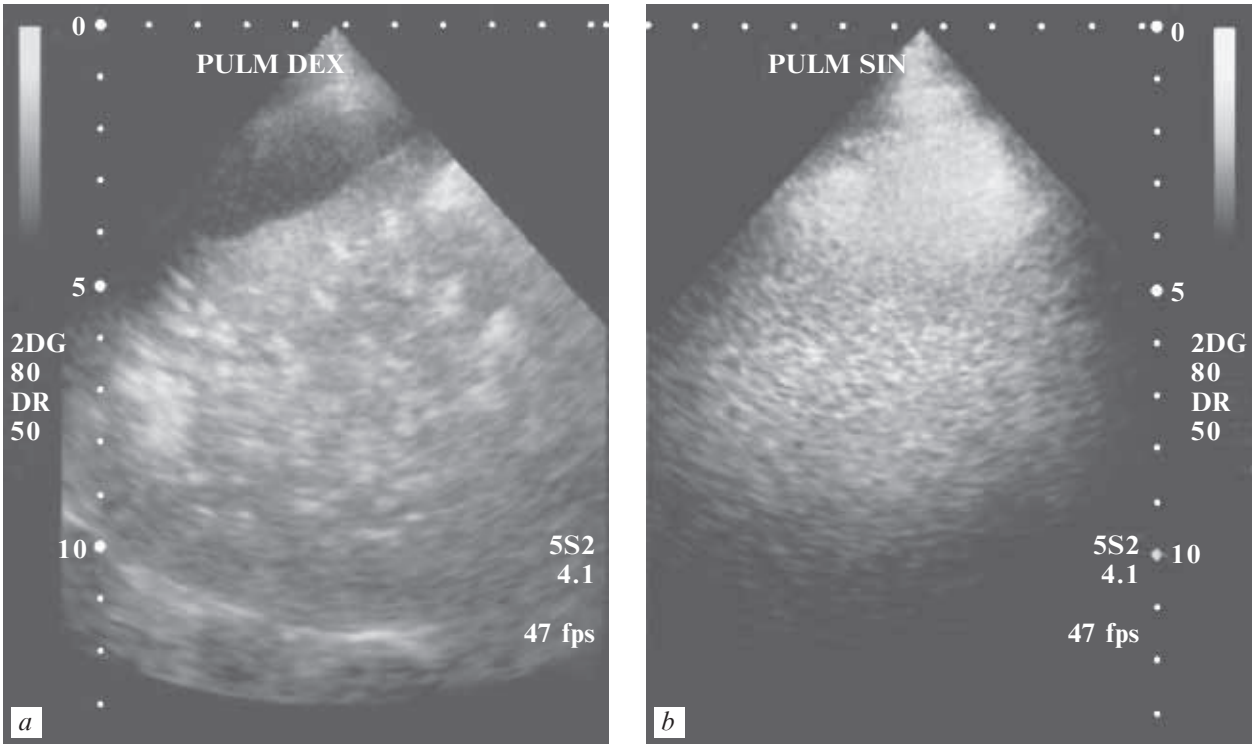


Fig. 1.6.2. Ultrasound picture at bacterial lung destruction (infiltrative form) *(a)*, healthy lung *(b)*

ities inflammatory shaft is traced. At positive dynamics, this shaft becomes thinner. Echografically airless areas first increase in size, a few small merge into larger, then in the center echogenic inclusions appear with a vague outline of round shape or a flat arcuate echogenic band. Arcuate strip is clearly visible in mutually perpendicular planes. Ultrasound also helps to visually assess the condition of prebulls parenchyma.

During a computer tomography bullae are well detected visually, ranging in size from 2 to 20 mm in diameter, round, with a density of up to 980–1008 N. Bull studying in soft mode visualization showed that their internal contours are always equal, there are no partitions. Adjacent to the bull areas with higher densitometry are lung parenchyma in a state of compression or scar tissue, formed as a result of the inflammatory process.

Lung abscesses. X-ray for an abscess are characteristic rounded shades, with sagging bottom contour and a high degree of shading; inner contour of the abscess is initially uneven, due to hanging in the lumen fragments of necrotic tissue.

At drainage of lung abscess into the bronchi on X-ray is determined the cavity with clear rough contours and the level of liquid in it — a basket symptom.

Depending on the location and size of the abscess, fistula size, informing the abscess cavity with a draining bronchus a different degree of emptying of the abscess is determined and, correspondingly, different X-ray picture (Fig. 1.6.3–1.6.5).

At ultrasound diagnosis in the case of abscess formation in the vacuum section of the lung liquid cavity is formed, containing echogenic slurry, air bubbles, and at draining it through the bronchus in the abscess cavity a large amount of air appears.

Ultrasound can also solve the emerging at X-ray examination difficulties in the differential diagnosis of lung abscess and pulmonary echinococcosis, especially in complicated forms of these diseases.

Abscesses on computed tomography are determined as rounded shadows of different diameters with a clear outline and matter density from -40 to + 20 N. The greatest value of computed tomography acquires at revealing of X-ray signs of multiple lung lesions. Computed tomography allows defining accurate lesion volume, revealing abscesses with a diameter of less than 2 cm, which is not possible with X-ray examination.

X-ray diagnosis of pulmonary-pleural form of BLD. At bacterial lung destructions, accompanied by pleural complications, X-ray picture is very diverse and presents three basic forms of complications — pyothorax, pneumothorax and pyopneumothorax. Total pyothorax is by X-ray looks like intense homogeneous darkening of an affected half of the chest, merging with mediastinum shadow. It is impossible to determine the pulmonary pattern, sinus is not differentiated. Intercostal spaces are expanded as compared with the healthy side, the shadow of the heart and mediastinum moves dramatically in the opposite direction (Fig. 1.6.6, 1.6.7).

At delineated pyothorax, there is an intense dimming, usually localized paracostal and above the diaphragm.

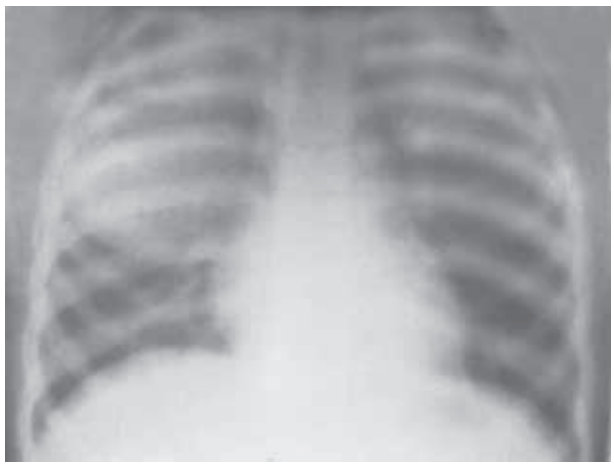


Fig. 1.6.3. Not draining abscess, direct projection

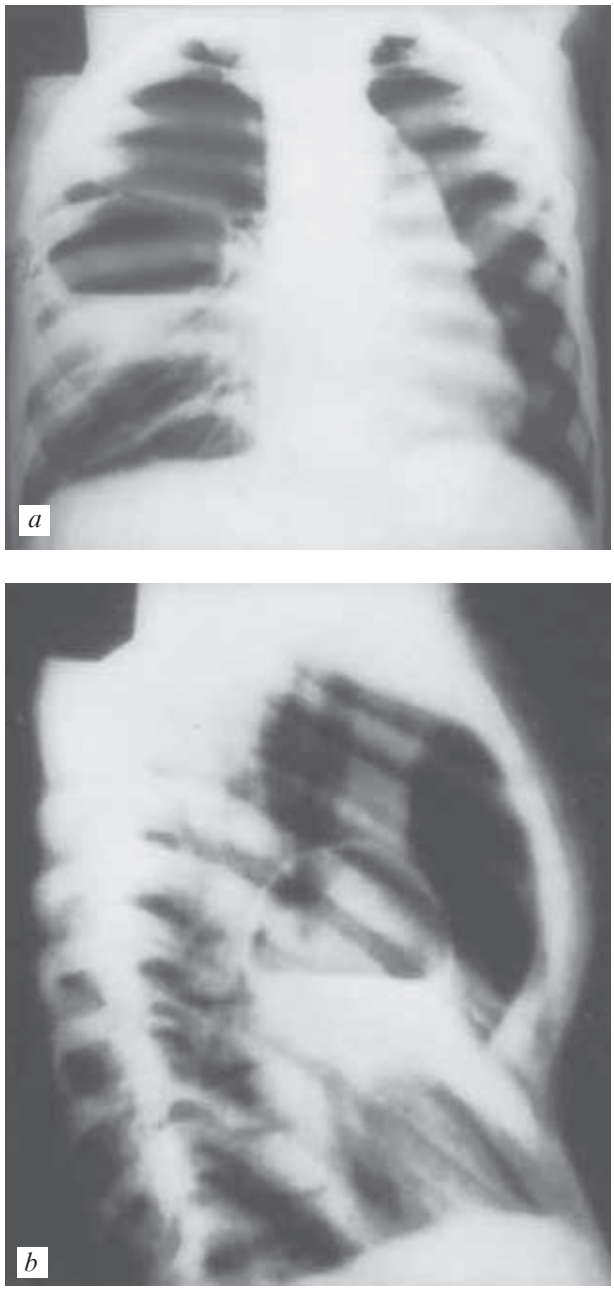


Fig. 1.6.4. Draining abscess, straight *(a)* and lateral *(b)* projections

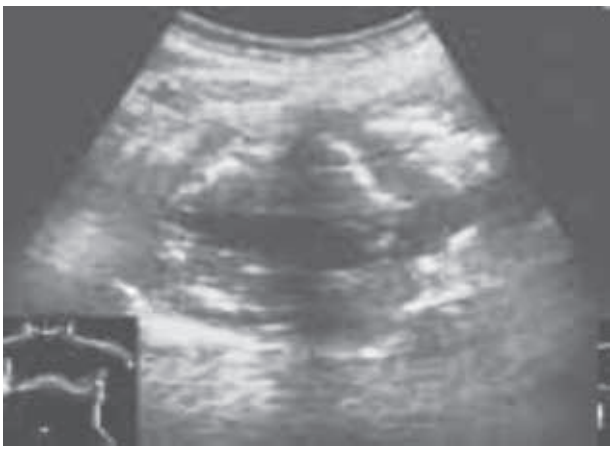


Fig. 1.6.5. Ultrasound image of lung abscess

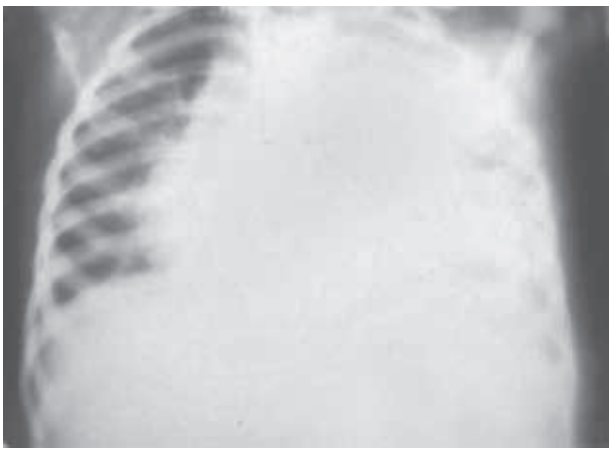


Fig. 1.6.6. Chest X-ray, direct projection: total left-sided pyothorax

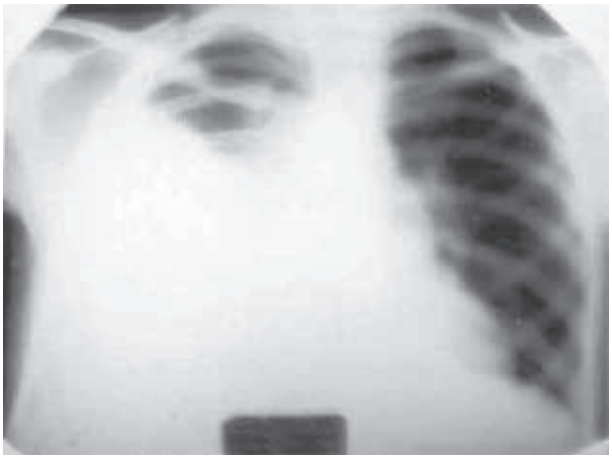


Fig. 1.6.7. Chest X-ray, direct projection: right-delimited pyothorax

Fibrinothorax should also be noted, which is still called mantle-like pyothorax and mantle-like pleuritis.

Pyopneumothorax is the second most common form of the BLD course. At pyopneumothorax there is the accumulation of air over clear liquid level, which is moved at changing the position of the baby's body. Against the background of darkening vascular pattern is not clear, mediastinal organs are displaced to the opposite direction.

During ultrasound examination complications from the pleura side are determined well before the X-ray examination, which is due to the possibility of higher resolution of ultrasound. Ultrasound picture is characterized by visualization of echogenic exudate with inhomogeneous internal structure with clear boundaries and location (Fig. 1.6.8).

At pyothorax in the initial stages in the pleural cavity on fluid background fibrin strands in the form of linear echo structures are visualized, that divide free space of pleural cavity into cells, resembling a honeycomb by structure. The main echographic sign of pyothorax pyopneumothorax — separation of two signals from the pleural sheets by echo negative site of uniform and inhomogeneous structure representing the contents of the pleural cavity. For fibrin thorax is common uniform echo negative effusion, freely distributed in the pleural cavity. Fibrinopurulent stage is characterized by heterogeneous pattern of pleural contents due to a variety of small signals, depending on the amount of suspended matter in the

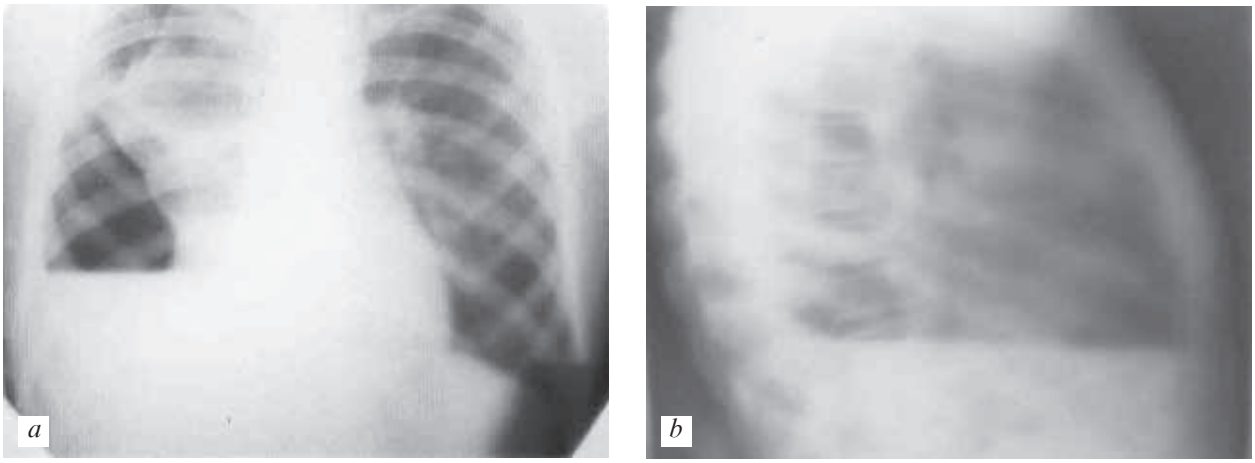


Fig. 1.6.8. Chest roentgenogram, straight *(a)* and lateral *(b)* projection: right pyopneumothorax

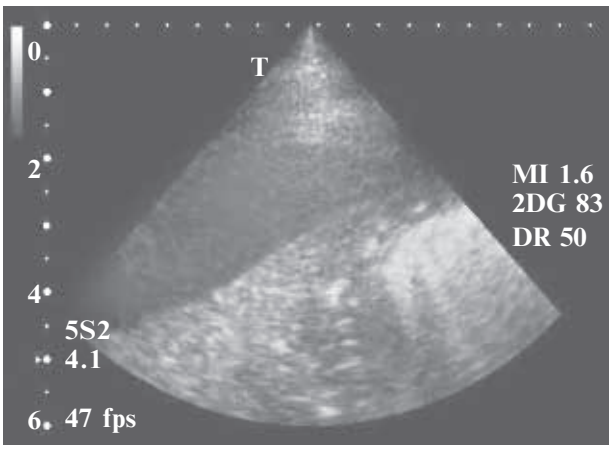


Fig. 1.6.9. Ultrasound picture of pleural effusion

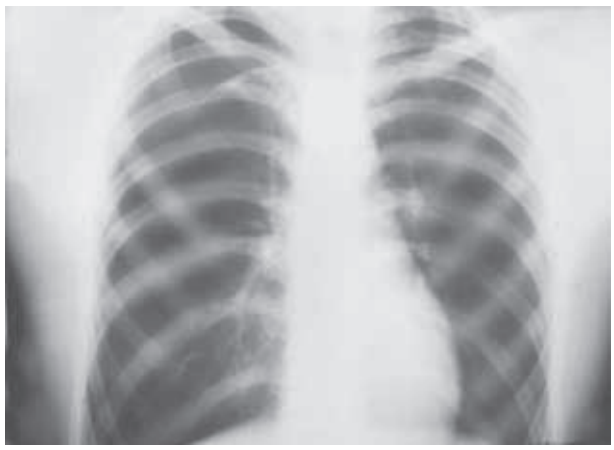


Fig. 1.6.10. Chest X-ray or direct projection: right pneumothorax

effusion; there is a moderate thickening of pleura leaves (Fig. 1.6.9).

Ultrasonography quite early identifies clusters of even a small amount of fluid in the pleural cavity, the presence of encapsulations. Unlike X-ray, ultrasonography gives the opportunity to see the volume and nature of pleural content that appears on X-ray as homogeneous shading. Highly informative in the case of pyothorax and pyopneumothorax is computed tomography.

The highest value it has in the diagnosis of multilimited, interlobar, paramediastinal fluid accumulation. On axial sections it is possible to identify the most minimal accumulation of fluid at any site. It enables more accurately than with ultrasound to determine the location for the puncture (Fig. 1.6.10).

At tension pneumothorax on the affected side there was a sharp transparency increase, pulmonary picture is missing, and the mediastinum is shifted to the opposite direction with the formation of "mediastinal hernia". The level of fluid in the pleural rug is not determined in a vertical position or in a study in later position. Lung becomes closer to the root, intercostal spaces are expanded, and diaphragm flattens due to its marginalization from top to bottom. In the absence of tension collapsed lung is less pronounced, displacement of the mediastinum was absent.

At pneumothorax lung ultrasonography is not informative because increased air in the pleural cavity resembles a picture of a healthy lung.

Very important in the diagnosis and treatment of dynamic control of lungs bacterial destruction are indicators of the general blood test. In 60% of patients the onset of the pathologic process is marked by reduction in the number of red blood cells to $2.1 \pm 0.7 \cdot 10^{12}$, suggesting a moderate anemia, which apparently preceded the disease causing secondary immunodeficiency due to chronic hypoxia. Usually patients with BLD suffer from leukocytosis due to absolute neutrophilia in the initial stages and the increased number of lymphocytes in the final stages of inflammation. In all cases there is a shift of leukocyte formula to the left, which indicates tension and intensity of the inflammatory response.

About 30% of patients at the early stages of the disease suffer from eosinophilia, while 70% of patients in the later stages of the disease have moderate eosinophilia ($6 \pm 1\%$), which is a sign of the positive dynamics of the process.

Treatment. Treatment of bacterial lung destruction like all septic diseases is aimed at three points of application (Table 8).

Thoracostomy with passive, active or combined aspiration to date remains the leading local treatment method of pleural complications of BLD in children. However, along with the positive aspects of the method, it is necessary to specify its shortcomings — early depressurization of the pleural cavity, the appearance of the chest wall cellulitis, tissue necrosis around the drainage, and pressure on the lung.

Thoracostomy with drainage is quite effectively. Conducting a rational adjuvant therapy in patients with various forms of BLD, including targeted antibacterial, immune treatment, intensive infusion therapy, as well as differentiated instrumental methods of treatment can significantly reduce the number of complications and virtually avoid of radical surgery.

PURULENT-INFLAMMATORY DISEASES OF BONES AND JOINTS

Purulent-inflammatory diseases of bones and joints include septic arthritis (metaepiphyseal osteomyelitis), acute hematogenous osteomyelitis (AHO), metaepiphyseal, chronic, atypical forms of osteomyelitis. Etiopathogenesis, classification, clinical features, diagnostic features, principles of treatment, prevention of complications are the objects of the study.

Specific objectives:

1. Learning the forms of hematogenous osteomyelitis, complications and consequences. Learning

Table 8

Scheme of Therapy of Bacterial Lung Destraction in Children

<table>
<tr><th colspan="2">Microorganism</th><th>Macroorganism</th><th>Focus of the inflammation</th></tr>
<tr><td colspan="2">Antibiotics
Bacteriophages</td><td rowspan="3">1. Immunity correction
2. Desensitization
3. Elimination of respiratory failure
4. Normalization of central and peripheral circulation
5. Stimulating therapy
6. Correction of acid-base balance and water-electrolyte metabolism
7. protease inhibitors
8. Vitamin therapy
9. Detoxification
10. Correction of anemia</td><td rowspan="3">1. Providing drainage of pulmonary and pleural cavities
2. Rehabilitation of tracheobronchial tree
3. Injection of medicinal substances into the lesion
4. Physiotherapy</td></tr>
<tr><td colspan="2">Injection methods</td></tr>
<tr><td>Intramuscularly
Intravenously
Per os</td><td>Intrafocal
Endolymphatically
Intrabronchial</td></tr>
</table>

the main clinical manifestations of septic arthritis (metaepiphyseal osteomyelitis) in children up to 3 years.

3. Differentiate acute forms of as a manifestation of sepsis, severe sepsis, and septic shock.

4. Interpret auxiliary methods: ultrasound, X-ray, CT, measurement of intraosseous pressure, cytological and bacteriological tests, biochemical parameters, and hemodynamic parameters.

5. Performing differential diagnosis of osteomyelitis with other diseases of the locomotor system.

6. Analyzing cause-and-effect relationship of inflammatory diseases of bones and joints, justifying and formulating a preliminary clinical diagnosis.

7. Learning the algorithm actions of the doctor with the inflammation of bones and joints.

8. Familiarizing with general principles of osteomyelitis treatment.

1. Topic actuality

Mortality from complications in the form of generalized sepsis forms ranges 10 to 75%, disability from chronic osteomyelitis — to 10%. Recognition of AHO within the first hours and days of the disease is an important diagnostic problem.

General information of the disease

A) Characteristic lesions of meta diaphyseal zones of long tubular bones with the development of subperiosteal abscesses and increase of intraosseous pressure, associated with thrombosis and necrosis in metadiaphysis. The process can be local or generalized (septic shock).

B) Early diagnosis and complex pathogenetic treatment, which consists of surgical treatment of osteomyelitic center, the impact on the macro-organism and microbial pathogens. Complications and consequences is the result of inadequate treatment.

C) The amount of intervention: a dissection of the periosteum, bone and joints puncture.

ACUTE HEMATOGENOUS OSTEOMYELITIS

The term "osteomyelitis" was first used by Reynaud (1831) and in translation from Latin means inflammation of the bone marrow. Indeed, the pathological process always starts in the bone marrow and then spreads to other elements of the bone, hitting spongiosa, cortex, periosteum, and in young children — the sprout area and epiphysis. Therefore, osteomyelitis, osteitis, periostitis, and chondrite should be understood by this term.

Depending on the routes of bone marrow infection differentiate hematogenous and exogenous osteomyelitis (S. Popkirov); the latter may occur after open fractures, gunshot wounds, after surgery, etc.

The occurrence of AHO is connected with penetration of microbes in bone marrow by hematogenic way.

Most often AHO affects actively growing long bones: tibia, fibula, humerus, radius and ulna. Lesion of the epiphyseal zones and epiphyses is typical-for children of 2–3 years. Multiple bone disease occurs in 9–30%. Boys suffer 2–3 times more often.

Etiology. The causative agent of AHO in 80–90% of cases is *Staphylococcus aureus.* In recent years the proportion of mixed and gram-negative flora has increased. Entrance gates at an early age may be purulent diseases of skin, mucous, navel, ear infections; in older age groups — carious teeth tonsils, infected wounds, and others.

Pathogenesis. Of a number many theories of AHO the most popular are the following:

1) Septic
2) Vascular (A. A. Bobrov)
3) Thromboembolic (Lexer)
4) Allergic (S. M. Derizhanov)
5) Nervous reflex (M. M. Elansky)

These theories give reason to believe that osteomyelitis is caused by autoinfection of sensitized organism against the background of reduced immunity. Peculiarities of blood flow and the structure of the bones in childrents play an important role in occurrence and clinical course. Under contributing moments they consider bone injury, previous infection, overcooling, vitamins deficiency, and other conditions that reduce the body's defenses.

The puculiar feature of process course at AHO is its development in a closed rigid bone tube.

N. G. Nikolaeva (1999) **identifies four phases i**n the dynamics of inflammation development:

Phase 1 — bone marrow edema (lasts 1–2 days);

Phase 2 — bone and brain abscess (3–4 day of the disease);

Phase 3 — subperiosteal abscess, and the pus through Volkmann's canal under pressure goes under the periosteum;

Phase 4 — soft tissue abscess — is characterized by necrosis of the periosteum and the release of pus in periosteal tissues (6th–7th day of the disease).

Prolonged disturbance of blood circulation in the affected bone leads to the formation of osteonecrosis (seizures). Depending on the degree of circulatory disorders sequestrations can be: total, central, cortical.

Sequestrum formation gives mechanical strength to bone and can be the cause of pathologic fracture.

Most appropriate ***classification*** was proposed by T. P. Krasnobaev, who distinguished three clinical forms of AHO:

1. Toxic or adynamic form — the disease fulvinant onset on the background of the common health and proceeds very rapidly with symptoms of *endotoxic shock*. Often there is collaptoid state with loss of consciousness, high temperature (40–41°C), vomiting. Punctate hemorrhages can be detected on the skin. Because of the severity of the condition it is difficult to establish local manifestations in the affected bone. The disease can result in death of the patient within the first 2–3 days. According to S. Stoyanov this form occurs in 10% of cases.

2. Septicopyemic (or severe form) occurs also with severe septic phenomena. However, this group of patients has clinical manifestations of bone disease. Onset is acute, the temperature increases to high values (39–40°C), intoxication increases, functions of vital organs and systems are violated. Pain syndrome is sharply pronounced due to increased intraosseous pressure.

3. Local or light form of AHO is different from the previous two because of predominance in the clinical picture of local changes. Against the background of relative well-being there is a acute pain in the affected limb. After 2–3 days of lesions local swelling and redness appear.

Persistent symptoms at AHO are pain in the affected organ, fever, swelling of the soft tissues of the affected limb. In most cases, there is a myogenic pain contracture, arthritis. Analgesics don't relieve pain, which differentiates it from pain of other origins. Identifying soreness in intramedullary disease stage (1–2 days) is advantageously carried out by palpation and percussion of the affected bone. Soft tissue swelling at the level of center occurs on the 2nd–3rd day. Hyperemia, floating appear at later periods of the disease.

Already at early stages leukocyte shift to the left is marked, increasing the number of neutrophils, stabs and young forms. Is noted increase in ESR — 25–60 mm/h or more. C-reactive protein level increases dramatically. Hypochromic anemia develops.

First radiographic signs of AHO as osteoporosis and spotted linear periostitis appear depending on the age only on the 2nd–4th week of disease (Fig. 1.6.11, 1.6.12).

The most informative and accessible method of early diagnosis of AHO should be considered bone marrow puncture. Upon receipt of pus diagnosis is not in doubt. In other cases, the results are evaluated by cytological studies. Through the needle of Waldman they measure intraosseous pressure. Pressure increase by more than 150 Mm Hg (at a rate of 60–80 Mm Hg) indicates osteomyelitis. The meth-

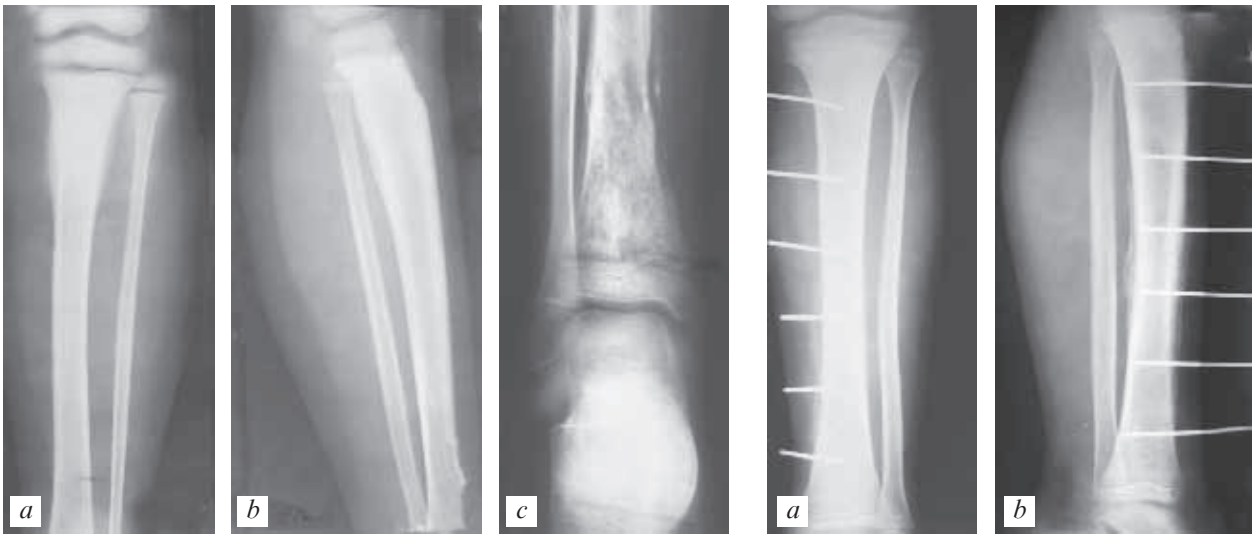


Fig. 1.6.11. Early radiographic signs of AHO: linear periostitis, osteoporosis *(a–c)*

Fig. 1.6.12. Linear periostitis, bone osteoporosis, needles drainage by Aleksyuk *(a, b)*

od is converted from diagnostic into therapeutic: through a needle or needles conduct focus decompression and conduct intralesional antibiotic therapy.

Most often it is necessary to conduct a *differential diagnosis* between AHO and trauma, as well as between purulent diseases of soft tissues, rheumatism and tumors.

From the moment of diagnosis the patient with AHO requires urgent rational and comprehensive treatment. We must begin to treat the disease within the first 2 days (in intramedullary stage of the process); thus we manage to cure up to 95% of patients.

Complex treatment of AHO is based on T. P. Krasnobaev's principles (1939):

1. The impact on the inflammatory focus

The surgical procedure volume at AHO depends on the phase of inflammatory process. Decompression of intraosseous focus is advantageously carried out by administering constant needles, constructed by K. L. Aleksyuk to the affected segmen (the needle with diameter of 2–2.5 mm, has lateral openings, and its end is designed as a drill) that allow not only gentle bone drainage, but also injecting antibiotics, antiseptics, inhibitors of proteolysis and othersdirectly into the inflammatory focus. The necessity of local antibiotic therapy at AHO is caused by the pathogenesis of the disease — because of local disturbances of blood supply only intraosseous injections of drugs are able to provide their local therapeutic concentration. Duration of intraosseous antibiotic therapy ~ 3–4 weeks, preparations are changed every 7–10 days according to the antibiogram (Fig. 1.6.13–1.6.15).

Fig. 1.6.13. Aleksyuk needle (K. P. Aleksyuk, 1969)

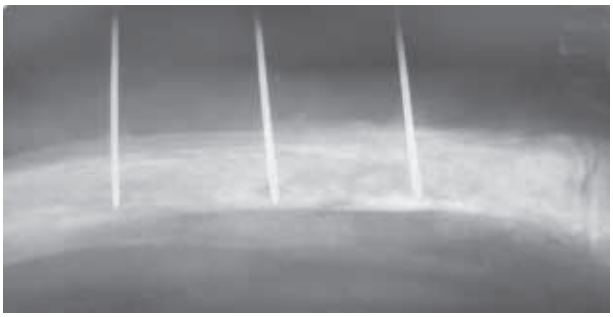


Fig. 1.6.14. Bone drainage with Aleksyuk needles

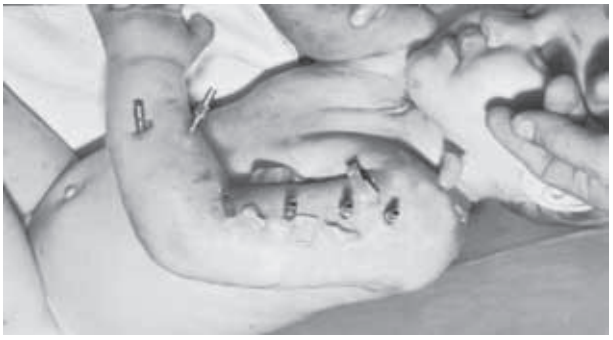


Fig. 1.6.15. Local treatment of acute hematogenous osteomyelitis: bone drainage with Aleksyuk needles, phlegmon autopsy

At subperiosteal phlegmon or phlegmon of soft tissues they make sparing cuts (up to 2 cm) and drainage is carried out; in case of arthritis in kids they are punctured, in older children perform microdraining or sparing arthrotomy.

The affected limb is applied with a plaster tire. Fixation aim is not only to ensure the rest of the affected segment of osteomyelitis, but also the prevention of complications such as pathological fracture, dislocation, for which deep plaster tire is made or they use functional means — stretching over the cuff, over "plaster boot", handling devices, at lesions of the hip joint in children — stirrups, cleol traction.

2. Impact on microorganism

Impact on the microorganism is carried out primarily with antibiotics.

In the first 7–14 days of the disease, i. e. during septicemia, antibiotics are advantageously introduced in combination: intramuscularly, intravenously, in the inflammation focus (intramedullary through previously injected needles), and after the elimination of septic phenomena only in the center. Such management is pathogenetically justified and provides antibiotics contact with infection, both in the blood and in the center.

Intralesionally, antibiotics are injected into the needle (needles) once a day at a daily dosage. Preference is given to osteotropic antibiotics. The course of treatment is 3–4 weeks. The main criterion in the selection of antibiotics is the degree of microflora susceptibility.

3. Impact on macro-organism

Defining the nature of the common treatment of patients with AHO pathogenetic components of diseaseshould be considered: a) immune deficiency — "immunity breakout" (S. Y. Doletsky, V. I. Pronyakov); b) pyosepticemia with focus localization of inflammation in the bones; c) sensitization of the organism; d) phenomena of toxicity, and often septic shock with differend degree of homeostasis disruption. Condisdering the fact, a proper attention in this period should be paid to increased immunoreactivity of the organism by replacement therapy.

For this purpose, taking into accont the causative agent, they use antimicrobial hyperimmune plasma, gammaglobulin, direct transfusion of blood from immunized donors (parents).

In accordance with the age of patients they appoint desensitizing therapy (pipolphenum, suprastin, diphenhydramine, and others.).

For the purpose of detoxification, children receive low molecular weight dextran (sorbilact, reopolyglukine, neokompensan et al.), vitamins C, B1, B6, cocarboxylase, ATP and others. At AHO with symptoms of toxic shock, it is advisable to use a short course of corticosteroids. Patients with severe forms of AHO

need regular monitoring of homeostasis and its correction.

Metaepiphyseal osteomyelitis (MEO) occurs mainly in children less than 2 years of age, in this group infants are mostly affected. Etiopathogenesis of the disease is somewhat different from AHO of older age. It is expressed in the fact that before the ossification of epiphyseal sprout zone of babies morphofunctionally immature, chondroblasts are randomly distributed, their ordering occurs in parallel Epiphysis lesion (from the center of growth in the direction of the periphery) and in accordance with the growth zone is gradually acquiring barrier properties. That is why, in the absence core due to morphofunctional immaturity of the growth zone inflammation penentrates it in the center, with the appearance of core penentration occurs in the peripheral part of the growth zone, and under conditions of ossification of epiphysis largest part Epiphysis lesion orderly growth zone performs barrier functions and inflammation spreads from the older not to epiphysis but to diaphysis. In older children mature sprout zone is a barrier to the inflammatory process.

Epiphysis lesion entails the destruction of the latter, the involvement in the pathological process of the joint and the occurrence of arthritis, which in turn leads to a pathological dislocation and paraarticular phlegmon. Discredited sprout zone partially or completely dies and further it causes the appearance of acquired malformations (shortening or lengthening, limb deformities); destruction of epiphyses leads to defects in the articular ends, the formation of destructive dislocations, and non-stability of joints.

By *clinical course* MEO has its own characteristics:

a) Most frequently purulent process is localized in femoral metaepiphysis, humerus metaepiphysis and shin bone metaepiphysis

b) MEO most frequently leads to severe deformations and bone growth

c) MEO rarely becomes chronic

g) MEO is always accompanied by arthritis. Disease in most cases begins acutely

In infants toxicosis is indicated with violation of the general condition of the child, increased body temperature, lethargy, pallor, and sometimes jaundice, enlarged liver and spleen, dyspepsia. Equivalent to pain relief is anxiety, which is amplified at moving, changing diapers. Affected limb takes forced position of "pseudoparesis" type, active mobility is sharply reduced, passive movements cause concern of the baby and cry, on 2nd–3rd day from the beginning of the disease is determined para-articular tissues pastosity, later — their swelling, hyperemia that is caused by metaepiphyseal localization of pathological process with involvement in inflammation of the joint and development of a key feature of AHO in babies — arthritis (Fig. 1.6.16, 1.6.17).

Early X-ray signs (3–10 days) include: thickening of soft tissues at the level of destruction, dilatation of the joint space, fuzzy or blurry contours of the pineal gland (Fig. 1.6.18, 1.6.19).

Differential diagnosis should be made with pyoinflammatory diseases of soft tissues of the extremities, limb injuries, rheumatism, generic paresis and paralysis of the limbs.

Treatment of children with MEO is carried out on the general principles of complex treatment of inflammatory diseases in infants and young children. However, it should be noted that at metaepiphyseal process localization should be done not only puncture decompression of affected metaphysis but also puncture from affected joint.

Great importance in the complex treatment of MEO has rational immobilization of the affected limb. For this purpose, depending on the age of the patient and the localization process, they use stretching on Schede, Dezo bandage, Vilenskiy spacer, different casts. At the Department of Pediatric Surgery

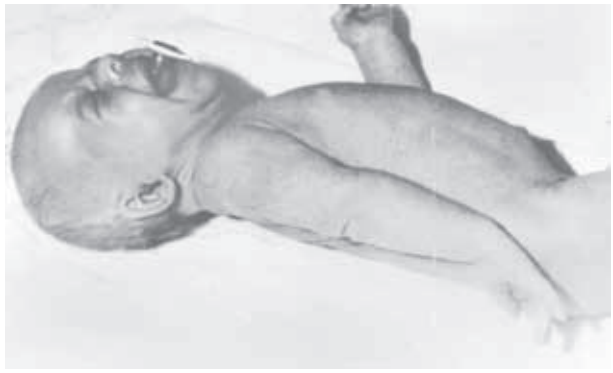


Fig. 1.6.16. "Pseudoparesis" at metaepiphyseal osteomyelitis

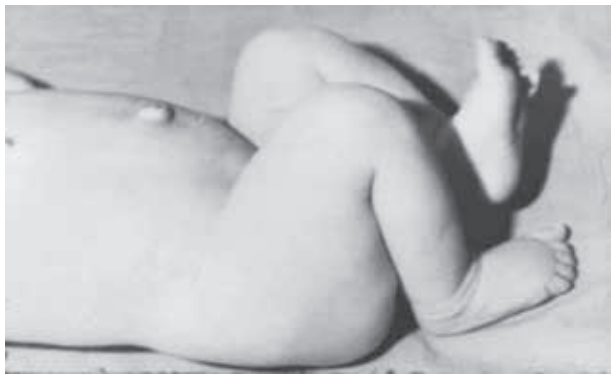


Fig. 1.6.17. Flexion contracture at hip metaepiphyseal osteomyelitis

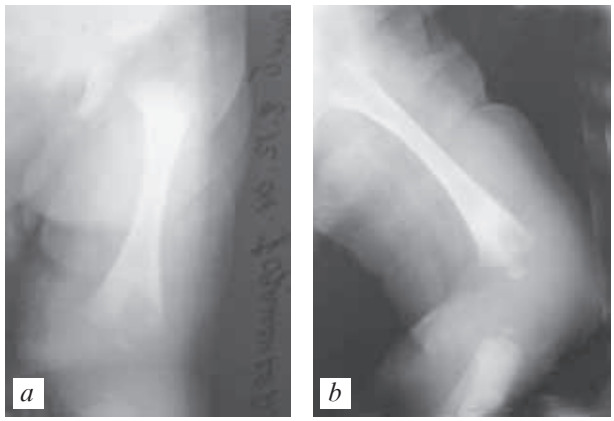


Fig. 1.6.18. X-ray diagnostics of metaepiphyseal osteomyelitis *(a, b)*

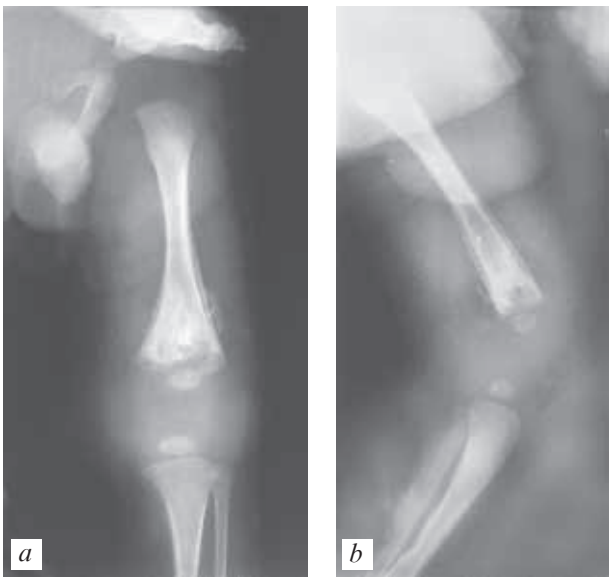


Fig. 1.6.19. X-ray diagnostics of metaepiphyseal osteomyelitis *(a, b)*

ONMedU for this purpose are developed original machines that are manufactured individually on prosthetic factory.

Patients who have had MEO must be followed up by a pediatric orthopedics for a long time. During this period of rehabilitation patients with epiphyses defects, germ plates need a long time (months or even years) to ensure the unloading of the affected area with the help of orthopedic tires, splints, articulated vehicles.

Chronic osteomyelitis is divided into chronic hematogenous osteomyelitis (primary and secondary) and chronic post-traumatic (exogenous) osteomyelitis (gunshot, burns, post-operative).

Secondary chronic hematogenous osteomyelitis (SCHO) should be considered as a complication of acute osteomyelitis. Most often, chronic osteomyelitis is a consequence of advanced forms of acute osteomyelitis, treatment of which was started in the late periods or performed poorly.

The transition of acute osteomyelitis in chronic is characterized by a marked improvement in the general condition of the patient, the disappearance of septicopyemia signs, pain in the affected area, persistent decrease in the temperature to subfebrile, partial recovery of limb function and others. Locally, in the affected area is noted sluggish inflammation with the presence of constantly functioning or sometimes closing fistulas, which are supported by large or small sequester (osteonecrosis) (Fig. 1.6.20).

In remission period the condition of patients may be satisfactory, and the disease does not manifest itself. During an exacerbation patients condition is deteriorating: disturb pains in affected limb, the body temperature rises, grow signs of intoxication. In the affected area appear infiltration, swelling and redness of the skin. Pus discharge starts from fistula, limb function is broken. Blood picture reflects the inflammatory process. Periods of remission alternate with exacerbations. Frequent occurrence and the presence of purulent focus often leads to the development of degenerative changes in kidneys, liver, myocardium, and sometimes to amyloidosis of the internal organs. In the diagnosis of chronic osteomyelitis X-ray examination is of paramount importance.

In the affected bone is X-ray detected abdominal formation, sequestrations, sclerosis and bone eburniation. During exacerbation appears periostitis.

All patients with fistulous form of chronic osteomyelitis make fistulography. It allows you to specify the amount of the pathological process in the bone and soft tissues.

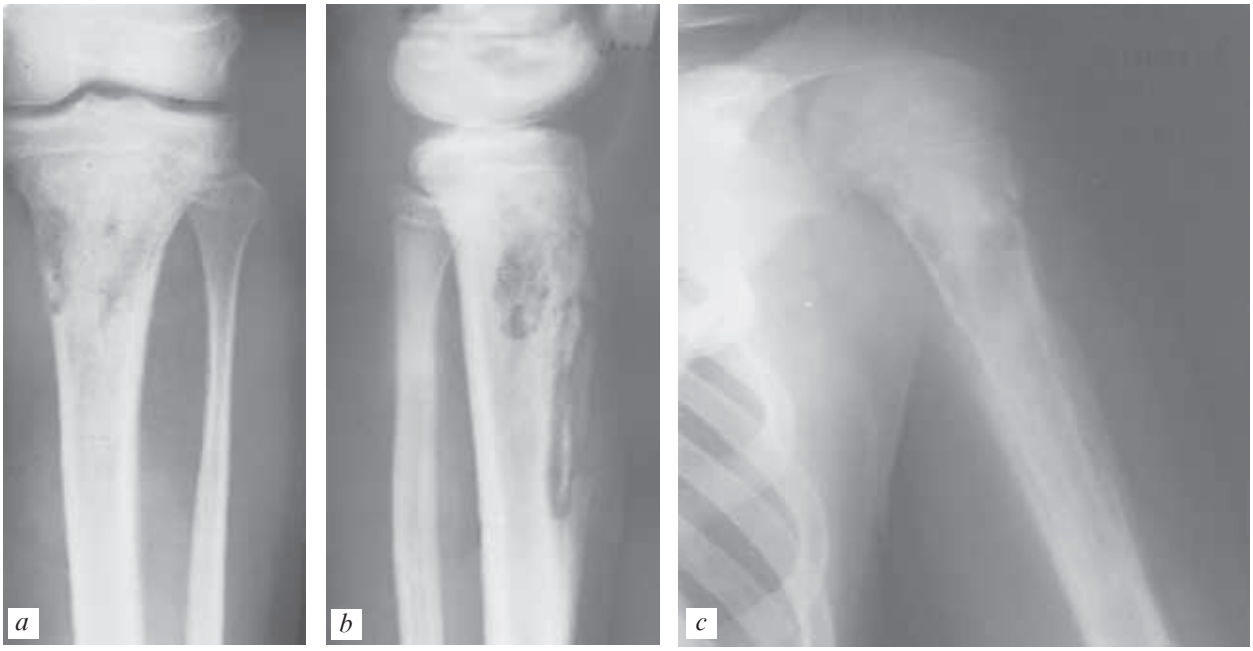


Fig. 1.6.20. X-ray at chronic osteomyelitis *(a–c)*

Primary chronic hematogenous osteomyelitis (PCHO) develops gradually, does not have the acute phase and is characterized by subacute course. The reason for this form of development is considered to be low virulence of microbes at a high reactivity of the microorganism. To primary chronic or atypical form of osteomyelitis include *Brodie abscess, Garre sclerosing osteomyelitis,Olie albuminous osteomyelitis, Popkirov antibiotic osteomyelitis and tumor osteomyelitis.*

At Brodie abscess occurs long asymptomatic anamnesis and there is no pronounced clinical picture — there is only a moderate swelling and pain over the lesion center.

X-ray presents focus of enlightenment, localized in the metaphysis, tibial, humerus, and femur bone. Focus form is rounded or elliptical; its perimeter is defined by a clear sclerotic rim. The cavity is usually "empty", does not contain any impurities. Nearby areas of the bone are not changed. Periosteal reaction is absent or poorly developed (Fig. 1.6.21–1.6.23).

Albuminous Olie osteomyelitis arises due to the fact that a weakened form cannot convert existing at the beginning of the disease protein-rich exudate in the pus. Teenagers are susceptible to disease more often. The process is usually localized in the distal part of a hip, during the development of destruction is accompanied with severe exudative reaction in periosteal tissues. Patients complain of persistent pain in the hip, then swelling joins gradually, increasing during 1–1.5–2 months. Rarely flushing of the skin appears. X-ray reveals regular and irregular cavernous formation with periosteal layers.

Popkirov antibiotic osteomyelitis may occur in patients during treatment with antibiotics of starting inflammatory process in the bone. It flows sluggishly, without pronounced pain, hyperthermia and intoxication. Laboratory tests also do not have any significant changes. Only moderate leukocytosis and increased ESR occur. Exudation processes, destructions and proliferations are not pronounced. Periosteal reaction can be mild or absent. Parallel to the development of destruction focus in the bone and the formation of small cavities with sequester are noted previous sclerosing.

Garre sclerosing osteomyelitis begins subacute and is characterized by pain in the limbs, especially at night, in violation of its function and mild fever.

X-ray signs of the disease are characterized by a pronounced fusiform thickening of bone diaphysis, against which may be visible lesions of diluting and they contain small sequestrums. Sclerosis has homogeneous character, bone structure is absolutely not revealed. Medullary canal is narrowed or completely uniformly obliterans. Periosteal reaction can only be determined in fresh cases, further it becomes invisible.

Tumor-like PCHO also does not have the acute stage of development, is revealed in 6–8 months after the onset of the disease due to the connection of functional disorders (lameness) or an increase in the limb size in affected area. Clinically moderate increase in local temperature may be determined. X-ray picture simulates osteoclasts, Ewing's sarcoma or eosinophilic granuloma. Crucial in the differential diagnosis is puncture biopsy.

The term of traumatic osteomyelitis combines various forms of inflammatory and necrotic processes, encountered by exogenous way in bone damage area (after open fractures and gunshots or after osteoplastic operations).

Treatment of chronic osteomyelitis (as well as of acute) is complex.

Impact on the focus during the exacerbation involves intralesional antimicrobial therapy (using reg-

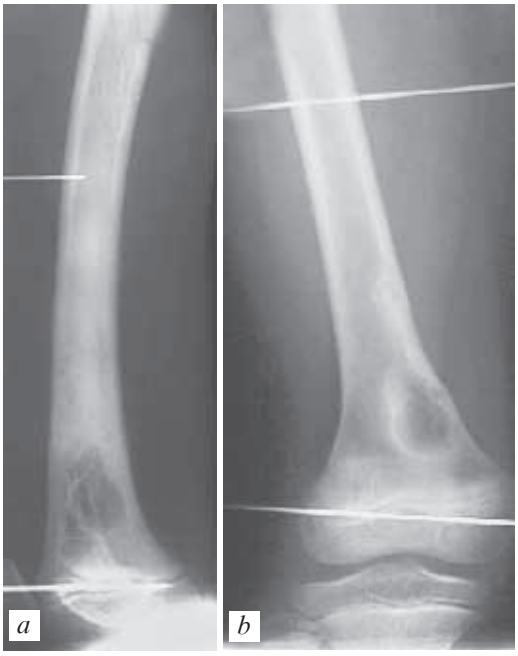


Fig. 1.6.21. Brodie abscess *(a, b)*

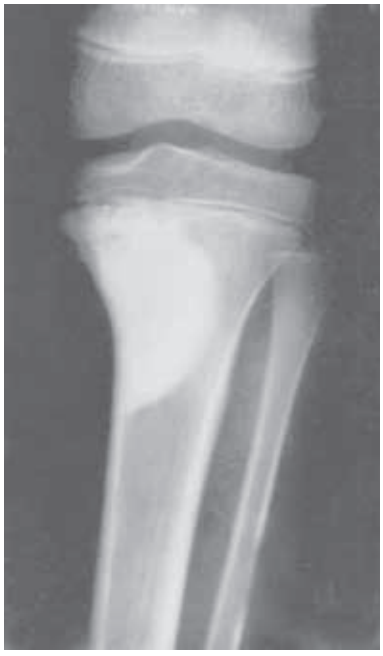


Fig. 1.6.22. Garre osteomyelitis

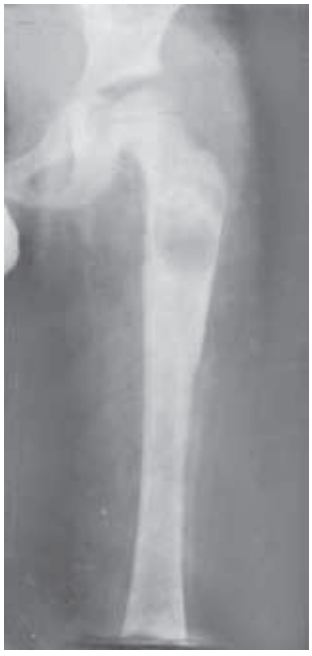


Fig. 1.6.23. Tumor-like osteomyelitis

ular Aleksyuk needles or catheters), if necessary — dissesion of subperiosteal or intermuscular abscesses.

In most cases, conservative treatment is the stage in preparation for radical surgery.

Surgical treatment is subject for all kinds of chronic osteomyelitis. Surgery involves radical necrectomy, postoperative cavity sanation (washing with antiseptics, laser treatment), multiple perforations of sclerosed bony walls, plastic of bone cavities with autologous bone marrow. Said automaterial is subjected to ultraviolet light, which greatly increases the osteogenic potential of bone-marrow progenitor cells.

Basic principles of children rehabilitation are based on complex sanatorium treatment.

QUESTIONS FOR THE FINAL CONTROL

1. When does the formation of long tubular bones epiphyses end and what are criteria of confirmation.

2. What is the normal intraosseous pressure?

3. What are the characteristics of diagnostic puncture and measurement of intraosseous pressure in children with suspected acute hematogenous osteomyelitis.

4. Features of long tubular bone structure.

5. What are the clinical forms of acute osteomyelitis.

6. Why does in infants and children up to 2 years of life occur metaepiphyseal lesion.

7. Name atypical forms of osteomyelitis.

8. What is the most common symptom of metaepiphyseal osteomyelitis.

9. Features of joints puncture in infants and how often they are held.

10. Features of immobilization and its duration in children with metaepiphyseal osteomyelitis.

11. List the complications that are associated with hematogenous osteomyelitis, and when they appear.

12. When is sanatorium treatment shown.

13. What are the most common surgical interventions at chronic osteomyelitis in children.

Practical Skills

1) Interpretation of acute hematogenous osteomyelitis X-ray.

2) The methodology of bone punctures.

3) The methodology of joints punctures.

PYOINFLAMMATORY DISEASES OF SOFT TISSUES

Stages of urgent medical measures at chronic inflammatory diseases of soft tissues. Toxic course patterns of inflammatory diseases of soft tissues and the development of septic complications. Necrotic abscess in newborns, mastitis, omphalitis, furunculosis, pseudofurunculosis. Lymphadenitis, adenophlegmon, mumps. Paraproctitis. Etiopathogenesis, classification, clinical features, diagnosis, treatment.

Specific objectives:

1. Master the list of inflammatory diseases of soft tissues, typical for newborns, developmental stages of sepsis.

2. Main clinical manifestations of chronic inflammatory diseases of the skin and subcutaneous tissue.

3. Conduct differential diagnosis of inflammatory diseases of soft tissues in children.

4. Interpret auxiliary diagnostic methods: thermometry, ultrasound, X-rays, blood tests, urine, cytology and microbiology testing.

5. Features of inflammation symptoms in newborns (incidence of process, fluctuation, inflammation of the navel, breast, etc.).

6. Analyze cause-and-effect relationship of inflammatory diseases occurrence in newborns, the development of severe forms.

7. Learn the sequence of physician actions in ordinary and toxic forms of chronic inflammatory diseases of soft tissues.

8. Familiarize with general principles of inflammatory diseases treatment of soft tissues.

1. Actuality of the problem

A. The rate of chronic inflammatory diseases of soft tissues in infants who require treatment in hospital makes up 30–50%. Among them, most frequently occur: phlegmon of newborns — 13%, omphalitis — 17%, mastitis — 11% paraproctitis — 6%.

B. Anatomical and physiological characteristics of skin and subcutaneous tissue, breast, navel formation and its blood supply.

2. Stages of urgent measures at chronic inflammatory diseases of soft tissues are shown in Table 9.

Due to the nature of skin structure and other soft tissues in the newborn, many purulent-inflammatory diseases gain such features that make the clinical picture of the disease completely different from that observed in older children and adults, causing the need for special diagnostic and treatment methods. Some of the inflammatory diseases can occur only at the neonatal period. One of these diseases — **necrotic abscess of newborns**.

At necrotizing phlegmon of newborns inflammatory changes in subcutaneous tissue are accompanied by necrosis, necrosis and rejection of the skin. The disease progresses rapidly, lesion area increases every hour, which leads to the development of sepsis. Areas of the skin that are most commonly affected: interscapular, lumbar region, chest and upper limbs. First, on skin appears a small area of hyperemia and edema, the skin is sealed. Lesion area increases very quickly, by the end of the first day of the disease the skin redness can capture an entire anatomical region. In the center of congested skin a recess portion is defined, which soon becomes cyanotic, and there are small fistulas with sero-purulent discharges. Inflammation of the subcutaneous fat causes throm-

Table 9

Stages of Urgent Measures at Chronic Inflammatory Diseases of Soft Tissues

Stage	Measures	Medical means and interventions
1	Adequate antibiotic therapy	Empirical approach — protected penicillin, cephalosporins of 3-4 generation, aminoglycosides
2	Detoxification, stimulation Vitamin therapy Physiotherapy events	Infusion therapy, plasmapheresis, immunotherapy. Vit B1, Vit B6 Quartz, magnetic and laser therapy, HBO
3	Surgeries phlegmon of newborns purulent mastitis	1–1.5 cm dissections at the site with healthy skin; in the center in a staggered manner. Bandaging after 4–6 hours. Radial dissections up to 1.5–2 cm, from the halos. Drainage.
	omphalitis: a) simple form b) phlegmonous c) necrotic	Decrease of exudation, sanation with hydrogen peroxide 3%. Sanation and drainage of the navel. Dissection up to 1–1.5 cm on necrosis site, drainage.
4	In the absence of effect and development of toxic forms	Therapies as in sepsis treatment.

bosis of vessels passing through the tissue that gives skin perfusion, and on the 3rd–4th day of onset necrotic skin begins to form and is rejected with formation of wide wound defect. Necrosis may extend to underlying muscles, cartilage, and bones (Fig. 1.6.24).

Depending on the severity of child's general condition they distinguish two forms of necrotizing phlegmon of newborns: toxic-septic and simple. In the first form pronounced symptoms of intoxication may appear before any changes in the skin. The general condition of the child within the first hours of the disease is regarded as a very severe: changes in child's behavior, he is restless in the beginning of disease, and then — lethargic, not eating, fever (39–40°C), there is vomiting, diarrhea, growing leukocytosis, phenomena of exsicosis and intoxication. Without intensive therapy this form in 2–3 days can result in death. At a simple form of the disease, which is less common, symptoms of the disorder in general condition, inflammatory changes, and intoxication are increasing gradually, and come to the local changes in the skin and subcutaneous tissues.

Differential diagnosis of necrotizing phlegmon in newborns is conducted primarily with erysipelas, distinctive symptoms of which are: clear edges of hyperemia (most often on the face, perineum, navel), which are distributed in the form of "tongues of fire", skin is edematous, with smooth surfaces with no cavities and areas of necrosis (see Fig. 1.6.24).

Complex treatment of necrotizing phlegmon in newborns includes surgery, antibacterial, immune detoxification, restorative therapy. Feature of the surgical treatment of this disease is the need to perform a variety of cuts, up to 1 cm over the entire surface of hyperemic skin to adjacent areas of unchanged skin. Incisions are made in a staggered manner with a distance of about 2 cm between them. Serosuppurative or sero-hemorrhagic fluid allocates from cuts, pieces of necrotic subcutaneous tissue. Wounds are washed up with an antiseptic solution and are covered with a wet bandage with a solution of antibiotics, drainage is not set. After applying the cuts every 3–4 hours it is necessary to carry out inspection of the affected area of the skin. If hyperemia continues to spread, additional cuts are applied. Application of a wide incision across the entire surface of hyperemia is incorrect treatment management and does not stop the progress of inflammation.

With the absence of necrotic changes of the skin incisions heal within 5–7 days, if there is necrosis of the skin it is necessary to carry out treatment for the

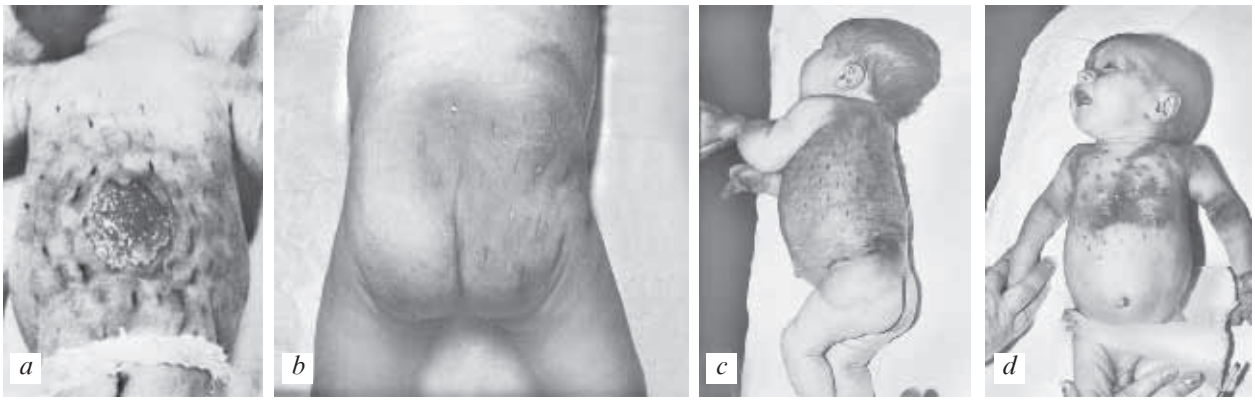


Fig. 1.6.24. Newborns phlegmon *(a-d)*

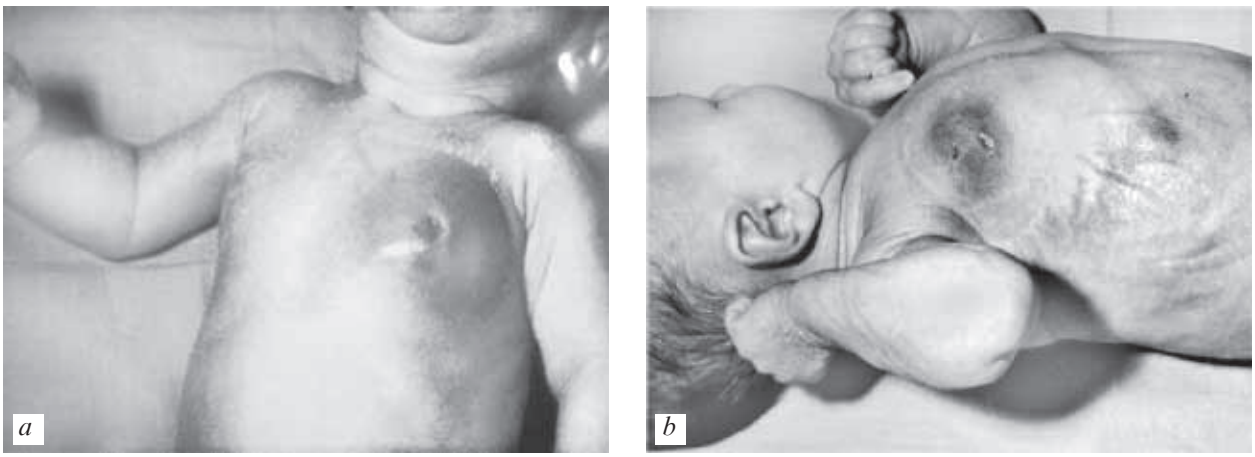


Fig. 1.6.25. Newborns mastitis *(a, b)*

rejection of dead parts. Formed defects of the skin, the area up to 25 cm^2, are covered with granulations and self epithelize. Therapy in such cases is aimed at stimulation of reparative processes and the prevention of secondary infection. At broader skin defects they indicate the use of various plastic methods.

One of the most common pyoinflammatory diseases in newborns is **mastitis**. The disease most often develops within the first two to three weeks of life and manifests in general and local inflammatory changes. ***Common symptoms*** of inflammation include violation of the general condition, anxiety, loss of appetite, increase in temperature to 38–39°C, inflammatory changes in the blood count. Locally increased, swollen one breast, skin hyperemia, local temperature rise, increased anxiety at palpation, which also detects the presence of a dense infiltrate in the projection of the prostate. With the progression of inflammation suppuration of infiltrate occurs, tenderness increases, softening and fluctuation parts appear. At late start of treatment the disease can take septic character with the transition of inflammation and necrotic changes in the tissue located near the chest. Mastitis coincides with physiological breast engorgement (mastopathy of newborns). To avoid diagnostic errors, it must be remembered that at physiological breast engorgement general condition of the child is not violated, the skin over the glands is not changed and the process is usually reversible. At mastitis on the stage of infiltration conduct ***conservative therapy*** (antibiotic therapy, local anti-inflammatory therapy, physiotherapy) (Fig. 1.6.25).

At the stage of suppuration *operative treatment* is indicated. Incision over the area of softening, 1–1.5 cm long, is carried out in the radial direction from the areola. Wound is drained by rubber strip, superimposing wet antiseptic bandage. At propagation of inflammation and necrotic changes beyond the breast additional cuts are performed. With prompt and adequate treatment function of the breast in the future does not suffer, but the postponement of surgical treatment at the stage of suppuration may cause purulent fusion of the breast anlage.

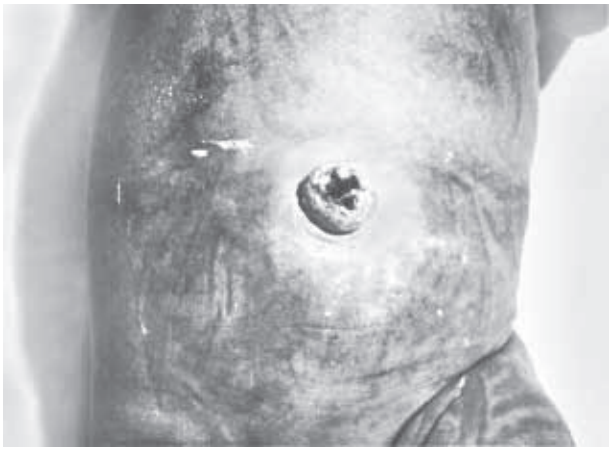


Fig. 1.6.26. Omphalitis

Inflammation of umbilical wound in the newborn is called **omphalitis**. The danger of this disease is that the inflammation may pass to the surrounding soft tissue by umbilical vessels in the peritoneal cavity with a development of contract peritonitis. Umbilical wound infection may become a source of inflammation in other organs and tissues, such as metaepiphyseal osteomyelitis, lead to sepsis.

In simple form of omphalitis general condition of the newborn is not violated, but the wound does not heal, is marked scant serous or sero-hemorrhagic discharge. Local treatment in a simple form of omphalitis (umbilical wound is treated with solutions of hydrogen peroxide and brilliant green several times a day) quickly leads to the regression of inflammation and wound healing (Fig. 1.6.26).

Phlegmonous form of omphalitis is characterized by hyperemia in navel area, in the center of which there are fibrinous deposits, covering the wound. Pus is oozing from the wound, the general condition of the child may suffer, which is manifested in anxiety, decreased appetite, and fever. At this stage of disease they indicate local treatment (bandages with an-

tiseptic solutions, physiotherapy — UHF, UFO), is appointed antibiotic therapy, restorative, detoxification and immune therapy is carried out.

The most severe form of omphalitis — *necrotic*, is characterized by widespread inflammation and necrotic changes over the surrounding tissues, abdominal cavity, and umbilical vessels. At this form skin necrosis can result in bowel loops eventrations, contsct peritonitis, liver abscess can develop. Condition of children is regarded as severe or very severe, there may be signs of sepsis. Umbilical vein thrombophlebitis can spread to the portal vein, which leads to stenosis and its development in the older age of portal hypertension syndrome. At necrotic form of omphalitis intense local and general treatment is supplemented by surgery: on the surface of all affected skin around the navel they apply cuts-notches, the edges of the wounds are bred, wash the wound with an antiseptic solution and draine with rubber bands, providing outflow of pus and necrotic changes in the tissues. Bandages are held daily, rubber bands are removed in a day — two, after which against the general and local anti-inflammatory therapy sores quickly epithelize.

Sometimes at symptoms of a simple form of omphalitis (serous discharge from the navel), despite long-term therapy, the amount of discharge is not reduced, but the inflammatory process is not progressing. In these cases, you need to think about such diseases as navel fistulas, which are formed during incomplete imperforate of embryonic ducts (bile duct and urachus). Children with suspected navel fistula need examination in a surgical hospital. At confirmation of diagnosis planned surgery is indicated.

Another quite common purulent disease of soft tissues in newborns — is **pseudofurunculosis**. Unlike furuncle, when the hair follicle inflames, at pseudofurunculosis inflammatory process affects sweat gland, inside of which a small abscess forms. Most often the process is localized on the occipital region of the head, scapular region of the back, buttocks. Multiple abscesses, the size of a few millimeters, located under the skin, the skin over them is hyperemic. Microabscesses may coalesce with the development of an extensive phlegmon of the subcutaneous tissue. The general condition of the child at pseudofurunculosis is determined by the size of the affected area. At extensive lesions child's behavior may alter, there is anxiety, fever, decreased appetite. Pseudofurunculosis treatment consists in opening of each microabscess with peaked scalpel and closing the wound surface with wet bandages with an antiseptic solution. Wound draining is not necessary. Antibiotic therapy is appointed.

Paraproctitis — an inflammation of perirectal tissue, most often is diagnosed in children within the first year of life. Pecularities of anatomical structure, often and easily developing diaper, rashes, scratches, furuncules and pyoderma of perianal area contribute to infection of perirectal tissue. Great importance in the development of paraproctitis in children is paid to the possibility of hematogenous or lymphatic introduction of infection. Of all possible locations of purulent process in perirectal tissue (subcutaneous, submucosal, ischiorectal, pelviorectal, retrorectal, paraproctitis) in children of 1 year old subcutaneous is often revealed. Acute paraproctitis is manifested in child's anxiety, which is enhanced at swaddling and defecation, the temperature rises to 39–40°C, the child refuses to eat. On examination hyperemia and swelling around the anus are determined, palpation of the area and a rectal examination help to clarify the location and extent of the inflammatory process. In acute paraproctitis over the place of the greatest fluctuations perform incision of crescent-shaped, up to 2 cm, by 1–2 cm from the anus. With blunt way revision of abscess cavity is made, its washing with antiseptic solution and drainage. Postoperative treatment is carried out by the general principles of treatment of pyoinflammatory diseases in children. Local treatment involves washing up the wound with antiseptic solutions and hip baths with the same solution 2–3 times a day.

In chronic paraproctitis, in addition to presence of inflammation in perirectal tissue navel fistula is formed, connecting the abscess cavity with skin of perianal area (external fistula) or rectal mucosa (internal fistula). If fistula holes are opened and on the skin and mucous, it is full adrectal fistula. In relation to the external sphincter fistula can pass inside of it, through its fibers, or outwards from the sphincter.

The first symptoms of chronic paraproctitis are similar to those in acute process. Often, chronic inflammation, especially in infants, is a complication of acute paraproctitis. For the examination the child should be prepared with the help of enema. On examination, we can determine the presence of fistula external opening with a dense structure, coming inside from the hole. At pressing a drop of pus is typically allocated. To identify the internal opening they use rectal examination (often fistula is located on the border of skin and mucosa of the rectum), injection of colorant material into exterior orifice with visual inspection of its release on the mucous membrane of the rectum, sensing the external opening of fistula. At complex adrectal fistulas or their high location they use fistulography and sigmoidoscopy. In chronic paraproctitis operative treatment is indicated, which is the complete excision of fistula motion with all its branches and pathological changed tissues in the course of fistula. Method of surgery depends on the location of fistula, its structure and length.

Lymph nodes are filters, stopping microbes and their toxins, which leads to a high incidence of **lymphadenitis** in children. The source of infection can be pyoderma, furuncle, toothaches, sore throat, and stomatitis. The most frequently submandibular, cervical, axillary and inguinal lymphadenitis is detected. During nonspecific lymphadenitis course two

stages are distinguished: the stage of infiltration and abscess formation stage. The first symptoms of lymphadenitis are increase in temperature, lymph node increase; it is tight, painful on palpation. At suppuration swelling of surrounding tissues, skin hyperemia, and fluctuation appear. In severe cases purulent process may extend beyond the capsule on the surrounding soft tissues — in such cases it is adenophlegmon. At the stage of infiltration conservative treatment, which includes antibiotic therapy, physiotherapy or other thermal treatment is used. With occurrence of softening area, which indicates lymph node suppuration, operative treatment is indicated (incision and drainage of abscess). After the operation conservative therapy continues.

Chronic inflammation of lymph node continues for more than 1 month, and is usually not accompanied by inflammatory changes of the node itself (the skin over a node is not changed, palpation is not very painful or painless), there is no general temperature and inflammatory changes in the blood count. Chronic lymphadenitis is poorly treated. It must be remembered that enlarged lymph nodes may be caused by tuberculosis, lymphogranulomatosis, and other tumors. Therefore, long-term treatment failure of chronic lymphadenitis dictates the need in lymph node biopsy.

Inflammation of the parotid gland **(parotitis)** in children is rare. The infection can get into the prostate through hematogenous or ascending path of the oral cavity. The process is usually one-sided, which allows making differential diagnosis with epidparotitis. At parotitis first complaints are pain during chewing and turning the head. Parotid salivary gland increases in size, becomes dense, painful, the skin over it is hyperemic. At abscess formation general condition worsens, a slight pressure on gland is accompanied by the release of pus from the ductless glands. Treatment of parotitis begins with conservative measures (dry heat and UHF locally, antibiotic therapy). At suppuration they indicate incision over the gland considering particularities of facial nerve branches in this area. One of treatment options may be gland puncture with pus aspiration and injection of antibiotics solution to abscess cavity.

Section 2
NEOPLASMS OF TISSUES

2.1. FEATURES OF CHILDHOOD ONCOLOGY. BENIGN NEOPLASMS IN CHILDREN

Specific objectives:

1. Master the diseases that cause tumors of soft tissues.
2. Recognize the major clinical symptoms of neoplasms of soft tissues.
3. Differentiate neoplasms depending on the species.
4. Interpret the principles of treatment of soft tissues neoplasms and their complications.
5. Recognize the main clinical symptoms of bone neoplasms.
6. Identify the characteristics of the course of certain diseases on the basis of clinical and radiographic features.
7. Differentiate benign bone tumors based on clinical examination and ancillary techniques (X-ray ultrasound, computed tomography, angiography, puncture and open biopsy).
8. Make a plan of examination of the patient and the algorithm of actions to identify and prevent complications of bone tumors (pathological fractures, false joints, metastasis, etc.).
9. Interpret the principles of treatment, depending on the type of bone tumors.

Pediatric oncology — a special section of oncology, which deals with the same problems as the general oncology but considers the developing organism.

Features of pediatric oncology

1. Malignant neoplasms in children appear 10 times less in comparison with adults. However, every year in the world a quarter million of children die from cancer, and in the structure of infant mortality cancer diseases take the second place.
2. 50% of childhood oncological diseases are tumors of the blood system (oncohaematology).
3. Most common tumors occur in children occur an early age (up to 3 years) and have congenital character (nephroblastoma, neuroblastoma, medulloblastoma, gepatobastoma, retinoblastoma), the so-called embryonal tumors.
4. In children tumors of connective tissue origin (sarcomas) dominate, which account for 84%, cancer occurs in 4–6% of cases.
5. Large proportion disembryogenetic tumor (choristoma, hamartoma, teratoma, embryonal tumors) occupies.
6. In older children tumors may arise as a result of physiological or pathological enhanced proliferation (osteosarcoma, thyroid cancer, etc.).
7. Proved transplacental transfer of malignant tumors from mother to child (Peller).
8. Children with various forms of immunodeficiency suffer from common systemic cancer diseases, which proves the theory of immunological control.
9. A rather frequent combination of malformations with tumors.
10. The inheritance of certain tumors is confirmed by bilateral tumors (retinoblastoma is associated with a dominant gene, xeroderma pigmentosum, and others).
11. Spontaneous regression of some tumors is typical for pediatric oncology, not only a complete spontaneous regression (neuroblastoma, hemangioma) also is found, but also the transition of a tumor from a malignant neuroblastoma to benign ganglioneuroma.
12. Most common tumors are usually located in hard to reach places.

Oncology of childhood is very different from adult oncology by nature of the process. Much less frequently (15-folds) in children are identified malignancies of epithelial origin (cancer), tumors of mesenchymal origin (sarcoma, embryoma) mainly form. Relatively rare the internal organs (lungs, gastrointestinal tract, bladder, reproductive organs) are affected — only 10%, while in adults — 60%. Large proportions have disembryogenetic tumors, which were formed as a result of tissue defects — namely choristoma, hamartoma, teratoma, true embryonal tumors.

Choristoma — tissue aberration, ectopia — appears from the choristia — the cleavage of tissue complexes and their inclusion into the adjacent tissues (dermoid, chondroma of the lungs).

Hamartomas are formed because of excessive development of any tissue. This is hyperplastic malformation, which occurs as a benign tumor, but in adulthood there is a high percentage of its malignancy. In this regard, they can be considered as a precancerous condition. Hamartoma can be solitary (some types of hemangiomas, fibrous dysplasia) and system (angiomatosis, chondromatosis).

Teratomas (or embryomas) — congenital tumors, which occur from improper formation of the three germ layers (ectoderm, mesoderm, endoderm). Typically, only one of tissue components is immature what may cause the malignancy of the tumor and its transformation into teratoblastoma, i. e. these are ectopic tissues with blastomatosis potencies.

True embryonal tumors arise during embryogenesis from immature tissue; subsequently they proliferate at an embryonic level and lead to kidney embriosarcoma, soft tissues, hepatoblastoma, etc.

Simultaneously immature embryonal tumors can ripen and even spontaneously regress. Spontaneous regression of malignant neoplasms is called Peregrine syndrome.

In many cases there is a correlation between malformations and tumors. Approximately 30% of malformations are accompanied by fetal tumors: Wilms' tumor with hemihypertrophy, Beckwitt–Wiedemann syndrome — a combination of omphalocele, macroglossia and gigantism with nephroblastoma, adrenal cancer, hepatoblastoma, Hirschsprung's disease with neuroblastoma.

Often there is a genetic tendency to appearance of neoplasms (polyposis, chondromatosis, and chondrodysplasia). It is registered transplacental transmission of matous blasts process.

The structure of the neoplasms in children

Benign tumors in children make 65%, malignant — 20%, tumor–like formations — 15%.

Among benign neoplasms frequency of pathology is the following:

— Angioma — 40% (hemangioma — 30%, lymphangioma — 10%);

— Pappilomas, polyps — 30%;

— Bone tumors — 15%;

— Nevi, teratoma, fibroma, lipoma, dermoid — 10%.

Benign tumors — *tumor benignum* — tumors that grow slowly and can exist for years without increasing. They have their own capsule. During growth, increasing, the tumor pushes the surrounding tissues, without destroying them. Histological structure of the tumor is slightly different from the tissue in which it developed. Therefore, benign tumors bear the names of own tissues, from which they evolved, with the addition of the suffix "oma" (lipoma, fibroma, myoma, osteoma, etc.). Removal of benign tumor with a shell leads to complete recovery of the patient.

In general, benign tumors are characterized by several features:

1. The tumor cells are mature, but there is atypism in location of cells and tissue structures.
2. Tumor growth happens on expansive type.
3. These tumors progress slowly.
4. No toxic components.
5. Tumors do not metastasize.
6. The main clinical syndromes are a syndrome of "plus–tissue" and dysfunction syndrome.

BENIGN NEOPLASMS OF SOFT TISSUES

Benign neoplasms of external integuments are quite common in childhood and require urgent attention. Some of them belong to the true tumors; the other part does not and makes interest in terms of differential diagnosis.

HEMANGIOMAS

Hemangiomas — a general and non-specific term that is traditionally used to refer benign tumors from vascular tissue, vascular birthmarks and vascular malformations.

Hemangiomas are the most common tumors in children, mainly localized on skin. By their nature, these formations are closer to malformations or congenital vascular hamartoma tissue than to true tumors.

Some features of hemangiomas force to refer them to cancer processes.

In most cases hemangiomas occur from birth; rarely occur during the first months of life. Usually the tumor affects skin and soft tissues but may also affect various organs. There are cases of discoveries of hemangiomas in the brain, liver, and mediastinum.

Classification of hemangiomas:

1. Simple, or capillary, hemangiomas — consist of many capillaries, are located superficially, look like spots or spots of bright red color with sharp edges and slightly protrude above the surface of the skin (Fig. 2.1.1).

Simple hemangiomas include papillary and stellate hemangiomas, observed in children of preschool and school age. Stellate hemangiomas are the formations from extended, convoluted blood vessels, which are located in the skin, in the center of which is the main supply blood vessel. "Strawberry" hemangioma refers also to capillary, which has rapid growth and intense red color, clearly limited, rising above the surface of the skin, easily compressed (Fig. 2.1.2).

2. Cavernous hemangiomas consist of blood–filled cavities, lined with endothelium and delimited by partitions of connective tissue. Integuments may not be changed. It is marked thickening and increase of the organ. Cavernous hemangiomas are usually of considerable size and have no clear boundaries. They distinguish diffuse and encapsulated hemangiomas. The latter grow slowly, within its own capsule. Diffuse hemangiomas have rapid infiltrative growth (Fig. 2.1.3).

3. Branched hemangiomas consist of coils of convoluted and intertwined arteries and veins of different caliber, capture the deep tissues, including muscle and bones.

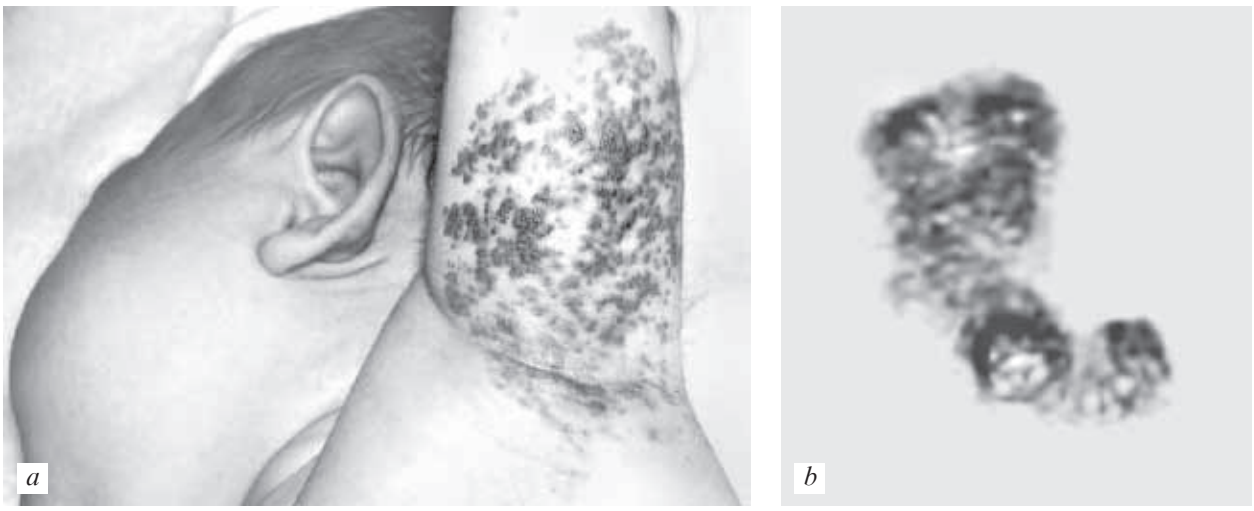


Fig. 2.1.1. Capillary hemangiomas *(a, b)*

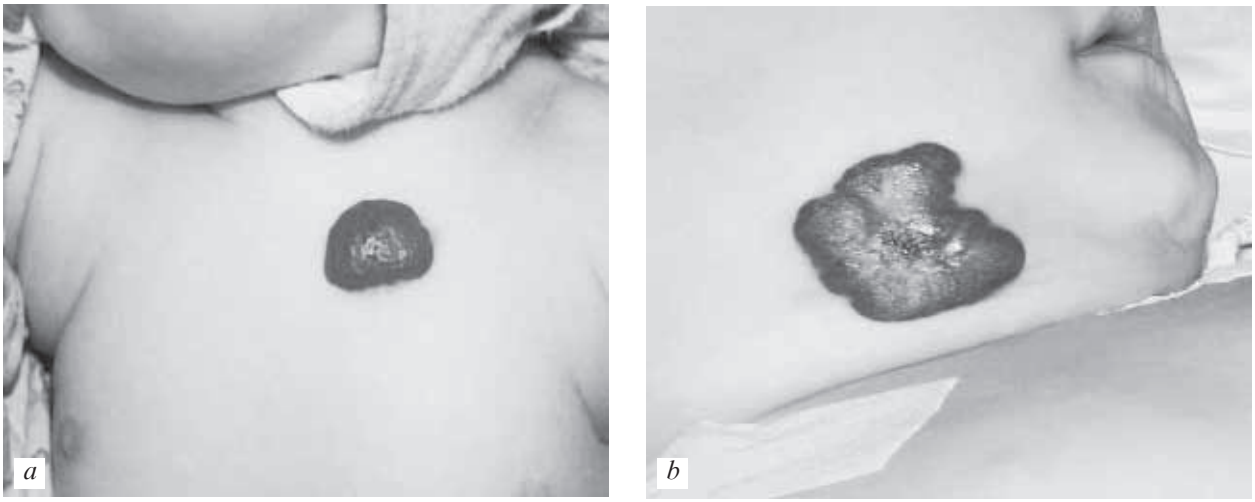


Fig. 2.1.2. "Strawberry" capillary hemangioma *(a, b)*

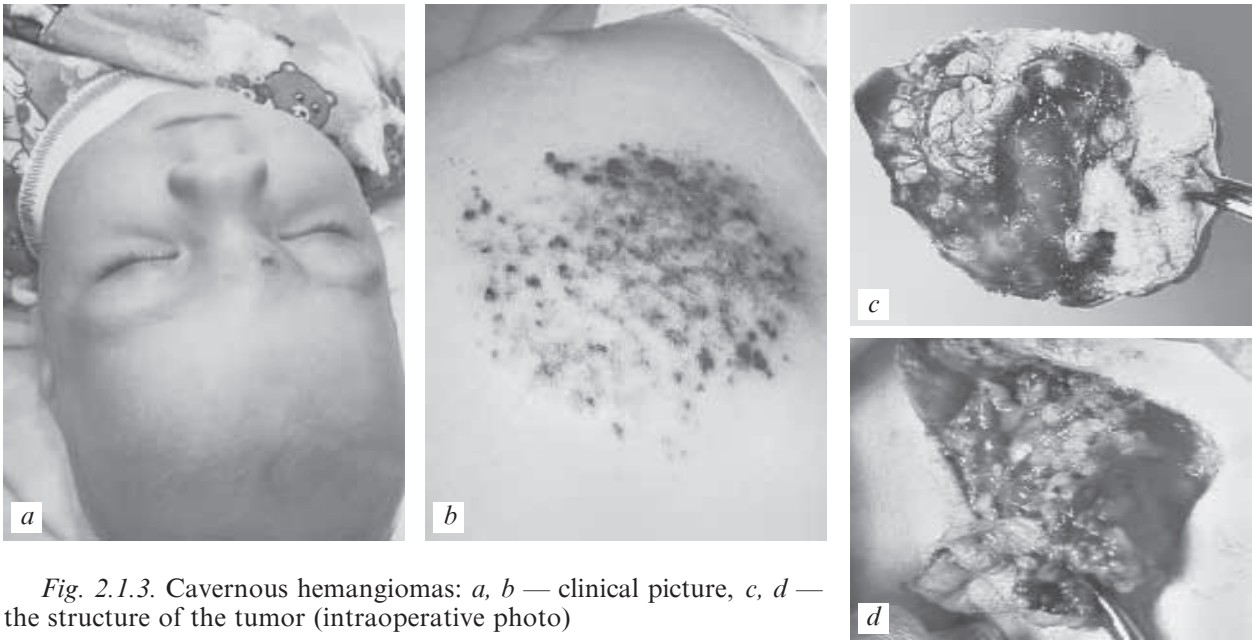


Fig. 2.1.3. Cavernous hemangiomas: *a, b* — clinical picture, *c, d* — the structure of the tumor (intraoperative photo)

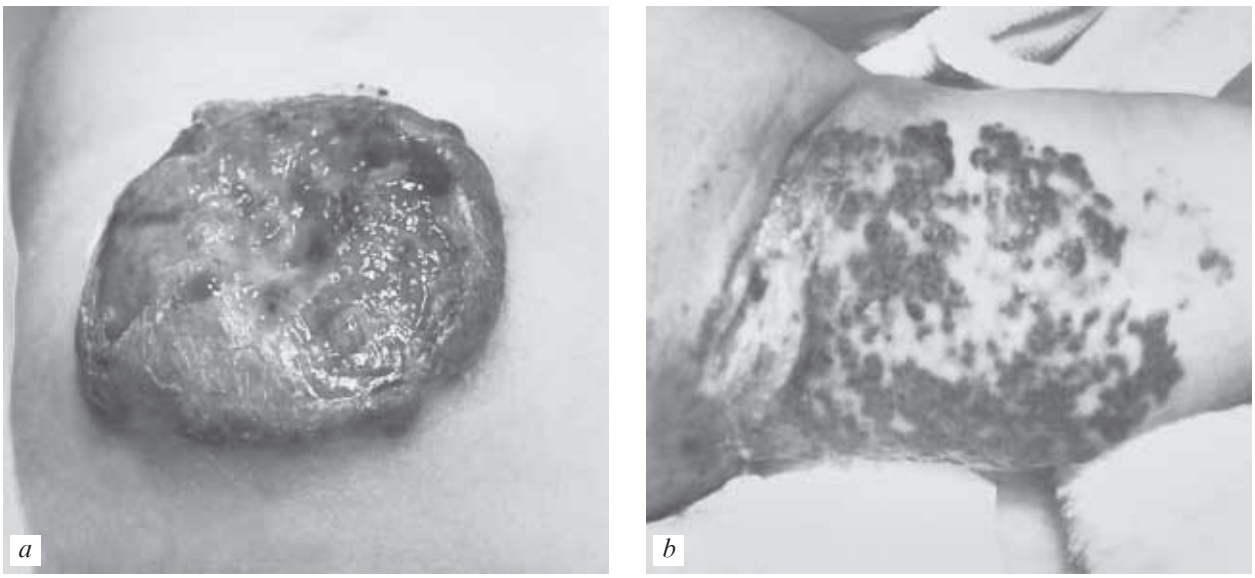


Fig. 2.1.4. Ulcerated hemangiomas *(a, b)*

4. Combined hemangiomas are the combination of the above hemangiomas.

5. Mixed tumors consist of tumor cells originating from the blood vessels and other tissues (angiofibroma, gemolimfangioma, angionevroma).

Clinical picture. Hemangiomas are characterized by tumor–like formations of various shapes and sizes with a bright red or slightly cyanotic color. Palpable tumor is usually painless, hotter than the surrounding healthy areas. A characteristic feature of hemangiomas is their rapid growth within the first months of life. In some cases the surface of hemangioma ulcerates, necrotic and exposes to infection (Fig. 2.1.4). Arising festering wounds often produce severe bleedings that may require emergency intervention.

Recognition of hemangiomas usually has no difficulties. They must be differentiated with other formations angioma–like (false hemangiomas).

So-called *flat hemangiomas* (angiomatous or vascular nevi) are spots, resembling spilled wine, located on the face and other parts of the skin and mucous membranes. They have different value, sometimes occupy half of the face; the color is from pale red to deep purple. Usually they are not raised above the skin, do not disappear when pressed, but only get pale in this case. Despite the lack of growth and superficial, these formations are causing serious concern for cosmetic reasons. Never disappear spontaneously.

Flat hemangiomas are often confused with other entities of pinkish color, localized on the midline of forehead, on the back of the nose or the occipital region (stained neonates). When the baby is asleep or in a relaxed state, these spots are barely visible or absent at the time, as if crying or straining their color becomes brighter. These spots usually disappear spontaneously at the age of 1 year. Do not require treatment.

Stellate hemangiomas (spider nevi) are common in children, aged 3–10 years old and usually are localized in the upper part of the face. Its appearance suites its name. It is found a small, size of a few millimeters, slick of pink color, from which a network of capillaries runs. Pressure on the central part results in emptying of capillary network. Progressive growth is not marked, but because of the location on the exposed skin, causes concern of parents for cosmetic reasons.

Pyococcus granulomas are not congenital and appear more frequently in children of middle–age and older. In fact, this granulation tissue is richly supplied with blood vessels, the appearance of which is sometimes associated with the introduction of infection at the point injury, but the true cause of the origin of these structures has not been definitively established. Clinically at first on the surface of the skin the discover in any part of a spot in the millimeter or a bit more, and within a few weeks it becomes a papilloma sized 0.5×0.5 cm or slightly more. Color changes and becomes dark purple. The surface is smooth at first, but during the formation is covered with crusts. Granuloma is easily injured and bleeds heavily, after it may disappear, but then after a few days it grows again.

Management and treatment. It is known that a significant part of hemangiomas has an ability to self-extinction, but this ability is not expressed in the same degree in different species of tumors. Therefore it is very important to solve the question of the beginning of treatment. Fundamentally this issue is resolved after a brief observation of the patient. If in the process of observation there is no tendency to a progressive increase or show signs of spontaneous disappearance of hemangiomas (flattening, blanching), the child is left under medical supervision. Examinations are better implemented monthly for the first 6 months, then once every 2–3 months or less. In case of doubt in the possibility of spontaneous disappearance of tumor patients are directed to treatment. It is, of course, necessary in cases of rapid growth of hemangiomas, with bleeding, as well as persistent, long–existing cavernous and branched forms. Individually they solve the problem of treatment with cosmetic purpose.

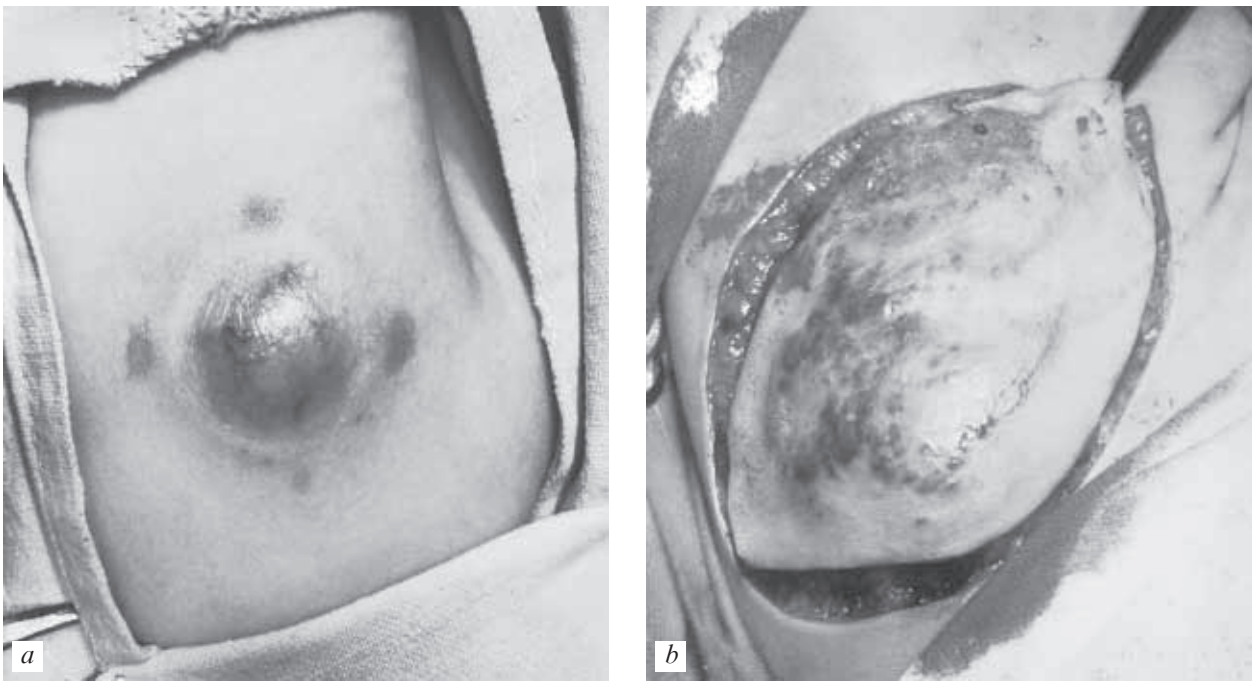


Fig. 2.1.5. Deleting of tumor within healthy tissue *(a, b)*

There are several methods of treatment of hemangiomas. An indication for the choice of a particular method depends on:

1. Type of tumor.
2. Its size and location.
3. The child's age and general condition.
4. The growth rate of the tumor.
5. Complications and functional disorders that arise in the process of its growth.

Treatment of hemangiomas may be operative and conservative. Surgical treatment is indicated in cases when tumor can be completely removed within healthy tissue without significant cosmetic damage (see Fig. 2.1.4). If you can not completely remove the tumor in a single step, resort to flash of the rest part in order to stop the blood flow in tumor and subsequent scarring. (Fig. 2.1.5).

If the application of surgical treatment of hemangioma is not possible, use conservative methods:

1) low temperatures (cryodestruction);
2) injection into the tumor and surrounding tissues of sclerosing agents (70% alcohol, hydrocortisone, etc.);
3) radiation therapy;
4) hormone therapy;
5) embolization of feeding vessels;
6) wave + cryodestruction;
7) compression therapy;
8) laser therapy.

Cryotherapy is more preferable at superficial hemangiomas in the open areas of the body. After cryotherapy on the spot of freezing an inflammatory reaction occurs. Inflammatory effects disappear by 7th–12th day, by that time there is no crust and becomes visible surface, covered with new epithelium. Application of this method is limited by the depth of germination of hemangioma (less than 5 mm), otherwise tumor growth continues in the deeply lying tissues (Fig. 2.1.6, 2.1.7)

Injection of sclerosing agents into the tumor shown in cases where its removal by surgery is not possible. For this purpose, use a 10% sodium chloride, 70% ethyl alcohol, 10% sodium sallicylate,

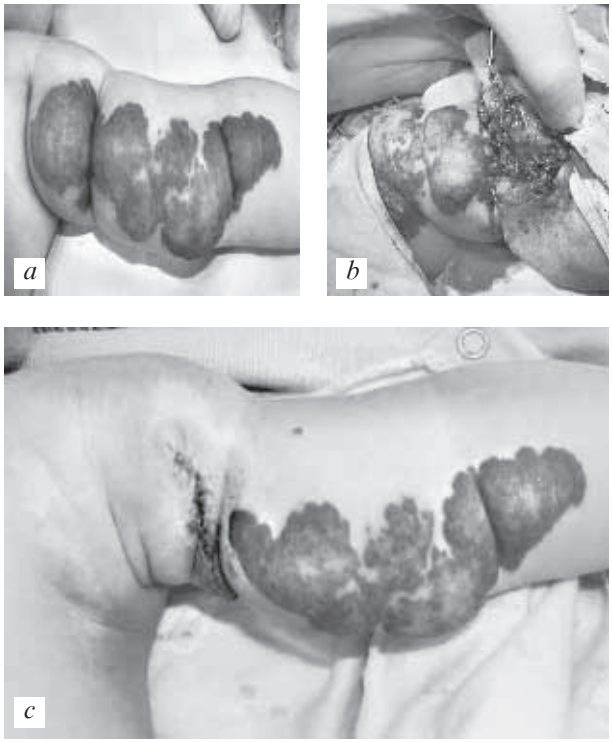


Fig. 2.1.6. Large hemangioma, management. *a* — before surgery, *b* — partial removal of the feeding vessels with stitching, *c* — after surgery

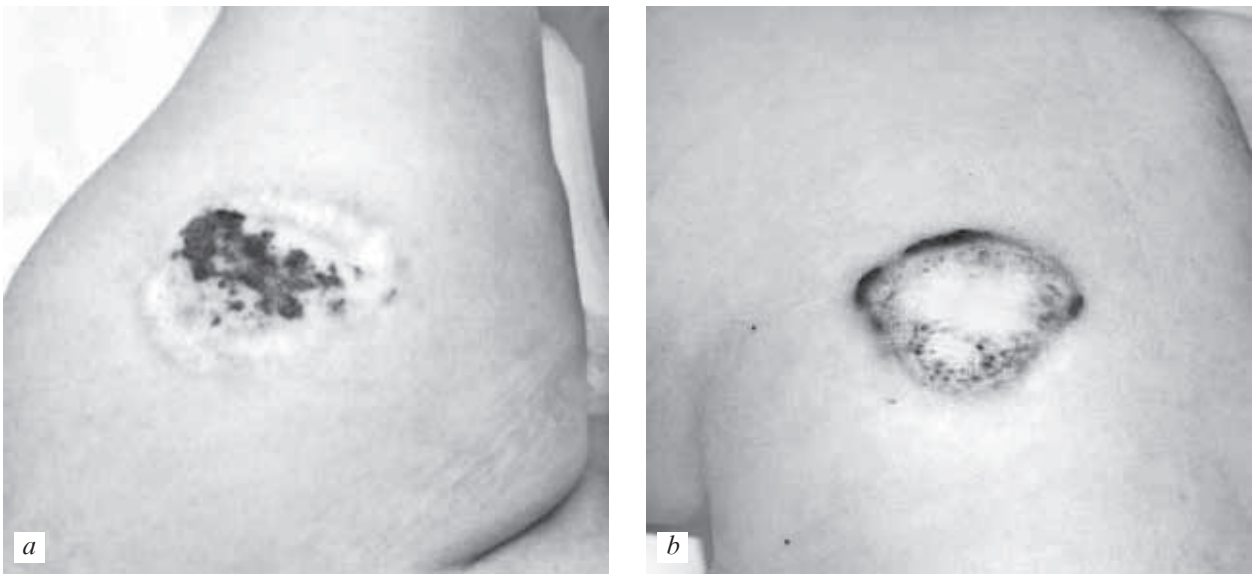


Fig. 2.1.7. Type of hemangioma after unsuccessful use of cryotherapy *(a, b)*

ethoxysclerol, sclerovein etc. Input of such substances in the tissue leads to aseptic inflammation and thrombosis in these vessels, that feed angioma, resulting in vascular emptying, the development of connective tissue and termination of tumor growth. Most widespread is injection of alcohol. For children they use 70% alcohol mixed with Novocain 2% in the ratio of 4:1. Treatment is repeated at intervals between sessions of 10–12 days.

Injections of alcohol are produced through healthy surrounding tissue slowly into the thickness of the tumor and under it. At superficial injection blanching of the skin occurs, which can lead to its necrosis. In such cases, they stop injection of substance and massage whitened site to restore circulation. Bleeding from the site of puncture is arrested by by finger pressuring.

During extensive hemangiomas, which are located on the face, neck and crotch, when it is impossible to carry out surgical treatment (Fig. 2.1.8), hormonal treatment is conduted. Prednisolone treatment of hemangiomas is conducted by short courses (24–28 days) in a day at a dosage of 2–4 mg/kg body weight, but not more than 40 mg a day. Prednisolone stops the growth of hemangiomas and causes hardening of angiomatous tissues.

Compression therapy (continuous compression or intermittent pneumocompression) — helps in emptying vessels, damage and endothelial cell proliferation with thrombosis. This method is used in large hemangiomas of extremities, the abdominal wall and the parotid gland (Fig. 2.1.9).

Embolization is an injection of thrombogenic material in the lumen of the blood vessels through the

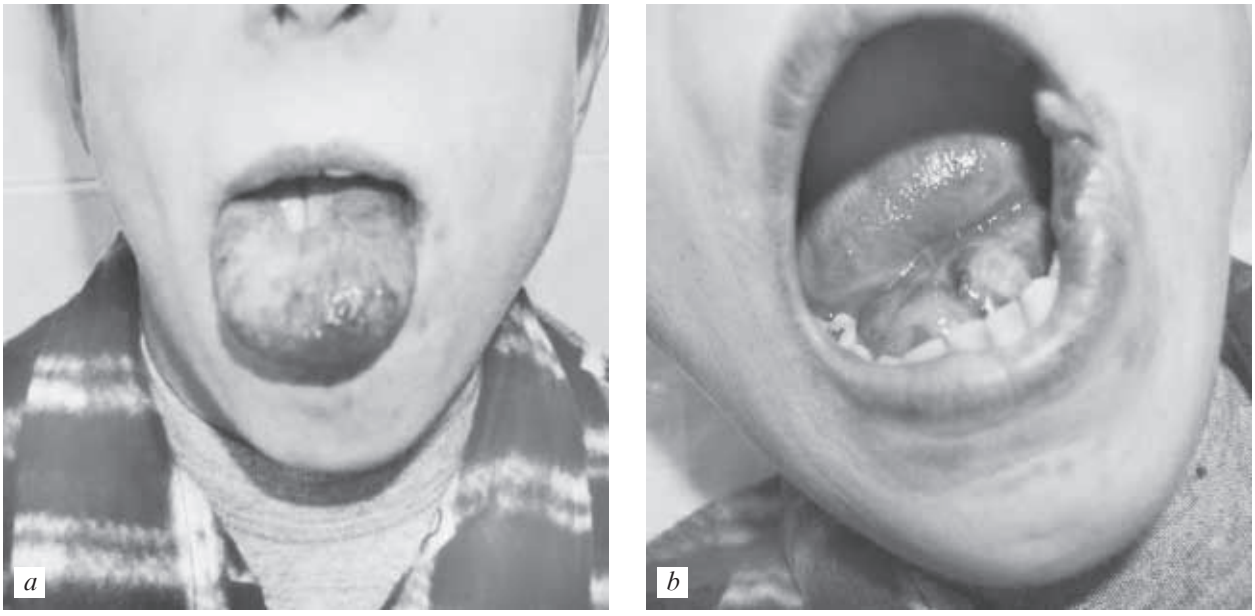


Fig. 2.1.8. Hemangioma of the tongue, cheeks and mouth floor *(a, b)*

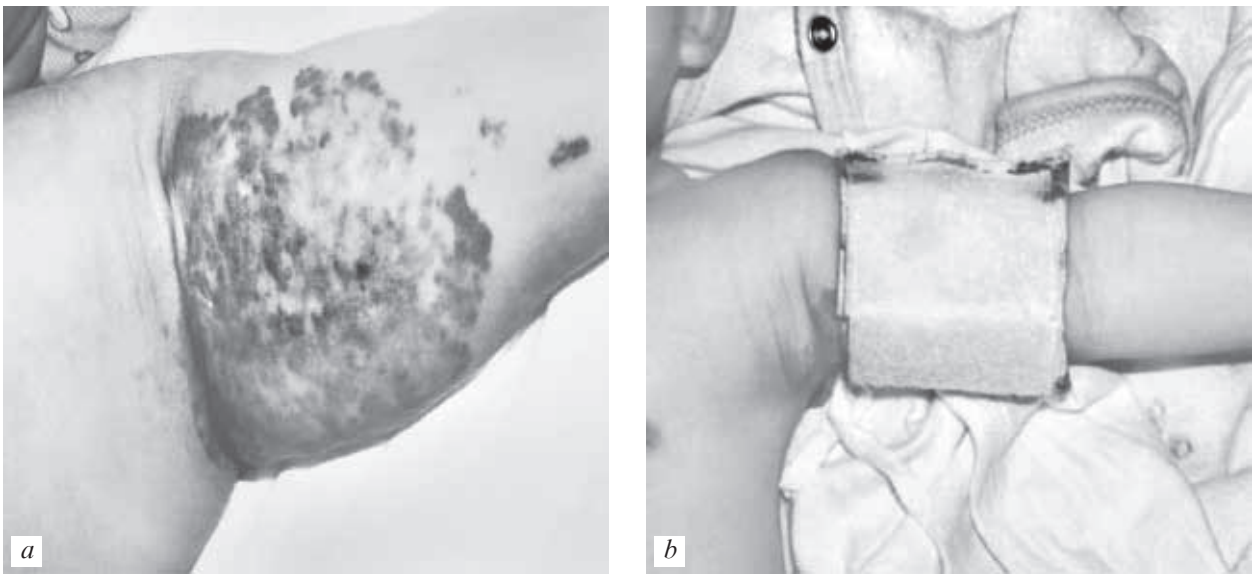


Fig. 2.1.9. Permanent pneumocompression of hemangioma with bandage *(a, b)*

arterial catheter, placed under the control of the tube and the X-ray contrast. It is used in cases where the hemangioma is not amenable to other treatments, the presence of severe complications (heart failure) and as preparation for surgery.

Laser therapy is used more frequently to treat wine stains, spider angiomas and strawberry hemangiomas. For the treatment of hemangiomas they use several types of laser with different principle — carbon dioxide (the action is based on the evaporation of water in the cells), argon (absorbed by hemoglobin in the blood vessels), its shorcoming is scarring of tissue, less intense during laser use on fluorescent dye.

Combined treatment is simultaneous or sequential use of surgical and conservative methods.

Complications of hemangiomas are divided into cosmetic and functional. They include bleeding, ulceration, infection, necrosis, airway obstruction, cardiac failure. "Capture" by hemangioma platelets can cause coagulopathy (Kasabach–Merritt syndrome) and/or intravascular coagulation. Periorbital hemangiomas can cause astigmatism, ptosis, strabismus and blindness. Depending on the location of hemangiomas, they can cause a variety of problems, associated with dysfunction of the organs and systems in the area where they are located.

LYMPHANGIOMA

Lymphangioma — congenital benign tumor that originates in the lymphatic vessels, and is composed of endothelial cells and the connective base. These tumors are classified as true benign tumors that arise from the disembryogenesis of lymphatic vessels. Lymphangioma occurrence is connected with abnormal growth or disruption of the normal development of the lymphatic system. These tumors occur less often than hemangiomas. The favorite localization of lymphangioma is neck, axillary region, cheek, lips, tongue, but they can be located in other parts of the body and internal organs.

Classification:

I. By histological structure:

1. *Simple* lymphangioma (capillary) is proliferation of lymph vessels in the skin and subcutaneous tissue. Clinically, they appear as thickening of the skin, slightly rough, mild delineated, sometimes in the form of nodules. Their typical localization is: cheeks, eyelids, lips, nose, less limb.

2. *Cavernous* lymphangioma is most common, represented by unevenly filled by lymph cavities formed from connective tissue spongy base. It is recognized by the presence of swelling, blurred outlines, soft consistency. Fluctuation is often marked. Skin can be soldered with the formation, but not changed. Displacability of the tumor is slight. Typical localization: neck, parotid region, cheeks, tongue, lips. Frequent inflammatory processes in the tumor are very typical. Cavernous lymphangioma can be spread over between fascial spaces, sprout vagina muscle.

3. *Cystic* lymphangioma in contrast to the cavernous represents one or several large cavities, not always interconnected. It is located under the skin, tumor forms protrusions of the right rounded shape, skin over them is not generally changed, but sometimes there is cyanosis due to radiographic fluid. Palpable borders of formation can be traced quite clearly, there are signs of fluctuation. Most often, these tumors occur in the neck, armpit (Fig. 2.1.10).

Lymphocele is often situated along the neurovascular bundle, can compress blood vessels, nerves, esophagus, trachea, which sometimes requires emergency care.

4. *Mixed* lymphangioma is usually hemolymphangioma. Prognostically these tumors have a more unfavorable course with frequent complications.

II. By localization:

1. *External* lymphangioma injure subcutaneous tissue, fascia, muscles;

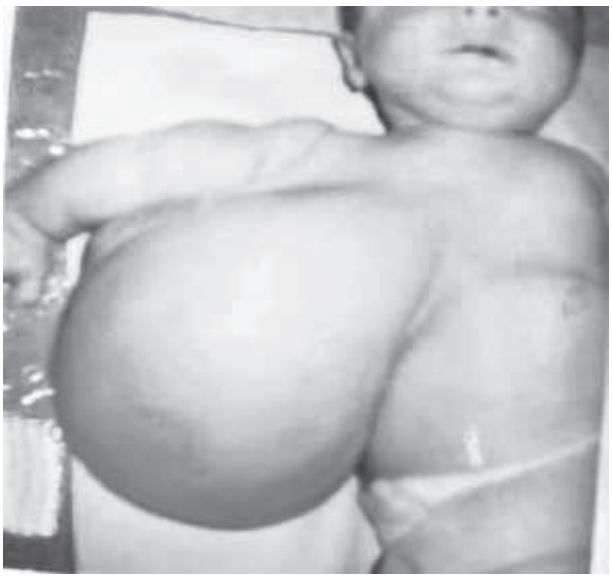


Fig. 2.1.10. Lymphocele of the armpit

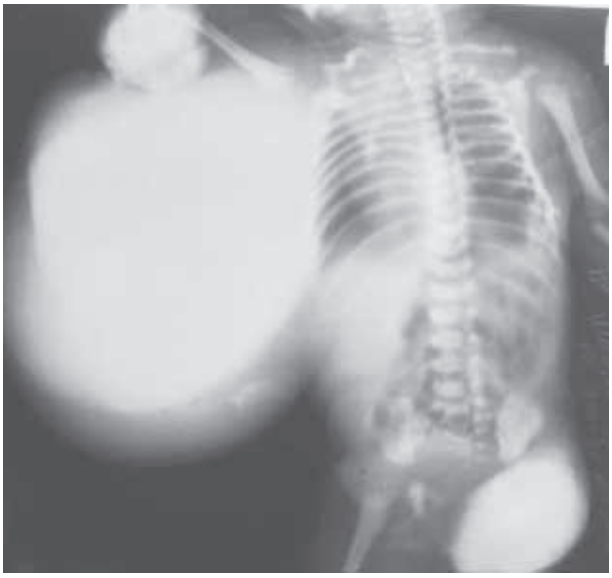


Fig. 2.2.11. X-ray of a child with cystic lymphangioma

2. Internal lymphangioma is situated in the abdominal cavity, proceed from the mesentery of the small and large intestines, omentum, sometimes the liver and spleen. Often they allocate cervical mediastinal location, when one part of the tumor is located in the neck, the other in the mediastinum.

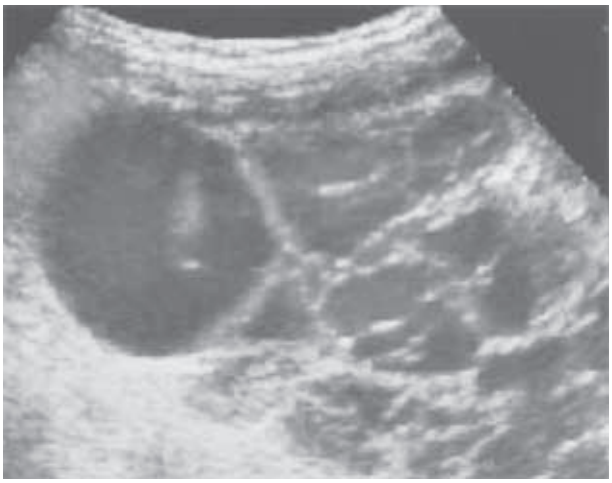


Fig. 2.1.12. Ultrasound of lymphangioma

In a number of children with lymphangioma of external localization there are various complications, including inflammation and suppuration, compression of adjacent organs, maceration, chylorrhea, bleeding in lymphangioma, cosmetic defect, acute respiratory failure. The diagnosis is made at the examination. X-ray detects the presence of soft–tissue formation (Fig. 2.1.11).

Ultrasound detectes mainly cystic formation, which contains the partitions of different thickness (Fig. 2.1.12).

More informative is CT with contrast enhancement of the tumor (Fig. 2.1.13), and MRI.

Treatment. Lymphangioma treatment includes surgical and non-surgical methods.

The surgical methods consists in:

a) removal of the tumor within healthy tissue;

b) artial removal of the deep component of the tumor with preservation of the skin part (in the case of demarcated capillary lymphangioma);

c) surgical treatment after puncture of lymphangioma with the evacuation of the contents.

Non-surgical methods aare following:

a) cryotherapy using liquid nitrogen;

b) laser therapy (carbon dioxide laser);

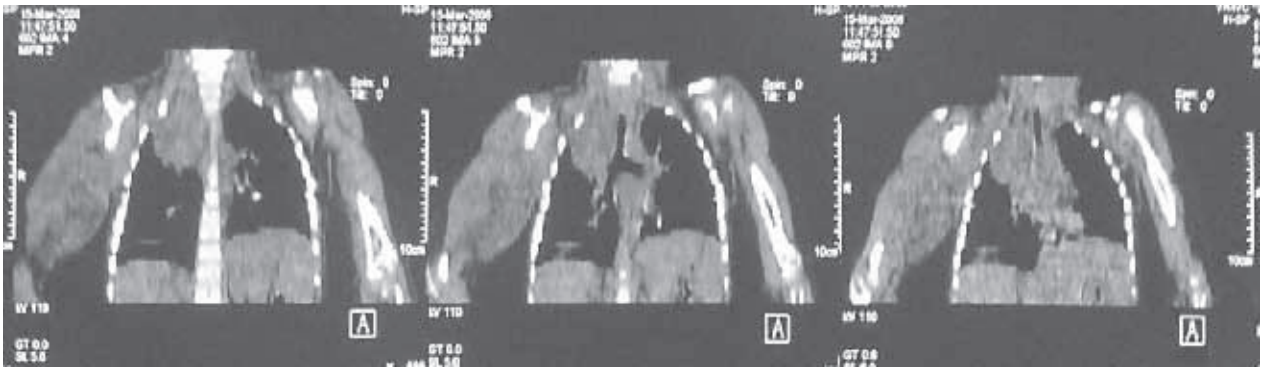


Fig. 2.1.13. CT of a child with lymphangioma

c) puncture method;

d) sclerotherapy (the use of bleomycin and OK–432).

The method of treatment of lymphangioma depends on several factors:

1. Type of lymphangioma (capillary, cavernous, cystic, mixed).

2. Tumor location (neck, mouth floor, axillary region).

3. The vastness of the destruction.

4. The presence of complications (stress, infection, abscess, hemorrhage, and compression of vital organs with violation of their function).

5. Anatomical location (involvement of the great vessels, nerves, etc.).

Most surgeons agree with the fact that the removal of lymphangioma is the best method of treatment. Unfortunately, this is not always possible. Removing some lymphangioma can be quite dangerous due to the risk of complications, such as recurrence, infection, damage to blood vessels and nerves, cosmetic deformation of fistulas.

A number of surgeons recommended in the absence of complications and the rapid growth of lymphangioma a conservative treatment and follow up management. This managements is based on the ability of spontaneous regression of some lymphangiomas.

Treatment of capillary lymphangiomas is surgical excision, cryotherapy, laser therapy.

Treatment of cavernous lymphangioma — removal, laser therapy. In the case of infection they use antibiotics.

Treatment of cystic lymphangioma is removal, emptying by aspiration (temporary procedure at the development of life–threatening conditions), drainage (in the case of infections and abscesses), sclerotherapy with the use of bleomycin or OK-432.

The use of bleomycin is contraindicated in children under 6 months and carries a risk of pulmonary fibrosis. The substance OK-432 is obtained by incubation Streptococcus pyogenes (human) with penicillin G. Relatively high efficiency of laser technology, especially with carbon dioxide laser.

BENIGN TUMORS AND TUMOR-LIKE LESIONS OF BONES

Among oncological diseases bone tumors in children account for 10–15%, and benign lesions among them are 10 times as much often. According to M. V. Volkov, treatment of bone neoplasms — a prerogative of not only oncologists but orthopedists because many tumors, including cancer, develop based on dysplastic cells of bone tissue.

Classification of primary tumors and bone dysplasias in children (according to the classification of WHO, Geneva, 1972):

1. Benign.

2. Malignant.

By histogenesis:

1. Bone-formed (osteoma, osteoidosteoma, osteoblastoma, osteosarcoma).

2. Cartilage–formed (chondroma, osteochondroma, chondroblastoma, chondromixoid fibroma, chondrosarcoma).

3. Giant cell (osteoklastoma).

4. Bone–marrow (Ewing's sarcoma, reticulosarcoma, myeloma).

5. Vascular (hemangioma, chylangioma, angiosarcoma).

6. Connective tissue (desmoplastic fibroma, lipoma, fibrosarcoma, liposarcoma).

7. Tumor–like (aneurismal or solitary cyst, fibrous dysplasia, fibrous metaphyseal defect, eosinophilic granuloma).

Clinical picture of benign bone tumors is characterized by slow progression, lack of "common tumor symptom" and metastases.

In the clinical picture lack of manifestations, lack of pathognomonic signs at intraosseous cells dominate, and these symptoms vary depending on the nature of the disease, its limitations and the presence of complications.

In 50% of children with intraosseous lesions at insignificant trauma pathological fracture occurs, which is the first differentiated sign of disease (chondroma, desmoplastic fibroma, solitary cyst, fibrous dysplasia).

Extraosseous tumors (osteoma, osteochondroma) are manifested by the symptom of palpable tumor, which is often determined visually. At intraosseous lesions swelling is detected at all eccentrically located and expansively growing tumors and tumor–like processes (osteoidosteoma, osteoblastoma, chondroma, aneurismal cyst), but the severity of symptoms depends on the size of the surrounding soft tissues and disease duration.

The pain is marked at osteoclastoma, osteoidosteoma, chondroblastoma, aneurismal bone and at compression of adjacent neurovascular elements. Long–lived process can cause deformation of limbs, paired bones.

General characteristics of benign bone tumors are their slow growth, painless, posed by strain of their bones, which are sometimes palpable, complications by pathological fracture. A random roentgenologic finding occurs often. However, some of them are exceptions from these rules. Most benign tumors are observed at young age, and some of them are more tumor–like lesions and have dysplastic nature.

Osteoma — one of the most mature morphologically benign bone tumors consisting of osteoblasts.

It is often diagnosed in childhood, sometimes is a random roentgenologic finding.

It is of 2 types: compact and spongy. Spongy osteoma is more often localized in tubular bones. As growth shifts away from the joint; throughout thinned cortical layer can be traced. The tumor has a trabecular structure with exophytic tumor growth.

X-rays presents always a "shadow plus", additional formation, related to bone with a broad base or foot (Fig. 2.1.14). Compact osteoma can be localized in the bones of the cranial vault, paranasal sinuses. Osteoma X-ray diagnosis is not difficult. Compact osteoma gives homogeneous structureless intensive shade (Fig. 2.1.15).

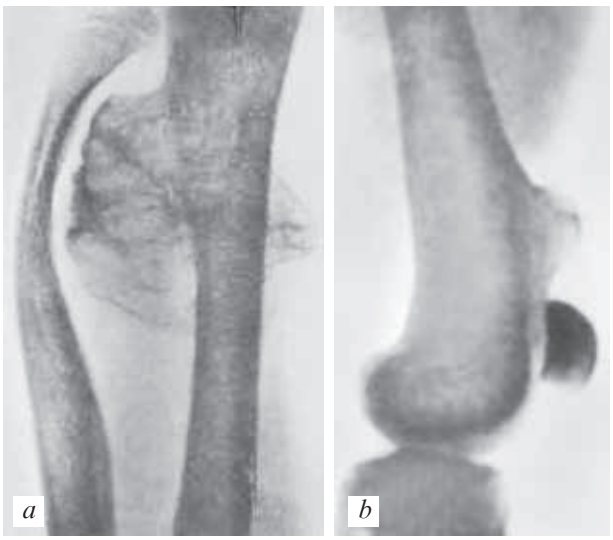


Fig. 2.1.14. Ostoma of the elbow bone *(a)*, femoral bone *(b)*

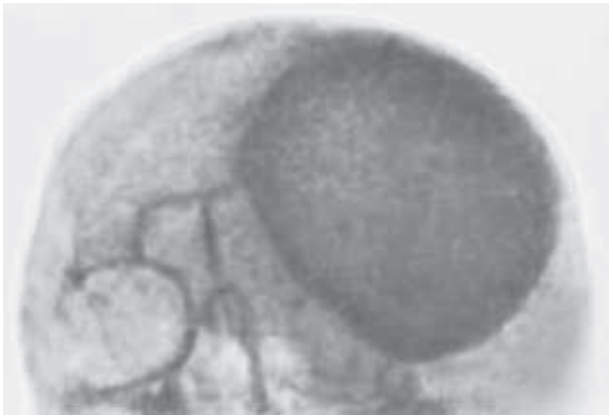


Fig. 2.1.15. Compact osteoma of calvarium

Clinical course of osteoma is favorable, tumor growth is slow. Clinical manifestations depend largely on the localization of osteoma. Compact osteoma of the skull that grow inward, can cause severe complications.

Differential X-ray of osteoma of limbs should be done mainly with axis myositis, subperiostal hematoma, osteochondroma, bone and cartilaginous exostosis. At axis myositis it is marked pain, no connection with the actual formation of bone, spotted incorrectly, fibrous structure of axis muscle. Subperiostal hematoma provides shade of fusiform, longitudinal size of which merges with the long axis of the bone.

In children axis venous sinus of the skull is taken for ostema by mistake — *sinus pericranium*, which is a variant of development.

The prognosis at osteoma is favorable. Osteoma does not become malignant, but requires radical surgery to prevent possible recurrence.

Treatment is operative — removal of the tumor with the healthy area of bone and periosteum.

Osteoblastoclastoma (giant cell tumor) is characterized by changing periods of active growth and destruction of bone tissue, stabilization periods with reparative processes in bone. Clinically it is manifested by pain and deformation of the tumor, is localized in 75% of cases at articular ends of long bones. This tumor can be quite aggressive, and in 12.8% of cases becomes malignant. The tumor occurs at a young age (up to 30 years), affects both epiphysis and metaphysis of long tubular bones. The first detailed description of this tumor belongs to Nelaton (1860). Osteoblastoclastoma is one of the most common bone tumors. There are no notable gender differences in the incidence of osteoblastoclastoma. There are cases of family and hereditary diseases. The age of patients with osteoblastoclastoma ranges from 1 year to 70 years old.

The second and third decades of life account for 58% of osteoblastoclastoma cases.

Osteoblastoclastoma is usually a solitary tumor. Sometimes double localization is marked, mainly in neighboring bones. Most often affects long tubular bones (74.2%), rarely — flat and small bones.

In the long tubular bones tumor is localized in epimetaphyseal department (in children — in metaphysis). It does not germinate articular and epiphyseal cartilages. In rare cases there is diaphyseal localization of osteoblastoclastoma.

*Pathomorphology: a*long with mononuclear oval cells of osteoblasts type they detect large multi–type giant cells of osteoclasts type.

They distinguish the following osteoblastoclastoma forms: lythic, active cyst and passive cyst.

Clinical picture. Clinical manifestations of osteoblastoclastoma largely depend on the tumor localization. The first sign is pain in the area of the lesion, develops bone deformity, pathological fractures are possible.

Signs of acquiring malignancy at osteoblastoclastoma: rapid tumor growth, increased pain, increasing the diameter of the focus of destruction or transition of alveolar–trabecular phase into lythic, destruction of the cortical layer over a large area, blurred contours of cell destruction, the destruction of the locking plate that previously limited the entrance to the medullary canal, periosteal reaction.

Conclusion about the malignancy of osteoblastoclastoma based on clinical and X-ray data should be confirmed by morphological study of tumor.

X-ray study. The tumor has the form of oval branch of enlightenment. An important X-ray evidence of all forms of osteoblastoclastoma that distinguish it from tubercular lesions of bone, is the lack of osteoporosis.

Affected bone segment appears asymmetrically swollen. Irregular cortical layer is thinned, often wavy, can deteriorate over a large area. At the site of interruption cortical layer is partly fibral or sharpened as a "sharpened pencil" which simulates in some cases "periosteal visor" at osteogenic sarcoma (Fig. 2.1.16). The tumor, destroying the cortical layer can extend beyond the bone in the form of soft tissue shadow.

They distinguish alveolar–trabecular and lythic phases of osteoblastoclastoma. In the first case destruction centers of bone tissue are determined, though separated by partitions.

Lytic phase is characterized by continuous branch destruction. Cell destruction is asymmetrically located concerning the central axis of the bone, but growing, can occupy the entire width of the bone. Is typical clear limitation of destruction branch from intact bones. Medullary canal is separated from the tumor by a locking plate.

Diagnosis. Diagnosis of osteoblastoclastoma of long tubular bones over time is complex (Table 10).

The greatest difficulties arise in the radial diagnosis of osteoblastoclastoma with osteogenic sarcoma, bone cysts and aneurismal cyst (Fig. 2.1.17).

For osteoblastoclastoma can be mistaken for monoaxis form of fibrous osteodysplasy of long tubular bones (Fig. 2.1.18).

Treatment of osteoblastoclastoma is operative. Sparing bone resection with removal of a tumor and bone grafting simultaneously (auto, homotransplants or replacement with methylmethacrylate) — the method of choice. At damage to spine with tumors they use radiotherapy.

Osteoid osteoma. Currently, there are two views about the nature of osteoidosteoma. Some authors consider osteoidosteoma a chronic purulent–necrotic cell osteomyelitis, other enroll osteoid osteoma to tumors.

A detailed study of this tumor by clinicians and radiologists began in 1935, after it was identified by Jaffe and called "osteoid osteoma". Five years earlier Bergstrand presented a description of the pathologic process — "osteoblastic disease" as a flaw of embryonic development. Osteoid–osteoma is found in young adults (11–20 years), men suffer twice more often. Usually osteoid osteoma is a solitary tumor localized in any department of bones (mostly long tubular bones).

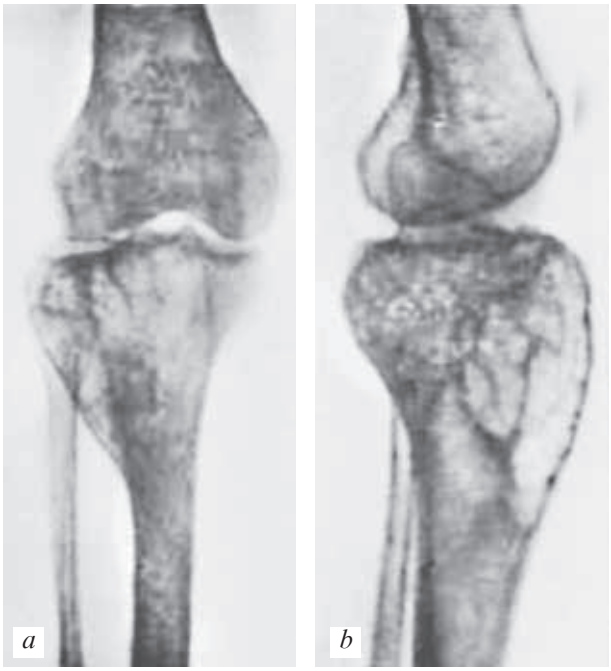


Fig. 2.1.16. Osteoblastoclastoma of tibia *(a, b)*

Clinical picture. Patients are disturbed by pain, especially at night. Pain is localized, reinforced by pressing the center. Skin covers are unchanged. At tumor localization in the lower extremities — lameness.

Preferably in the diaphysis or metadiaphysis of long tubular bones is determined oval shaped with clear contours cell destruction of bone tissue, which is less than 2 cm in diameter. Around the focus of destruction is determined area of osteosclerosis especially pronounced in cases of intracortical location of

Table 10

Differential Diagnosis of Osteoblastoclastoma

Tumor characteristics	Osteoblastoclastoma (lythic phase)	Osteogenic osteoblastoclastic sarcoma	Bone cyst
Age	20–30 years old	Before 20–26 years old	2–14 years old
Localization	Epimetaphysis	Epimetaphysis	Metadiaphysis
Bone shape	A pronounced asymetric thickening	A slight distention of a transverse part	Fusiform thickening
Contours of destruction focus	Clear	Non-clear, vague	Clear
Condition of bone marrow channal	Closed by closing plate	At the boarder of the tomour is open	
Cortical layer	Thinned, curve, interrupts	Thinned, destructed	Thinned, even
Sclerosis	Atypical	Present	Atypical
Periostal reaction	None	Pronounced, mostly by type of "periostal peak"	None
Condition of epiphysis	Epiphysial plate is thinned, curve	At initial stages the site of epiphysis remains intact	Intact
Next diaphysial department of the bone	Intact	Osteoporotic	Intact

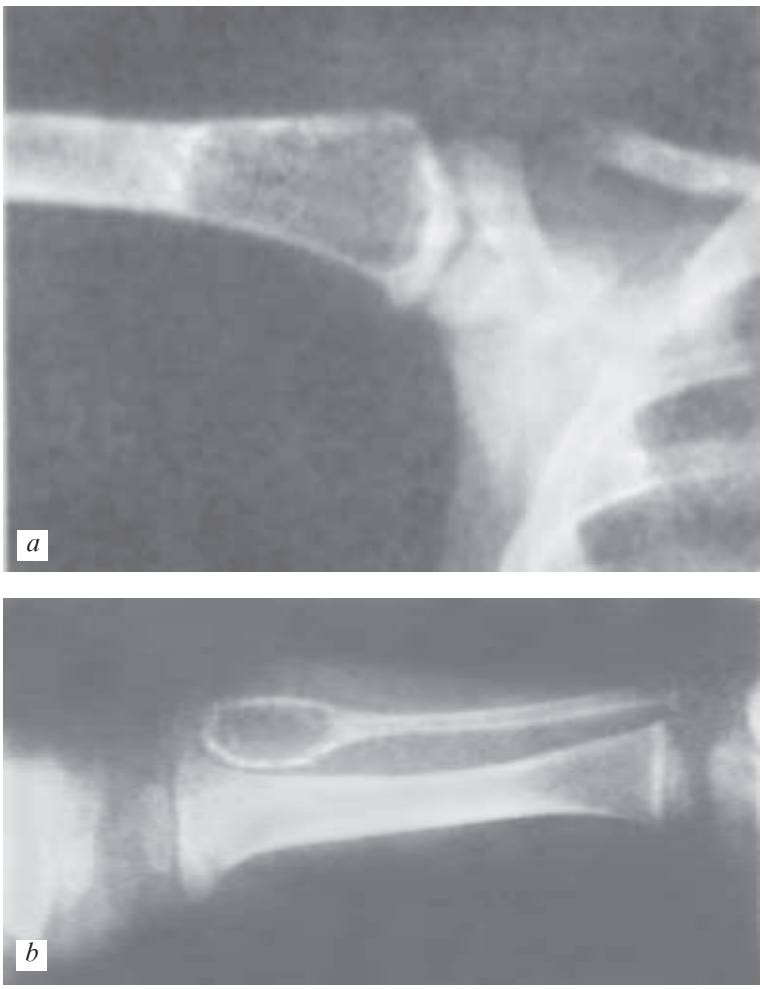


Fig. 2.1.17. One-chamber simple cyst of the humeral bone (*a*); aneurismal cyst fibular bone *(b)*

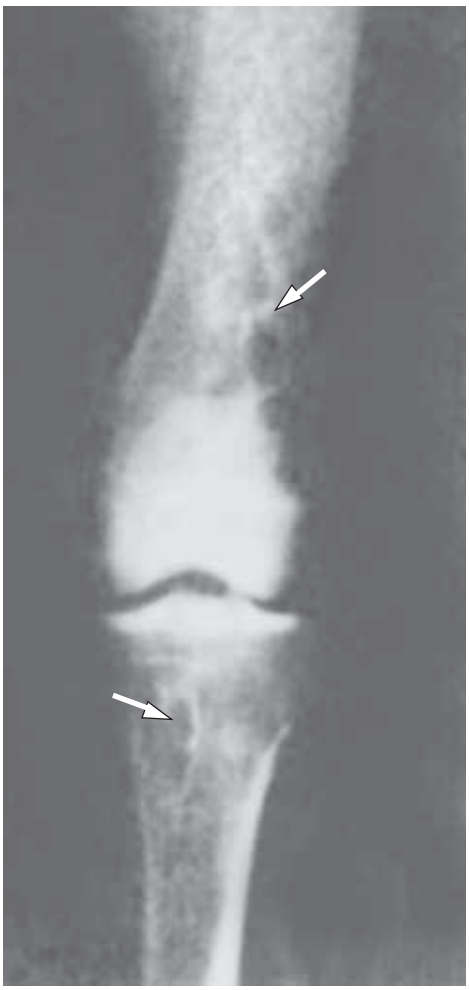


Fig. 2.1.18. Fibrous dysplasia of the femoral and tibial bones (arrows)

cell destruction. Area of sclerosis due to periosteal and to a lesser extent, endosteal unilateral change causes deformation of long bones. The massive bony growths prevent cell degradation detection on X-ray, therefore, to clarify the nature of affection and a clearer focus detection ("of this tumor nest") is recommended tomography.

At localization of cell destruction is marked in spongy substance a narrow bezel of sclerosis. Inside the cell of destruction can be seen inclusion bone, which Walker (1952) calls "tiny round sequesters" and considers them osteoid osteoma typical.

Differential diagnosis of osteoid osteoma is conducted with Brodie bone abscess.

Treatment is operative. After radical removal usually osteoid osteoma does not recur.

Bone–cartilaginous exostosis looks like additional formation to the bone, which appears on its surface. Occurs in the bones that going in the development a cartilage stage and cause disturbances in process cartilaginous plates. With few exceptions, clinically asymptomatic, but occasionally comes malignization.

Chondroma — a tumor of childhood and adolescence. Affects more often short tubular bones cysts (about 70%) and the foot, then pelvic bones, processes of vertebrae, sternum. In long tubular bones chondroma is localized in metaephisar ends. In short tubular bones of hand and foot chondromas are more often multiple, and bilateral lesions are marked. In the flat bones, especially in long bones, there are solitary chondromas. Chondromas should be treated as potentially malignant tumors. Chondromas are divided into enchondroma and ecchondroma.

Enchondroma — tumor, located inside the bone. In the center of bloated single cell there is homogeneous enlightenment of irregular round or oval form with clear contours. In its homogeneous background detect there are single shadows of centers of cartilage filling.

Ecchondroma — tumor originating from bone and growing towards the soft tissue. Against the backdrop of sealing of the soft tissues they detect calcification areas of different size and intensity.

The boundaries of the tumor and its foundation are identified hardly.

Clinical picture. More often phalanx of fingers and foot, shank and metacarpus bones affects, rarely — femoral and humeral bone. The joints are usually not changed. But at larger tumors pronounced deformation of bones occurs, which mechanically prevents movement in joints (Fig. 2.1.19).

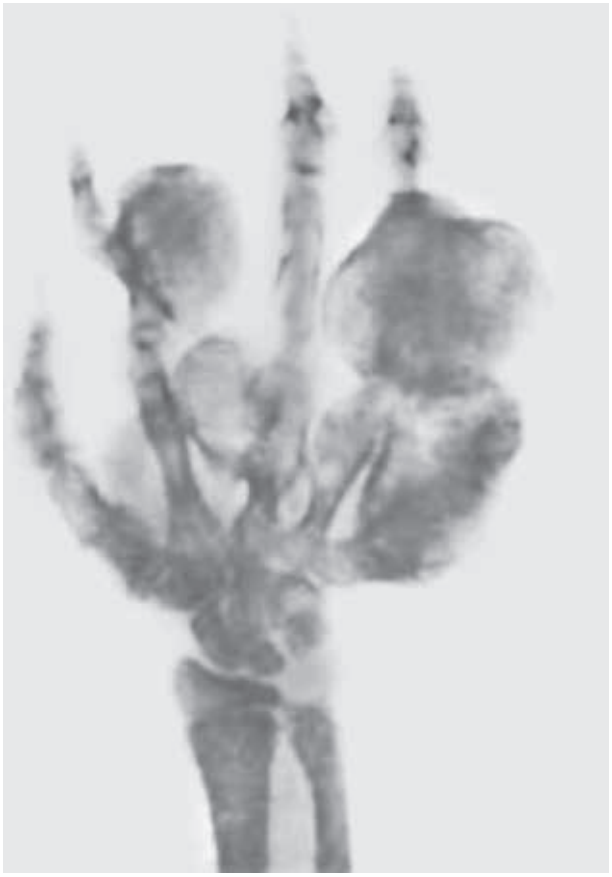


Fig. 2.1.19. Multiple chondromas of phalanx

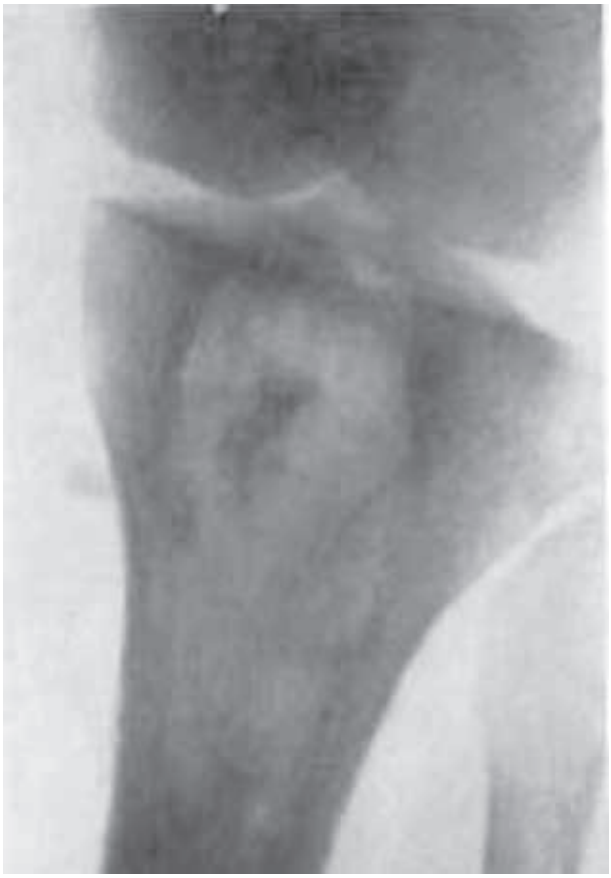


*Fig. 2.1.20. C*hondromas of the tibia

Characteristics: swelling which gradually develops, in close proximity to the joint — arthralgia, synovitis phenomenon (Fig. 2.1.20).

Enchondroma at malignancy turns into chondrosarcoma: thus accelerating the growth of the tumor, pain, tumor calcification and accumulation of the isotope occur at scintigraphy. To clarify the diagnosis tumor biopsy is recommended. Chondromas treatement is only surgical.

PIGMENTED TUMORS

Pigmented tumors include nevi and melanoma. Nevus is a benign neoplasm, melanoma is a malignant tumor. These tumors — a violation of the proliferation of melanocytes. Histologically, nevus cells lose their normal dendrites, but have other characteristics of melanocytes.

Nevi (birthmarks) are benign neoplasms, the occurrence of which the majority of researchers explain by migration in the embryonic period of melanoblasts from neuroectodermal tube in the basal layer of the epidermis.

Studying pathology of the skin is interesting in connection with the fact that in th site of pre-existing congenital or acquired benign nevi a terrible malignancy melanoma often develops.

It is well known that almost all people have various nevoid neoplasms, both congenital and acquired. More than 90% of the population have pigmented skin neoplasms. The average number of nevi at each person is 20, ranging from 3 to 100, and the number usually increases with age. If we compare these number with statistics of the incidence of melanoma (1.8 per 100 000 population), we obtain the hundreds of a percent to the number of patients who visit doctors with nevi.

Nevi may be located in the epidermis or dermis or both of these layers (Fig. 2.1.21).

Histological characterization of benign nevi

Border of the nevus — flat or towering brown spot no more than 1 cm often locates in the center of the foot, hands and genitals. They may disappear spontaneously.

Dermal nevi are usually localized at the upper third of the thickness of the dermis, may have any localization, often occur in older children. Color varies from light to dark brown, sometimes they contain hair.

Complicated nevi are such nevus cells located in the dermis and the epidermis. Color is from brown to black. Tendency to localization in the palms, soles, genitals and limbs.

Ordinary blue nevi is located within the dermis, in close contact with the melanin–bearing macrophages. They do not contain true nevus cells. It is black and more often located on the face, scalp and dorsum of hands. Its variety — blue cell nevus — common on the buttocks and sacrococcygeal region.

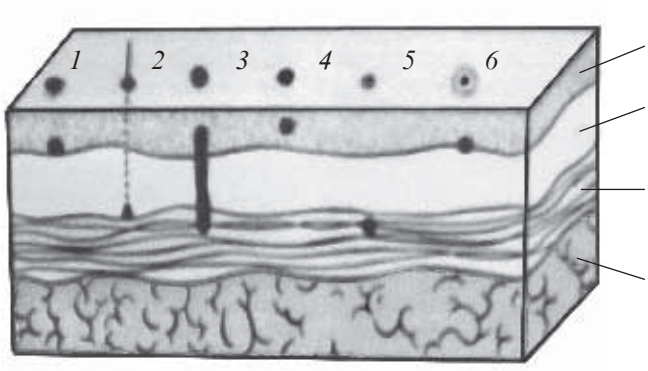


Fig. 2.1.21. Localization and appearance of nevi (K. U Ashcraft, T. M. Holder "Pediatric Surgery", T III, p. 241, 1999):
1 — boundary
2 — dermal
3 — complicated
4 — common *blue*
5 — spitz
6 — galonevus

Spitz nevus is usually localized inside inside dermis. Its cells are highly infiltrative and have diffuse pigmentation. Bright feature is mitotic activity. The most frequent localization is head and neck, often — on the cheeks, rearer — in the trunk and buttocks.

Galonevus is a pigmented border nevus, surrounded by lymphocytic infiltration, which creates a zone of depigmentation that gives the appearance of a nevus halo effect. They are located on the trunk.

Not all nevi may potentially develop into melanoma, so they are considered to be divided into melanoma dangerous and melanoma non-dangerous.

1. Nevi without potential of malignization:
— Epidermal nevus
— Epidermal nevus syndrome
— Becker's nevus
— Follicular keratosis
— Syndrome of basal-cell nevus
2. Combined nevi:
— Ott nevus
— Ito nevus
— Blue nevus
3. Nevi with malignization potential:
— Dysplastic nevus syndrome
— Large nebo cellularity nevus
— Small nebo cellularity nevus
— Sebaceous nevus

Among melanocytic nevi the following two types are the true precursors of malignant melanoma: dysplastic melanocytic nebo cellularity nevi. In addition, the penchant for development of basal cell carcinoma has also a border nevus sebaceous.

Border nevus (Fig. 2.1.22, *a*) is evenly a yellow–brown to black spot, usually no more than 1.5 cm. The spot appears within the first years of life, has clear boundaries, not above the surface of the skin, and grows with a child. In childhood it is not dangerous. As the child grows this nevus and changing, becoming complex.

Complex nevus (Fig. 2.1.22, *b*) is similar with a boundary, but is distinct by raising above the surface of the skin, looks like a papule, the contours are smooth, uniform color. Further, with the growth of the child, nevus border may remain the same, but, more often again changing and turning into intradermal nevus.

Intradermal nevus (Fig. 2.1.22, *c*) is domed or papilloma formation with pedicellate, rarely can be in the form of blackberry or clam prominent node on a broad basis. The surface is often covered with hair. Nevus diameter is about 1 cm. Color is from light brown to black, in some cases may be reddish or whitish, sometimes translucent. This variant of nevus occurs in adolescents and adults, the presence in a young child is a bad symptom.

Galonevus is a pigmented formation, similar in appearance with complex nevi, around which there is rim of depigmentation (i. e. plot light staining), the appearance of the rim is due to autoimmune reactions of the organism to nevus cells. Usually occurs in adolescents and pregnant women, and only is sometimes congenital, having the typical symptoms, but sometimes it is necessary to differentiate it with melanoma.

Blue nevus is divided into simple and cellular. Simple blue nevus appears as a single nodule with diameter up to 1 cm, the color of which varies from light gray to black with a bluish tinge, surface of the node is smooth, hairless, texture is density elastic. Usually located on the face, neck and upper limbs, sometimes on the body.

Cellular blue nevus is a kind of blue nevus, characterized by an abundance of cellular elements. It has a large size (an average 3 cm in diameter) and pronounced pigmentation, appears as a node of blue color with a smooth or rough surface. Almost 50% of cases are localized on the buttocks and lumbosacral region, rearer — on the back of the hands, feet. Do not contain nevus cells, but in structure has special melanocytes, capable to transform into melanoma, so we took it to the first group.

Congenital nevi (Fig. 2.1.22, *d*) — pigmented lesions which are laid in utero, however, during the birth of a child may not be visible due to the low pigmentation, within the first year of life appear almost always, they are divided by size into small — less than 1.5 cm, middle — from 1.5 to 20 cm and a huge — more than 20 cm in diameter. The typical localization of congenital melanocytic nevi is lower torso, upper back, arms, chest, upper (proximal) parts of hands and feet. Giant nevi may have a form of "cowards", "bathing suit" or resemble "animal skin". It is believed that up to 12 years, if possible, such nevi should be removed. Children with congenital nevi are subject to monitoring by oncologist. This group of nevi is the most dangerous as for transformation into melanoma.

Spotted nevus (Fig. 2.1.22, *e*) — appears at birth or later, looks like a light brown flat spot with irregular contours, against which there are small dark brown spots, raised above the skin surface. Spotted component in structure doesn't differ from lentigines (doesn't contain nevi cells), but the small dark blotches that are not seldom seen during children getting older, are borderline or complex nebo cell nevi, occasionally is dysplastic nevus. This, the second component is melanoma dangerous.

Dysplastic nevus (Fig. 2.1.22, *f*) occurs as an independent form and as a form of transformation from other forms. Signs of such nevi are irregular shape, fuzzy contours, irregular surface and color, the inhomogeneous density, signs of ulceration, seals. Is regarded as a transitional form to melanoma. It is subject to mandatory removal with histological examination.

Mongolian spot (Fig. 2.1.22, *g*) is present from birth in 80% of children of negroid and mongoloid races, in caucasian children occurs in 1% of cases. It is a well delimited round or oval spot in the lumbosacral region, has a uniform dull grayish–bluish or brown color. It can reach a diameter of 5–10 cm to 7–13 years disappears, does not contain nevus cells, and doesn't provoke melanoma.

Warty nevus (Fig. 2.1.22, *h*) is a so called malformation of the skin, also known as verrucous nevus, verrucous epidermal nevus, usually located on the head, rarely on the body, is an accumulation of pigmented nodules of various sizes, usually no more than 5–8 mm, located along the course of the nerve trunks exists since birth or early childhood, rarely comes later. Is common at 0.1–0.5% of adults. Does not contain nevus cells, i. e., not a real nebo cell nevi, so melanoma non dangerous, but sometimes in adults, from its elements can appear skin cancer.

Nevus of the "coffee with milk" type (Fig. 2.1.22, *i*) is not a typical nebo cell nevi, may be congenital or appear later, single spots (at least 3) may be at healthy people, multiple lesion is characteristic of the Recklinghausen disease (neurofibromatosis), as the only manifestation, so and in combination with other symptoms. It looks like a flat spot, which is not raised above the skin surface with light brown color and irregular shape, sized 1–20 cm or more.

Freckles (efelids) — pigment spots on the face, which appear in the active insolation period (spring and summer) and whiten in the dark time of the year (winter). Appears as small reddish-brown flat spots, a number from a few to hundreds.

Youthful lentigines — pigment spots, very similar to the transition pigmented nevi, but unlike the nevus, lentigo has no nevus cells, intense pigmentation is caused by increased level of melanocytes, lentigo is not dangerous concerning malignancy. The items are anywhere on the skin or mucous membranes, often on lips, especially the lower, the genitals.

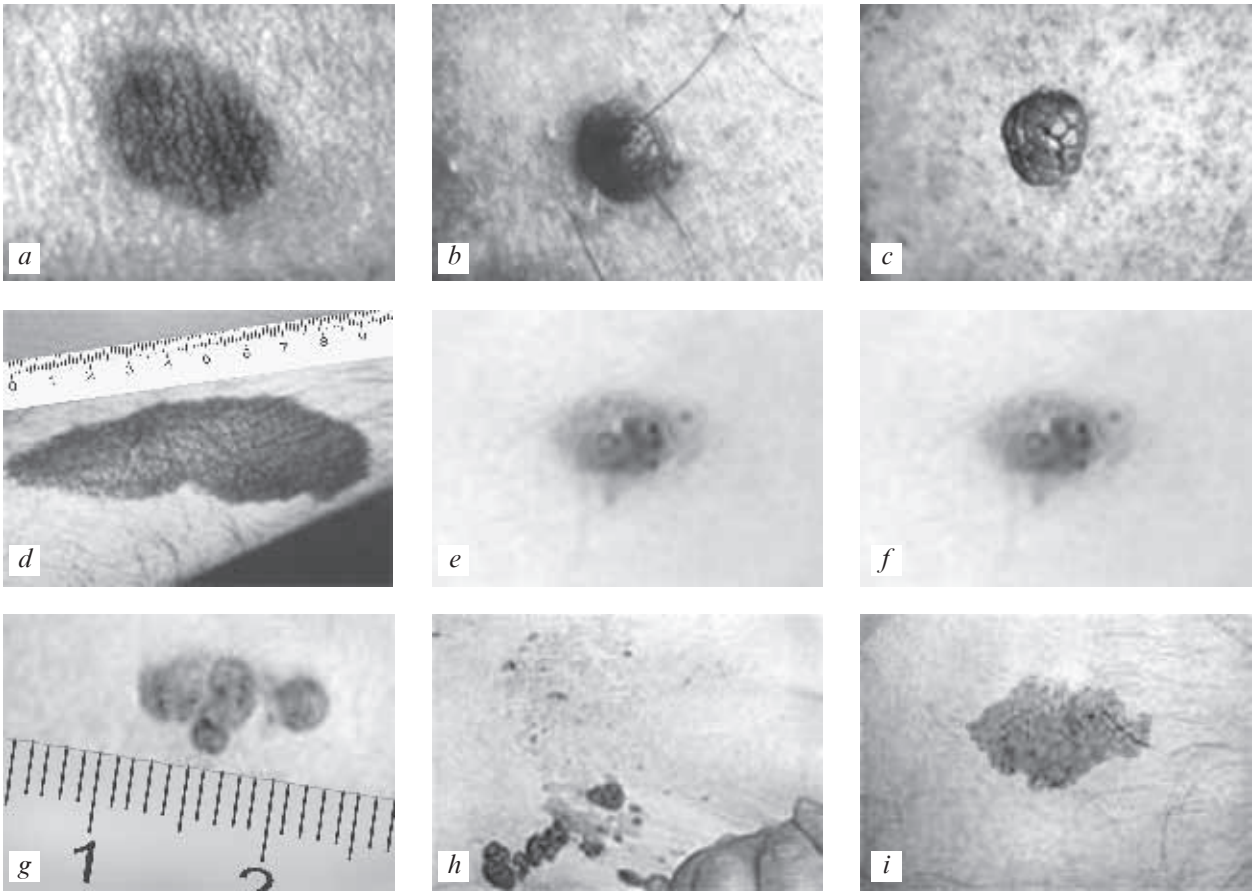


Fig. 2.1.22. Types of nevi: *a* — border nevus; *b* — complex nevus; *c* — intradermally nevus; *d* — congenital nevi; *e* — spotted nevus; *f* — dysplastic nevus; *g* — mongolian spot; *h* — warty nevus; *i* — nevus of the "coffee with milk" type

Other variants of nevi: epithelioid (Spitz nevus), Ota nevus, Ito nevus, Becker nevus — in children are rare.

The vast majority of melanocytic nevi of epidermal and dermal melanocytic origin do not need treatment.

Common indications for removal of nevi are:

— Cosmetic, since it is often found that the patient's desire is a sufficient argument for the removal of nevus, especially as during histological research in such nevi dysplastic changes are often revealed; signs of atypia often should be expected in the "ugly" melanocytic nevi.

— The availability of constant irritation. So first of all nevi should be removed, being constantly exposed by mechanical irritation, periodically increasing and/or changing the color, as well the elements, situated below the belt, bra straps or collar.

— Localization in the places, poorly accessible for self-control, such as the scalp and perineum.

— Unusual change or rapid growth of the nevi that does not match the rate of growth of the body, as well as nonsimultaneous changing of all nevi with a sudden change in color, sizes or topography of the individual entities.

— The presence of elements of high risk of malignancy, including cellular blue nevus, border nevus in children older than 13–15 years, dysplastic and/or congenital nevus loose, a large number of nevi, protruding above the skin at presence in a personal family history of melanoma, sun-induced freckles.

— Some features of localization, this applies to heavily pigmented nevi in acral sites and mucous membranes, especially if they are congenital and may be precursors of acral melanoma.

— Localization at the sites of the nail bed and the conjunctiva, where the presence of nevi should cause suspicion on melanoma and its precursors.

The require surgery if there is at least one of seven signs of malignancy of melanocytic nevi:

1. Rapid increase in the area or height of formation.
2. Increase in the intensity of pigmentation, especially if it is uneven.
3. Local regression.
4. Appearance of dark corolla or elements — satellites.
5. Inflammatory reaction in the nevus.
6. Itching.
7. Erosion and bleeding.

It should be noted that the change of the nevi at the period of puberty from the border (in the form of a flat spot) to the complex and intradermally (in the form of a papilla on the leg) without features of rapid growth and signs of dysplastic nevus is natural and non-dangerous. But, on the contrary, the presence of flat nevi in adolescents and young adults, particularly in the soles, palms and genitals is melanoma non-dangerous.

The main type of treatment of pigmented nevi is surgical excision with histological examination (under the microscope). Removal by other methods is possible, for example by laser, cryotherapy, but with only cosmetic purpose and only after a mandatory inspection of the oncologist. Such variations of removal do not remain the living tissue of the nevi, which excludes histology, and it is impossible to provide 100% absence of cancer.

TERATOMAS

Teratomas are embryonal cell tumors. It is a tumor that develops from three embryonic layers: ectoderm, mesoderm and endoderm. The name "teratoma" is derived from the Greek *teratos* — a monster, a freak and *onkoma* — tumor formation.

Teratoma (synonyms: a complex tumor embryoma, mixed teratogenic tumor, tridermoma, monodermoma, parasitizing fruit) — a tumor, consisting of multiple tissue types, derived one, two or three germ layers, the presence of which is not peculiar to the organs and anatomical areas of the body, in which cancer develops.

Teratoma tumors occur primarily in children within the first years of life and at least — as a teenager, and make up about 6% among tumors of childhood.

In newborns and infants with tumor diseases teratomas are found in 22–25% of cases.

Localization of teratoid tumors in children is various: sacrococcygeal region — 38%, ovaries — 31%, retroperitoneal space — 12%, testicles — 6%, mediastinum — 4%, others — 9%.

Teratoma is a structurally heterogeneous tumor containing cystic and solid parts, may contain organs and organ systems (organoid teratomas) (Fig. 2.1.23).

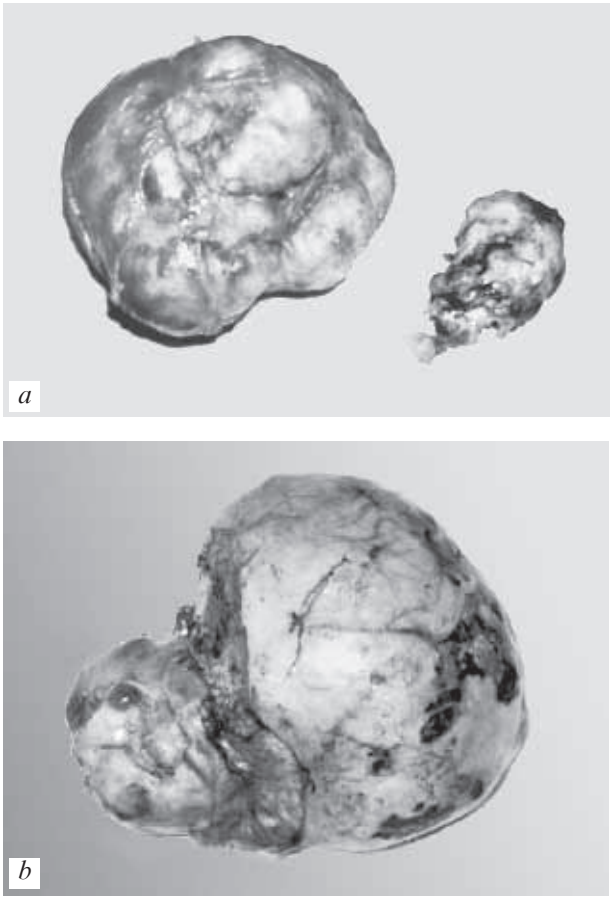
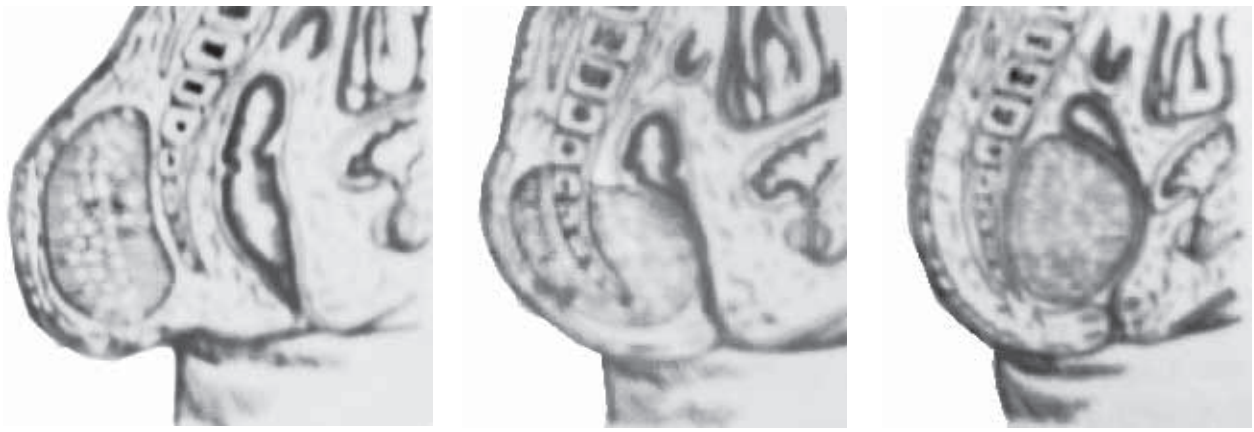


Fig. 2.1.23. Macroscopic view of teratomas *(a, b)*



a *b* *c*

Fig. 2.1.24. Localization of sacrococcygeal teratomas (G. A. Bairov Urgent surgery of children. p. 413, 1999): *a* — an external location, *b* — external-internal location; *c* — the internal layout

As usually skin, teeth, CNS tissue, mucous membrane of respiratory tract and intestine, the cartilage are foound in teratomas. Benign teratomas contain only mature tissues (epithelium, cartilage, bones, teeth, etc.). Malignant teratomas can contain malignant embryonal carcinoma cells, yolk sac tumors, choriocarcinomas or a mixture of malignant cells. Benign cells sometimes coexist with malignant or tumor is composed entirely of malignant cells. The risk of malignancy of sacrococcygeal teratoma increases after 6 months of age.

Clinical manifestations of teratoma are varied and are largely determined by the location of the tumor (Fig. 2.1.24).

A common feature of teratomas in children is that they are usually benign and rare spreading in the nearby lymph nodes, lungs, liver or bones.

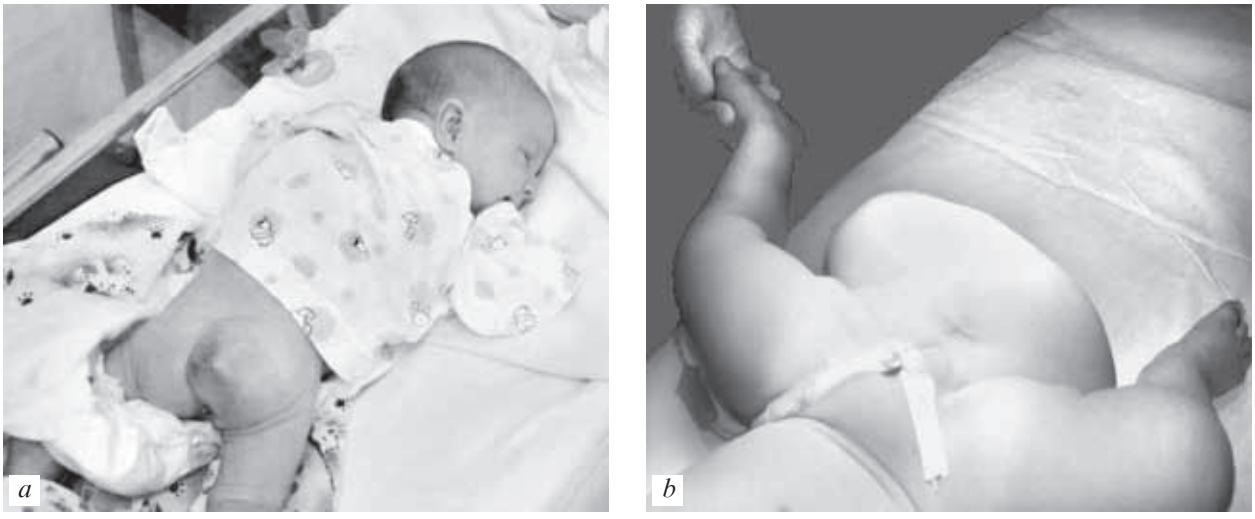
At external location of teratoma in sacrococcygeal region, it can be easily identified at birth because they localize on the buttock on average sacrococcygeal line (Fig. 2.1.25).

During external-internal location of teratoma except serving tumor there is another part of it, located between the sacrum and the rectum. During the growth of the inside part of the teratoma disorder symptoms of pelvic organs as disturbances in urination, constipation or fecal incontinence occur. Such teratoma is often fused with the coccyx, growing outwards and can be accompanied by disintegration of the skin, infection connection and bleeding.

Teratoma by touch can be smooth or bumpy (during malignant degeneration), the skin over it is unchanged or necrotic (dead skin) during large sized tumors.

The consistency of the tumor during benign teratoma is generally milder than during malignant. Skin temperature over malignant teratoma is usually raised, and vascular pattern is well marked. Unlike benign teratomas, malignant tumor types rarely reach large sizes.

During benign teratomas general condition of the child suffers a little if the tumor is not compressing the surrounding organs and tissues. Some terato-



a *b*

Fig. 2.1.25. Teratoma in sacrococcygeal region *(a, b)*

mas, which include immature tissue, produce alpha-fetoprotein that has prognostic significance. Increasing of alpha-fetoprotein in cases when teratoma contains immature cells indicates a high risk of malignancy.

In children with cancer there are always pronounced local symptoms. There is skin pallor, weight loss, retarded physical development, fever, increased ESR. The rise of the alpha-fetoprotein level thousand times is marked at teratomas, containing yolk sac carcinoma and embryonal carcinoma. Children are often restless, which is associated with pain.

Differential diagnosis

To differentiate teratoma of sacrococcygeal region is nesessary with meningomyelocele, pilonidal and dermoid cysts. In addition besides of visual inspection it is necessary to conduct a number of studies: X-ray, CT and MRI.

Treatment

In all the children with sacrococcygeal teratoma the tumor must be fully removed along with the coccyx. Formation of an anatomically normal zone is provided by stitching perianal sphincter to pre sacral tissues (Fig. 2.1.26).

Complications of teratomas

Teratomas may become malignant, at larger sizes trophics of teratomas violates and ulcers may appear (see Fig. 2.1.26, *b*), infection, bleeding, compression of organs during internal or external-internal position with symptoms of intestinal obstruction, urinary retention.

Urgent surgical treatment is indicated during development of the following complications:

1. Compression of the rectum or urethra by the tumor.
2. Rupture or sharp shell thinning at cystic forms of tumors.
3. Ulceration or skin necrosis, abscess of individual cystic cavities.
4. The rapid growth of the tumor, suspected malignant degeneration.

Ovarian teratoma has a clinical course without symptoms for a long time. The first sign of the dis-

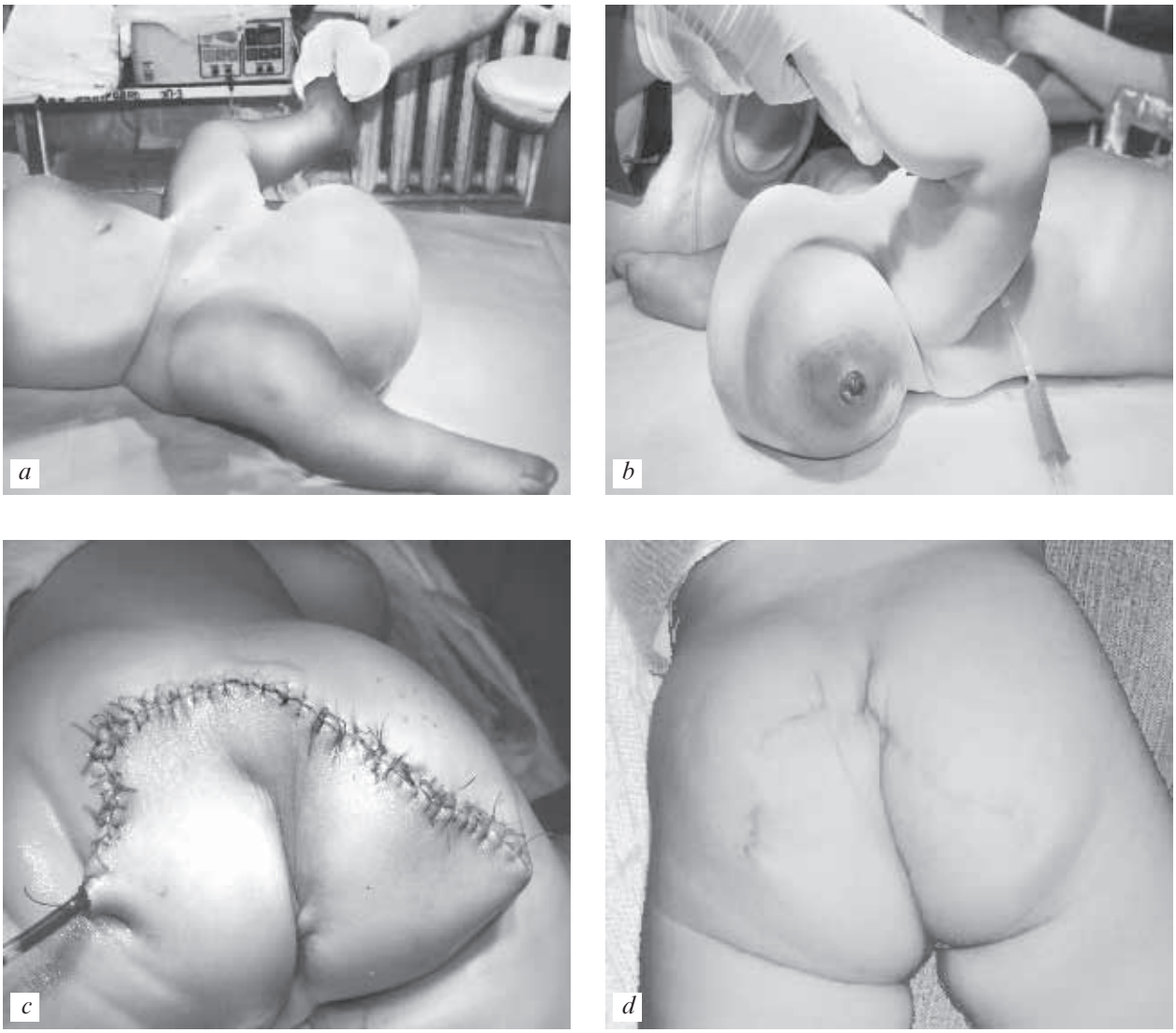


Fig. 2.1.26. Formation of an anatomically normal zone: *a, b* — types of teratoma before surgery, *c* — view of the perineum and buttocks immediately after surgery, *d* — view of the perineum and buttocks after 6 months after surgery

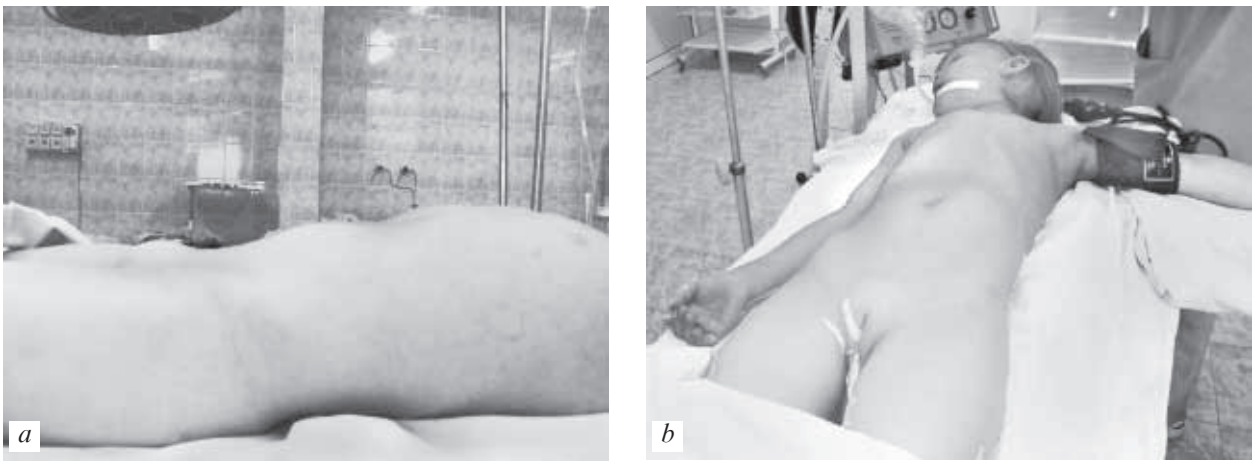


Fig. 2.1.27. The increase and the asymmetry of the abdomen in a girl with ovarian teratoma *(a, b)*

ease is usually an increase in the size of the stomach (Fig. 2.1.27).

Sometimes children complain of abdominal pain.

Torsion of the ovarian teratoma's pedicle can cause, except pain, vomiting, and muscle tension of abdominal wall (the so-called syndrome of "acute abdomen"). Also at torsion of ovarian teratoma in the absence of teratoma's pedicle, necrosis of the fallopian tube and ovary is possible (Fig. 2.1.28).

Retroperitoneal tumors cause symptoms, which are provoked by compression of the surrounding organs and tissues: recurrent pain, dilatation of vessels on the anterior abdominal wall, the presence of the tumor during abdominal palpation.

Diagnosis of ovarian teratomas is based on ultrasound, CT of the abdomen and pelvis, MRI, determination of the level of alpha-fetoprotein level (Fig. 2.1.29).

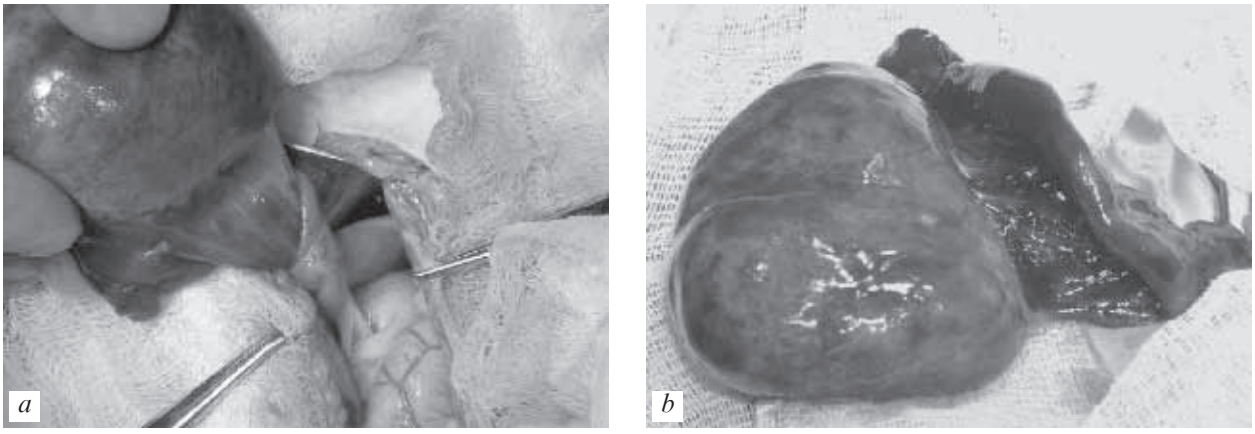


Fig. 2.1.28. Torsion of ovarian teratoma: *a* — without tube necrosis, *b* — with necrosis of the tube

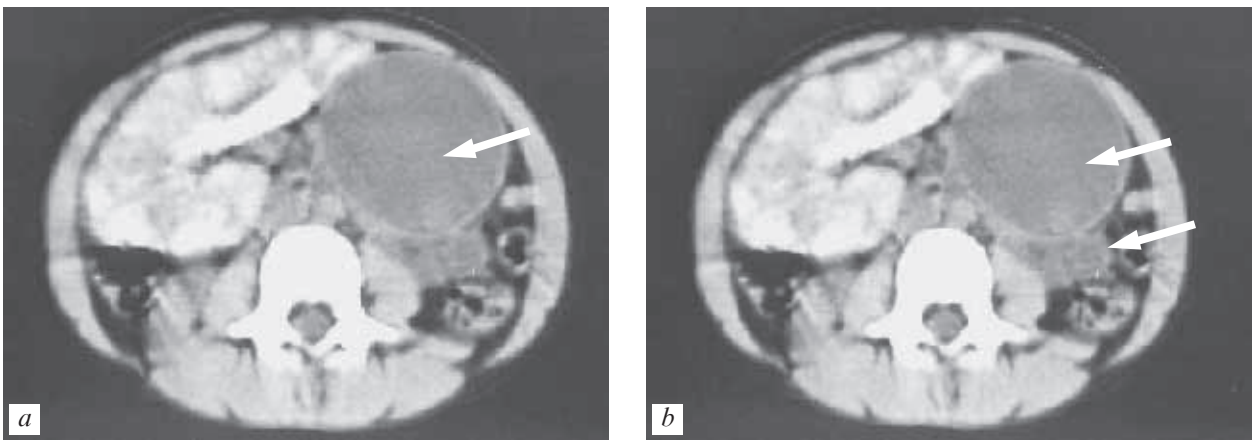


Fig. 2.1.29. CT of teratoma of the left ovary *(a, b)*

Treatment

Treatment of ovarian teratoma is only surgical, involves removal of the tumor with subsequent histological examination.

Patients with teratomas of external location (neck, testicle, etc.) usually have only local symptoms.

Prognosis

If the tumor is composed of mature benign tissue, surgical treatment (complete removal of ovarian teratoma) is the method of choice and is accompanied by a good separation results.

Recurrence after complete removal of benign teratoma is rare, sometimes recurrent tumor is malignant in nature. Malignant teratomas are extremely aggressive and have a high mortality rate.

2.2. MALIGNANT NEOPLASMS IN CHILDREN

Specific objectives:

1. Studying the diseases which cause tumors of soft tissues.
2. Recognize the main clinical manifestations of malignant tumors of soft tissues.
3. Differentiate tumors depending on the species.
4. Identify signs of malignant transformation and tumor complications (bleeding, inflammation, compression of neurovascular tumors, etc.).
5. Interpret the principles of treatment of malignant tumors of soft tissues and their complications.
6. Recognize the main clinical manifestations of bone neoplasms; distinguish the signs of malignancy disease course.
7. Identify the characteristics of the course of certain diseases on the basis of clinical and radiographic features.
8. Differentiate malignant bone tumors based on clinical examination and ancillary methods (X-ray, ultrasound, computed tomography, angiography, puncture and open biopsy).
9. Make a plan of patient's examination and the algorithm of actions to identify and prevent complications of bone tumors (pathological fractures, false joints, metastasis, etc.).
10. Interpret the principles of surgical treatment, chemotherapy, radiation therapy, depending on the type of bone tumor and benign or malignant course.

MALIGNANT TUMORS OF SOFT TISSUES

MELANOMA

Melanoma is a malignant tumor that forms at melanocytes cell degeneration, which normally are found predominantly in skin and produce under ultraviolet radiation a coloring matter — a pigment melanin. The number of these cells and the pigment produced by them determines the color of human skin. Melanocytes are rich in nevi, called birthmarks in everyday life, so melanoma can also develop from these fairly innocuous formations, which are available in most people.

Melanoma occurs about 10 times rarer than skin cancer, and is about 1–4% of the total number of malignant tumors. In Europe, during the year the tumor is detected in 4–6 people per 100,000 population. The highest incidence is observed in Australia and the USA. Epidemiologists say that every 5–10 years the number of cases of melanoma among Caucasians doubles. A similar trend is occurring in our country.

Melanoma is usually painless, flat formation on the skin (flat melanoma) or nodule (nodular melanoma). Color of melanoma may be different: black and blue, brown or pink. Sometimes the tumor can have multiple colors (non-uniform distribution of pigment).

Flat melanoma develops with the same frequency both on the intact skin and from the pigment nevus. It is a plaque of irregular shape with scalloped contour, focus of discoloration, mosaic coloring, and keratosis on the surface. On average, in a few years there is a node on the plaque, that marks the transition of the tumor in a very aggressive form.

Nodular melanoma, which makes 10–30% of all melanomas of the skin, is the most aggressive type of tumor. Normally it is a node, rarely — a polypoid mass on the skin. For several months there is doubling of node volume, its early ulceration and bleeding.

The most common localization of melanoma is the skin of back, neck, head and limbs. However, melanoma can develop under the nail (about 8% of all melanomas of the skin). Usually a dark spot forms under the nail plate in this case, making it very difficult for timely diagnosis.

Melanoma is curable at an early stage, so we need to pay attention to ourselves and do not miss the first "alarm", indicating a possible malignant nature of formation on the skin. It is the thicker the more dangerous. At an early stage (thin and flat tumors) surgical excision helps to get rid of the tumor for a period of 5–10 years in more than 90% of the patients. In contrast, no more than 50% of patients with tumors thicker than 4 mm, and particularly ulcerated have a five year survival without a return of the disease. Chances of recovery decrease sharply at trying self-removal (cutting, tying "pedicle" of the tumor, burning by various chemical substances, etc.).

There is a kind of "alphabet of melanoma", describing a number of signs of degeneration of birthmarks identified by the first four letters of the Latin alphabet:

A (asymmetry) — asymmetric: the form of "good" birthmarks is more often symmetrical.

B (border irregularity) — edges of birthmarks are usually smooth and crisp. Irregular, scalloped contour is more typical for melanoma.

C (color) — benign nevi are colored more or less evenly. Uneven color of different parts of the tumor is more typical for degenerative birthmarks.

D (diameter) — diameter of birthmarks more than 6 mm: the larger the birthmark is, the greater

the probability of its degeneration. Nevertheless, 1 mm melanomas occur. (Fig. 2.2.1)

Malignant transformation is indicated by various changes of pre-existing birthmarks. It is found that the pigmented lesions on the skin which regularly change shape and color turned out to be melanoma 4 times more often than those remained unchanged. Therefore, to the first four letters of "melanoma alphabet" one can add the fifth one.

E (evolving) — the appearance of any external changes of birthmark, which most often is:

— color change (decrease or a sharp increase of pigmentation);

— infringement or complete absence of skin picture in the nevus area, "lacquer" surface or peeling;

— appearance of inflammatory areola around birthmarks (red in the form of corolla);

— changes of configuration on the periphery, the contour blurring of nevus;

— increasing the size of nevi (especially those aged over 30 years) and its seal;

— itching, burning, and tingling in birthmarks area;

— cracks, ulceration in the field of birthmarks, bleeding;

— the loss of available hair on the birthmark;

— a sudden disappearance of birthmark (especially after tanning in the sun or in a solarium).

Complications of melanoma. The main complication of *melanoma* is metastasis (i. e. spread and damage to other organs and tissues).

Melanoma spreads through the blood vessels (the hematogenous route), with metastases can settle anywhere in the body: the liver, lungs, bones, brain, as well as the lymphatic system, and the lymph nodes are affected.

Treatment of melanoma. *Melanoma* is excised surgically, with the capture of a healthy skin about 2–3 cm, with subcutaneous fat and muscle.

Other methods of treatment include radiation therapy, immunotherapy, laser destruction, and cryosurgery.

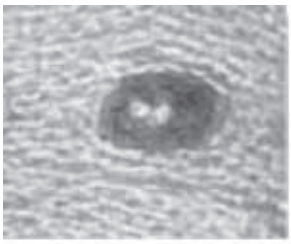
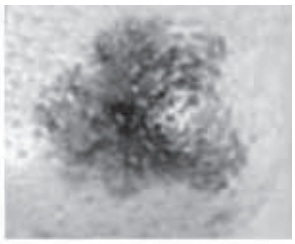
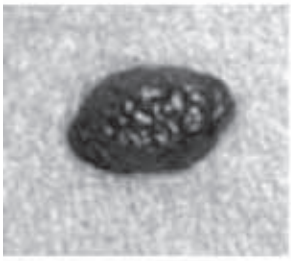
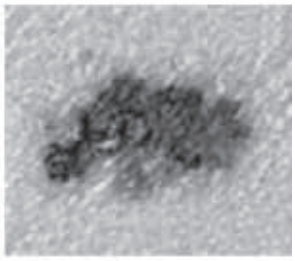
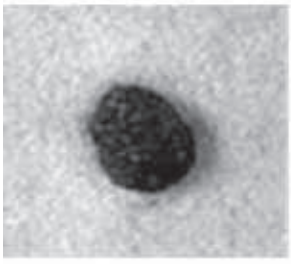
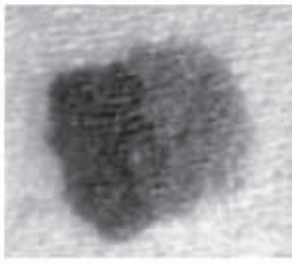
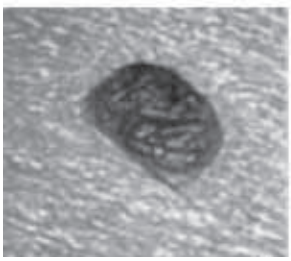
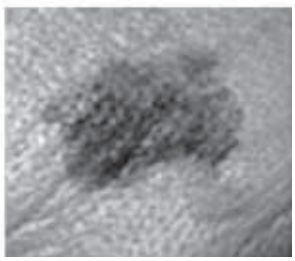
Normal Mole	Melanoma	Sign	Characteristic
		Asymmetry	When half of the mole does not match the other half
		Border	When the border (edges) of the mole are ragged or irregular
		Color	When the color of the mole varies throughout
		Diameter	If the mole's diameter is larger than a pencil's eraser

Fig. 2.2.1. Signs of benign nevus degeneration

RHABDOMYOSARCOMA

Sarcomas are malignant tumors that develop from connective tissues of the body, in particular, from the muscles, fat tissue, membranes that line the joints and blood vessels.

Rhabdomyosarcomas develop from rhabdomyoblasts — cells that form the skeletal muscle during maturation.

About 85% of rhabdomyosarcomas are diagnosed in infants, children and adolescents. Most often, these tumors arise in the head and neck (40%), urinogenital (27%), the upper and lower extremities (18%) and trunk (7%).

Rhabdomyosarcomas are divided into three types:

Embryonal rhabdomyosarcoma is detected most frequently and are usually located in the head and neck, bladder, vagina, prostate and testicles. This type of rhabdomyosarcoma is usually diagnosed in infants and young children.

Alveolar rhabdomyosarcoma is found in the large muscles of the trunk and limbs and usually affects older children and adolescents.

Pleomorphic rhabdomyosarcoma occurs mostly in adults and rarely in children.

Rhabdomyosarcoma is approximately 4% of the total number of tumors in children. Over 90% of rhabdomyosarcomas are diagnosed up to 25 years and 60–70% — at the age 10 years. They observe two major peaks of disease — early (at the age of about 2 years) and late — (from 15 to 19 years). Males get sick more often than females in ratio 1.4–1.7 : 1.

Diagnosis. One third of patients with rhabdomyosarcoma are diagnosed at early stages, when complete removal of the tumor is possible. However, during the detailed examination some metastases are found in 80% of cases, for treatment of which it is necessary to use chemotherapy.

Many cases of rhabdomyosarcoma occur in areas where they can be easily detected, for example, an eyeball or in the nasal cavity. Protrusion of the eyeball or runny nose makes to see a doctor, which helps early to suspect tumor and conduct a survey. If rhabdomyosarcoma occurs on the surface of the body, it can be easily detected without special investigations.

Rhabdomyosarcomas in the field of the testis usually develop in young children and are revealed by their parents during the washing of the child. Tumors of the urinary tract cause difficulty in urination or bleeding, so these symptoms cannot be ignored.

Rhabdomyosarcoma is harder to detect on the limbs and trunk in older children, so they cause the symptoms (pain, swelling), occurring during the game.

Currently, there are no methods of screening (before symptoms detection) of rhabdomyosarcoma. Families with multiple cases of malignant tumors, especially in children should be kept under special medical supervision.

The first sign is usually the appearance of local induration or swelling that does not cause pain or other problems. It conserns in particular the rhabdomyosarcoma of trunk and extremities.

At the location of the tumor in the abdomen or pelvis vomiting, abdominal pain or constipation may occur.

In rare cases, rhabdomyosarcoma develops in the bile ducts and leads to jaundice.

If you suspect a tumor a type of biopsy (taking a piece of tissue for microscopic examination) is performed, which specifies option of rhabdomyosarcoma.

After extensive survey they specify degree of distribution of tumor and the treatment plan.

Depending on the tumor spread and radicalism of surgery, patients with rhabdomyosarcoma are divided into 4 groups:

• Group 1 (16%) — patients (children or adults) with localized process (without visible metastases) with a fully deleted tumor.

• Group 2 (20%) — patients with localized but not completely deleted microscopic tumor, the presence of regional lymph nodes and the absence of distant metastases.

• Group 3 (48%) — patients with incompletely deleted tumor, visible to the eye, and the absence of distant metastases.

• Group 4 (16%) — children and adults with distant metastases at the time of diagnosis.

To determine the stages of rhabdomyosarcoma the TNM system can be used, which takes into account the prevalence of the process, but does not take into account the results of the surgery.

The symbol T is the size of the tumor, N — regional lymph nodes involvement and M — the presence of distant metastases.

Treatment

Surgery. The first step in the treatment of rhabdomyosarcoma should be the *complete surgical removal* of the tumor, which is carried out under general anesthesia.

If for any reason the entire tumor can't be removed, then most part of it is excised. In addition, the tumor is removed as much as possible within the healthy tissue that is specified during the study under the microscope.

During the operation nearby (regional) lymph nodes are also removed. When placing rhabdomyosarcoma in the head and neck areas, the complete removal of the tumor is often impossible without breaking the vital functions or serious cosmetic defect. So the operation can be delayed until a pronounced effect of chemotherapy or radiation.

Chemotherapy. All patients with rhabdomyosarcoma usually receive chemotherapy. If, for any reason, chemotherapy is not granted, the probability of relapse is significantly increased, even after complete removal of the tumor.

In the case of residual rhabdomyosarcoma is recommended radiation therapy along with chemotherapy. Upon reaching the pronounced effect the second attempt of complete removal of the tumor is taken.

The main drugs for the treatment of rhabdomyosarcoma are vincristine, dactinomycin, and cyclophosphamide. In patients with metastatic tumor chemotherapy with these drugs is not as effective and is being sought of new anticancer drugs and their combinations.

Radiation therapy. Radiation therapy is an effective method of influence on tumor cells remaining after surgical removal of rhabdomyosarcoma.

Radiation therapy is usually appointed through 6–9 weeks after completion of chemotherapy in the remaining area of the tumor. In the case of rhabdomyosarcoma in the area of localization shells or medulla of the brain and spinal radiation therapy may be assigned immediately to the use of chemotherapy.

Bone marrow transplantation. Bone marrow transplantation allows using high doses of chemotherapy. Anticancer drugs do not have the electoral actions and kill and injure not only the tumor but also fast-growing normal cells.

This leads to anemia as a result of reducing the number of red blood cells, increased susceptibility to infections due to low white blood cell count and bleeding due to low platelet count after high-dose chemotherapy.

Transplantation of bone marrow, taken from the patient before chemotherapy, can solve these problems. After completion of chemotherapy in advance bone marrow is again returned to the patient that allows to quickly restoring its function. Instead of bone marrow transplantation can be used fence and subsequent transfusion of peripheral stem cells, which leads to a similar effect.

Prognosis. In general, more than 70% are successfully treated from rhabdomyosarcoma. Many factors affect on the survival of patients, such as the incidence of tumors at diagnosis, localization, radical surgery and histological variant.

Survival rates ranges from 30% in group 4 to 90% in group 1. During localization of rhabdomyosarcoma at the head and neck areas, vagina and testes prognosis is favorable.

In patients with alveolar rhabdomyosarcoma prognosis is worse than during embryonic.

The best results are in patients, receiving treatment in specialized units.

MALIGNANT BONE TUMORS

OSTEOGENIC SARCOMA

Osteosarcoma is the most common primary bone tumor in children, 6th by frequency among all malignant tumors of childhood. The tumor originates from primitive bone-forming mesenchyme, is characterized by production of osteoid during malignant proliferation of spindle cell stroma. The peak of incidence occurs on the second decade of life. Boys have higher incidence of the disease, whereas at earlier age girls mostly suffer (having at that time the bone age higher than boys).

Etiology. Tumor development has some connection with the rapid growth of the bone. Children with osteosarcoma are usually taller than age norm, and the disease affects the most rapidly growing parts of the skeleton.

The development of bone tumors is often associated with trauma, but rather injury attracts the doctor and makes to hold X-ray.

The only agent of the environment known as a stimulator of bone sarcomas is ionizing radiation. The interval between the influence of this factor and the occurrence of osteosarcoma can be from 4 to 40 years (on average 12–16 years old).

Among people, who suffer from Paget's disease 2% suffer from osteosarcoma, often with multiple lesions of bones.

The presence of benign bone tumors (osteochondroma, enhondroma, etc.) increases the risk of osteosarcoma.

Clinical picture. The main clinical feature of osteosarcoma is pain over the affected area. Dull pain, constant with a gradual increase in intensity. The characteristic symptoms are nocturnal pain. Soft-tissue component may be present in 3/4 patients. Limb is increased in volume, often looks swollen. Pain and increased volume lead to functional disorder. Duration of anamnesis makes an average 3 months.

Typical are lesions of metaphases of long bones. The most frequent localization (about 50% of cases) — the area of the knee, hip and distal part of the proximal part of the tibia. Often the proximal part of the humerus and femur also is affected, and the middle third of the femur. The defeat of the flat bones, especially the pelvis in children occurs in less than 10% of cases.

Osteosarcoma has a great tendency to develop hematogenous metastases. By the term of diagnosis 10–20% of patients already have macro metastases in the lungs, revealed by X-ray. But about 80% of patients at diagnosis have micro metastases in the lungs, undetectable by X-ray, but visible on computed tomography. As the bones do not have developed lymphatic system, early dissemination of osteosarcoma in regional lymph nodes is rare, but if it takes place, it is a bad prognostic sign. Other areas of metastasis are bones, pleura, pericardium, kidneys, central nervous system.

Osteosarcoma has local aggressive growth, can spread on the epiphysis and the nearby joints (knee and shoulder joints are most severely affected), extending along the articular structures through articular cartilage, pericapsular space or straightway, because of a pathological fracture, and form do not adjacent to its foci — satellites — "skip"-metastases.

Diagnosis. Complete X-ray examination.

This study allows suspecting the presence of osteosarcoma in a patient, and reveals the presence of soft tissue component, pathological fracture, determines the size of the tumor, and the optimal level of the biopsy.

Radiographic signs of osteosarcoma (Fig. 2.2.2):

— metaphyseal localization in the long tubular bones;

— the presence of sclerotic and lythic lesions in the bone, the presence of vascularization;

— pathological foci of osteoformations in soft tissues;

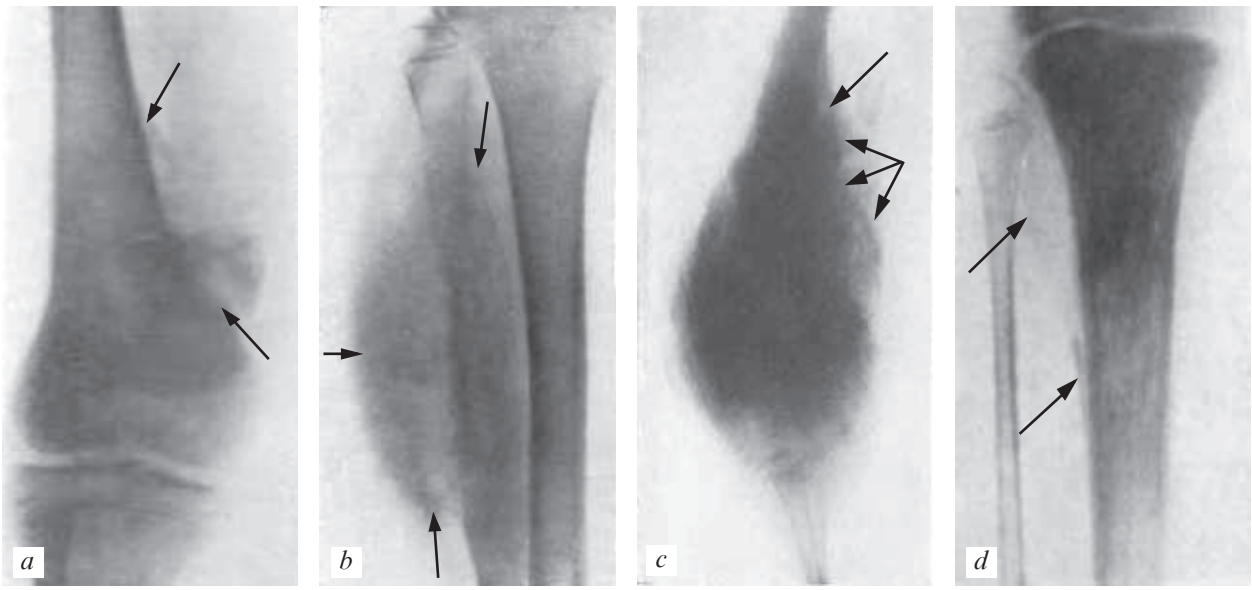


Fig. 2.2.2. Radiographic signs of osteosarcoma: *a* — osteogenic sarcoma of the femur, *b* — a tumor of the fibula, *c* — osteogenic sarcoma of the humerus, *d* — the defeat of the tibia

— violation of the integrity of periosteum with formation of "visor" or "Kodmen's triangle";

— Pin periostitis — "spicules" (swelling of the periosteum in the form of needles, placed perpendicular to the bone surface);

— X-ray of lungs reveals macro metastases.

Morphological examination of the tumor

One of the frequent causes of the impossibility of sparing surgery is unsuccessful biopsy with local colonization by tumor cells, the development of pathologic fracture. Therefore, a biopsy should be performed by the surgeon and preferably to carry out trepan biopsy than the knife one (for maximum protection of tissues, adjacent to the tumor areas from contact with the biopsy specimens).

Bone scan (BS) with Te-99 allows revealing other centers in the bones, although increasing accumulation of the isotope is not specific. During the BS in the dynamics on the change of percent of accumulation of the isotope in the outbreak before and after chemotherapy, can quite accurately judge about effectiveness of chemotherapy. A significant decrease in the percentage of accumulation of the isotope in the outbreak precisely enough correlates with a good histological tumor response on chemotherapy.

Computed tomography (CT) of the source allows revealing the exact location of the tumor, its size, the ratio of the tumor to the surrounding tissues, its extension on the joint. Lungs CT allows revealing micro metastases which were not revealed by X-ray.

Magnetic resonance imaging (MRI) is the most accurate method of staining of the tumor, revealing its relation to the surrounding tissue, the neurovascular bundle, and allows determining the dynamics of the process during chemotherapy, its effectiveness and, consequently, the amount of planned operations. Currently, MRI is performed with contrast, containing gadolinium, which accumulates at the periphery of the tumor, clearly limiting it.

The largest cancer hospitals of the world use an advanced method — DEMRI — dynamic capture of contrast material, identified on MRI. During using a computer is performed quantification (in%) of tumor cells, accumulating contrast before and after chemotherapy, thereby defining the histological tumor response to treatment in the preoperative period.

Angiography is carried out before the operation. Using this method detecter whether vessels are free from the tumor or not, which determines the volume of the operation. At presence of tumor emboli in the blood vessels, organ-sparing surgery is impossible.

Treatment. Until recently, the treatment of osteosarcoma was limited by operation (e. g., amputation of limbs). But, the survival rate was very low — 15–20%. Significant progress has been achieved in the treatment of the last two decades as a result of adjuvant chemotherapy, and technical means of diagnosis.

The most important components of treatment of osteosarcoma.

1. Preoperative chemotherapy is always the first stage of treatment and has certain advantages:

— Improving the system tumor — control — the suppression of micro metastases in the lungs.

— Better control of the primary lesion, which extends the capabilities of reconstructive surgery on the limbs.

— The ability to objectively evaluate the histological tumor response on chemotherapy, which often determines the prognosis of the disease and the management of further treatment.

In modern programs of osteosarcoma treatment medications are used: high–methotrexate (12.8 g/m^2), adriblastin, ifosfamide, platinum drugs (carboplatin, cisplatin), and etoposide.

2. Operation is the second required stage of the treatment.

Developing the capacity of modern surgery and means of control of primary tumor can improve quality of life by applying organ preserving surgery. So the common practice in the world is implantation of joints endoprosthesis, as well as replacement of resected bone by auto- and allotransplant. However, conducting organ preserving operations may not be possible for certain reasons:

— cancerous neurovascular bundle;

— pathological fracture (now is not an absolute contraindication for limb salvage);

— improperly chosen place of biopsy;

— infected biopsy places;

— too young age of the patient (as there is a large reserve of growth of the limbs), but for the upper limbs it does not matter;

— prior operations with colonization of soft tissues;

— large sizes of the tumor with a significant involvement in the process of soft tissues.

Presence of metastases at the modern treatment is not a contraindication for organ preserving surgery.

If it is impossible to make organ preserving surgery is performed amputation with subsequent prosthetics of limbs. Amputation at the modern prosthetics leaves great opportunities for an active lifestyle and even sports.

3. Post-surgery chemotherapy is conducted taking into account histological tumor response on chemotherapy (continued treatment, or changing the therapeutic regimen, or cessation of therapy).

Question of conducting radiotherapy is being discussed in the national literature. However, in most major foreign clinics that question has not been solved in favor of this method of treatment. As an example, a 20-year experience with preoperative radiotherapy followed by surgery at Children's Hospital St. Jude (Memphis, USA): 84 patients received preoperative radiotherapy, 61 did not receive radiotherapy. 5-year survival rate (without HT) was less than 20% in both groups.

Currently, the development of metastases does not predetermine fatal disease. Level of modern thoracic surgery allows removing large number of metastases, spending subsegmental resection. During this they also use a pre-operative chemotherapy with the definition of histological tumor response. As chemical preparations of the 2nd line carboplatin and etoposide are used.

The use of these drugs and radical removal of metastases can reach a 5-year survival in 55% of patients with secondary pulmonary metastases (COSS96).

During the ineffectiveness of standard chemotherapy they use high-HT with colony-stimulating factor and peripheral stem cell transplantation.

Currently, attempts are made applying other drugs, in addition to chemotherapy:

Liposomal muramyl tripeptide (a synthetic analogue of a lipophilic muramyl dipeptide). It is encapsulated on liposomes and has selective affinity to lung macrophages and circulating monocytes, activating them and giving them tumors properties.

Verapamil and cyclosporin A — according to researchers, reduces the expression of P-glycoprotein in the tumor, thereby improving the digestibility of drugs by tumor cells. The good effects of these drugs together with ifosfamide and etoposide during common forms of osteogenic sarcoma are marked.

EWING'S SARCOMA

Ewing's sarcoma is the second most frequent malignant tumor of bones in children (after osteosarcoma), was first described by J. Ewing in 1921 and was named after the author. This tumor is rare in children under 5 years and adults over 30 years. Most often, this tumor occurs in adolescents aged 10 to 15 years. There are cases of non-bones Ewing's sarcoma with lesions of soft tissues.

Currently, there are several risk factors associated with the occurrence of Ewing's sarcoma.

Sex. Ewing's sarcoma is slightly more common in boys than girls.

Age. 64% of cases of Ewing's sarcoma occur between the ages of 10 to 20 years.

Race. Ewing's sarcoma is most commonly observed at the white population.

Clinical picture

1. Pain. The first symptom is pain, which, in contrast to the inflammatory process, do not subside at rest (increased at night, independently from the fixation of the limbs).

With the growth of the tumor a nearby joint function suffers, and then palpated tumor develops, often with pathological fracture (a late sign).

On the third–fourth month from the onset of the disease because of the pain at first disturbs, and then stops the movement in near joint.

2. Raising the temperature of the patient.

3. The presence of tumor formation.

In the area of a growing tumor there is swelling, redness, dilated subcutaneous veins and local increase of the temperature.

4. There are possible abnormalities in the affected limb.

5. Sometimes there is fever.

X-ray diagnosis:

— during microfocal destruction (localization in meta-epiphyseal sections of the tubular and flat bones), X-ray reveals many small round or oval, poorly defined areas of bone loss;

— during lamellar destruction (loss of diaphysis of tubular bones) is a longitudinal bundle of cortical substance on several plates;

— much less frequently macrofocal destruction occurs, accompanied by the formation of large outbreak of round or oval shape, like swelling of the bone, in which can be partitions which gives it a kind of cellular cystiform.

Endosteal reaction is also expressed in differend degree from sclerotic rim on the edge of destruction foci (Fig. 2.2.3), up to a sharp compression of the bone against which is barely differentiated medullary canal. It is believed that for Ewing's sarcoma is characterized periostitis. The peak shape of periosteal proliferations can be linear layered, bumpy, fringed, needle (spicules shaped). However, in flat bones with-

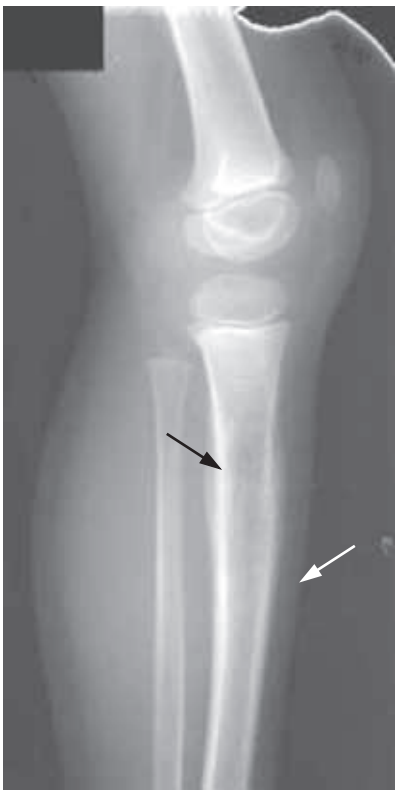


Fig. 2.2.3. Ewing's sarcoma of the tibia in a child (X-ray)

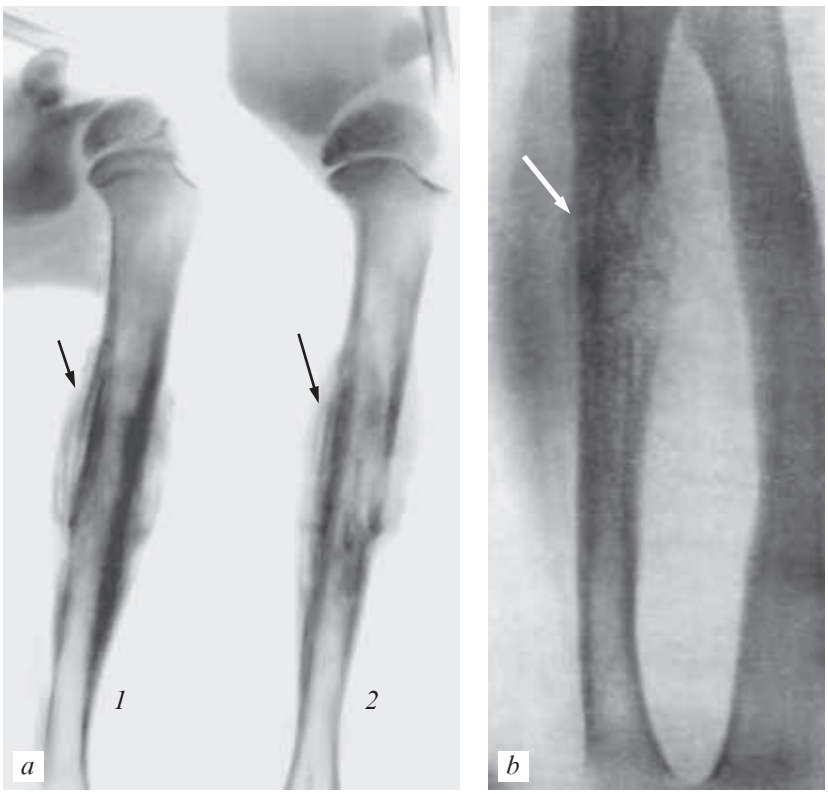


Fig. 2.2.4. X-ray: *a* — the humerus and the shoulder joint (*1* — straight; *2* — axial projections), *b* — of the ulna at Ewing's tumor: multilayered periosteal reaction is noted throughout the shaft

out periosteum, and metaphyses this reaction may be absent. More common is linear periostitis — jet bony growths are deposited along the diaphysis by parallel layers (Fig. 2.2.4). During the needle periostitis they look like needles, placed perpendicular to the bone (Fig. 2.2.5).

— A biopsy of the tumor. Sufficient amount of material can sometimes be obtained from the soft tissue component. If this is not possible, the material is obtained from the site of the bone, bordering with medullary canal

— X-rays and CT of lungs

— Aspiration biopsy or bone marrow from multiple locations (as Ewing's sarcoma has a tendency to metastasize in the bone marrow)

— Bone scan allows revealing other foci in the bones, as there may be multiple bone metastases.

— Computed tomography of the outbreak most accurately determines the size of the tumor and its connection with the surrounding tissue, the neurovascular bundle, the spread of tumor on bone marrow canal

— Angiography

— Ultrasonography

Treatment

— Multicomponent chemotherapy (using drugs: vincristine, adriamycin, ifosfamide, cyclophosphamide, actinomycin, vepeside in combination). In modern treatment programs apply preoperative and postoperative chemotherapy, taking into consideration also the histological tumor response to treatment. Good tumor response on chemotherapy is the presence of less than 5% of live tumor cells

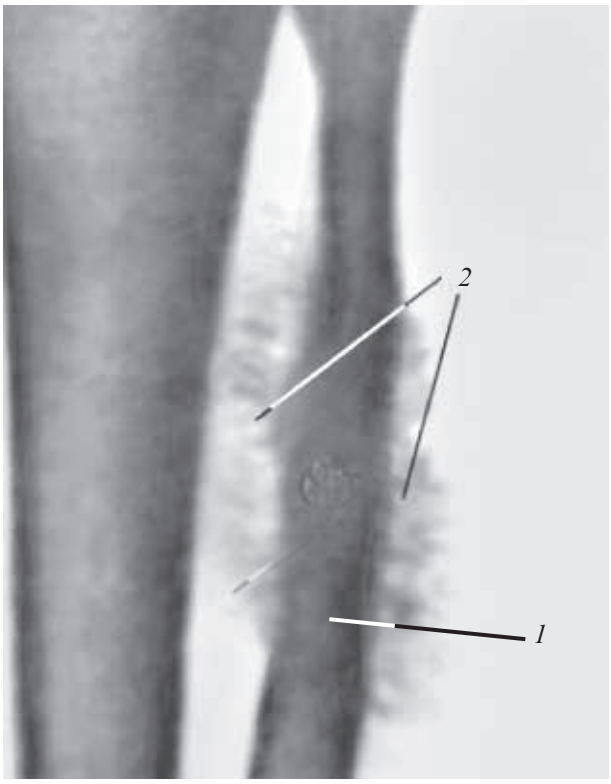


Fig. 2.2.5. X-ray of the proximal of shin bones during Ewing's tumor (straight projection): *1* — diaphysis of the fibula is thickened and deformed, medullary canal in the area of destruction is not differentiated, *2* — massive needle (spicules shaped), periosteal proliferation

— Radiation therapy on the locus in high doses. During the development of lung metastases radiation therapy on the lungs is made

— If possible, radical removal of the tumor (including bone and soft-tissue component.) Radical resection is possible with the outbreak of the fibula, the bones of the forearm, ribs, clavicle, and scapula

Operation improves local tumor control. In combination with intensive chemotherapy and radiation therapy the risk of local recurrence is significantly reduce. Reducing the frequency of local recurrence is marked even after non-radical operations. Modern surgical technique allows organ preserving operations at femoral lesions, shoulder bones, and resection of the pelvis.

Patients with a poor prognosis, particularly with bone metastases and bone marrow with survival less than 10%, in recent years is prescribed more intensive treatment — chemotherapy of drugs megadoses with total body irradiation and autologous bone marrow transplantation or peripheral stem cells. This therapy can cure more than 30% of patients with advanced process (during metastases in the bone and bone marrow). In patients with good sensitivity of the tumor even better results of treatment can be achieved (a 7-year survival rate is about 50%).

NEPHROBLASTOMA AND NEUROBLASTOMA

Specific objectives:

1. Master the characteristic symptoms of "palpable tumor" in the stomach during nephroblastoma and neuroblastoma.
2. Distinguish nephroblastoma and neuroblastoma, depending on their clinical needs and stage of the disease course.
3. Interpret ancillary data of research techniques (ultrasound, X-ray, urography, pneumoperitoneum, CT, puncture biopsy).
4. Conduct differential diagnosis of nephroblastoma, neuroblastoma, hydronephrosis, polycystic, a doubling of the kidneys, liver tumors, tumors of the adrenal gland, lymphogenous tumors).
5. Explain the principles of integrated treatment of neuroblastoma and nephroblastoma depending on the stage of the disease.

Nephroblastoma in children

Nephroblastoma *(Greek nephros kidney + blastos germ, the embryo +–oma; synonym: Wilms' tumor, embryonal carcinosarcoma, embryonal nephroma),* diesontogenetic malignant tumor of the kidney in children, it is a three-component embryonal tumor, containing epithelial, blast and stromal elements (adenomyosarcoma). It accounts for about 8% of all childhood tumors. Most often (75%) in children aged 1 to 5 years, accounting for 97% of all renal tumors in this age. In most cases the neoplasm is detected sporadically, but in 1% of cases has family character. In rare cases extrarenal tumor location is observed: in the pelvis, ovary, uterus, retroperitoneal fat, and the groin area. It is described in details by a German surgeon Wilms (M. Wilms) in 1899 year.

The tumor occurs with equal frequency in children of both sexes, mostly in the age of 2–3 years, although described cases of nephroblastoma in newborns and adults. Sometimes bilateral lesions develop. Macroscopically, the tumor is a node, sometimes very large, clearly separated from the renal parenchyma (Fig. 2.2.6).

During microscopic examination in nephroblastoma they usually find tubules, sockets, pseudo glomerule, mesenchyme, muscle, cartilage, bone and other tissues and structures in various combinations.

Depending on the predominance of mesenchymal or nephroblastic components distinguish three main morphological variants of nephroblastoma:

— typical nephroblastoma, mesenchymal and nephroblastic (epithelial) components are presented in equal amounts;

— nephroblastoma with a predominance of epithelial component;

— nephroblastoma with dominant mesenchymal component.

Along with typical nephroblastoma tubular, cystic, fetal rhabdomyoma and anaplastic variants secrete. Among nephroblastoma they also identify mesoblastic nephroma, clear cell bone metastasis and malignant rhabdoid sarcoma, which have characteristic histological and clinical features.

Nephroblastoma metastasizes through the blood and lymphatic system: specific metastasis to the lungs, liver, lymph nodes, rarely in other organs. Sometimes the tumor grows into the inferior vena cava and the right atrium.

Classification. There are four stages of nephroblastoma:

Stage I — the tumor is localized in the kidney and doesn't germinate its capsule;

Stage II — the tumor extends beyond the kidney, but does not germinate own capsule, no metastases.

Stage III — the tumor grows its own capsule, perirenal cellular tissue or psoas and adjacent organs, there is regional lymph nodes, tumor rupture before or during surgery.

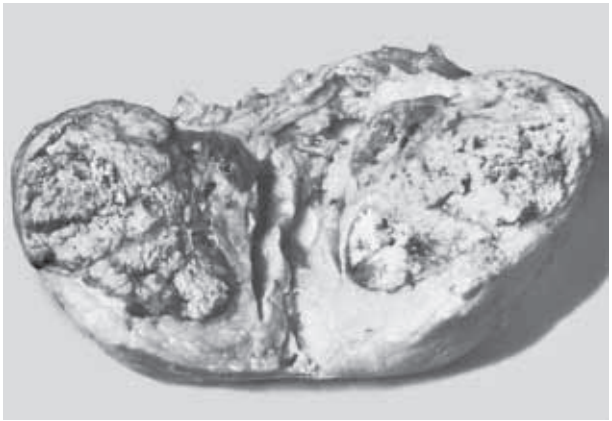


Fig. 2.2.6. Sample of the kidney during nephroblastoma: on the cut is seen a large tumor node with areas of necrosis, clearly separated from the renal parenchyma

Stage IV — the presence of distant metastases (into the lungs, liver, bones and other organs).

Some authors call bilateral nephroblastoma V stage.

Clinically, the disease is often asymptomatic; the tumor is detected incidentally during abdominal palpation. Pain, drowsiness, lack of exercise, poor appetite, elevations in body temperature, abdominal discomfort occur in the later stages of tumor development, as well as hematuria, usually observed in the renal pelvis location of the tumor. Sometimes nephroblastoma is combined with birth defects: aniridia, hemihypertrophy, cryptorchidism, pseudohermaphroditism, gonadal dysgenesis, Bekuit's–Wiedermann syndrome, duplication, etc. The discovery of such anomalies, as well as cases of nephroblastoma in relatives of the child is signs of possible development of nephroblastoma.

Diagnosis. Methods of early diagnosis are the inspection and palpation of the abdomen, allowing identifying its increasing or asymmetry, and palpate tumor. Diagnosis of retroperitoneal neoplasms is difficult for traditional methods, due to the anatomical and topographical features, the long absence of clinical symptoms or absence of specific symptoms. So by the time of palpation, abdominal pain, fever, anorexia, hypertension and/or hematuria is detected, 80% of children have distant metastases.

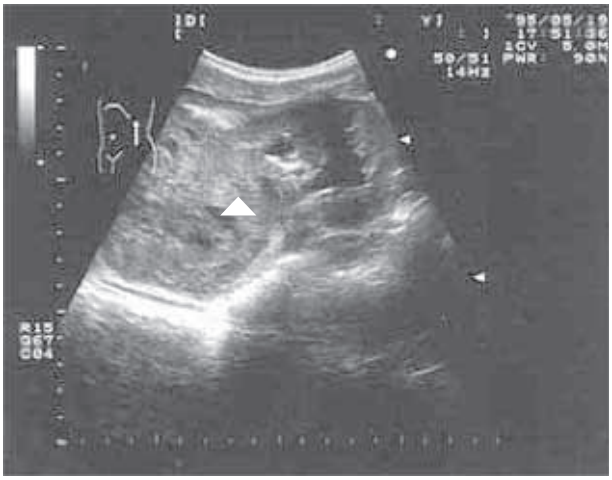


Fig. 2.2.7. Ultrasound scanning of kidney tumors (nephroblastoma)

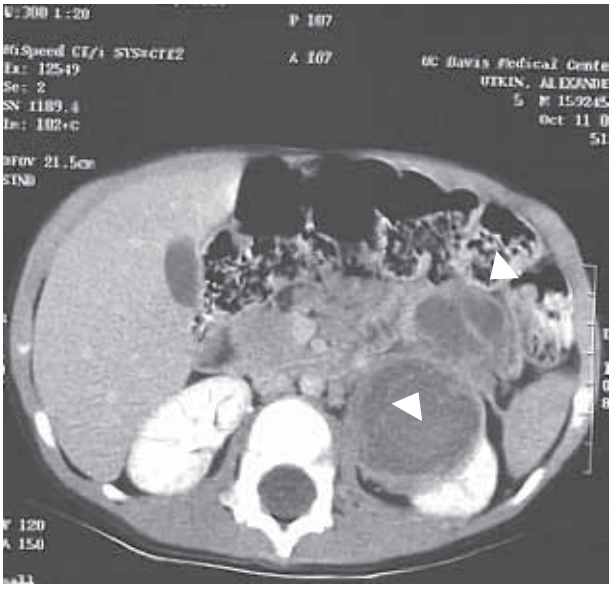


Fig. 2.2.8. X-ray computed tomography of kidney tumor. It is clearly seen as a part of the kidney is replaced by tumor mass

The leading in correct diagnosis is visualization.

Already the survey X-ray of the abdomen in the projection of the kidney homogeneous shadow, "forced bending" of the spine, intestinal loops shift to the opposite side from the tumor and fuzzy contours of the psoas on the affected side are detected.

On excretory urograms during Wilms tumor renomegaly, blurring of its contours, changing of its position and deformation of the kidney collecting system are determined. In some cases at large sizes of the tumor and almost complete renal parenchyme failure, its function on urograms is not detected (a "silent" kidney). In these cases, as with bilateral renal disease and combination of tumor with malformations of the kidney, it is obligatory to conduct angiographic study.

On a series of angiograms symptoms of Wilms tumor are clearly defined — accumulation of contrast material in the tumor as "lakes", dilatation, deformity and amputation of segmental arteries, the violation of their dichotomous branching, and quick reset of arterial blood in the venous system on abnormal arteriovenous shunts.

Very informative for surgeons is opacification of the inferior vein during cavagraphy, since in some cases it is possible to identify its location according to the tumor, the presence in vein of tumor thrombus or its secondary deformation due to compression by enlarged metastatic lymph nodes.

Lung metastases are detected with chest X-ray in five different views (straight, two lateral and two oblique photographs), bone lesions — by radioisotope study of the skeleton.

The main methods of diagnosis are ultrasound (Fig. 2.2.7), traditional X-ray (including excretory urography), X-ray CT (Fig. 2.2.8), MRI and angiography.

Ultrasound examination can also identify tumor metastases into the liver and retroperitoneal lymph nodes, and in time, during dynamic monitoring study, to detect recurrent disease. With the help of ultrasound it is possible to differentiate congenital and acquired kidney cystic formation from tumors, establishing quite accurately the size of neoplasms and its relation to the nearest organs and monitor the effectiveness of the treatment.

Differential diagnosis. Nephroblastoma must be differentiated primarily from malformations of kidneys (hydronephrosis, renal polycystosis and kidney multi cystic, kidney doubling, horseshoe kidney, etc.) and retroperitoneal extrarenal entities (neuroblastoma, rabdomioblastoma, angiosarcoma, teratoma). In some cases it is necessary to carry out differential diagnosis with tumors of the liver and abdominal lymphoma.

Nephroblastoma *complex treatment:* surgical (transperitoneal nephrectomy), radiotherapy (preoperative and postoperative tumor bed), chemothera-

Table 11

Nephroblastoma Treatment Scheme According to the Stage

Stage	Preoperative chemotherapy	Preoperative radiotherapy	Surgery	Postoperative radiotherapy	Postoperative chemotherapy
I	– (+)	–	+	–	– (+)
II	+	–	+	– (+)	+
III	+	– (+)	+	+	+
IV	+	– (+)	– (+)	+	

py (preoperative and postoperative). During choosing a method of treatment they take into account the stage of the disease, the morphological structure of the tumor, the age of the child.

Schematic diagram of nephroblastoma treatment depending on the stage of the disease is shown in Table 11.

During tubular and cystic variants of nephroblastoma, unlike anaplastic, prognosis is favorable. Due to used methods of treatment survival of patients with nephroblastoma for 3 or more years reaches 80–90%.

NEUROBLASTOMA

Neuroblastoma is a malignant tumor that was first time described in 1865 by Virchow, and called "glioma". In 1910, Wright proved that it developed from embryonic neuroblasts of sympathetic nervous system and gave its real name. It is the most common extracranial solid neoplasm, observed in children, and makes 14% of all childhood tumors (L. A. Durnov, 1986; Putnam et al., 1983 and others).

Neuroblastoma (a common name of malignant tumors of sympathetic nervous system) — the most mysterious tumor of childhood, both with clinical and biological point of view. In the structure of all cancer diseases neuroblastoma makes 7–11% of the total number of malignant tumors in children, ranking fourth plact after acute leukemia, CNS tumors and malignant lymphomas. The frequency of neuroblastoma is 0.85–1.1 per 100,000 children under 15 years. The age distribution of neuroblastoma throughout childhood is heterogeneous and the frequency of diagnosis decreases as children get older. In first year children neuroblastoma is the most common malignant tumor, its incidence in this age group is 6.1 per 100 000 children under one year. The incidence of neuroblastoma at age from 1 to 5 years old is 1.7 per 100 000 children, aged 5 to 10 years — 0.2 per 100,000 children in this age group.

Neuroblastoma belongs to a group of embryonic tumors, such as hepatoblastoma, nephroblastoma, embryonal rhabdomyosarcoma. All of them are characterized by manifestation at the early age, have similar cytomorphological characteristics, typical for embryonic tumors. Neuroblastoma has a number of specific, unique features of its biological behavior, not typical for other malignant tumors.

1. An ability for spontaneous regression.

In clinical practice, a well-known example of infants with the classical picture of the 4S stage neuroblastoma (usually with massive liver damage) malignant process involution is observed at about 4 months of age. Only a very small part of such patients require a minimum chemotherapy or radiation therapy for "induction" of the process of regression. Still unknown any marker that defines a turning point in the progression of the disease to regress. It is unknown why some patients up to 1 year with 4S stage of the process do not have tumor regression.

Also the factors that cause this process in groups of patients with neuroblastoma, who do not belong to the category 4S stage are not defined.

2. An ability for differentiation ("maturation").

Another surprising feature of neuroblastoma tumor cells was observed during its culture research: culture of cells, derived from aggressively growing tumor in the cultivation process got features of differentiating neural tissue. Different agents are able to induce this process in vitro: retinoic acid, a so-called nerve growth factor, some chemotherapy agents, papaverine. However, to date there were no reports about successful induction of the differentiation process in vivo.

The first clinical case of "maturing" of ganglioneuroma into benign after 10 years after diagnosis and without treatment was described in 1927. According to the German authors (F. Berthold) the differentiation of malignant and benign tumors of neuroblastoma is rare (1 : 1150), although convincing.

3. An ability to rapid development of aggressive and rapid metastasis.

International Classification of Neuroblastoma Stage (Brouder et al., 1988).

Stage and its characteristics:

I. Localized tumor, located in the field of the original development; neoplasm is completely removed, with or without microscopic signs of its residues, confirming the absence of lesion of lymph nodes on both sides of the spine.

IIA. Unilateral tumor with removal of largest part of it; microscopically — no lymph nodes lesions on both sides.

IIB. Unilateral tumor, deleted or most of it, microscopically — there is unilateral lesion of lymph nodes

III. Tumor extends to the opposite side, with or without metastatic lesions of regional lymph nodes, unilateral tumor with metastases in opposite lymph nodes; median tumor with metastases in lymph nodes on both sides.

IV. Disseminated tumor with metastases in distant lymph nodes, bones, lungs, and other organs.

IVS. Localized primary tumor, determined in stage I and II with metastases to the liver, skin and/or bone marrow.

Pathomorphological classification

Neoplasms develop from sympathetic ganglia. They differentiate three types of tumors, differing from each other by level of differentiation.

Ganglioneuroma consists of mature ganglion cells and a benign tumor is in its nature. It often calcifies. Kemshead et al. (1985) suggest that all ganglioneuromas are mature neuroblastomas. The literature describes cases of spontaneous or therapeutic maturation of neuroblastoma to ganglioneuroma (Everson and Cole, 1966).

Ganglioneiroblastoma is an intermediate form of neoplasm, lying between ganglioneuroma and neuroblastoma. Mature ganglion cells and undifferentiated neuroblastomas occur in different parts of the tumor in different proportions.

Neuroblastoma is undifferentiated form of neoplasm, consisting of small round cells with dark-spotted kernels. Rosettes and characteristic neurofibrils can often occur. Hemorrhages and areas of calcification are found in tumor.

Clinical picture of the disease depends on the location of the primary tumor, the presence and location of metastases, the number of vasoactive substances, produced by the tumor.

They distinguish symptoms of the primary tumor, parneoplastic syndrome (associated with the overproduction of catecholamines) and metastatic disease.

The main complaints are pain (at 30–35%), fever (25–30%), and weight loss (20%). Presence of complaints and its number mainly depends on the stage of the disease.

The defeat of the cervicothoracic of sympathetic trunk early causes Horner's syndrome that makes it possible to do a relatively early diagnosis of tumor, arising from these departments. Localization of the tumor in the posterior mediastinum can cause compulsive cough, respiratory distress, deformities of the chest wall, may cause dysphagia and in babies — frequent regurgitation. Bone marrow lesion leads to anemia and hemorrhagic syndrome. Is characteristic symptom of the "glasses" with exophthalmos during lesion of retrobulbar space in children with 4 stage of disease. Skin metastases have bluish-purple color and dense texture.

During localization process in the retroperitoneal space palpation reveals bumpy, stony hardness, almost non-displaced tumor (early fixation of the tumor is due to fast growing into the spinal canal through the intervertebral holes). Spread of the tumor from the thoracic cavity in the retroperitoneal space through the diaphragmatic opening causes the same symptom of the "hourglass" or "dumbbell".

Fundamentally neuroblastoma can occur anywhere in the body, which has sympathetic innervation, but the typical source of tumor growth during neuroblastoma is sympathetic nerve trunk along its entire length, and the adrenal medulla. Approximately 40% of tumors arise in the adrenal glands, 30% come from the lumbar sympathetic trunk, 15% — from the thoracic, 3% — from the pelvic paraganglia, 1% — from cervical spine.

According to different authors a share of nonstandard locations or neuroblastomas with unknown primary site accounts from 5 to 15%.

The most typical clinical picture during retroperitoneal (most common) tumor localization: a dense practical unmoved tumor. Very often there are pronounced common symptoms: weight loss, fatigue, bone and joint pain, anemia, and fever.

Quite often a child is examined for bone pain with diagnoses: arthritis, epiphisitis — rheumatoid or nonspecific character, and up to that time while the common symptoms of the disease will not develop.

Increase in blood pressure may be connected either with excessive secretion of catecholamines tumor, or with compression of renal vessels. Intractable diarrhea, a symptom, although rare, can long conceal the real cause of intestinal disorders.

Neurological symptoms can be expressed in children whose tumor is localized paravertebrally with penetration into the spinal canal, or during primary intraspinal localization.

The initial symptoms of neuroblastoma have no specificity, consider various pediatric diseases.

- Neoplasms in the abdomen
- Swelling
- Weight loss
- Anorexia
- Bone pain (metastases)
- Anemia
- Fever

Symptoms related to the excess of catecholamines — facial flushing, sweating, tachycardia, hypertension, headache, rarely chronic diarrhea. During the localization of the head and neck may be the first symptoms of the presence of palpable tumor nodules and the development of Horner's syndrome, during development of tumors in the chest, may be a violation of breath, dysphagia, compression of the veins. The first sign of the presence of tumor in the abdominal cavity may be the presence of palpable tumor masses, neoplasms of the pelvis can manifest in violation of the act of defecation and urination. Neuroblastoma, growing through between vertebral holes with compression of the spinal cord (tumors in the form of a "dumbbell"), causing characteristic neurological symptoms such as flaccid paralysis of the limbs and impaired urination with a strained bladder. Most tumors are localized in the retroperitoneal space, mainly in the adrenal glands, at least — in the mediastinum on the neck.

Neuroblastoma has a tendency to metastasize to certain areas such as the bone marrow, lymph nodes. Rare tumors occur in the skin and liver, as an exception, the brain is affected. Establishment of initial diagnosis occurs in cases of large tumors (about 70%).

Clinical symptoms associated with metastatic disease are very diverse. In newborns the first sign of the development of metastases is the rapid enlargement of the liver, may result in the formation on the skin bluish nodes and bone marrow. In older children, the development of metastases can cause bone pain and

swollen lymph nodes. The disease may have symptoms that are typical for leukemia, in children anemia and hemorrhage in the mucous membranes and skin develop, which is caused by pancytopenia, caused by bone marrow affection by cells of neuroblastoma.

Diagnosis. According to international standards neuroblastoma diagnosis can be determined by histological examination of biopsy material, obtained from the primary tumor (or from metastases) or during detection of lesions of bone marrow in combination with an increased level of catecholamines or their derivatives: vanillylmandelic (CMC), homovanillic (HVA) acid and dopamine in the blood or urine. CMC and HVA levels are increased by 85% of patients and dopamine level — by 90% of patients with neuroblastoma. The level of catecholamine excretion did not affect the prognosis, while a high ratio of HVA: CMC (directly proportional dependence) indicates the presence of poorly differentiated tumor and is associated with a worse prognosis.

Tumor markers. Neuroblastoma belongs to the category of children tumors when tumor markers are used as for diagnosis, monitoring in treatment process as prognostic factors. Diagnosis and differential diagnosis of neuroblastoma is based on the determination of the level of the daily excretion of catecholamines with urine, their precursors and metabolites (vanillylmandelic and homovanillic acids).

Research has shown that the detection of neuroblastoma at the preclinical stage, as determined by vanillylmandelic of homovanillic acid in the urine of 6 months old children, and timely initiation of treatment significantly increase survival. These data indicates that neuroblastoma cells at preclinical stage predominantly secrete adrenaline and noradrenalin and therefore are more differentiated. Normally cystathionine is not detected in urine; the presence of cystathionine in urine indicates the presence of neuroblastoma.

Strictly specific and readily identifiable are metabolites of catecholamines: CMC of HVA and dopamine (DA). The level of these substances is identified in daily urine and in blood serum of a child. Diagnostically significant is the increase in the level of CMC, HVA and DA 3 times in comparison with the age norm. It is interesting that method for the determination of catecholamines derivatives is more sensitive approximately by 15% than in blood serum.

In the case of false-negative determination results of metabolites of catecholamines (which occur in about 15% of patients) assistance in the diagnosis will be determination of concentration in blood serum neuron-specific enolase (NSE), enzyme found in neurons.

In 1981 K. Tapia showed that tumors of neurogenic origin have a high level of NSE. Increased level of this enzyme in the blood serum is not specific for neuroblastoma, but also for other tumors of neuroectodermal origin (PNET, Ewing's sarcoma), during Wilms' tumor, some types of leukemia. The level of NSE in serum has prognostic value, since the value of NSE is over 80–100 ng/ml indicates an extremely unfavorable prognosis.

Another biochemical marker of neuroblastoma is ferritin. A number of patients with neuroblastoma have increased ferritin level, and its amount in serum is directly proportional to the tumor mass volume.

It is known that ferritin has some biological effects, adversely affect on immunity of the patient and, therefore, the high level of ferritin is also associated with a worsening forecasts in patients with neuroblastoma.

Lactate dehydrogenase (LDH) enzyme does not belong to the specific markers for neuroblastoma, but its level in serum has prognostic value for these patients: its increased level is often observed in advanced stages of the disease, which probably explains the connection of increased LDH level with poor prognosis. LDH level only reflects the proliferation rate of tumor and consequently at common processes with a known unfavorable prognosis is registered a high level of LDH.

Other markers of neuroblastoma are ganglioside GD2, neuropeptide Y, and chromogranin A. Identification of these markers by immunohistochemistry is a parameter for diagnosis of neuroblastoma, and their impact on the prognosis of the disease is being studied.

Scheme for determining the extent of cancer spread.

1. Site of the primary tumor — ultrasound, CT, MRI.
2. Chest cell — X-ray, CT.
3. Abdominal cavity — ultrasound, CT.
4. Bone scintigraphy with Te 99 and subsequent X-ray of the revealed centers of isotope hyperfixation.
5. Scintigraphy with ^{131}I metaiodinebenzylguanidine (MJBG).
6. Aspiration biopsy of the bone marrow (from 4–8 places).
7. Trephinebiopsy of bone marrow with histological and immunohistochemical studies.
8. Biopsy of lesions, suspected on tumor metastasis.

Visualization techniques: urography, computed tomography (CT), MRI, bone scintigraphy or an X-ray of the skeleton, chest X-ray, liver scintigraphy, angiography (Fig. 2.2.9, 2.2.10).

During X-rays and CT scan are identified characteristic calcifications in the tumor tissue (which is a good prognostic sign), and the number of which increases with the positive response to chemotherapy (see Fig. 2.2.11).

Treatment, depending on stage of the tumor:

- Surgery
- Radiation therapy
- Chemotherapy
- Bone marrow transplantation

The younger the child, the more favorable the prognosis. To predict the development of the disease and evaluate treatment efficacy is necessary to determine periodically the level of catecholamines in the serum. Favorable prognosis takes place when the tumor is diagnosed at an early age and at an early stage of development.

At treatment of neuroblastoma there are used all three methods of cancer treatment: chemotherapy, radiation therapy and surgery.

The treatment of patients with localized 1 and 2A stage consists in radical removal of the tumor. The

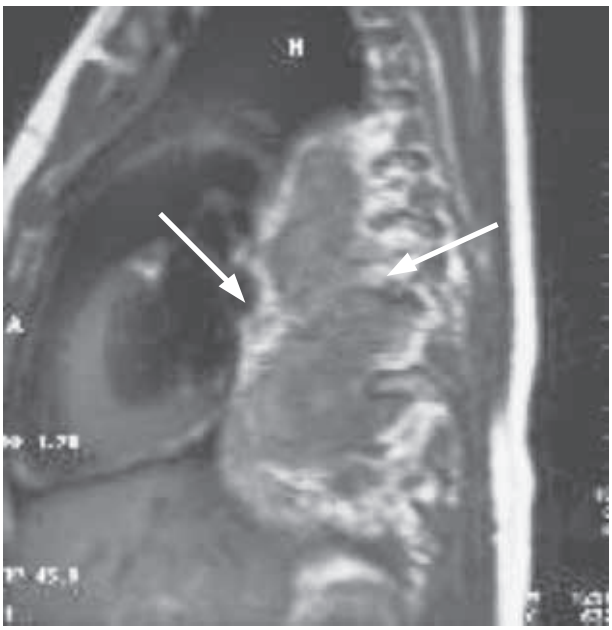


Fig. 2.2.9. Posterior mediastinum neuroblastoma with tumor penetration into the spinal canal (MRI)

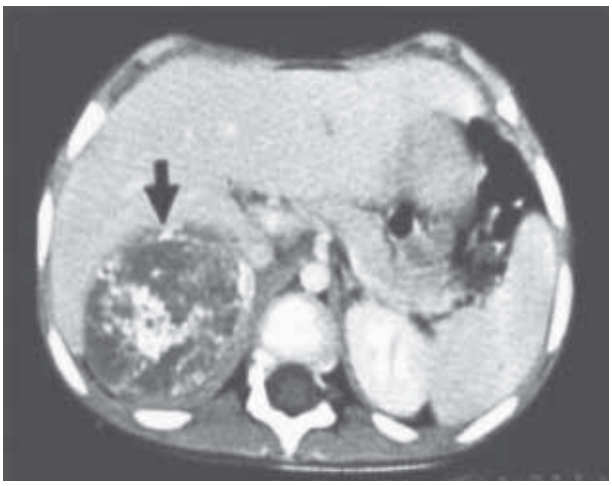


Fig. 2.2.10. Retroperitoneal neuroblastoma (CT)

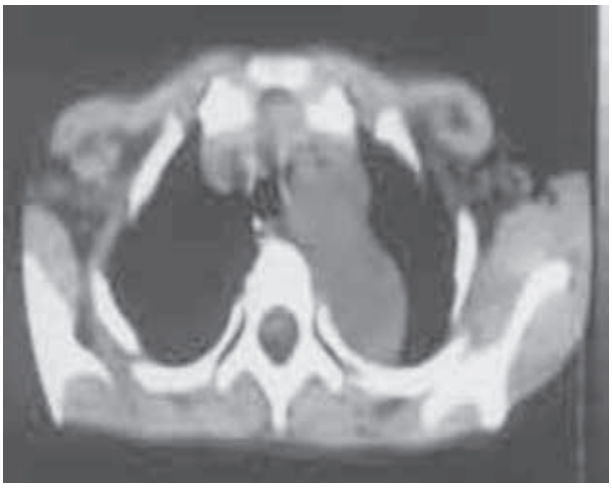


Fig. 2.2.11. Pposterior mediastinum neuroblastoma (CT)

presence of microscopic residual tumor, according to many authors, almost never leads to recurrence or metastasis, which distinguishes neuroblastoma from most other solid tumors. It is important to produce the biopsy of regional lymph nodes on both sides of the tumor for ascertaining the stage of disease. Patients with stage 2B must also be used chemotherapy and radiation of focus and the involved lymph nodes.

Patients with 3 stage of disease already in the debut have unresectable tumor, so the preoperative chemotherapy is necessary, which leads to a significant reduction in tumor size, sometimes even to the possibility of its radical removal. In the case of incomplete removal, radiation therapy on tumor bed can help deal with the residual tumor. Modern programs of chemotherapy and improved surgical techniques (microsurgery) make it possible to cure up to 60% of patients with 3 stage of neuroblastoma.

Patients with 4 stage of disease, which constitute the majority of patients, have a worse prognosis; 5-year survival rate even with using the current program of chemotherapy is not more than 20%.

Prognostic role for patients with 4 stage of neuroblastoma is the localization of metastases: children with metastases only in the distant lymph nodes have a much better prognosis than patients with bone metastases. In the treatment of this group of patients with a very unfavorable prognosis even the use of mega doze chemotherapy (melphalan, vepeside), and total body irradiation with autologous transplantation of bone marrow does not produce the expected results. There are studies underway on the application of a new class of chemotherapeutic agents (topoisomerase 1 inhibitors, such as topotecan and irinotecan) and immunotherapy in patients with resistant forms of neuroblastoma and in the progression of the disease.

In modern standard chemotherapy regimens there are used platinum drugs (cisplatin, carboplatin), epipodophylotoxins (VP-16, VM-26), dacarbazine, adriblastin, cyclophosphamide, ifosfamide, vinkalkaloids (vincristine, vindesine).

According to different authors survival of patients with neuroblastoma as a whole is about 50% (49–55%), in stages: 1 — 100%, 2 — 94%, 3 — 60% (67–57%), 4 — 10–20%, 4S stage — 75%.

MEDIASTINAL TUMOR

Mediastinal tumors are observed with equal frequency in men and women, are found mainly in young and middle age. Most of them belong to congenital tumors. Benign tumors of mediastinal significantly predominate over the malignant.

Clinical symptoms of benign neoplasms of mediastinal depends on many factors — from the growth rate and size of the tumor, its location, the level of compression of the adjacent anatomical structures, etc. A progress of the neoplasms of the mediastinal has two periods — the period of asymptomatic and clinical manifestations. Benign tumors develop asymptomatic for a long time, sometimes years or even decades.

There are two main syndromes at a pathological mediastinal — compression and neuroendocrine. Compression syndrome due to a significant increase in pathological formation is characterized by a feeling of fullness and pressure, dull pain behind the breastbone, dyspnea, cyanosis of face, swelling of the neck, face, increasing saphenous veins. Then there are signs of impaired functions of the different organs as a result of compression.

There are three types of compression symptoms: organ (the displacement and compression of the heart, trachea, bronchi, esophagus), vascular (compression of brachiocephalic and superior vena veins, the thoracic duct, the displacement of the aorta) and neurogenic (compression with conduction of the disturbance of the vagus, the phrenic and intercostal nerves, sympathetic trunk). Neuroendocrine syndrome is manifested by joint disease, reminiscent of rheumatoid arthritis, and also large and long bones. There are various changes in heart rate, angina.

PRIMARY MEDIASTINAL TUMORS

THYMOMAS

Thymomas (Fig. 2.2.12) are observed at any age, prevails in the age of 20–30 years. The most common primary tumor of the mediastinum (20%).

Distinguish lymphoid, epithelial, spindle or mixed thymomas.

The clinical picture of thymomas is very diverse. Almost in half of the cases progress is asymptomatic; usually tumor is detected accidentally during preventive X-ray examinations or in connection with the appearance of symptoms of compression of the anterior mediastinum. With a significant compression appears a feeling of tightness behind the breastbone, discomfort and pain, dyspnea, swelling of the neck veins, facial puffiness and cyanogens. At children particularly pronounced respiratory problems due to compression of the relatively narrow, pliable trachea. Asymptomatically developed tumors at the time of detection can reach large sizes. Thymomas can be combined with myasthenia

Malignant form, featured high invasiveness into the surrounding tissue, occurs almost as often as benign (35–50% of patients). Characterized by dissemination of thymoma on the pleura, herewith hematogenous and lymphatic metastases are rare. Thymoma is diagnosed in 15% of patients with myasthenia; herewith myasthenia is diagnosed in 50% of patients with thymoma. There are cases of thymomas of the posterior mediastinum.

DISEMBRYOGENETIC TUMORS

Dermoid cysts and teratomas occur in 5–8% of patients with tumors of the mediastinum. These tumors arise from the violation of embryogenesis. Dermoid cysts evolve from ectodermal elements, have thick walls of coarse fibrous connective tissue. In the cavity of these cysts there is often a brown, viscous liquid, elements of the skin, hair. Calcinates are revealed in 10% of patients. Teratomas arise from multiple germ layers and have in their composition several different structures of tissues, and the degree of differentiation of the cells can be benign or malignant (in 10–20% of patients). Although these diseases are congenital, it is usually diagnosed in patients only in adulthood with the appearance of pain or "compression syndrome". In 95% of patients these tumors are located in the anterior mediastinum.

Teratoblastomas (Fig. 2.2.13) are found almost as often as thymoma, and constitute 11–17% of mediastinal tumors. Malignancy is detected in 25–30% of cases.

Dermoid cyst (Fig. 2.2.14) is a tumor of epidermal origin. The disease is most often detected in adolescence, half of the entire tumor contains calcifications. Dermoid of mediastinal take the form of solid or cystic formations. With festering of dermoid cyst the content becomes liquid, pyoid. A progress of mediastinal dermoid cyst is long. Pathognomonic symptom is coughing up of pasty mass and hair (the breakout of cyst in the bronchus), is rare.

Rapid growth is typical for malignancy, although also reason for the rapid increase in the vol-

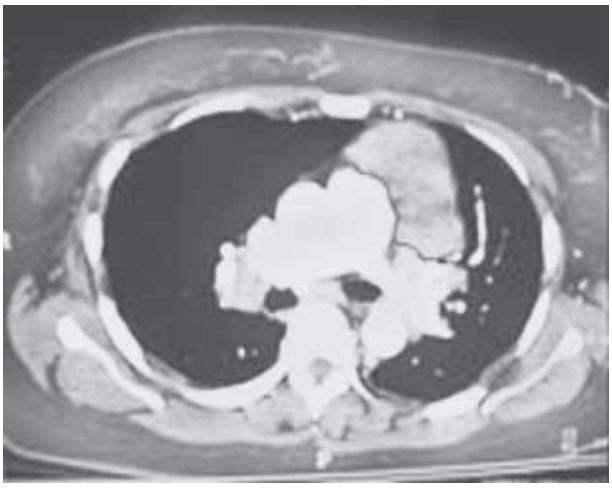


Fig. 2.2.12. CT scan of patient with thymoma

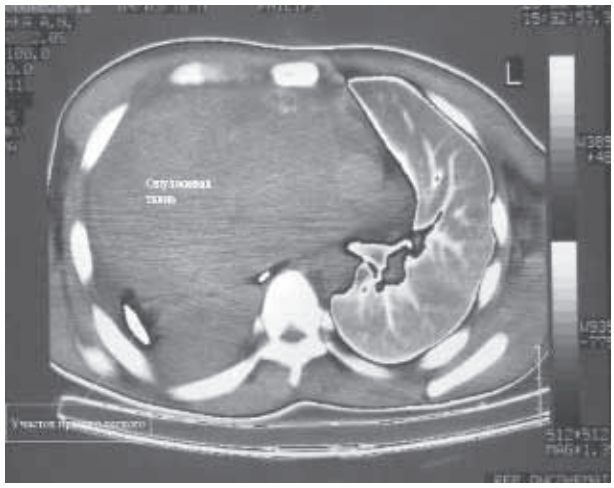


Fig. 2.2.13. Teratoblastoma of anterior mediastinum

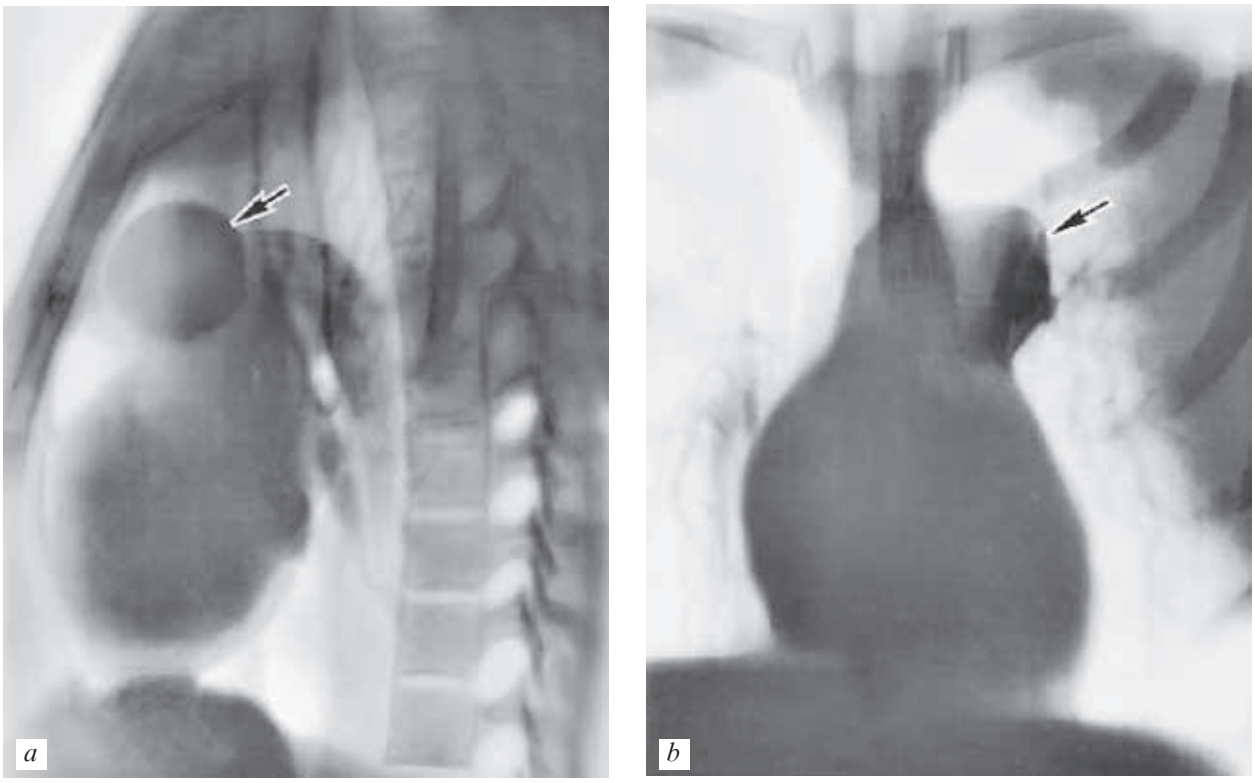


Fig. 2.2.14. Dermoid of the anterior mediastinum (*a* — lateral, *b* — a direct projection)

ume of the tumor can be a hemorrhage in its tissue. Sometimes when malignancy metastases in the lungs and regional metastases can be observed. Operative treatment is removal of a cyst or tumor, removal of mediastinal teratomas is advisable due to their tendency to malignancy. At teratomas, with no signs of malignancy, operative intervention yields good long-term results.

MESENCHYMAL TUMORS

Mesenchymal tumors are found in all three departments of mediastinum, but more often — in the anterior mediastinum. They occur in 4–7% of patients with tumors of the mediastinum, evolving from adipose, connective, muscle texture, and from endothelium of the vascular wall.

Benign tumors of mesenchymal origin occur mostly asymptomatic; with the growth can become very large, causing the symptoms of compression of the organs of the mediastinum.

Treatment of surgery — removal of the tumor.

Lipoma (Fig. 2.2.15) and liposis are more common in the lower one side of the mediastinum. They can spread from the mediastinum in caudal or cranial direction. From this group of tumors are seen mostly lipomas. A favorite localization — right cardiodiafragmal corner. X-ray defines homogeneous formation, adjacent to the shadow of the heart. In the differential diagnosis of a tumor of the lung, pericardial cyst is most useful artificial pneumothorax and pneumo mediastinography. Using the latest one, X-ray reveals that paraplasm (lipoma) is surrounded by the gas and has no connection with the pericardium and diaphragm. On the other hand, omental fat can penetrate into the lower parts of the posterior mediastinum.

Thanks to its soft consistency, lipoma does not impact on surrounding organs and often random finding. Lipomatosis may be iatrogenic due to corticosteroid therapy. Lipo- (fibro-) sarcomas are very rare tumors, usually located in the posterior mediastinum, which can cause the displacement of adjacent organs.

Fibroma does not cause complaints as long as it reaches a significant size. The presence of pleural effusion can sometimes be a sign of fibro sarcoma (usually located in the posterior mediastinum) and fibroids.

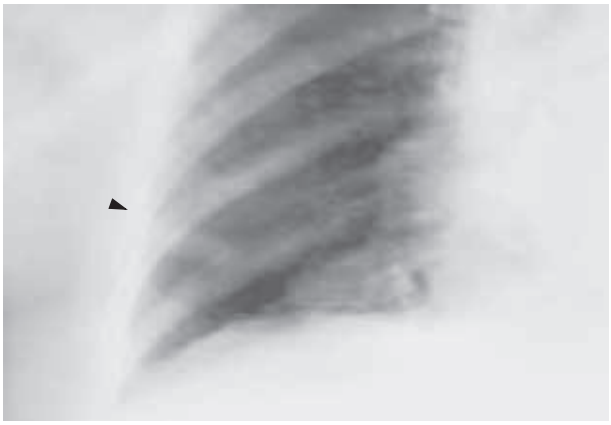


Fig. 2.2.15. Abdominal-mediastinal lipoma

Hemangioma (cavernous hemangioma, hemangioendothelioma, hemangiosarcoma) — in the mediastinum is rare, in 2/3 of cases is located in the anterior mediastinum. They have different shapes, are surrounded by a connective tissue capsule, can be multiple. Often contain flebolits.

Lymphangioma (hygroma) — most of them occur in children. They develop from the lymphatic vessels, grow in different directions with the formation of knots. They may spread to the neck, because significant displacement of neighboring organs, there are cavernous and cystic variants. They are located in the lower anterior mediastinum, if are not complicated by chylothorax, usually asymptomatic.

NEUROGENIC TUMORS

Most commonly found tumors of the postmediastinum, usually located on the top of its department. They develop from the branches of the vagus nerve and the intercostal nerves, sympathetic trunk and spinal membranes, multiple neuromas can be detected in the neurofibromatosis (von Recklinghausen disease). Most often asymptomatic; with growth in the spinal canal clearance neurological symptoms appear.

Neurogenic tumors are at the first place (20% of all tumors of the mediastinum) in frequency among all tumors and cysts of the mediastinum. They arise at any age and are often benign. Their favorite localization is postmediastinum.

Neurofibromas develop from nerve fibers and their membranes. Neurolemmoma is from the lemotsitis (Schwann cells), ganglioneuroma — from the sympathetic trunk and contains both the ganglion cells and nerve fibers. These tumors usually occur in children. Paragangliomas and mediastinal pheochromocytoma originate from elements of chemosensory nerve apparatus and is analogous to the sinocarotid tumors zones by the structure, contains chromaffin cells, often have hormonal activity, manifested by hypertension and frequent crises. Half of the patients with these tumors are malignant.

Malignant neurogenic tumors are also neuroblastoma (sympaticogonioma, simpatoblastoma, ganglionitis — roblastomas, and neurogenic sarcoma). Distinguishing features of neurogenic tumors are back pain, symptoms of spinal cord compression with growth of the tumor in the spinal canal to the type of "hourglass", the development of paresis and paralysis. X-ray neurogenic tumors are characterized by an intense round shadow in the field of the vertebral costal grooves with sharp edges, sometimes visible uzuration of vertebrae, ribs. Malignant tumors are frequently revealed by a hemorrhagic effusion into the pleural cavity on the affected side. Before surgery obtaining the cellular elements of the tumor is usually not possible because of the difficulty of the tumor puncture of the posterior mediastinum.

Treatment: surgical removal of the tumor through thoracotomy access. With the growth of the tumor according to the "hourglass" additional resected shackles of several vertebrae is conducted and a fragment of a tumor from the spinal canal is removed.

THE COELOMIC CYST OF THE PERICARDIUM

Cysts of the pericardium are malformations (7% of mediastinal tumors). More often their location is right or left (rarely) cardiophrenic corner. (Fig. 2.2.16).

The true cyst does not communicate with clearance pericardium. When communicating with the cavity of the cyst with the cavity of pericardium, the diverticulum occurs. Course of the disease is asymptomatic, as a rule, it is revealed in a survey of prophylactic fluorography. At large sizes of the cysts or prelum of the heart in the diverticulum disturbances of the cardiac activity, arrhythmia, and cardialgia may arise. Operative treatment — removal of the cyst.

BRONCHOGENIC AND DUPLICATION CYST OF MEDIASTINUM

Bronchogenic and enterogenous cyst of the mediastinum (Fig. 2.2.17) (7–8% of mediastinal tumors). Cysts appear during fetal development and formed from the dystrophic rudiments of intestinal or bronchial epithelium. Bronchogenic cysts can be localized in the mediastinum, and deep in the lung tissue.

In the mediastinum they are often closely adjacent to the trachea and major bronchi, usually behind the bifurcation of the trachea. Clinical symptomatology occurs when cysts reaching large size. The most characteristic symptoms of the respirato-

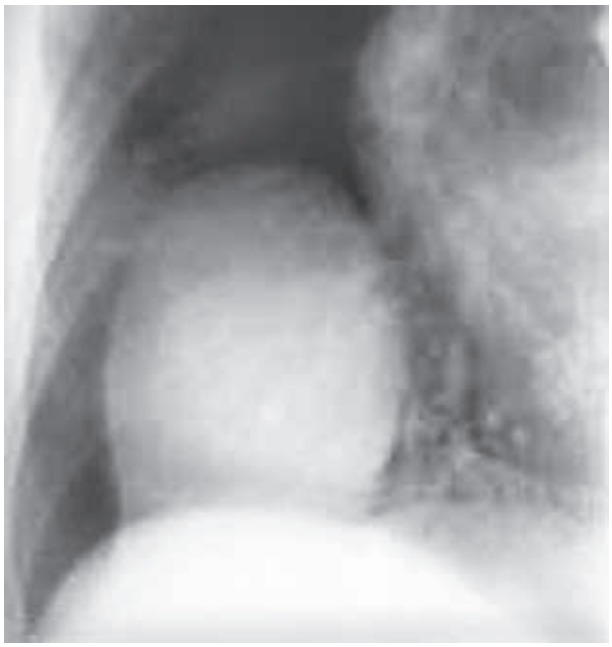


Fig. 2.2.16. The coelomic cyst of the pericardium

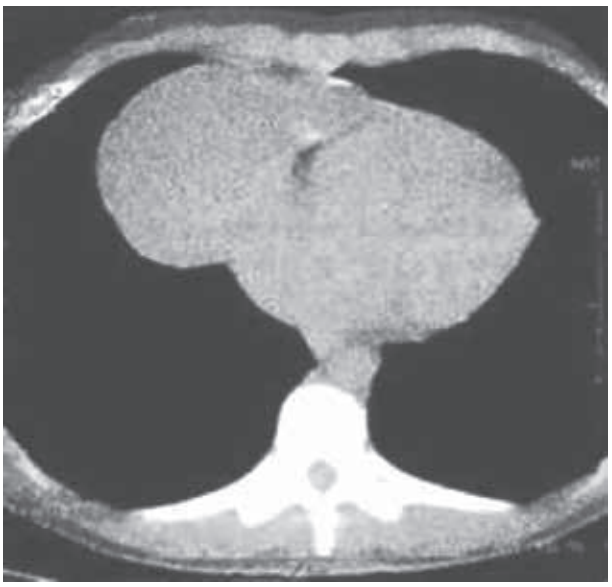


Fig. 2.2.17. Bronchogenic cyst of the mediastinum

ry tract suppression — dry cough, dyspnea, breathing crowing. X-ray revealed rounded shadow adjacent to the trachea.

Especially clear it is seen at pneumo mediastinography. Rarely cyst has a connection with clearance of the airways and then with X-ray in its clearance is seen the fluid level, and at the introduction of the of liquid contrast material into the bronchial tree, it fills the cavity of the cyst. Surgical treatment consists in excision of the cyst. Indications to it arises from the danger of suppuration cyst, perforation of its wall with the development of mediastinitis, pneumoempyema, bleeding.

From the cyst wall cancer growth may start. Enterogenous cysts evolve from the dorsal departments of the primary intestinal tube, are located in the lower posterior mediastinum, adjacent to the esophagus. According to the microscopic structure a cyst may resemble the stomach wall of the esophagus, small intestine. Due to the fact that the inner shell of some cysts lined with gastric epithelium that produces hydrochloric acid, there may be wall ulcerations, ulceration bleeding and perforation of them. In addition, duplication cyst may be exposed by suppuration followed their breakthrough into the cavity of the pericardium, pleura, hollow organs. The most informative diagnostic method — pneumo mediastinography. At the same time expose a link of pathological formation with the trachea, bronchus, and esophagus. In connection with the real possibility of developing complications conduct surgery — removal of the cyst.

LYMPHADENOPATHY

Lymph node enlargement of the mediastinal is observed at lymphoma and metastases of carcinomas and non-neoplastic diseases (tuberculosis, sarcoidosis, and so on).

LYMPHOMAS

Lymphomas occur in 35% of patients with tumors of the mediastinum and in 20–25% of patients with all malignancies of the mediastinum. Lymphomas evolve from the mediastinal lymph nodes. The most frequent localization of lymphoma is anterior mediastinum, although this type of tumors can affect the lymph nodes of any part of the mediastinum. All three types of lymphoma: lymph sarcoma, clasmocytoma and lymphogranulomatosis are characterized by malignant course. Symptoms of the disease are caused by intoxication first, general malaise, fatigue, subfebrile or febrile body temperature, weight loss, itchy skin.

With large packets of lymph nodes dry cough, chest pain, symptoms of compression of the mediastinum appear. In the diagnosis the most informative are X-ray, mediastinoscopy, and parasternal media with lymph node biopsy. Surgical treatment is indicated only at the early stages of the disease, when the process is locally affects a certain group of lymph nodes. At the later stages of the disease radiation therapy and chemotherapy are used.

The lesion of the mediastinal lymph nodes at lymphoma can be both isolated and in combination with lymphadenopathy of other locations, as well as lesions of various organs. Aggressive lymphomas are characterized by tumor invasion into surrounding anatomical structures (vessels, trachea, bronchi, pleura, lung, esophagus, etc.).

SECONDARY TUMORS OF THE MEDIASTINUM

Secondary tumors of the mediastinum are metastasis of malignant tumors of the thoracic or abdominal cavity in the lymph node of the mediastinum. Most often arise at cancer of the lung, esophagus, and proximal stomach. Clinically occur most often asymptomatic. At the first place come symptoms of the underlying disease and general signs of the cancer process. Only when metastases get large sizes there arise various compression syndromes — most often superior vena cava syndrome, Horner's syndrome. Principles of diagnosis are similar to those during primary tumors of the mediastinum. The presence of tumor metastasis in the mediastinum significantly reduces the efficacy of surgical treatment and its feasibility.

Prognosis is usually poor.

The nature of lymphogenous metastasis of malignant tumors is associated with features of the lymph flow from the affected organ. Enlarged lymph nodes due to metastatic tumors of intrathoracic and extrathoracic locations are only in 20% of cases. The most common cause is bronchogenic cancer of the lung, in which, in the later stages, metastatic lesion of the lymph nodes is more than 80% of cases. The involvement of lymph nodes in bronchiogenic carcinoma is not a one-way — through cross lymph flow, right tracheobronchial lymph node groups affected

even when left-sided tumor. Less often a source of metastases is malignant tumors of the gastrointestinal tract (esophagus, stomach, and pancreas), breast, kidneys, testes, prostate, thyroid and larynx). In accordance with the nature of the lymph flow, at cancer of mammary glands one should focus on the retrosternal lymph nodes, at tumors of the gastrointestinal tract, kidney, testes, and prostate — on the back of mediastinal groups of the lymph nodes, at cancer of larynx and thyroid gland — on the lymph nodes of the upper mediastinum. When lymphatic dissemination of extra thoracic tumors, metastases are affected on the lymph node over in the direction to the mediastinal groups, lesions of cervical and retroperitoneal lymph nodes are typical. Tumor lymphangitis in the lung is observed with breast cancer, stomach, pancreas, thyroid, larynx, and usually associated with lymphadenopathy roots of the lungs. As compared to cancer, sarcomas rarely metastasize to the mediastinum through the lymphatic pathways.

QUESTIONS FOR THE FINAL CONTROL OF CHAPTERS 1, 2

1. What are the frequency pyoseptic diseases in newborns, their structure and main cause of appearance?

2. What anatomical and physiological features skin structure and subcutaneous tissue in newborns contribute to the spread of the inflammatory process?

3. Identify clinical manifestations of newborns necrotic phlegmon depending on the form.

4 Medical management at necrotic phlegmon of newborns.

5. Forms and clinical manifestations of omphalitis in newborns.

6. Differential diagnosis of omphalitis and fistulas navel simple forms.

7. Treatment features of omphalitis depending on the form of the disease.

8. Complications and consequences that may arise in connection with omphalitis in a child.

9. When arises and what is the reason of newborns mastitis development?

10. Features of surgical intervention in newborns mastitis.

11. Causes of paraproctitis in newborns.

12. Operative interventions, which are performed at paraproctitis depending on its cause.

13. What directions consists treatment of septic diseases in newborns of?

14. What is the basis of empirical approach to the prescription of antibiotics?

15. Bacteriological examination at pyoseptic diseases. How to conduct it?

16. Name the components of therapeutic effect on children at purulent-septic diseases and dependence.

17. What is the systemic inflammatory response syndrome?

18. Name components of toxic form of the disease according to the international classification of sepsis.

19. What indicators of clinical and laboratory tests (blood pressure, pulse, general analysis and biochemical blood tests) show hemorrhagic syndrome and bleeding in the abdomen.

20. What are the main clinical manifestations of bleeding from the upper and lower parts of the digestive system.

21. What are the main causes of bleeding from the digestive system?

22. Assess the gastric contents and stool depending on the height of the source of bleeding.

23. What are the general principles of treatment and bleeding termination.

24. What are the features of clinical course control of bleeding from the digestive tract? Identify the indications for surgical treatment.

25. Describe indications for conservative and surgical intervention.

26. Give the main list of surgical diseases in children, which cause inflammation of organs of abdominal cavity.

27. What are the clinical signs and local symptoms of abdominal cavity organs inflammation?

28. What clinical signs and symptoms are typical for abdominal cavity organs inflammation in newborns?

29. Describe eatures of clinical picture of acute appendicitis in young children (under 3 years).

30. What are the indicators for conservative treatment and characteristics of peritonitis in the newborns?

31. Describe features of preoperative preparation for abdominal cavity inflammation.

32. What clinical signs and auxiliary diagnostic methods are specific for destructive forms of cholecystitis?

33. Specify general principles of cholecystitis treatment and determine the indications for surgical methods of treatment.

34. What are the main causes of acute pancreatitis?

38. Formulate the indications for conservative and surgical treatment of acute pancreatitis, its scope and methods.

39. What is the classification of acquired intestinal obstruction in children?

40. Identify the main causes of intussusception, peritoneal commissures, obstructive and dynamic obstruction.

41. List the major symptoms of acute intussusception.

42. What are methods of diagnosis and treatment of acute intussusception?

43. Features and terms of preoperative preparation with acquired intestinal obstruction.

44. Describe the roentgenologic stages of acquired intestinal obstruction.

45. Identify the main signs of viability at compressed intestine.

46. What methods of surgery are executed at intussusception?

47. Give the general principles of conservative therapy for early adhesion — paretic obstruction.

48. Summarize the main stages of the surgical treatment and approaches at early and late intestinal obstruction.

49. Define the classification of dynamic obstruction.

50. List the main stages of conservative treatment of the dynamic obstruction.

51. List the main mechanisms of injury of the abdominal cavity organs.

52. Give classification of injuries of the abdominal cavity at trauma.

53. List main symptoms of parenchymal organs damage.

54. List main symptoms of hollow organs damage.

55. What additional methods of investigation are most informative at damage to parenchymal and hollow abdominal cavity organs?

56. What are indications for emergency surgery for the ongoing bleeding resulting from damage to hollow and parenchymal abdominal cavity organs?

57. What indicators for clinical and laboratory tests (blood pressure, pulse, general analysis and biochemical blood tests) show hemorrhagic syndrome and bleeding in the abdominal cavity?

58. What clinical symptoms are observed at perforation of the esophagus?

59. What is the most informative method of X-ray to diagnose esophageal damage?

60. What methods of examination are used in the trauma of the chest cavity organs?

61. What is the indication for surgical intervention in hemopleura?

62. What methods are used in the diagnosis of esophageal damages?

63. What is important to identify a patient with traumatic damage to the urinary system?

64. What changes can be detected by palpation of the lumbar region of a patient with kidney damage?

65. What is a justification of previous diagnosis of traumatic damage to the kidneys based on?

66. What methods of diagnosis will help in detecting kidney trauma?

67. What are features of children examination with damage to the bladder and urethra?

68. What should a specialist identify at a patient with traumatic damage to the bladder?

69. What principles is the list of damage for the differential diagnosis of traumatic damages of pelvic bones and organs based on?

70. What are the most reliable diagnostic methods that help in detecting urethral trauma?

71. List features of the physical examination of children with the scrotum trauma.

72. What is the most important to determine at forming treatment strategy at patients with traumatic damage to the urinary system?

73. List features of childhood traumatology.

74. List main clinical symptoms of fractures, diagnosis, fracture displacement cases in children.

75. Immobilization method of treatment fractures in children. Advantages and disadvantages of the method.

76. Functional method of treatment fractures in children. Advantages and disadvantages of the method.

77. Surgical treatment of fractures in children.

78. Fractures of the extremities in children. Classification, clinical symptoms, diagnosis, treatment. The value of early rehabilitation.

79. Fractures of spine in children. Classification, clinical symptoms, diagnosis and treatment.

80. Fractures of pelvis in children. Classification, clinical symptoms, diagnosis and treatment.

81. When does formation of epiphyses of long bones stop, what is the confirmation criteria?

82. What is normal intraosseous pressure?

83. What are the features of diagnostic puncture and intraosseous pressure measurements in children suspected of acute hematogenous osteomyelitis?

84. List features of long tubular bones structure.

85. What are the clinical forms of acute hematogenous osteomyelitis.

86. Why does metaepiphyseal destruction happen in infants and children under 2 years of life?

87. What are the atypical forms of osteomyelitis?

88. What is the most common symptom of metaepiphyseal osteomyelitis?

89. List features of puncture joints in children and how often the procedure is conducted?

90. List features of immobilization and its term in children with metaepiphyseal osteomyelitis.

91. List the complications associated with hematogenous osteomyelitis, and indicate when they appear.

92. When is spa treatment recommended for osteomyelitis?

93. What is the most common surgery for chronic osteomyelitis in children?

94. What is the frequency of purulent and septic diseases in neonates, their structure and the main reasons of appearance?

95. What anatomical and physiological features of the skin structure and subcutaneous tissue in newborns facilitate the dissemination of the inflammatory process?

96. What are the clinical manifestations of necrotizing phlegmon of newborn depending on the form?

97. Medical management at necrotic phlegmon of newborn.

98. Forms and clinical manifestations of omphalitis in newborns.

99. Differential diagnosis of omphalitis simple form and the fistulas navel.

100. Features of the omphalitis treatment depending on the type of disease.

101. Complications and consequences that may arise from the omphalitis at a child.

102. When doest appear and with what is connected the development of mastitis at newborns?

103. List features of the operative interference with mastitis newborns.

104. List causes of appearance paraproctitis in newborns.

105. 84. Surgeries that are performed at paraproctitis depending on its root causes.

106. What does treatment of purulent and septic disease of newborns consist in?

107. What is the basis of the empirical approach to prescription of antibiotics?

108. Bacteriological study at purulent and septic diseases. How to conduct it?

109. What are the methods of therapeutic effects on the body of a child with purulent septic diseases and specify on what it depends.

110. What is a systemic inflammatory response syndrome?

111. List the clinical manifestations of hemangiomas and limphangiomas.

112. What are the methods of conservative and surgical treatment of hemangiomas and chylangioma.

113. What are the clinical manifestations of pigmented tumors?

114. Medical management at pigmented tumors.

115. The clinical picture of atheroma and dermoid cysts.

116. Features of the dermoid cyst removal.

117. List the clinical manifestations of soft tissue malignant tumors (rhabdomyosarcoma, teratoblastoma).

118. Surgical and combined treatment of soft tissue malignant tumors.

119. Supporting methods of diagnosis of tumors in children. Their role in the differential diagnosis.

120. Features of clinical course of benign tumors and tumor-like bone diseases.

121. Methods of surgical treatment of benign tumors and tumor-like bone diseases.

122. Conservative treatment of bone cysts.

123. Clinical manifestations of osteogenic sarcoma and Ewing's sarcoma.

124. The principles of combined treatment of malignant tumors of bone.

125. Clinical picture of benign bone tumors.

126. Methods of benign bone tumors treatment.

127. Operative treatment of bone cysts.

128. Methods of diagnosis of malignant bone tumors.

129. Clinical manifestations and treatment of nephroblastoma in children.

130. Clinical manifestations and treatment of neuroblastoma in children.

131. Clinical picture and diagnosis of mediastinal tumors.

132. Treatment of children with malignant and benign tumors of the mediastinum.

133. What are the main causes and clinical manifestations of bleedings from the upper parts of gastrointestinal system?

134. What are the main causes and clinical manifestations of bleedings from the lower parts of gastrointestinal system?

135. Specify classification and causes, major clinical manifestations of portal hypertension in children.

136. What are the methods of diagnosis of gastrointestinal bleeding in children and the basic principles of medical care at the hospital and hospital stages.

137. Clinical features of acute appendicitis in children under 3 years.

138. Differential diagnosis of acute appendicitis in children under 3 years.

139. Describe clinical features of acute appendicitis in older children.

140. What are the causes and the main clinical symptoms of peritonitis in newborns?

141. What are the causes and main clinical symptoms of invagination in children?

142. List the indications for conservative and surgical treatment of invagination.

143. Specify classification and main clinical symptoms of adhesive intestinal obstruction.

144. X-ray diagnosis of acquired intestinal obstruction (invagination, dynamic, adhesive).

145. What are the causes and symptoms of major clinical dynamic obstruction in newborns and older children?

146. The main symptoms of parenchymal organs damage.

147. The main symptoms of hollow organs damage.

148. What injuries of the chest lead to respiratory failure?

149. What are the causes and major clinical manifestations of esophageal injury in children?

150. Name the method of diagnosis and clinical manifestations of renal damage in children.

151. What are the causes, methods of diagnosis and clinical manifestations of lesions of the bladder in children?

152. Specify classification of destructive pneumonia in children.

153. Destructive X-ray diagnosis of pneumonia in children.

154. Describe the phases and stages of acute hematogenous osteomyelitis in children.

155. Name the clinical manifestations and diagnosis of metaepiphyseal osteomyelitis in children.

156. Describe the principles of treatment of acute hematogenous osteomyelitis in children.

157. Features of omphalitis treatment depending on the form and stage of disease.

158. Describe clinical manifestations and treatment of nephroblastoma in children.

159. Describe clinical manifestations and treatment of nephroblastoma in children.

160. What are the main clinical manifestations of bone benign and malignant tumors in children?

161. Name tumors and cysts of mediastinum.

Section 3
MALFORMATIONS IN CHILDREN

3.1. MALFORMATIONS ACCOMPANIED BY RESPIRATORY FAILURE

Specific objectives:

1. To learn the classification of respiratory system, esophagus and diaphragm malformations.
2. Recognize major clinical manifestations of malformations, accompanied by respiratory failure.
3. Differentiate malformations.
4. Interpret auxiliary methods: esophagus probing, ultrasound, fibroesophagogastroduodenoscopy, radiography (plain, contrast), bronchography, bronchoscopy, computed tomography, angiography, etc., laboratory and biochemical tests, hemodynamic parameters (P, AT, Nt, Hb), immunological.
5. Demonstrate examination of the child with malformations of the esophagus, diaphragmatic hernia and lobar emphysema.
6. Identify peculiarities of defects.
7. Offer algorithm of actions and management of patients with malformations.
8. To interpret the general principles of treatment defects.

General questions of the frequency of malformations, teratogenesis, heredity, factors of occurrence, course of birth, diagnostic methods, general methods of preoperative preparation, transportation and providing specialized medical care.

MALFORMATIONS OF THE RESPIRATORY SYSTEM

Malformations of lungs include a large group of pathological conditions and bronchopulmonary system caused by disturbance of the normal development of the lungs and bronchi during embryogenesis.

Summary data of lungs embryogenesis. Building of tracheopulmonal system begins in embryo in the late 3rd — early 4th week of development. Respiratory organs appear as bulging medial caudal foregut segment consisting of primary esophageal and tracheal germ.

Tracheal tube is growing down rapidly and gives beginning to two anlages of bronchi. Primary bronchial anlages rapidly dividing and on 5–6 weeks of fetal development appears branching of bronchial tubes of 2nd order and the formation of three parts of the right and two parts of the left lung.

At 6th–8th weeks of age in embryo main arterial and venous lung collectors are formed.

At the 3rd month primaries of glands, cartilage and muscle components of the walls of the bronchi appear. The beginning of the 4th month of development is characterized by the formation of individual segmental bronchi. During the five-month development occurs qualitative restructuring of terminal bronchi — bronchial lumen dilatation and regeneration of bronchial epithelium. By the middle of the 5th month pulmonary circulation begins to function as a closed system.

In the period from 7th month before birth there is a new structure — alveolus. However, lung alveolisation continues after birth all newborn period. In the period of 8–9 months bronchial tree is topographicaly shaped, and an intensive process of differentiation its distal branches — terminal and respiratory bronchi, alveolar ducts and sacs occures in lungs. There is a formation of functional lung units — acinus.

On the 20th–24th week the synthesis of surfactant begins.

By the 7–8 months development lungs are already so differentiated that can function as an organ of the external respiration and gas exchange.

CONGENITAL LUNG CYSTS

Background. Congenital lung cysts are found in 3–5% of the total number of patients with nonspecific lung diseases, and according to Yu. F. Isakov, V. I. Heraskin, E. A. Stepanov — up to 6% of all chronic lung disease. Right-side localization occurs twice as often. In violation of embryogenesis single or multiple lung cysts arise depending on the period of endogenous and exogenous factors influence. Usually cystic congenital anomalies vary in terms of their development and can be central (at the gate of lung or mediastinal) and peripheral by localization. They result from the separation of small groups of

cells that turn into isolated non-functioning tissue mass. If the cyst is located centrally, it is usually rare, and its course is asymptomatic until it is accompanied with infection. These formations are usually round, and if there is connection with respiratory tract, can be filled with air. If such connection is absent, cysts on X-ray will look like airless parenchymal formations. Peripheral cysts are formed due to the violation of development on 6–16 weeks of gestation and, unlike central, are mostly multiple and small. If after birth there are several cysts, sometimes respiratory distress syndrome occurs, which can even lead to death of the child.

Adrift cysts are divided into uncomplicated and complicated by infection, stress, a breakthrough in the pleural cavity (Fig. 3.1.1).

The clinical course of uncomplicated cysts is asymptomatic, observed in 15% of patients. The most common clinical manifestations of uncomplicated cysts are:

— Lagging of chest half in breathing on the affected side;

— Percussion over the lung field — tympanitis or reduction of sound;

— Heart borders are shifted in healthy side.

Among the most common complicated cysts festering (60%), stress (20%) and a breakthrough in the pleural cavity (5%) occur. Complications usually develop after children infections and flu. In the clinical picture of cysts that have suppurated, the main symptoms are intoxication (fever, hectic temperature curve, pale, flabby, shift of blood formula to the left). It is noted the correlation between the child's age and size of the cyst. The most common symptom is a dry or wet cough. Percussively blunting can be detected, and during auscultation — weakened breathing and wheezing. Intense cysts are developing as a result of the valve mechanism.

Interference that occurs in the bronchi, allows the air that enters the cyst cavity at the time of breathing, stay there. Increasing of the cysts leads to healthy areas compression and displacement of the mediastinum. Clinically tension of cysts manifests in increased anxiety, refusal of food, vomiting reflex and increasing respiratory failure. X-ray study in congenital cysts is critical. Plain X-ray of chest organs can detect air cystic formations with clear contours. The presence of fluid in the cavity indicates at partial drainage of bronchus.

Differential diagnosis. Uncomplicated cysts should be differentiated from bullas at bacterial destruction of lungs, which at repeated X-ray studies often change shape and size, their contours are very clear, and eventually they may even disappear. Multiple congenital cysts may resemble X-ray diaphragmal hernia. In such cases contrast study of the gastrointestinal tract should be conducted. Congenital cysts sometimes have to be differentiated from lungs abscess, parasitic cyst (mostly echinococcal) and lung tumors. In such cases computed tomography, anhiopulmonography can help and in case of echinococcosis — epidemiological history, serological tests and ultrasound.

Treatment of congenital lung cysts is usually surgical.

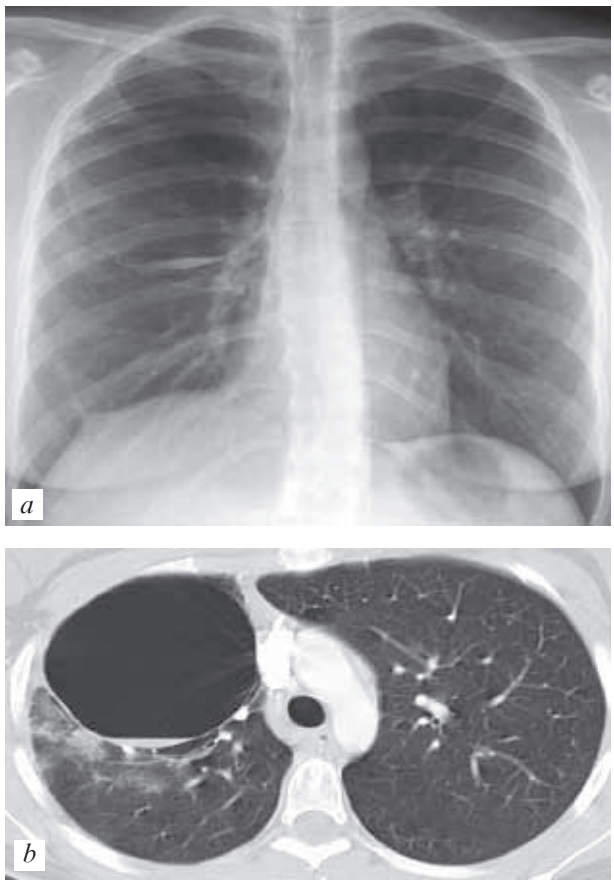


Fig. 3.1.1. Right-sided intrapulmonary bronchogenic cyst (X-ray, CT) *(a, b)*

At intense cysts with mediastinal shift in the preoperative period, percutaneous puncture of cysts and their drainage to reduce intrathoracic pressure are conducted.

The amount of surgery depends primarily on the size of cysts and cystic formations spread to nearby tissues. At central location of the cyst, its large size and pronounced perifocal changes segmental lung resection or lobectomy are preferred.

Agenesis and lung aplasia

Lung agenesis is a malformation at which there are no structural units of the lung: bronchi, vessels, parenchyma — while lung aplasia — just a rudimentary underdeveloped lungs with bronchi. Vessels are absent, and lung tissue is either absent or is a dense structureless area of the lung parenchyma. Both anomalies are extremely rare. According to Klimovich I. T. (1965), agenesis is found only in one child in 144 patients with lung malformations.

Unilateral agenesis is compatible with life, but is often combined with other severe congenital anomalies similar to those detected at VATER-syndrome (vertebral defects, anal atresia, tracheodigestive fistula, renal dysplasia), but other malformations can occur. Children with bilateral agenesis are not viable.

At unilateral agenesia following symptoms are found: respiratory asymmetry, shortening of percus-

sion sound, breathing is weakened and is carried out on the opposite side.

Cardiac impulse is shifted toward destruction. If inflammation of the lungs is added, condition deteriorates, dyspnea, cyanosis occur. Most of these children die within the first year of life due to inflammatory diseases. Typically, heart is rotated clockwise, which manifests in the form of dextrocardia.

On plain X-ray eclipse of appropriate half of the chest with a shift of mediastinal organs in the same direction appears.

Differential diagnosis should be carried out at lung atelectasis, aplasia, hypoplasia and bronchus foreign body.

Surgical treatment is not performed and is aimed at correcting other problems. Thus the main objective is to save the maximum number of respiratory units. At the inflammatory process a patient must be prescribe massive antibiotic therapy.

Lung hypoplasia

This even underdevelopment of all elements of the lung parenchyma and bronchi reaches 85% among congenital malformations. Respiratory underdevelopment of the lungs leads to a significant decrease in its total. There are simple and cystic forms of hypoplasia.

At simplest form, the changes most often occur in the lower lobe of the left lung. Often simple hypoplasia is combined with other malformations.

Functionally healthy lung is hypertrophied, so that functional disorders are not very noticeable. If a patient with a simple hypoplasia lives up to 20–25 years without violations of the breath, the further course of simple hypoplasia is asymptomatic. Most complications occur in early childhood, which is associated with the development of inflammation in the affected lung. Usually, if complications have appeared once, they will progress. Inflammatory changes in the affected lung determine the nature of major complaints. The most common is cough with a significant number of muco-purulent sputum, but sometimes there can be even determined coughing up blood, shortness of breath, palpitation on exertion. These patients suffer from catarrhal diseases which, moreover, are very long lasting and very difficult to treat.

On examination, the patient can note a slight asymmetry of the chest wall and even its slight retraction on the side of defect. At percussion it is possible to note a slight heart shift toward the damage.

Auscultatory, breath may be unchanged or slightly weakened. During inflammation wheezing can be detected.

In the diagnosis physical, functional methods, radiobronchiological survey, anhiopulmonography are used.

X-ray detects the signs typical for lung volume reduction: reduction of hemithorax, narrowing of the intercostal spaces, mediastinal organs shift to the affected side. At bronchographic picture distinguish two simple variants of simple hypoplasia. At first state only the presence of 3–6th row of bronchi, and at the second — from large bronchus very thin branches depart which in their appearance and caliber remind 8–10th row bronchi of normal lungs.

Diagnostic bronchoscopy enables to define a small shift of the trachea toward amended lung, and the phenomenon of subacute or chronic bronchitis.

Methods of treatment: conservative and operative, depending on the amount of damage, the presence of complications, the characteristics of the disease, condition of pulmonary breathing function.

Surgical treatment: typical resection, segmental resection, extirpation of bronchi, combined resections.

At cystic hypoplasia, in contrast to simple, is observed not only reduction in volume of underdeveloped lung but also cystic dilated bronchi. In some cases, they occupy only part of the organ, limited to one or two segments. Histologically cystic hypoplasia is characterized by the absence of cartilage plates in the walls of cysts that distinguishes this condition from acquired bronchiectasis.

Such processes frequently arise in patients with cystic hypoplasia, and eventually become chronic. In such cases, there are phenomena of respiratory failure, initially during physical exercises, and eventually disturb the patient in a calm state. Cardiopulmonary failure may develop. The course of disease at cystic hypoplasia is very diferent. In cases where the course of cystic hypoplasia is "asymptomatic", diagnosis is established only during prophylactic fluorography. However, in these patients the frequency of acute respiratory viral infections is somewhat higher than in children with healthy lungs. These signs do not cause significant concern, so do not bring the patient to the doctor.

In most cases, cystic hypoplasia is the focus of constant existence of inflammatory processes. Depending on the severity of clinical manifestations of cystic hypoplasia forms can be mild, moderate and severe.

At mild form of the disease, patients complain of cough with a small amount of phlegm. Inflammatory processes are rare, normal life and performance are not violated.

Average form is characterized by cough with discharge of about 150 ml muco-purulent sputum. Pneumonias in such patients occur quite often, two or three times a year, mostly one-sided. During remission these patients suffer from shortness of breath at exertion.

Clinical manifestations of severe forms can arise from childhood. Patients suffer from cough with sputum discharge in large quantities — up to 200 ml per day or more. Sputum is purulent with an unpleasant smell. In this form may be marked episodes of hemoptysis and even bleeding from the lungs. Quite often pneumonia arises (4–5 times a year) from the side of underdeveloped lung. Wheezing, shortness of breath even at slight exertion. Over time appear manifestations of cardiopulmonary failure.

The results of physical examination depend on the severity of manifestations of cystic hypoplasia.

These patients look pale, with yellowish gray skin. Fingers can take the form of drum sticks. Above altered lung, chest wall lags in breathing, can

be reduced in volume. Shortening of percussion sound is observed and diverse weezing at auscultation.

Instrumental methods. Plain X-ray in two projections can set cystic hypoplasia, and tomography clarifies its identity. However, the most informative method of investigation of this pathology is bronchography. A characteristic and pathognomonic sign is a deformation of segmental and subsegmental bronchus with thinning borders of their walls.

Surgical treatment is carried out in cases when inflammatory process that develops in the cavities of hypoplasia lobe, does not tend to cease, and there is a great risk of purulent-destructive process. Operation is aimed at removal of non-functional cystic or extirpation of the affected bronchi.

Lung sequestration

It is characterized by separation of the lung tissue areas, usually in the form of cysts located inside or outside the lung, which has isolated blood supply system (abnormal artery, which departs from the aorta or its branches). Lung sequestration is quite a rare pathology that among patients with bronchopulmonary malformations makes 0.15–6.4%.

Interpartial and intrapartial lung sequestration are distinguished (Fig. 3.1.2). Interpartial exists as an independent area of the lung, often modified cystic parenchyma, within a well-functioning lung. Intrapartial sequestration can be located in the pleural cavity, mediastinum, neck and even in the abdomen. Sequested area of the lung parenchyma in morphological relation is always inferior and abnormal. The range of these changes may be significant, from cystic degeneration with the formation of one or more content filled mucous cysts that are lined with bronchogenic epithelium, to fibrotic changes.

Clinical picture. Recognition of sequestration lung is quite difficult due to lack of pathognomonic clinical signs. Usually the course of the disease has the course of chronic recurrent inflammation in the lungs. Disorders, caused by sequestration, primarily arise in the case when the purulent inflammation begins in sequestrated lung.

Then, depending on the course of pathological changes will manifest fairly typical local and general symptoms. Interpartial sequestration often covers the lower proportion of the left lung and lies in backmedial department. There it is connected with arterial vessel, which starts from the lateral wall of the descending thoracic aorta.

In case of inflammatory changes in pulmonary sequestration, patients complain of fever, discomfort in the chest. At the onset (sometimes for quite long) cough does not occur, and if does, with discharge of a small amount of mucous expectoration. Only in case of abscess rapture which was formed in the bone of sequested lung area, in one of the bronchus a large number of sputum is observed, that always attracts the attention of the patient. With the discharge of phlegm general condition of the patient improves, the temperature decreases or becomes normal. Over time, due to existance of bronchus drainage, clinical

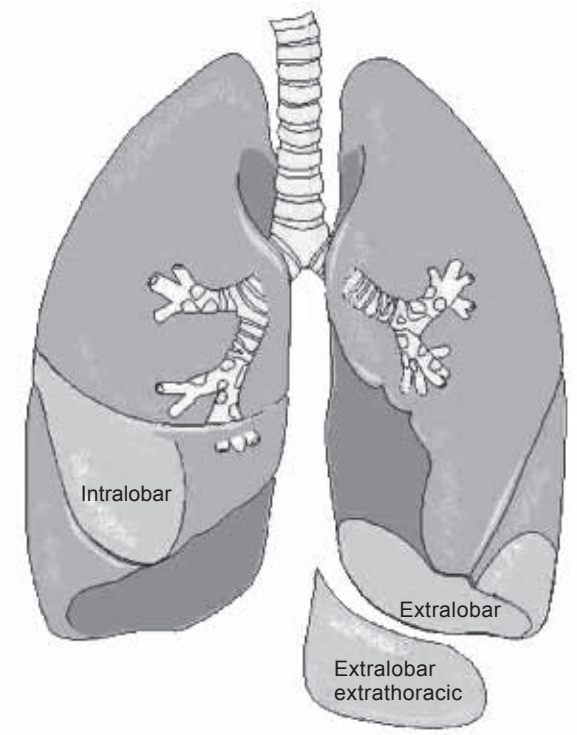


Fig. 3.1.2. Congenital pulmonary sequestration (scheme)

manifestations of the disease acquire chronic lung disease signs, chronic abscess formation. At physical examination of these patients, in addition to general signs of pulmonary inflammation, it is imposible to establish certain pathognomonic signs of lung sequestration.

Diagnosis. X-ray detects homogeneous shadows of round or oval forms with clear contours, or cystic cavities with fluid level on the background of unchanged lung tissue. Targeted tomographic studies can detect aberrant arteries that depart from the thoracic or abdominal aorta. Bronchography assists in detecting only the displacement of neighboring bronchi, but if occurs purulent process with recanalization of cysts, they can be contrast. A clear reflection of abnormal blood vessels can be obtained during aortography (Fig. 3.1.3).

Reliably it is possible to confirm lung sequestration using CT and MRI studies, and abnormal vessels that come from aorta to sequested lung area can be detected. This is very important because during surgery it is possible to injure the unrecognized vessel, leading to massive blood loss and even death of the patient.

Intrapartial sequestration has no direct connection with the lung, can be found on the neck, chest or abdomen.

Cysts, which are formed in the pulmonary sequestration, have no connection with the bronchi and so infect very rare.

There is no independent, enough pathognomonic clinical picture for extrapulmonary sequestration. Typically it does not become the cause of painful disorders and anxiety. Often the diagnosis of intrapartial sequestration is random. If you suspect this de-

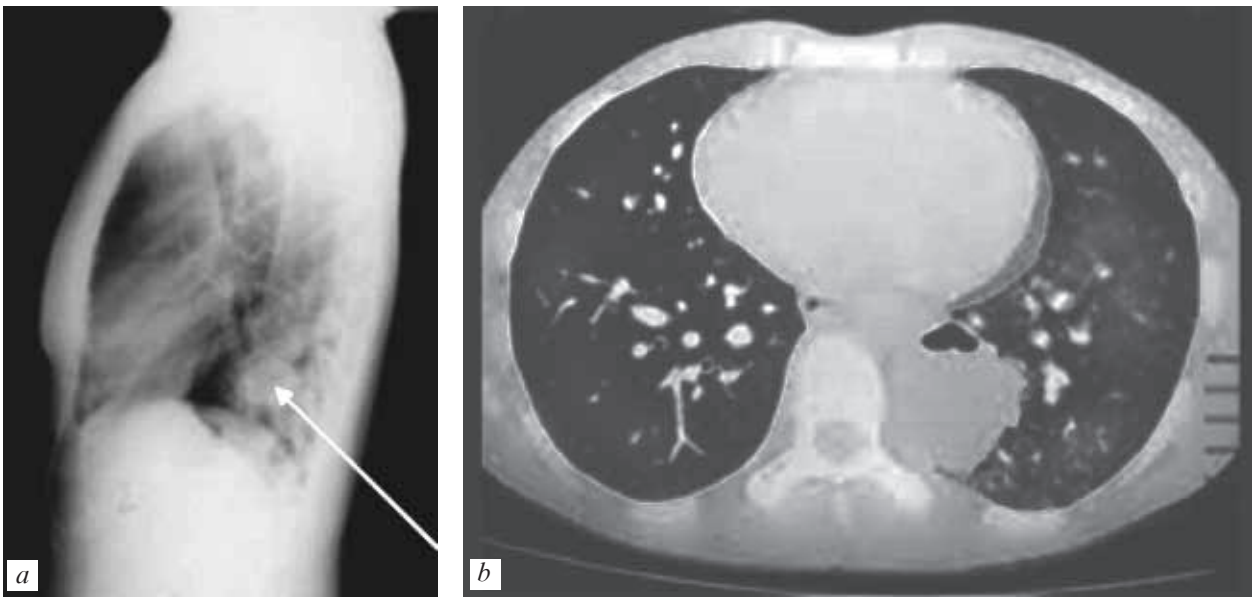


Fig. 3.1.3. X-ray *(a)* and CT *(b)* of left-sided pulmonary sequestration

fect, perform selective contrast aortography to identify anomalous artery.

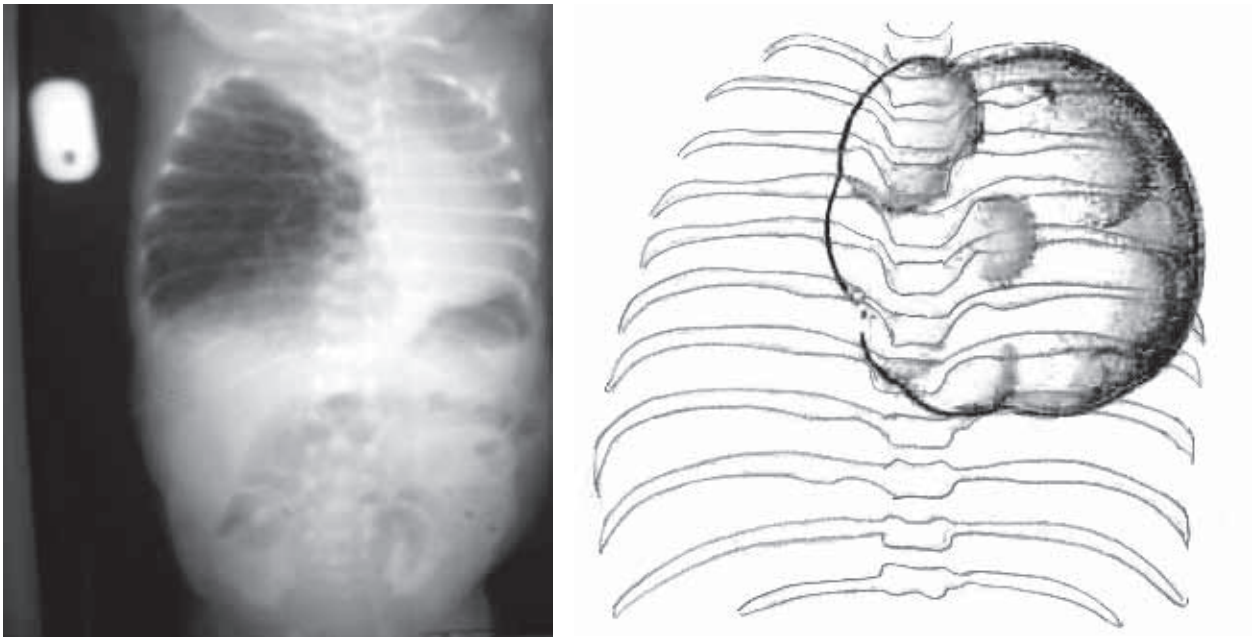
Treatment. Formation of purulent lesions is usually a cause of targeted therapeutic measures regarding interpartial sequestration. In this case surgery is conducted. It is necessary to conduct removal of lung lobe, which includes sequestration site. During the operation should focus on treatment of pulmonary ligament, bandaging and suturing aberrant vessel to prevent profuse bleeding.

Congenital partial emphysema

Background. Congenital partial emphysema is a rare malformation of respiratory system, early diagnosis of which consequences of the disease depend on. It is characterized by sharp increase in volume and airiness of one of the lung parts. *The etiology* of congenital emphysema consists in partial aplasia of smooth muscles of terminal respiratory bronchioles, the absence of intermediate generations of small bronchi or respiratory aplasia department lobe.

The valve mechanism formed in the partial or segmental bronchi, leads to disruption of normal ventilation of the lung by "air trap" type. This disruption of normal ventilation is combined with disorders of the cardiovascular system, circulatory system. Most often this defect occurs in the upper lobe of the left lung (Fig. 3.1.4).

Clinical manifestations depend on the form of partial congenital emphysema: compensated, subcompensated, decompensated.



a *b*

Fig. 3.1.4. Congenital lobar emphysema *(a, b)*

Compensated form is diagnosed in children over 1 year. Developing without acute episodes of emerging disease. The course is slow, asymptomatic. A child has shortness of breath, frequent catarrhal diseases, deformation of chest with bulging of the affected part is observed. This form can be found by chance during X-ray, but sometimes these patients visit the doctor due to moderate respiratory distress during physical exercises that develops over time.

Subcompensated form appears much earlier, at the age of 1–3 months, the child has gradual increase of respiratory disorders or attacks of cyanosis, which quickly pass. It is characterized by periodic course — by intervals of several weeks and more — often without reasons and at different times asphyxia attacks and difficulties while breathing. Such attacks are stopped on their own. Patients most often associate such attacks with carrhal diseases, respiratory tract infections. Over time, a sick child has deformation of the chest with increasing of intercostal intervals on the affected side. Progressing chronic respiratory failure: shortness of breath at movement, and then during rest. The real threat for these patients is pneumonia of a healthy lung. X-ray reveals defect in development.

Decompensated form of congenital emphysema is diagnosed in infants within the first days of life. This form is characterized by manifestation of the syndrome of intrathoracic pressure. A child has increased respiratory failure with long expiration, dry cough, skin cyanosis. Short "light" welfare intervals are changed by episodes of severe asphyxia with loss of consciousness and convulsions. Examination noted the asymmetry of the chest. Breathing is not auscultated, shift of cardiac dullness to the opposite side.

On the X-ray on the affected side diaphragm dome is flattened, available shift of mediastinal organs to the healthy side, increased volume and transparency of one of parts.

Treatment of congenital emphysema of newborns is only operative. At decompensated forms of interventions, surgery is performed urgently, at subcompensated form — term of surgery depends on the child's condition and course of disease, at compensated one — treatment is planned. In all cases, the choice of surgery is to remove the altered, maldeveloped lung lobe.

DIAPHRAGM DEFECTS

Diaphragmatic hernia

Diaphragmatic hernia is a malformation that results in a possible exit of abdominal organs into the chest through natural and abnormal openings in the diaphragm septum (Fig. 3.1.5).

According to Mark D. Stringer et al. (2006), the incidence of diagnosed diaphragmatic hernia in infants is approximately 1 in 2000–5000 live births. Still there is a high level of overall mortality at this defect (14–30%), despite the optimization of pre- and postnatal diagnosis, surgery and post-operative care for a child. Most often deaths of these patients is associated with lung hypoplasia, pulmonary hypertension and visceroabdominal imbalance.

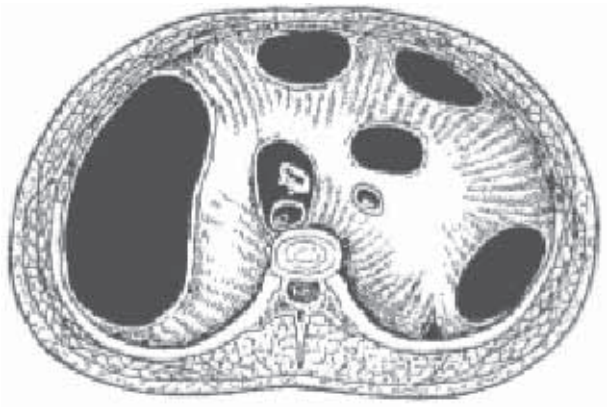


Fig. 3.1.5. Sites of failure of the diaphragm to completely close during development

Classification of diaphragmatic hernial by S. Y. Doletskyy:

1. Hernia of diaphragm:
— True hernia.
2. Protrusion of bounded dome area.
3. Full protrusion of the dome (relaxation):
— False hernia.
4. Fissure posterior defect (Bochdeleck's).
5. Absence of one diaphragm (aplasia).
6. Hiatal hernia:
— Oesophageal;
— Paraesophageal.
7. Hernia of the anterior diaphragm:
— Front;
— Phrenopericardial.

Clinical picture depends on the type of hernia, hernia gate size, volume and type of hernia contents. Terms of symptoms manifestations are very different — from pronounced since birth to asymptomatic, latent hernia that due to such course are diagnosed by chance in about 15% of cases.

All diaphragmatic hernia symptoms can be divided into three subgroups.

1. Violation of respiratory function that occurs due to movement of abdominal organs, leading to compression of lung on the affected side, the shift of mediastinum, partial compression of contralateral lung. In children hypoxia, which worsens during feeding, is observed. Respiratory disorders manifest in cyanosis, attack-like cough, respiratory insufficiency.

2. Violation of the digestive tract function can occur in two options:

a) infringement of hollow organs at hernia gates, which can lead to intestinal obstruction (pain, vomiting, discharge absence of feces and gases) and eventually to the development of peritonitis. Most often infringes Bochdeleck's hernia;

b) at hiatal hernia there are conditions for reflux (regurgitation, vomiting), aspirations, occurrence of peptic esophagitis with symptoms of bleeding in emesis, anemia, eventually formed cicatricial stenosis may develop.

3. Violation of the cardiovascular system, which is caused by a shift of mediastinum, heart and its rotation, and inflection of large vessels, performed by shortness of breath, palpitations, cyanosis, enlargement of the liver, changes in the electrocardiogram.

Also at diaphragmatic hernia common disorders in the organism of the child will be marked. These patients generally lag in development, hypotrophic, anemic.

Severity of these symptoms depends on the type of hernia and age. The most severe clinical picture takes place in newborns and in children over 1 year clinical picture is less pronounced.

For each type of hernia major symptoms can be identified:

1. Diaphragmatic hernia — respiratory and cardiovascular system disorder.

2. Oesophageal hernia — disorders of the gastrointestinal tract.

3. Anterior diaphragmatic hernia — abdominal pain.

False diaphragm hernia

Defects of the diaphragm can be divided into three groups: posterior defect, great dome defect and the absence of one of the domes of the diaphragm. Fissure defect is located in the area of the lumbar-rib department of diaphragm. Usually there are two versions of this defect — at the first gap is located on a wall surface, at the second gap is separated from the chest wall with slices.

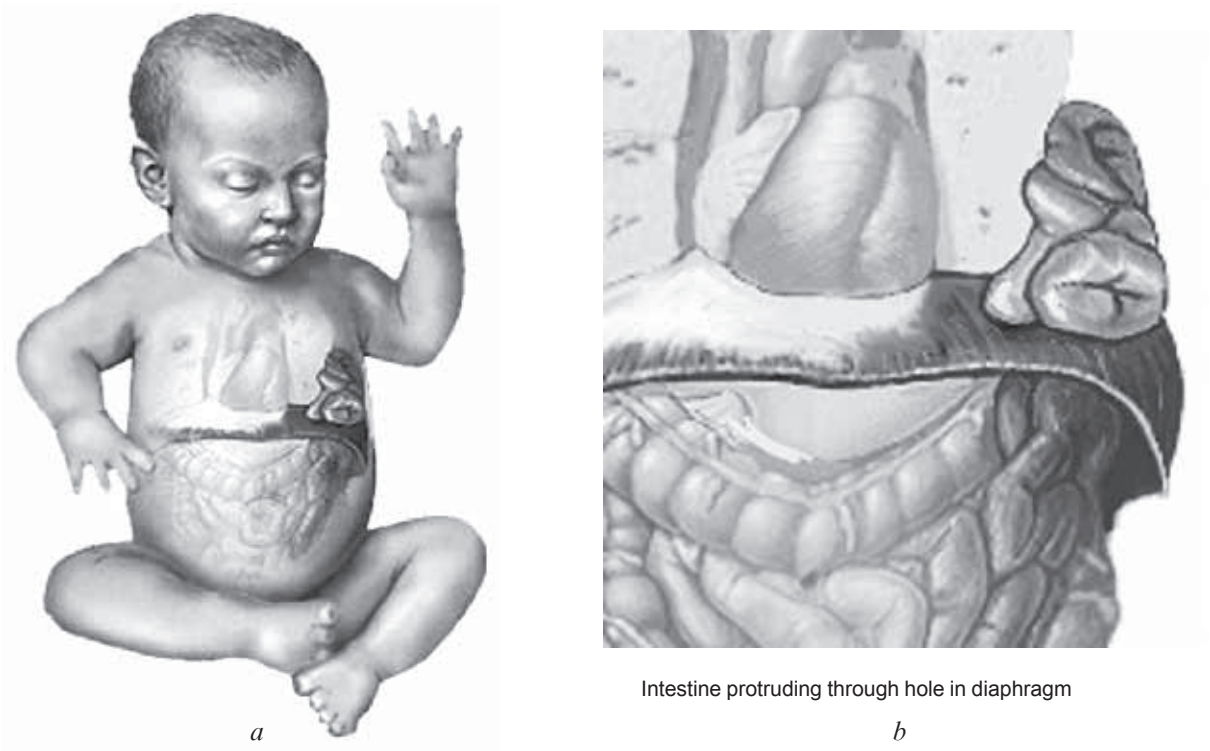
The leading symptoms include cyanosis, dyspnea, signs of hypoxia. On examination respiratory excursions are enlarged, asymmetric chest, explodes from lesion, stomach is hollow. Percussively on the corresponding side it is marked blunting or tympanitis, auscultatively — weakened breathing, peristaltic sounds, heart tones are shifted to a healthy side. Quite often one can observe the development of severe asphyxia (Fig. 3.1.6).

The patient's condition progressively worsens during crying, shifting the child and feeding. At flatulence there are signs of intestinal obstruction, accompanied by asphyxia. S. Ya. Doletskiy called such state asphyxia jamming, emphasizing the increasingly important role of acute respiratory failure symptoms.

True hernias of the diaphragm

True hernia of the diaphragm is characterized with asymptomatic course. Often it depends on the level of displacement of abdominal cavity organs into the chest. In some cases, the hernial sac is so stretched that contains all the intestines, moved to pleural cavity. In these situations collapsed lung, heart and blood vessels mediastinal shift with a significant violation of their functions can happen, violations of the gastrointestinal tract are possible. With small sizes of hernia protrusion the course is asymptomatic.

Diagnosis of diaphragmatic hernia should begin with fetus. Proper identification of the defect and its severity rating in the neonatal period can greatly im-



a *b*

Fig. 3.1.6. Congenital diaphragmatic hernia (scheme) *(a, b)*

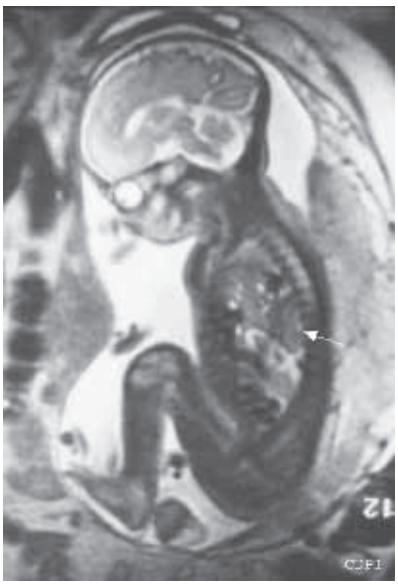


Fig. 3.1.7. Sagittal MRI of the fetus with left sided diaphragmatic hernia

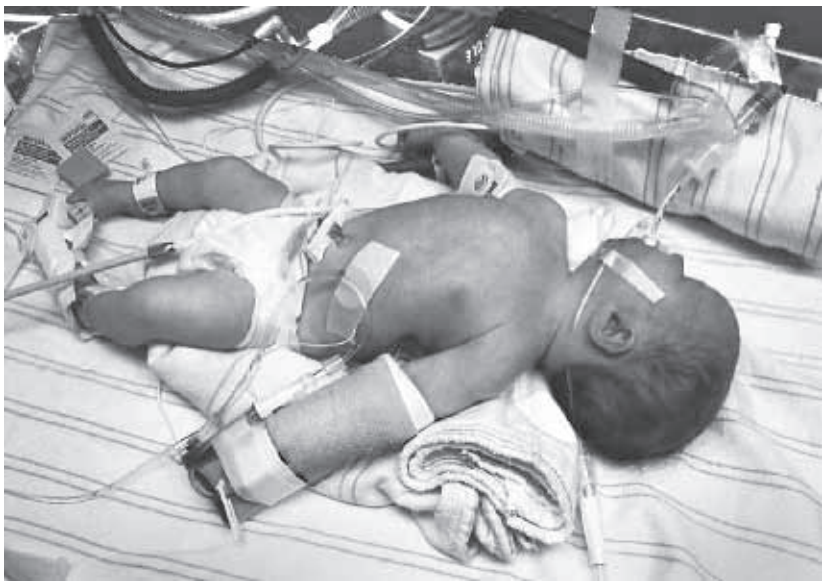


Fig. 3.1.8. Patient with diaphragmatic hernia

prove the treatment and care of the patient. Prenatal ultrasound can detect diaphragmatic hernia on the 14th week of gestation, if necessary, can be made MRI of the fetus in order to detail the severity of the defect (Fig. 3.1.7). At establishing diaphragmatic hernia in the fetus should also carefully look for other anomalies because, according to the literature, they can occur in 50% of cases. The most common diaphragmatic hernia are combined with the abnormalities of the gastrointestinal tract and cardiovascular system, as well as chromosomal abnormalities. Thus, at prenatal diagnosis of diaphragmatic hernia amniocentesis should be done to rule out chromosomal abnormalities.

In the postnatal period the most common methods of diagnosis are plain X-ray, X-ray with contrast agent and ultrasound. At diaphragmatic hernias an observational X-ray on the affected side detects more abnormal enlightenment of oval or spherical shape, transparency of which depends on the degree of filling of intestinal loops with air.

Differential diagnosis is made with swelling and hemorrhagic syndrome, lobar emphysema, lung cyst, spontaneous pneumothorax (Fig. 3.1.8).

Treatment. Earlier, the death rate during diaphragmatic hernia reached about 50%, today, with the possibility of care after these children in neonatal hospitals, it decreased to 30% or less.

If the diagnosis was established prenatally, it is important to carefully plan birth. Support of a newborn in such cases should be focused on breathing and hemodynamics.It is necessary to enter a nasogastric tube to decompress the stomach and prevent the dilatation of the intestine within the chest, if necessary, adjust artificial respiration. Today worldwide is common expectant practice regarding surgery. Surgery is performed when the child's condition becomes stable (Fig. 3.1.9).

The volume of surgery is determined by the essence of malformations.

At false diaphragmatic hernia defect closure or alloplasty is used, at true — diaphragmoplasty (Fig. 3.1.10).

Approximately half of the observations of vomiting appears from the first hours of life and very often emesis contains blood, they may look like coffee grounds. Recurrent aspiration pneumonias are typical. As a result of reflux esophagitis hemorrhagic syndrome develops that causes chronic anemia. Older children complain of mild pain that occurs after a meal, in a prone position.

X-ray studies in the Trendelenburg's position detect cardia above the diaphragm and gastroesophageal reflux during esophagoscopy can be detected peptic esophagitis.

Repair of diaphragmatic hernia is shown at Fig. 3.1.11, 3.1.12.

Paraesophageal hernias are characterized by that cardio is located fine, but stomach or intestines protruding in the hernial ring, which can cause strangulation. The angle between the esophagus and the stomach remains, which causes the absence of reflux. The failure may not have clinical manifestations and be a random finding. Some patients have vomiting, almost always without admixture of blood, but also found hidden bleeding include strangulation, which may even lead to anemia. Mostly in newborns complications have respiratory character, may have shortness of breath and cyanosis.

The diagnosis is confirmed by X-ray, determined cystiform formation, sometimes with a fluid level which increases during breathing.

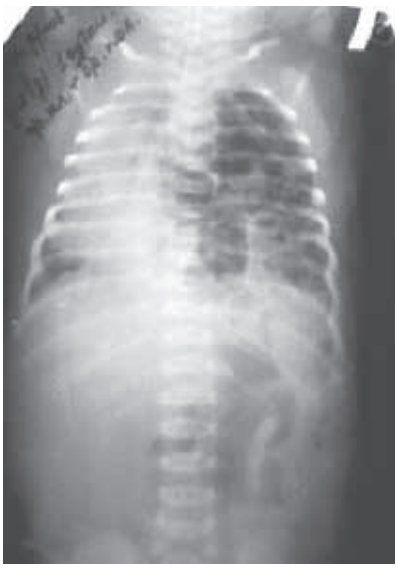


Fig. 3.1.9. X-ray image of the fetus. Patient with a left-sided diaphragmatic hernia

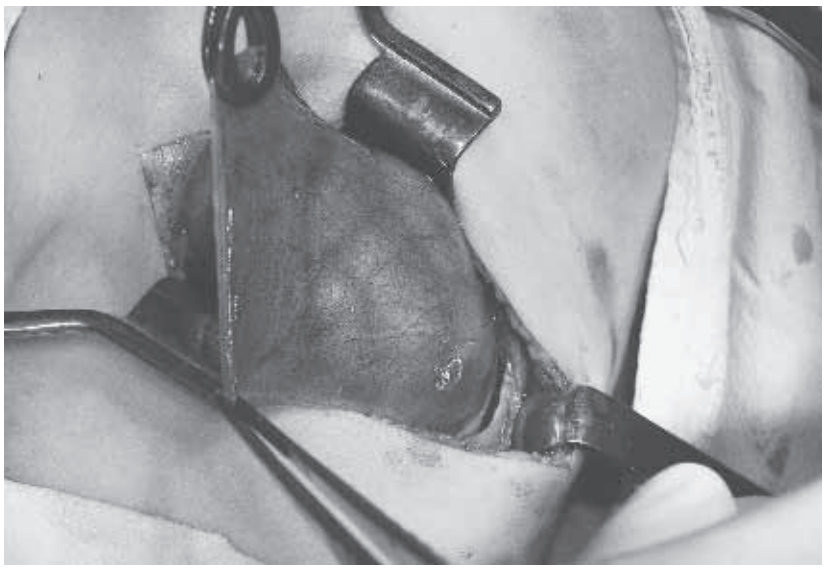


Fig. 3.1.10. Intrasurgical picture of diaphragmatic hernia

Hernia of the anterior part of diaphragm

Parasternal hernia, at which hernia ring is a Larrey's slot, are usually with asymptomatic course. Sometimes children complain of chest rumbling outside, moderate pain sensations, sometimes a dry cough. Physical examination reveals no absolute cardiac dullness, weakened heart tones, chest auscultative may be determined by intestinal peristalsis. Diagnosis is based on radiopaque study of the gastrointestinal tract.

Phrenopericardial hernias are manifested acutely since birth (permanent cyanosis, vomiting, anxiety), due to partial cardiac tamponade by bowel loops, which are located in the cavity of the pericardium. In some cases, displaced abdominal cavity organs into the heart bag so violate function of the heart that may even cause its arrest. Radial symptoms are poorly understood.

Penetrating into the cavity of the pericardium, intestinal loops cause heterogeneity of heart shadows that is stored in any projections, while during the front part of diaphragm hernia additional formation is found in a side view and goes forward. Phrenopericardial retrograde hernia can only cause changes in the mediastinum. Characteristic cardiac curves are not defined, and there is no typical cardiac pulsation, median shadow contours are smoothed, straightened and look like tents.

Treatment. At hiatal hernia surgery is performed. At paraesophageal hernia after lowering the stomach and cutting of hernial sac suturate diaphragm legs behind esophagus. At expressed gastroesophageal reflux simultaneously with legs suturing perform antireflux surgery.

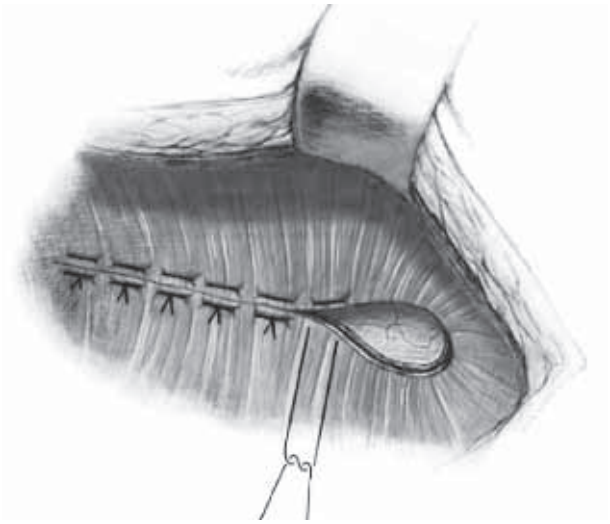


Fig. 3.1.11. Repair of diaphragmatic hernia by direct suture

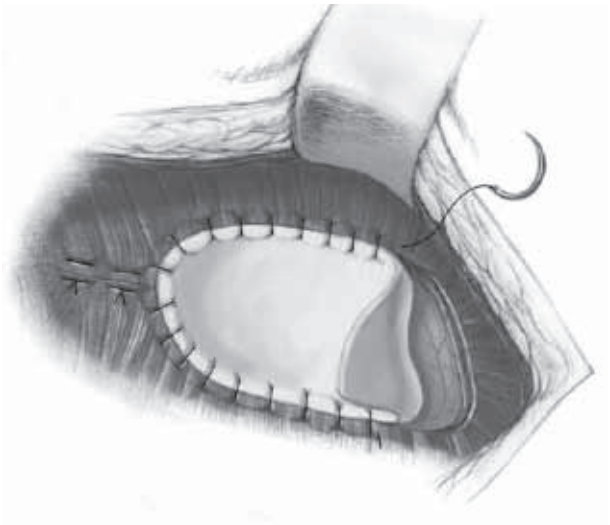


Fig. 3.1.12. Repair of diaphragmatic hernia with prosthetic patches

3.2. MALFORMATIONS OF ESOPHAGUS

Malformations and diseases of the esophagus occur at different ages beginning from the newborn period to a later age. The main clinical syndrome is a syndrome of respiratory failure due to aspiration syndrome. The most common defects and diseases of the esophagus are following (Fig. 3.2.1):

Esophageal atresia

Esophageal atresia is a severe malformation, during which the upper segment of esophagus ends blindly. The lower segment of the organ is most commonly connected with trachea. Esophageal atresia is often associated with other malformations — congenital heart disease, gastrointestinal tract, urinary tract, etc. In 5% of cases esophageal atresia occurs during chromosomal diseases. Population morbidity — 0.3:1000. The combination of male and female — 1:1.

Development of the defect is associated with disturbances at the early stages of embryogenesis. It is known that trachea and esophagus arise from a single anlage — the head end of the foregut. In the earliest stages trachea is widely connected with esophagus. Their separation occurs at 4–5 weeks of embryogenesis. During the wrong direction and growth rate of the trachea and esophagus, as well as the processes of vacuolization in solid phase, which the esophagus passes together with other entities of the intestinal tube in a period of 20 to 40 days, may develop esophageal atresia. From history of pregnancy polyhydramnios and risk of miscarriage within the first trimester are typical.

Anatomic forms of esophageal atresia can be both without connections with trachea (complete absence of luminal, esophageal aplasia), and with tracheoesophageal fistula.

Classification

On the basis of anatomic variants six types of esophageal atresia are distinguished (Fig. 3.2.2).

The walls of the blind ending segment of esophagus are hypertrophic, and in its distal segment thinned.

Clinical picture and diagnosis. Clinical symptoms appear soon after birth. They quite typical, which creates conditions for early diagnosis and adequate surgical treatment. The earliest, and at the same time stable symptom is an abundant and continuous discharge from the mouth and nose of newborn frothy viscous mucus (false "hypersalivation"). A part of mucus aspirate, cyanosis occurs. After the extraction of content a temporary improvement comes, and then foamy discharge and cyanosis re-appear. This allows suspecting obstruction of esophagus before the first feeding.

The second major symptom is detected with the first feeding of the child or drinking liquids. Food or liquid immediately stands back with full atresia and tracheoesophageal fistula, getting to airways, causes choking and coughing with severe respiratory failure and the appearance of cyanosis. Regurgitation during eating and drinking differs from vomiting by the fact that it occurs after one–two sips.

The appearance of these symptoms requires immediate check of patency of esophagus. For this purpose, use sensing (catheterization) of esophagus by

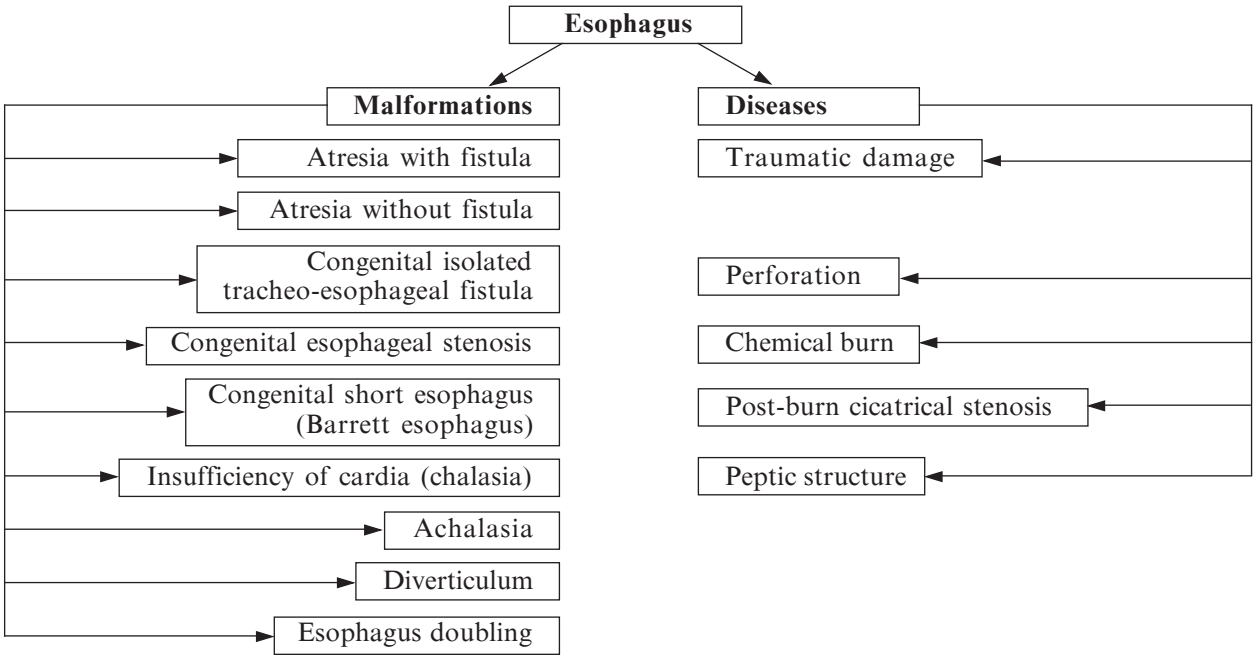
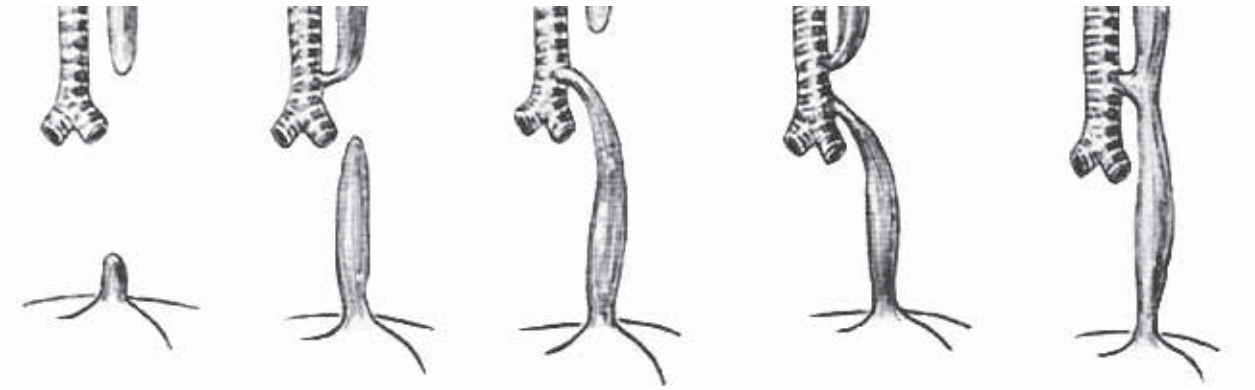


Fig. 3.2.1. Malformations and diseases of esophagus



a *b* *c* *d* *e*

Fig. 3.2.2. Gross's anatomical classification of esophageal atresia: *a* — the complete absence of esophagus, instead there is a connective strand; *b* — the esophagus forms two isolated blind sacs; *c* — the upper segment of esophagus ends blindly, and the lower is connected by fistulous course with trachea above its bifurcation; *d* — the upper segment of esophagus is connected by fistulous course with trachea and the lower segment ends blindly; *e* — the upper and lower segments of esophagus are connected with trachea by fistulous course.

catheter number 8–10, which is inserted through the nose. During atresia catheter can not be conducted deeper than 10–12 cm from the edge of the gums, because it rests on a blind segment of esophageal atresia. Quite informative is Elephant probe: in catheter which is in esophagus and held to the end, 10–15 cm^3 with a syringe is injected air. During atresia blown air immediately with the noise comes back through the patient's mouth and nose.

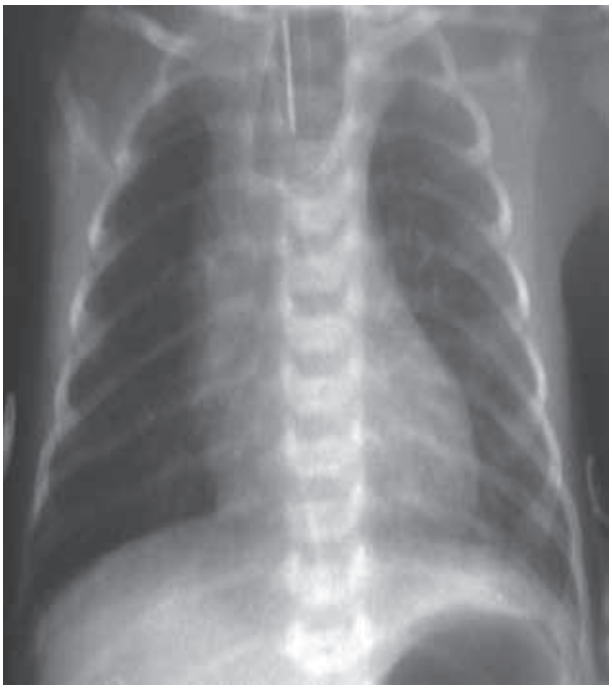


Fig. 3.2.3. X-ray of Esophageal atresia (with nasogastral tube)

In a few hours of life in a child with esophageal atresia, in cases where there is esophageal-tracheal fistula develops respiratory distress syndrome, aggravated by inhalation of gastric contents.

During child's examination reconfiguring of the upper part of the abdomen is marked: its retraction or flattening, typical for complete atresia, or bulging in the case of esophageal atresia with tracheal fistula. Very quickly respiratory disorders develop, such as apnea, disorder of the respiratory rhythm, intermittent and then permanent cyanosis, in the lungs moist rales appear. Strengthening is typical for these disorders after each meal and liquid. After the birth of a child with esophageal atresia within 24 – 28 hours is probable meconium stool, and then constipation appears.

Within 2–3 days after birth, the baby's condition is deteriorating; severe complications develop, the most important of which is the aspiration pneumonia due to spill of any food or mucus in the respiratory tract through the fistulous passages. With full atresia no less severe complication occurs — dehydration and exhaustion. Respiratory disorders lead to respiratory acidosis, polycythemia, and increased hematocrit.

The final diagnosis of esophageal atresia and refining its shape are possible with X-ray (Fig. 3.2.3, 3.2.4). X-ray examination begins with an overview image of the chest, and then transferred to the X-ray of the abdomen. In the presence of distal tracheo-esophageal fistula air in the stomach and intestines is detected.

Subsequently contrast study of the esophagus in the upright position is conducted in order to prevent aspiration of contrast material. It is advisable to use as a last-soluble iodine preparations, or by using radiopaque probe in esophagus. X-rays are taken in the frontal and lateral projections.

After the end of X-ray contrast agent is thoroughly drained. On the X-ray filling with contrast medium blind segment of the esophagus or its penetration directly into the lungs (with esophageal — tracheal fistula) reveals. However, with full esophageal atresia contrast material may be in the lung tissue due to regurgitation, but in much lesser quantity. In order to prevent this phenomenon used amount of contrast should not exceed 2 ml.

Treatment. Already in maternity hospital preoperative preparation should be started, including mouth-nasal aspiration every 15–20 minutes, oxygen supply, complete exclusion of oral feeding. Transportation must be carried out by a specialized team as soon as possible. The total preoperative duration is determined by the severity of homeostatic disorders and hemodynamic, respiratory failure, the degree of dehydration. At clear evidence of aspiration, respiratory failure, and even more in pneumonia or atelectasis should as soon as possible resort to direct laryngoscopy and tracheal catheter aspiration. In case of ineffectiveness of the latter, under anesthesia bronchoscopy or tracheal intubation is performed with thorough aspiration. The patient is placed in incubator, which provides a continuous flow of oxygen, aspiration of oropharyngeal contents, warming. Infusion, antibacterial, symptomatic therapy are provided.

The choice of method of surgery is determined by the shape of atresia and patient's condition. At the most common form of atresia with distal tracheoesophageal fistula at patients with low operational risk (full-term, with no concomitant defects of the vital organs and symptoms of intracranial birth trauma) should be started with a thoracotomy, division of tracheoesophageal fistula. If diastasis between the ends of the esophagus does not exceed 1.5–2 cm, impose direct anastomosis. With a large diastasis segments of esophagus cervical esophagitis and gastrostomy by Kader are imposed. At non fistula forms because of the significant diastase surgery of gastrostomy and esophagostomy is performed. At patients with high operative risk surgery is started with gastrostomy. The second stage of operation is performed after improvement, after 2–4 days.

Repair of esophageal atresia is shown at Fig. 3.2.5.

A child in the postoperative period within the first year of life requires constant dispensary supervision. May occur dysphagia, complicated by obstruction in anastomosis area what requires urgent esophagoscopy. Therefore, children within the first year of life require feeding with homogenized food supply. Failure of cardio- and gastroesophageal reflux, often complicating the postoperative period, is clinically manifested by nocturnal regurgitation, recurrent pneumonia, and require prompt diagnosis. Due to the recurrent nerve injury of children in the next 6–12 months hoarseness is possible.

At children with esophago- and gastrostomy at the age of 2–3 months to 3 years a second stage of operation — esophagoplasty with colonic graft is performed.

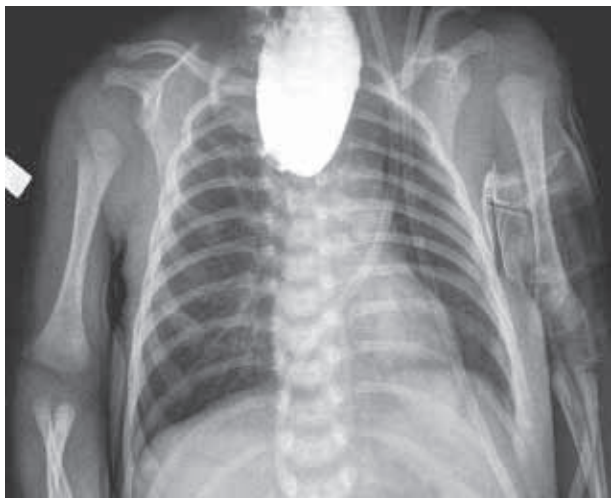


Fig. 3.2.4. X-ray of esophageal atresia (with contrast)

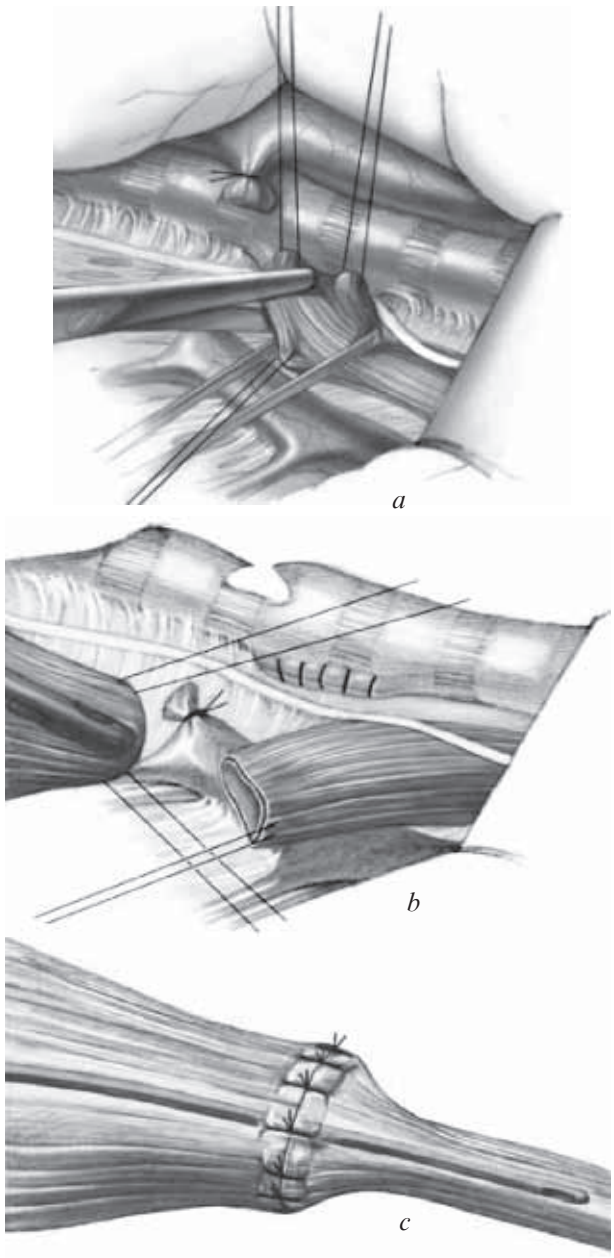


Fig. 3.2.5. Repair of esophageal atresia *(a–c)*

3.3. CONGENITAL INTESTINAL OBSTRUCTION

Congenital intestinal obstruction is among the most common morbid conditions which call for emergency surgery. It occurs in children at any age, though most frequently in the neonatal period. It is caused by various malformations which can be conventionally separated into four groups: malformations of the intestinal tube; malformations of the intestinal wall; malrotation of the intestine; defective development of other abdominal organs.

These malformations originate during organogenesis (the first 3–4 weeks of intrauterine development) when one of the processes of the formation of the intestinal wall and lumen and growth and rotation of the intestine are distorted.

The digestive tube goes through a "solid" stage during its development, with the proliferating epithelium closing the intestinal lumen completely. The process of vacuolation which follows next terminates in the restoration of the lumen of the tube. In certain cases, however, the last phase is disturbed and the intestinal lumen remains closed. When recanalization is disturbed only in a small area the lumen is closed with a thin membrane (membranous atresia) (Fig. 3.3.1).

An opening of various size forms in the membrane (membranous stenosis) in cases in which recanalization has already started. In closure of the lumen for a considerable distance, the atresia has the character of a fibrous strand. This form of atresia may result from deficient development of the corresponding branch of the mesenteric vessel. Atresia may be multiple ("sausage" form).

The malformations are encountered most frequently in the regions of "complex embryological processes", namely, in the major duodenal papilla, at the junction of the duodenum and the jejunum, and in the distal segment of the ileum.

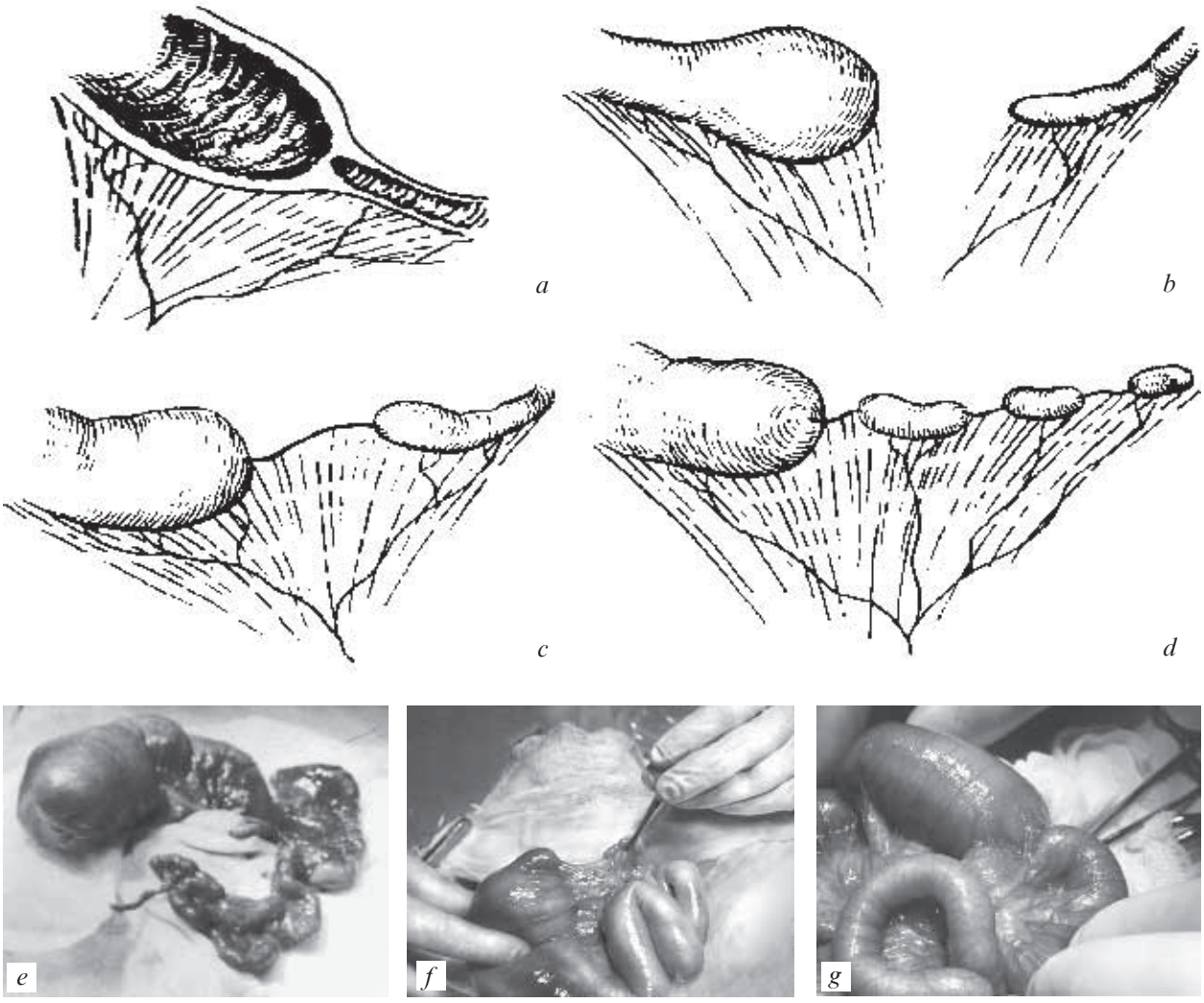


Fig. 3.3.1. Atresia of the intestine and its variants: *a* — membranous form; *b* — complete atresia with separation of the blind ends; c — atresia in the form of a fibrous strand; *d* — multiple atresia ("sausage" form): *e, f, g* — view of affected organs

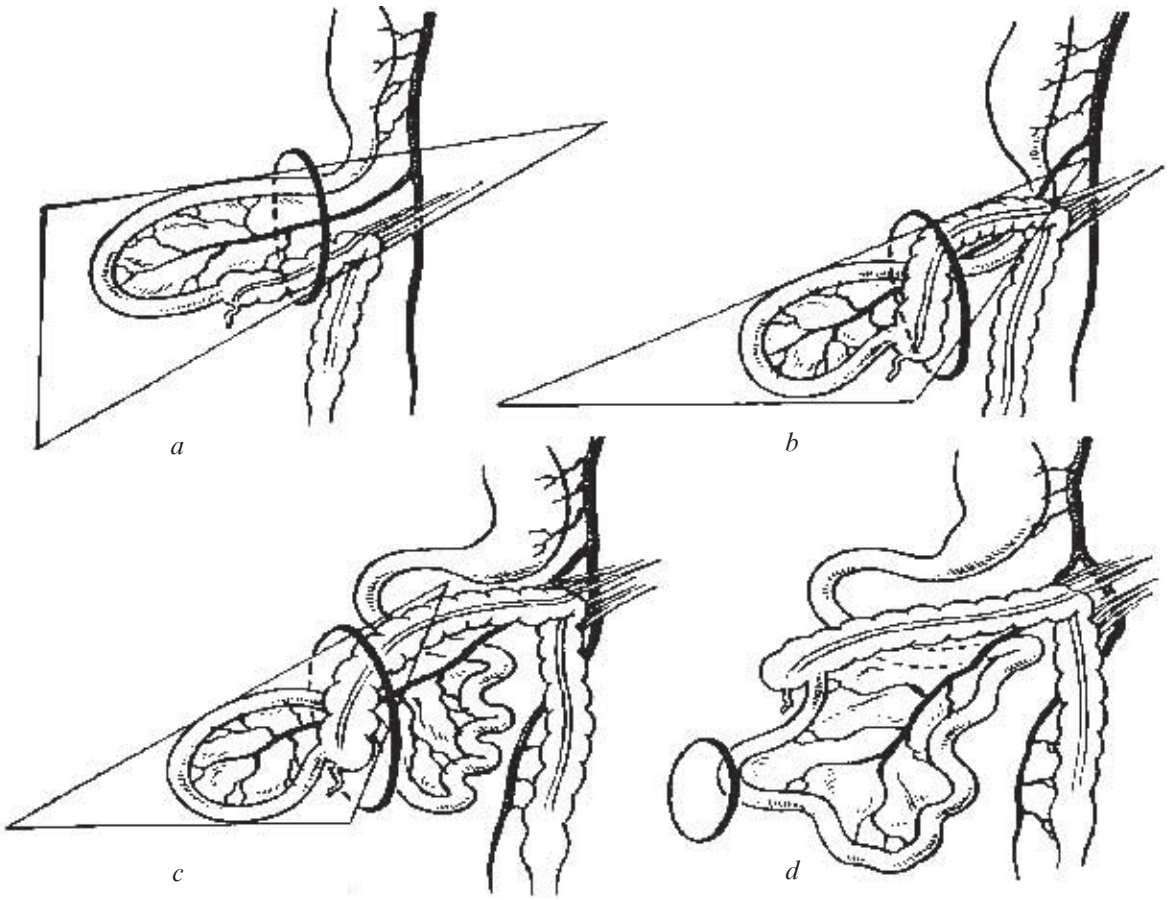


Fig. 3.3.2. Normal rotation of the intestine: *a* — stage of "physiological" umbilical hernia; *b* — the first stage of rotation; rotation by 90°, the large intestine is on the left, the small intestine is on the right; *c* — second stage of rotation: rotation by 270°, the small and large intestine have a comon mesentery; *d* — the third stage of rotation is concluded by fixation of the intestine, the small and large intestine acquire separate mesenteries

The intestine changes from the primary embryonic position (phase of "physiological embryonic hernia") to the normal position by rotation (Fig. 3.3.2). Delay of rotation in the different stages may result in various malformations which cause intestinal obstruction. When this happens in the first stage, the baby is born with incomplete rotation of the intestine. The midgut (stretching from the duodenum to the middle of the transverse colon) in this case remains fastened at one point, at the origin of the superior mesenteric artery.

The loops of the small intestine are in the right half of the abdominal cavity, the caecum is in the epigastrium, under the liver or in the left hypochondrium, the colon is on the left. This type of fixation creates the conditions for torsion about the root of the mesentery and the development of acute strangulation intestinal obstruction.

When the second stage of rotation is disturbed, the caecum located in the epigastrium is fastened by embryonic strands in front of the duodenum and compresses it.

Compression of the duodenum may be combined with torsion around the superior mesenteric artery, this is known as Ladd's syndrome. Atypical position of the vermiform process together with the caecum makes the diagnosis of acute appendicitis in older children and even in adults difficult (Fig. 3.3.3).

In disturbance of the third stage of rotation, fixation of the intestine changes which results in the formation of defects in the mesentery and various pockets add recesses predisposing to strangulation of the intestinal loops. Internal hernias (Fig. 3.3.4) are variants of these anomalies.

Meconium ileus is a condition of peculiar origin, it results from congenital cystic fibrosis of the pancreas. Due to the deficient enzymatic activity of the gland, the meconium is abnormally viscous and blocks the lumen of the ileus (Fig. 3.3.5).

All types of congenital intestinal obstruction are subdivided into high and low according to the level of the obstruction, into acute, chronic, and recurrent according to the course, and into complete and incomplete according to the extent of closure of the intestinal lumen.

Main symptoms of congenital intestinal obstruction are vomiting with pathological impurities and lack of meconium stool. Other numerous symptoms are of specific type of obstruction. Diagnosis is based on an assessment of the main symptoms and objective examination data. On examination the general condition is evaluated, the presence of stigma embryo

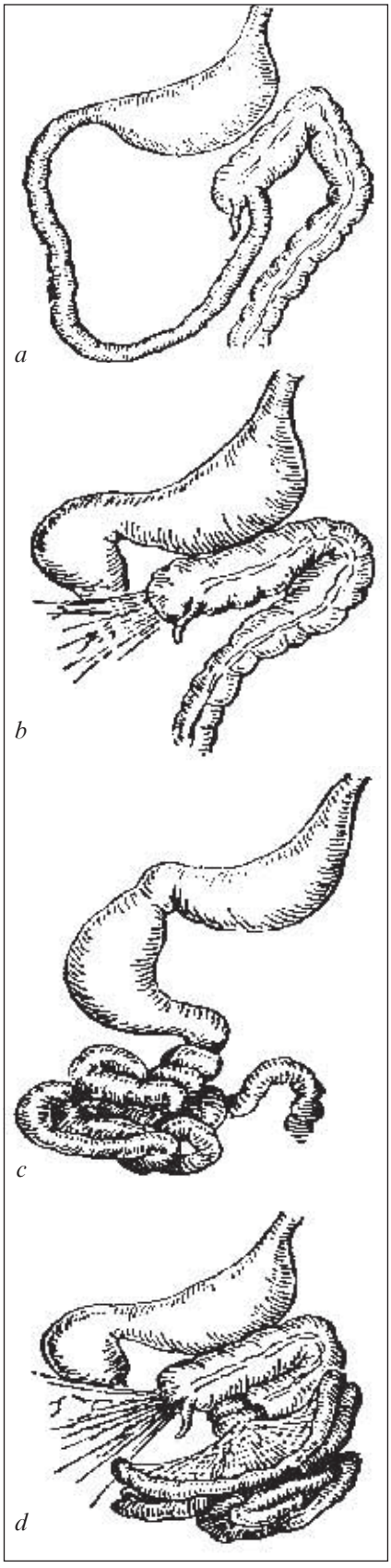


Fig. 3.3.3. Main variants of malrotation of the intestine: *a* — complete absence of rotation; *b* — compression of the duodenum by strands of the mesentery; *c* — torsion of the midgut; *d* — Ladd's syndrom

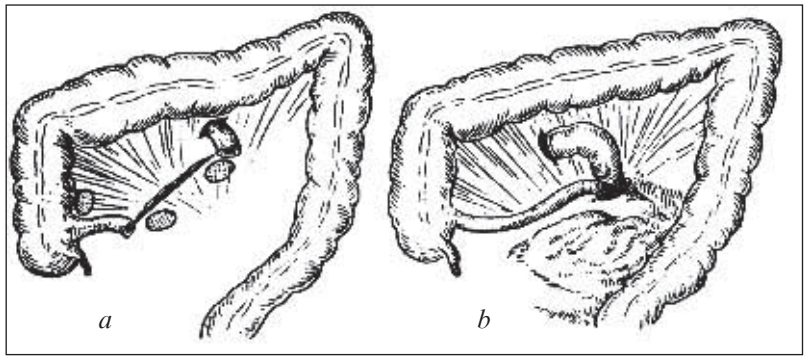


Fig. 3.3.4. Internal hernias (schematical representation): *a* — location of internal hernias; *b* — appearance of hernia and incomplete according to the extent of closure of the intestinal lumen

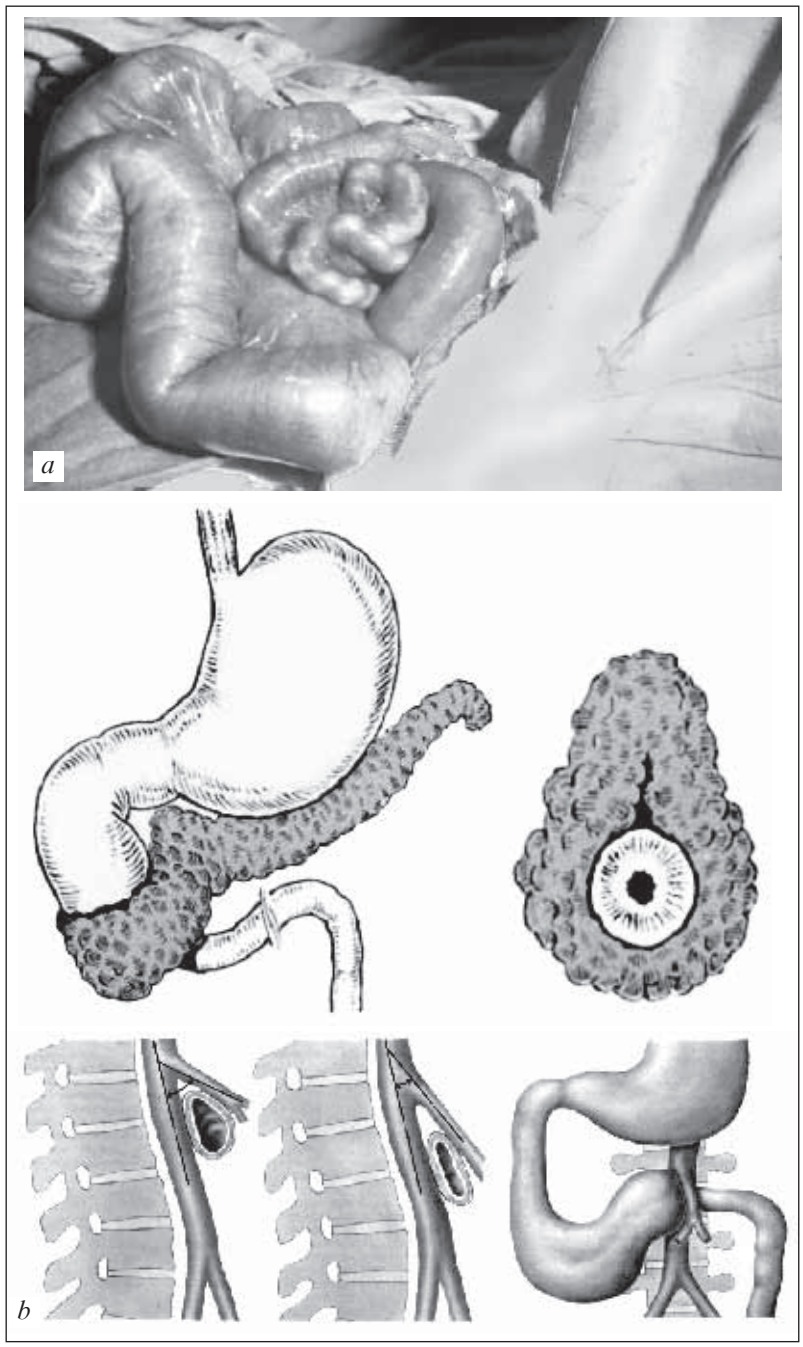


Fig. 3.3.5. Malformations that lead to compression of the intestinal tube (meconium ileus annular pancreas, aberrant vessel) *(a, b)*

fetopathy, the degree of prematurity, the display of toxic-exsicosis.

High congenital intestinal obstruction is a set of intestinal lesions, causing the impassability of the gastrointestinal tract at the level of the stomach, intestine, 12-duodenal ulcer of primary sections of jejunum. During the first day an independent stool or after the enema. Character of vomiting and stool depends on the level of obstruction. If the obstruction is located above the ampulla of Vater, the vomit will be without bile, there may be colored stool. At obstruction below ampulla of Vater in the vomitting will be admixture of bile, stool is not colored, scanty.

Considering the level of obstruction — secreting sections of the intestine — a newborn child is suffering from the loss of water and electrolytes, and therefore exsicosis develops. In addition, excessive vomiting, typical for high obstruction, leads to aspiration pneumonia.

Low intestinal obstruction that occurs at all levels, ranging from 15 cm of jejunum, including small and large intestine.

Low intestinal obstruction develops at the level of suction intestine, and therefore is characterized by the development of intoxication, and translocation peritonitis, due to bloating and high standing of the diaphragm dome — respiratory disorders.

Clinical picture is characterized by the absence of stool and gas from the first day of life; even abdominal distention, increasing over time; green vomiting with or stool from the end of second, beginning with the third day of life.

The *method of examination* in cases of suspected congenital intestinal obstruction consists of the following moments:

(1) study of information concerning the course of pregnancy and delivery;

(2) study of the disease history (the time of appearance of the symptoms and the dynamics of changes in them);

(3) estimation of the general condition (signs of exicosis, and toxicosis, degree of prematurity, combined maldevelopmental anomalies, manifestations of birth injury and infection);

(4) examination of the abdomen (distension, asymmetry, Vaal's symptom, tenderness, peritoneal signs);

(5) intubation of the stomach (determination of the amount and quality of the gastric contents);

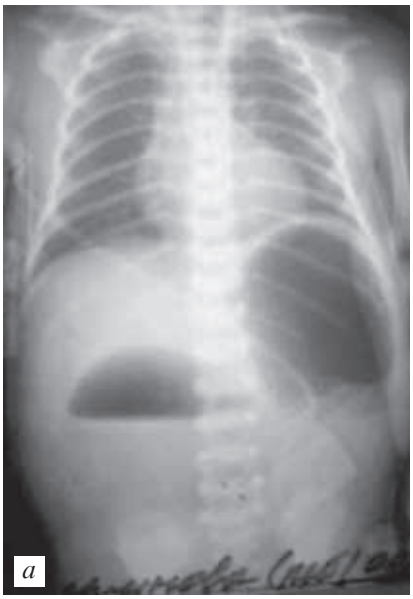
(6) rectal examination;

(7) X-ray examination.

During complete obstruction of the stomach the survey X-ray reveals the individual level, corresponding to the stomach and the lack of pneumatization of the lower abdomen. During complete obstruction at the level of the duodenum two levels of liquid and two gas bubbles are defined, corresponding the stomach and duodenum, their size and location are variable, the lower abdomen are deprived of pneumatization during complete obstruction (Fig. 3.3.6, 3.3.7).

Differential diagnosis. High intestinal obstruction must be differentiated from pylorostenosis, pylorospasm, and the pseudo-occlusion syndrome. In contrast to duodenal obstruction, pylorostenosis occurs in later periods and is marked by vomiting of curdled milk without admixtures of bile or green matter. Pylorostenosis has typical X-ray and endoscopic pictures. Pylorospasm is characterized by inconstancy of signs and good effect of spasmolytic therapy. The pseudo-occlusion syndrome occurs characteristically in premature babies. The differential diagnosis is based on the X-ray picture (retention of evacuation of the contrast medium) and the efficacy of non-operative measures (neostigmine methylsulphate, siphon enemas, irrigation of the stomach).

Low intestinal obstruction is differentiated from dynamic obstruction (Fig. 3.3.8). As distinct from congenital obstruction, intestinal paresis develops more gradually in babies at the end of the first



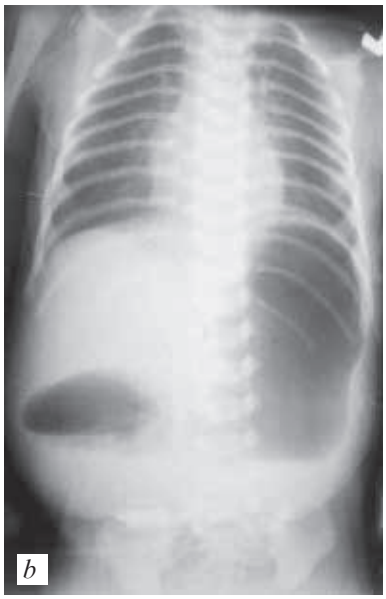


Fig. 3.3.6. Complete obstruction of duodenum *(a, b)*

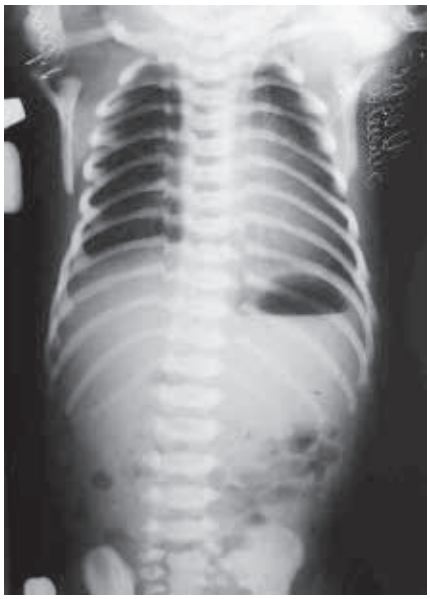


Fig. 3.3.7. Survey X-ray during partial duodenal obstruction

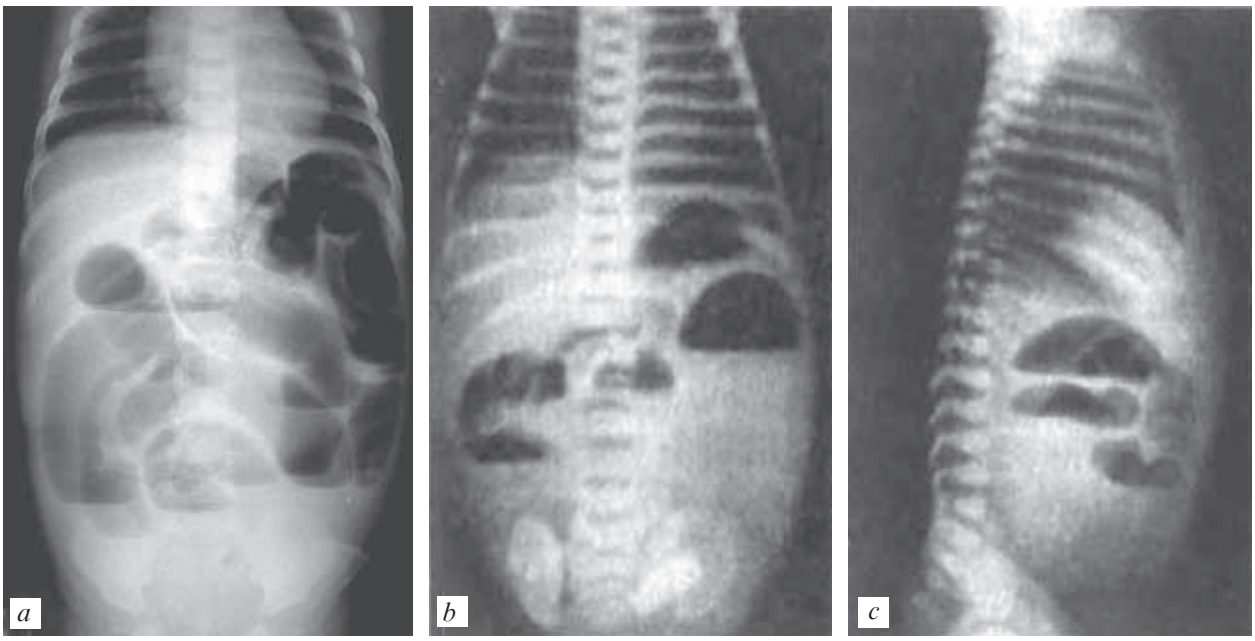


Fig. 3.3.8. Low intestinal obstruction *(a–c)*

month of life and older who suffer from some diseases (severe pneumonia, otitis, intestinal infection, sepsis). Paresis is marked by uniform distension of the abdomen, absence of peristalsis, and by signs of severe toxicosis. X-ray examination reveals abnormally large amounts of gases in the intestine.

Treatment. Surgery is indicated in congenital intestinal obstruction. Preoperative management is an important stage; its specific features are determined by the type of the obstruction (high or low), the duration of the disease, and the child's age.

The character of the operation depends on the anatomical variant of the malformation. Access is gained by median, transrectus or transverse laparotomy. The opened abdominal cavity is inspected, taking careful note of the presence and character of the exudate, the size and appearance of the intestinal loops, the size of the mesentery and whether it is attached normally. In high obstruction, the intestine appears collapsed and only the duodenum and the initial portion of the jejunum are drastically distended above the obstruction. An atretic duodenum is often larger than the stomach, the pylorus is stretched out and outlined as a hardly noticeable groove (double-cavity reservoir). Low obstruction is characterized by drastically distended afferent intestinal loops which diameter is 10–12 times as much as that of the collapsed intestine. With the location and anatomical variant of the malformation determined, the main stage of the operation is begun. Ladd's syndrome is the most common cause of high congenital obstruction. After envagination of the intestine into the wound, it can be seen that it is twisted one to three times around the root of the mesentery. With the volvulus corrected, the caecum is usually found high in the left half of the abdominal cavity while the wide planar strands radiating towards it from the right lateral part of the abdomen compress the duodenum and cause its obstruction. Ladd's operation consists in correcting the volvulus and dividing the peritoneal strands. The caecum is transposed into the left half of the abdomen, calculating on its taking the normal position in the right lower abdominal quadrant in 6 to 12 months. In other cases with incompleted torsion surgery consists in removing the obstruction by a simpler method (division of the strands, closure of the defect in the mesentery, correction of the volvulus).

The operation of choice in atresia of the duodenum and its compression by annular pancreas or an aberrant vessel is posterocolonic (interior duodenojejunostomy. Duodeno-duodenoanastomosis is established in high duodenal atresia (above the level of the major duodenal papilla); some cases with membranous atresia and stenosis are managed by duodenotomy and subsequent excision of the membrane and transverse closure of the intestine with sutures.

In atresia of the jejunum and ileum, the atresia intestine is resected together with the distended blind end and 5–7 cm of the collapsed distal segment, which is functionally deficient, is also removed. The continuity of the intestine is restored by side-to-side or end-to-end anastomosis established by single-row Halsted's sutures applied with atraumatic needles. The anastomosis should be at least 2.0–2.5 cm wide.

In atresia of the terminal segment of the ileum, the abnormal part of the intestine is resected and the end of the small intestine is anastomosed with the side of the colon.

Meconium ileus is managed by Mikulicz's operation consisting in extra-abdominal resection of the intestinal segment jilled with the abnormally viscid meconium and establishment of double ileostomy which is closed in 3 or 4 weeks. Typical resection of the small intestine is recently used in the management of meconium ileus. Pancreatin is administered through the enterostomy in the postoperative period to liquefy the meconium.

The results of treatment depend on timely diagnosis and the combination of developmental anomalies in the child. Complications in the form of intes-

tinal necrosis (in volvulus), perforation and peritonitis (in low intestinal obstruction), and irreversible disorders of homeostasis develop in late diagnosis.

CONGENITAL PYLOROSTENOSIS

Impaired patency of the pyloric part of the stomach is a genetically determined malformation and results from hypertrophy and degeneration of the pyloric sphincter. The factor underlying these changes is defective laying down of the nerve elements in the muscle and, as a consequence, its stable spasm.

Clinical picture. The first manifestations of the disease usually appear from the end of the second or the beginning of the third week of life. Its main sign is projectile vomiting between feedings. The vomited material is congestive in character: its amount is greater than the dose of a single feeding and it contains curdled milk with a sour smell.

The child starts loosing weight, signs of dehydration appear, and the baby urinates seldom while the stool is small in amount. In the acute form of the disease the signs develop rapidly and violently, so that the patient's condition grows worse in a week. Disorders of water-electrolyte and acid-base balance predominate in this case.

The findings of laboratory tests bear evidence of pachyhaemia (increased haemoglobin and haematocrit) and a picture of hypochloraemic alkalosis and hypokalaemia. The haemorrhagic syndrome may develop in severe cases as a consequence of intravascular coagulation of blood.

In the subacute form, the signs progress gradually: the baby regurgitates and vomits at first once or twice and then more frequently as a result of which hypotrophy develops. No gross water-electrolyte disorders occur in this form.

On examination of the patient attention is focused on the degree of hypotrophy and exicosis, epigastric distention, and increased peristalsis of the stomach by an "hourglass" type (Fig. 3.3.9).

Ultrasound of the stomach and pylorus (diagnostic confidence are lengthening of pylorus over 16 mm, diameter over 14 mm, thickness of its layer over 4 mm and narrowing of the channel less than 4 mm) (Fig. 3.3.10).

Fibrogastroscopy is used lately in making the diagnosis of pylorostenosis (Fig. 3.3.11). A distended and plicated antral part of the stomach and a drastically narrowed (to a pin-point size) lumen of the pyloric canal which fails to open after atropinization and inflation with air (in contrast to pylorospasm) are found. Moreover, fibroscopy makes it possible to perform oesophagoscopy and determine the severity of reflux oesophagitis which is a frequent adjunct to pylorostenosis.

X-ray and endoscopic methods of examination are applied to make the exact diagnosis. The size of the stomach, the presence of fluid in a fasting stomach, the time of the first evacuation of the stomach, the condition of the pyloric canal, and the time of full evacuation of the stomach are studied during X-ray

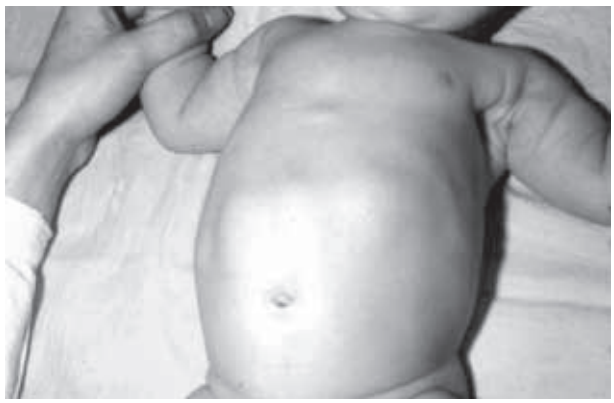


Fig. 3.3.9. The "hourglass" sign in a child with pylorostenosis

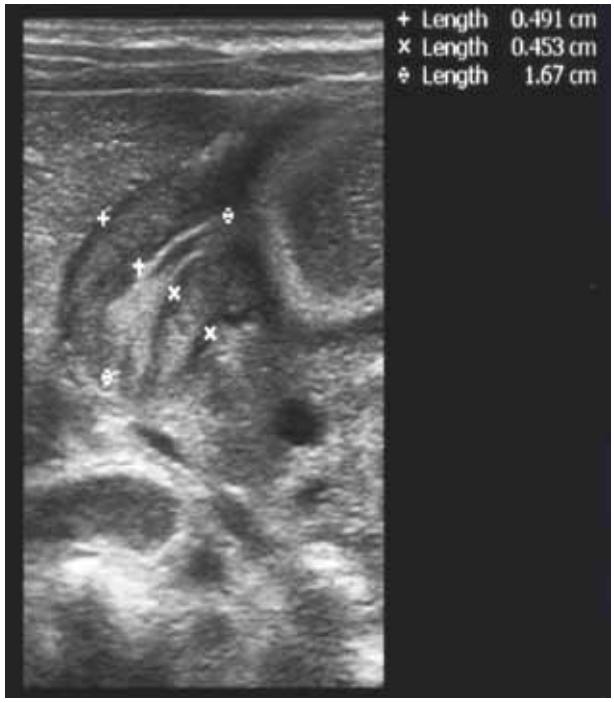


Fig. 3.3.10. Ultrasound diagnosis of pyloric stenosis

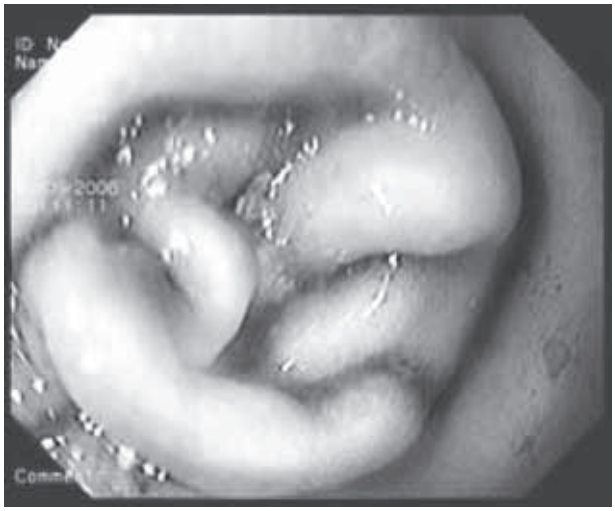


Fig. 3.3.11. Endoscopic picture of pyloric stenosis

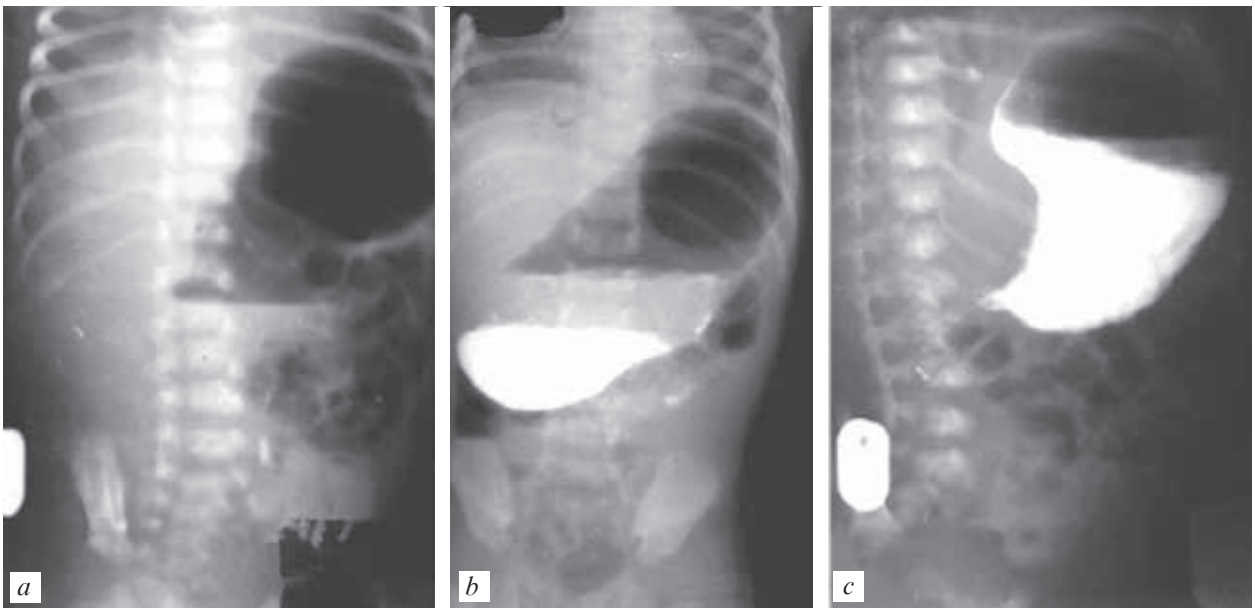


Fig. 3.3.12. Radiographic signs of pyloric stenosis *(a–c)*

examination (Fig. 3.3.12). A distended stomach filled with gas and with a fluid level is found during scout examination. Segmental peristalsis of the stomach and absence of primary evacuation into the duodenum are demonstrated 15–20 minutes after a contrast medium (5 per cent aqueous barium suspension in 50–70 ml of breast milk) is given; a constricted pyloric canal is seen on a lateral view. Examination of the patient in 3.6, and 24 hours reveals retention of the contrast medium in the stomach.

Differential diagnosis is made with pylorospasm, the adrenogenital syndrome (Debre–Fibiger syndrome), and partial duodenal obstruction above the major duodenal papilla. It is based on the difference in the X-ray and endoscopic pictures, the time of appearance of the first symptoms of the disease, and the results of biochemical tests. In pylorospasm, evacuation of the contrast medium from the stomach is disturbed for more than 2–3 hours, and antispastic therapy produces a good effect.

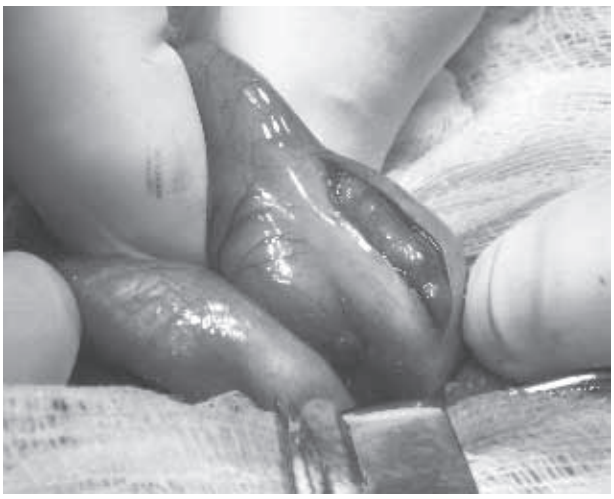


Fig. 3.3.13. Pyloromyotomy by Fred–Weber–Ramshtedt

The adrenogenital syndrome is characterized by an admixture of blood in the vomit, hyperkalaemia and hyponatraemia; evacuation of the contrast medium from the stomach does not suffer. The appearance of symptoms from the first days of life and typical X-ray findings are specific of high intestinal obstruction. Fibrogastroduodenoscopy helps in the diagnosis.

Treatment. Pylorostenosis requires surgical treatment. The operation is preceded by preoperative management for correcting dehydration, alkalosis, and hypokalaemia. The operation is conducted under general anaesthesia, though local anaesthesia is also possible. Access to the pylorus is gained through a superomedian or transrectus incision or through a transverse incision in the superior abdominal quadrant.

The Fredet–Ramstedt operation is performed (Plate II). The seromuscular coat of the pylorus is cut in the vascular zone and the cut edges of the muscle are drawn apart with an instrument until the mucous coat of the pylorus prolapses into the wound for the whole distance of the incision. The thickness of the muscular coat on section ranges from 0.5 to 1. 0 cm, whereas normally it is 0.1–0.16 cm thick (Fig. 3.3.13).

The most responsible moment is division of the pylorus at its junction with the duodenum because the muscle is thinned out here and the overhanging mucosa may be injured. To check the intactness of the mucous membrane, the stomach is compressed and its contents moved towards the duodenum. This simple manipulation allows perforation to be recognized in time. If perforation is found, the defect is closed with one or two sutures. Should this prove difficult, the site of pyloromyotomy is sutured and a cut is made on the contralateral side.

As the result of the operation, the anatomical obstruction is removed and passage of the pylorus restored. Three hours after operation the child is giv-

en a drink of 10 per cent glucose solution, in another 4 hours 10 ml of drawn off breast milk is given at intervals of 2 hours for 10 feedings daily (no night feeding). In the following days the amount of milk is increased by 100 ml daily (by 10 ml for each feeding). From the 5th day, when the child already receives 50 ml of milk every 2 hours, its amount is increased to 70 ml and the interval between the feedings is prolonged to 3–3.5 hours, after which the common type of feeding is given.

The lack in proteins and fluids within the first postoperative days is made up parenterally by intravenous infusion of plasma, 10 per cent glucose solution, and Ringer–Locke solution. Microenema is prescribed every 4 hours (15 ml of 10 per cent glucose and 15 ml of Ringer–Locke solutions). The prognosis is favourable.

HIRSCHSPRUNG'S DISEASE

It was believed for a long time that Hirschsprung's disease was extremely rare (1 : 20,000–1 : 30,000). In recent years it is found more frequently, which is evidently due to earlier and exact diagnosis. Its incidence among boys is 4 to 5 times as much than among girls. The hereditary (genetic) nature of this severe malformation has been proved at the current stage.

It was believed for many decades that Hirschsprung's disease was caused by congenital malformation of the muscular components of the large intestine, the presence of mucosal folds in its distal part, kinking of the elongated sigmoid colon and its congenital atonia, altered sympathetic tone, etc.

Changes of the histostructure of the intramural nerve apparatus in a definite segment of the large intestine are the principal element in the pathogenesis of Hirschsprung's disease (O. Swenson; Yu. Isakov). Morphological studies showed considerable changes of ganglia not only in Auerbach's (intermuscular) but in Meissner's (submucous) plexus in the constricted zone. The results of these studies allow this form of megacolon to be characterized as congenital agangliosis of an area of the large intestine, a condition in which peristalsis cannot be produced in intestinal areas devoid of Auerbach's plexus or in those with its deficiency.

Severe changes and even death of the muscular layers of the aganglionic zone aggravate the peristaltic disorders and make them permanent. In accordance with these disorders, the zone is aganglionic morphologically and aperistaltic clinically.

Absence of peristalsis in this intestinal segment results in stasis of the faecal material above the site of the affection, which in turn causes dilatation and hypertrophy of the proximal parts of the large intestine. Hypertrophy develops as a consequence of intensive peristalsis of the proximal segments to propel the contents through the aperistaltic aganglionic area. The diameter of the dilated intestine may be very large. The aganglionic segment, in contrast, is narrowed.

Study of the pathological anatomy (Fig. 3.3.14) of the large intestine in congenital agangliosis showed prevalent involvement of the rectosigmoid segment (70 per cent) and the perineal and ampullar parts of the rectum (20 per cent). The occurrence of aganglionic zones in two places and in the proximal parts of the large intestine is much rarer. Total affection of the large intestine with agangliosis is known.

Clinical picture and diagnosis. Failure to pass stool (chronic constipation) is the principal sign of Hirschsprung's disease. The disturbed activity of the gastro-intestinal tract is manifested by constipation from birth or the first months of life as a result of which chronic faecal toxicosis gradually develops. Meteorism occurring, like constipation, within the first days and weeks of life is a constant sign of the disease.

With the gradual chronic retention of faeces and gases, the sigmoid colon and later the proximal segments of the colon become dilated as a consequence of which the abdomen becomes larger. The high position of the diaphragm leads to the formation of a barrel chest.

In prolonged and persistent retention of faeces and gases, the abdomen grows still larger and acquires the form of a "frog" abdomen (Fig. 3.3.15*)*. The abdominal wall is thin. The distended intestinal loops are outlined and their intensified peristalsis can be seen now and again. The characteristic "clay" sign is found by abdominal palpation in most cases (clearly defined depressions felt through the skin of the abdominal wall form from pressure with the fingers on the intestine overfilled with faeces). It must be pointed out that the child's general condition also suffers in such a case, he is retarded physically and mentally, and anaemia and cachexia develop. These signs are more pronounced in older children. Inflammation of the intestinal mucosa, manifested by diarrhoea, occurs sometimes as the result of dysbacteriosis.

The initial manifestations of constipation and their subsequent character and persistence are determined to a great measure by the length of the aganglionic segment, the character of feeding, and the compensatory capacities of the intestine.

Severe neglected forms of Hirschsprung's disease are rarely observed nowadays. Elaboration of methods for its early diagnosis even in the neonatal period was conducive to this. Three forms of the clinical course of Hirschsprung's disease are distinguished: severe (acute), moderately severe (subacute), and mild (chronic).

The *severe,* or *acute, form* of congenital agangliosis is manifested from the first days of life as low intestinal obstruction. Meconium is retained or passed in very small amounts, gases are not passed. Abdominal distension increases progressively, intestinal peristalsis is visible, and copious vomiting occurs. Percussion produces tympany due to the sharp meteorism. Enemas given regularly prove ineffective and the passage of gas is poor. Children are often admitted to the hospital for suspected high intestinal obstruction.

The discrepancy between the clinical signs characteristic of high intestinal obstruction (vomiting of bile) and the X-ray evidence of impaired evacuation

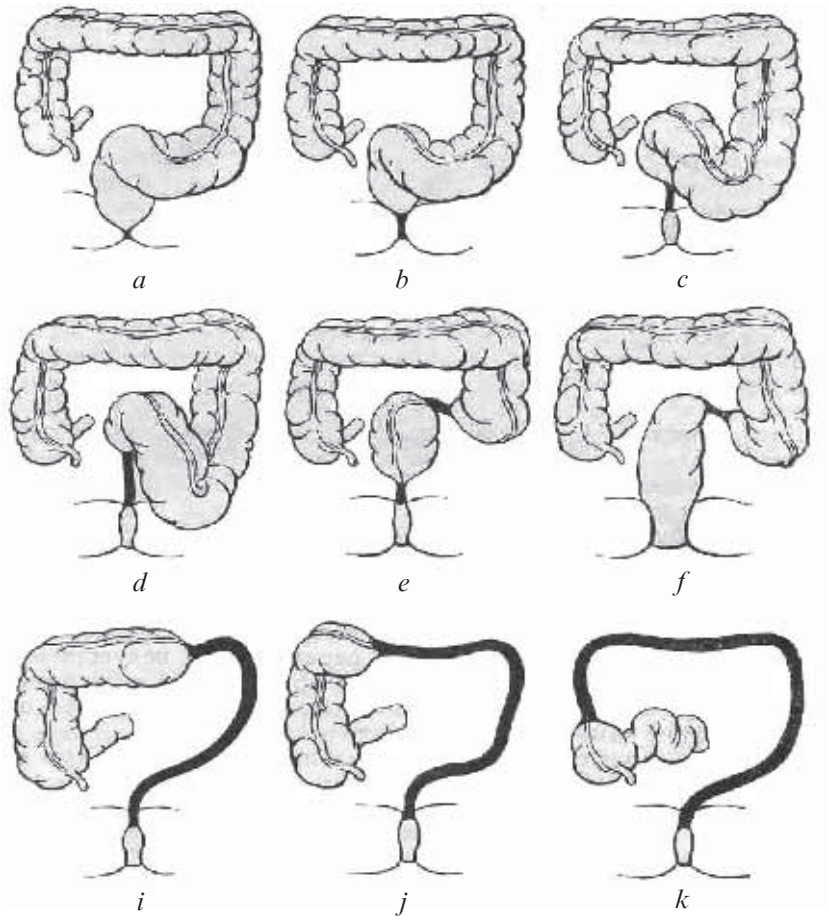


Fig. 3.3.14. Anatomical forms of Hirschsprung's disease (scheme) *(a–k)*

of the large intestine (distension of the loops thorough clinical and X-ray examination (Fig. 3.3.16).

Hirschsprung's disease in the newborn is managed by non-operative methods as a rule, and only in rare cases when they prove ineffective and the patient's condition grows worse rapidly and toxicosis increases a faecal fistula of the ascending colon is established.

The *moderately severe*, or *subacute, form* is usually an intermediate stage between the severe and mild forms and develops when the aganglionic zone is smaller in length and forms no kinks. The general condition deteriorates slowly but continuously. Constipation becomes more and more stubborn. Non-operative measures produce a temporary effect. Siphon enemas are resorted to more often to evacuate I lie intestine. The child's condition changes depending on the degree of stool retention. Most children are physically retarded, lose weight, nod signs of toxicosis and anaemia develop.

The *mild,* or *chronic, form.* Patients with this form hardly differ from healthy babies within the first days and sometimes weeks of life. Tim stool may occasionally be retained and mild abdominal distension and vomiting occur in attendance, but the general picture causes no alarm, the more so since stool is passed after a small enema or introduction of a colonic tube. But the child's condition grows worse when artificial feeding is given, and stool is passed only after a finalising enema. Faecal stones form in prolonged Coprostasia due in inadequate care. They are so large in neglected cases that are mistaken for an abdominal tumour. The general condition gradually diteriorates, which is linked with chronic faecal toxicosis. Anaemia Mini hypotrophy are, however, mild. The abdomen is usually distended, enlarged transversely, and spread out. Peristalsis of the dilated loup of the large intestine is seen in occasional cases. Digital rectal examination shows increased tonus of the sphincter muscle.

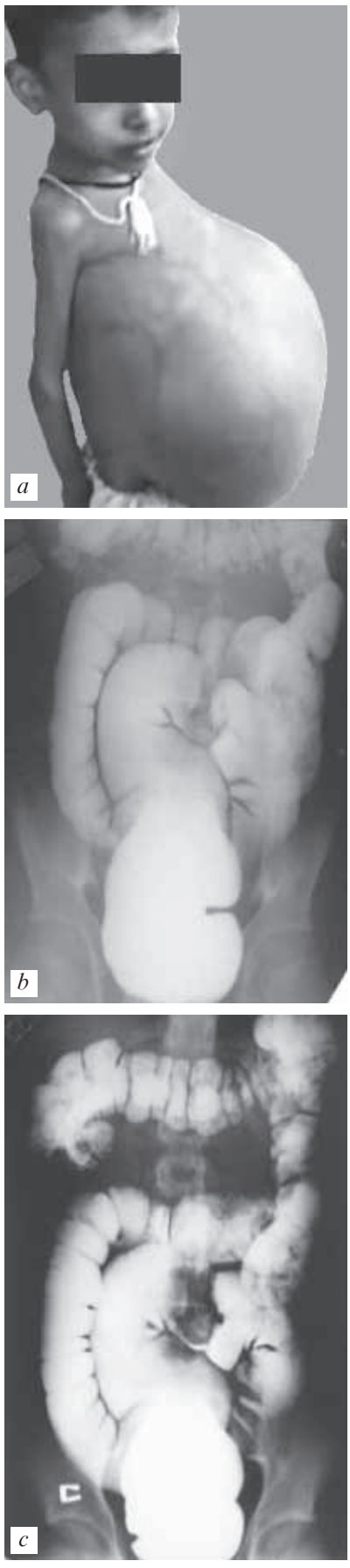


Fig. 3.3.15. General appearance of patient — the "frog" abdomen, X-ray picture *(a–c)*

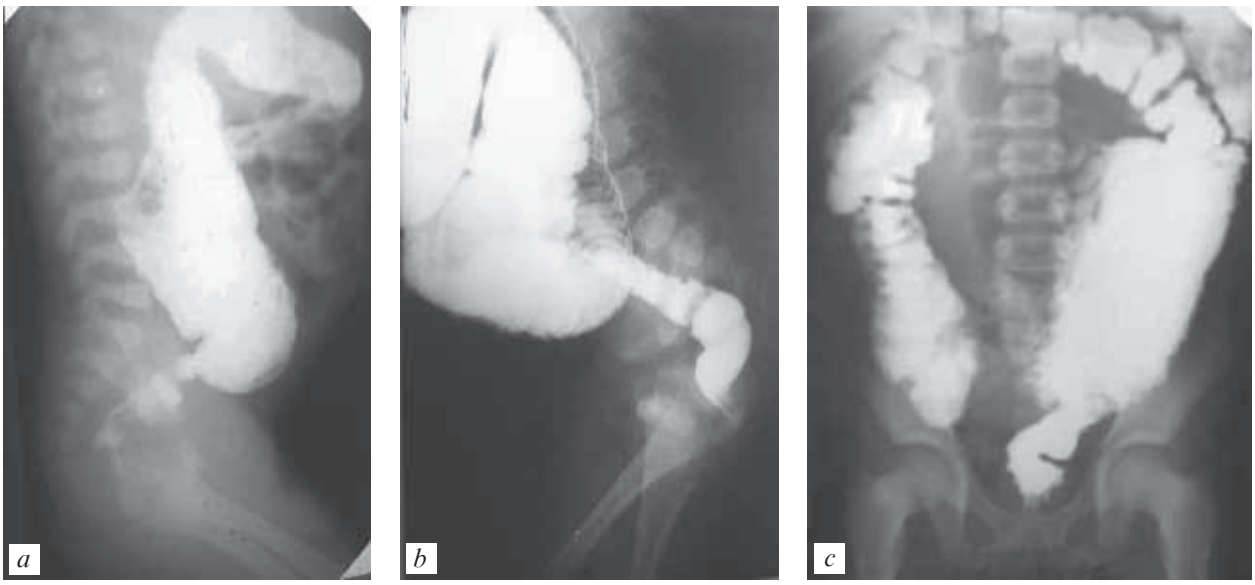


Fig. 3.3.16. X-ray picture of Hirschsprung's disease (irrigogramms) *(a–c)*

A chronic course of Hirschsprung's disease is due to a short aganglionic zone in which the functional disorders are mild. Forms with an aganglionic zone of a greater length (from the descending colon to the rectal ampulla) are, however, also encountered and the disease gradually develops. The final diagnosis is made only from the results of X-ray examination. Much importance must be payed to functional methods of examination of the rectum (measurement of intraintestinal pressure, study of the condition of the sphincter ani external and internal muscles, etc.).

X-ray examination is undertaken after the intestine is completely evacuated of faecal material. Scout radiography of the abdominal cavity usually reveals dilated and distended loops of the large intestine and a high position of the dome of the diaphragm. Radiocontrast study by means of an enema (irrigography) is the only method demonstrating the most characteristic signs. A barium suspension in a 1 per cent common salt solution is used as the contrast medium; 80 ml is a sufficient amount for the newborn and infants, older, children are given up to 500 ml by enema. A narrowed zone in the large intestine with a suprastenotic dilatation proximal to it (Fig. 3.3.17) is an authentic X-ray sign of Hirschsprung's disease.

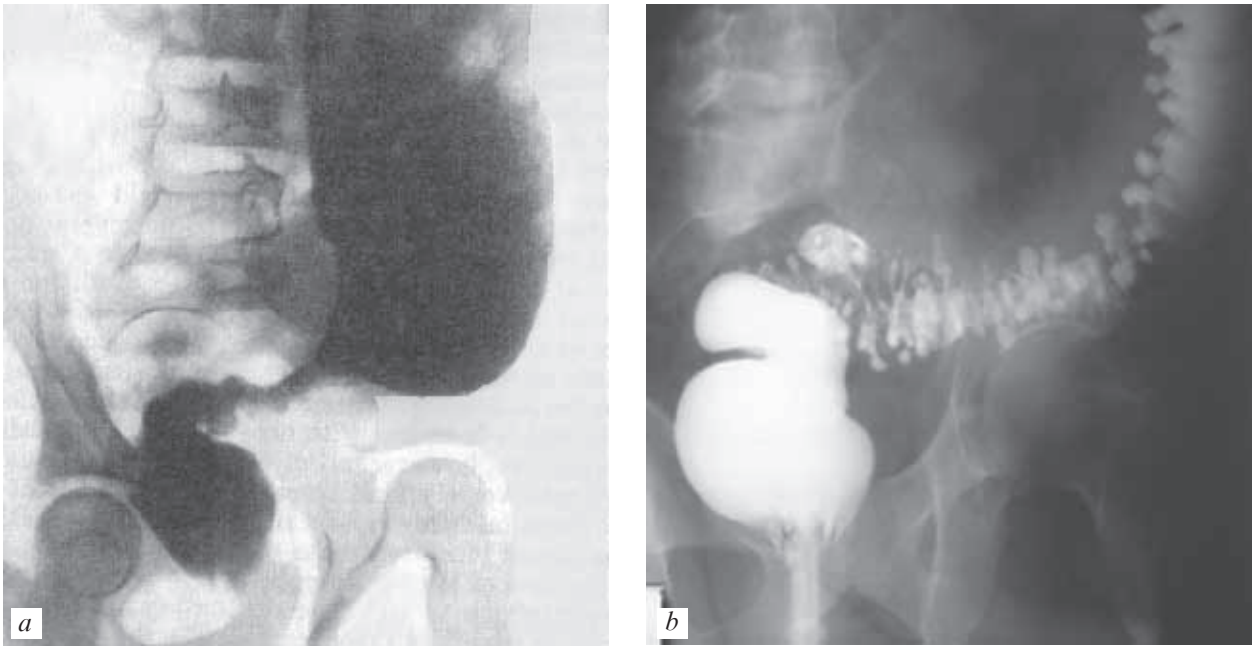


Fig. 3.3.17. Hirschsprung's disease. X-ray of the large intestine (bariu in suspension enema). A constriction continuous with a dilated part of the large intestine is seen in the rectosigmoid zone *(a, b)*

The most characteristic changes are demonstrated on lateral view X-ray showing not only the localization of the narrowed aganglionic zone but the degree and length of the narrowing. Evacuation of the intestine is delayed after X-ray rectal examination. This is one of the main diagnostic signs of Hirschsprung's disease in the newborn babies and infants, in whom the difference in the diameter of the stenosed and dilated parts of the intestine is still negligible.

Faecal stones can sometimes be demonstrated with X-ray. In contrast to Hirschsprung's disease, in other types of megacolon a constricted aganglionic area is not found while the large intestine in distended and in some cases also elongated.

Since Hirschsprung's disease is attended by other malformations, particularly often by diseases of the urinary system, urological examination along with irrigography must be conducted in all cases.

The *differential diagnosis* of Hirschsprung's disease is made with developmental defects and some diseases attended hy constipation.

Congenital elongation, dilatation or atonia of the large intestine differs from Hirschsprung's disease in having a later onset (from 2–3 years) and less pronounced signs. With a change in the diet, stool is passed spontaneously at regular intervals, which never happens in children over 12 months of age suffering from Hirschsprung's disease. The abdomen is usually normal in shape and size. This general condition is never disturbed like in Hirschsprung's disease. A wide rectal ampulla filled with faeces is found on digital examination of the rectum.

Secondary megacolon consequent upon congenital or acquired cicatricial stenosis of the rectum is revealed by examination of the anal region and digital rectal examination. Sigmoidoscopy is performed in some cases.

Common constipation caused by anal fissures, haemorrhoids, and diseases diagnosed from careful analysis of the medical history and the findings of examination of the anal region. Such constipation is managed by removing the cause.

The practical importance of differential diagnosis consists in early recognition and treatment of Hirschsprung's disease since it is the most severe sickness. Biopsy of the rectum is resorted to in some cases; it reveals an absence of nerve ganglia in Hirschsprung's disease.

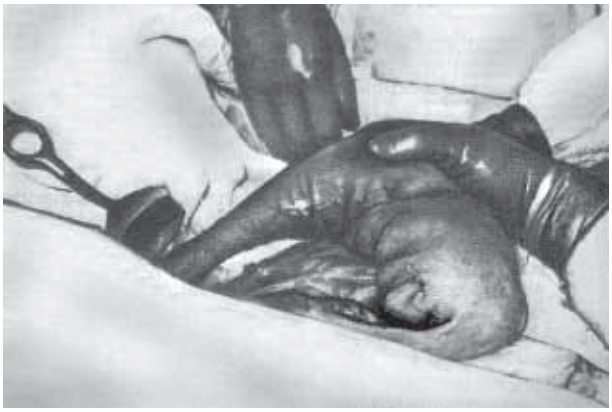


Fig. 3.3.18. Hirschsprung's disease, photo taken during operation, showing a narrowed portion of the rectosigmoid region and a suprastenotic dilatation of the sigmoid colon

One should have in mind that he intramural apparatus is still immature by the time of birth and matures finally in the next months of the child's life. In view of this rectal biopsy is advisable only after the age of 12 months.

Treatment. There is full agreement today that Hirschsprung's disease must be treated surgically. Abdominoperineal resection of narrowed segment and part of the distended large intestine is the radical and pathogenetically substantiated operation. The best time for its performance is between the age of 2–11 years.

Much significance is attached to *non-surgical management* in period preceding the radical operation. It is usually conducted home and its object is regular evacuation of the intestine (Fig. 3.3.18).

An appropriate diet is prescribed according to the child's age with food activating intestinal peristalsis (oatmeal and buckwheat porridge, prunes, beetroot, carrots, apples, honey, etc.). Sour clotted milk, sour fermented milk, fresh fruit have a beneficial effect. Massage of the abdomen and a complex of exercises strengthening the abdominal muscles are necessary. The parents must be instructed in these simple manipulations carry them out daily for 10 to 15 minutes before each feeding.

Various types of enemas (a common cleansing enema, hypertonic, siphon, etc.) are the principal measures in non-operative treatment of Hirschsprung's disease.

A 1 per cent common salt solution of room temperature is used for a siphon enema. Warmed water must not be used because in improper evacuation of the intestine the broken-up faecal material rapidly absorbed and toxicosis and oedema of the brain develop which may cause acute deterioration of the condition and even death. The volume of the siphon enema is determined by the child's age (from 0.5 to 2–3 litres for infants and 3 to 10 litres for older children). After the enema, a colonic tube is advanced through the narrowed zone into the dilated part of the large intestine and left for 1–2 hours. Laxatives are not advisable, oral vegetable oils (peach-kernel, sunflower, olive oil) are preferable (1 teaspoonful for children of nursery age, 1 dessertspoonful for those of preschool age, and 1 tablespoonful 3 times daily for schoolchildren).

Intensive non-operative measures, even if applied in an in-patient clinic, fail to produce the needed effect in some cases and the child's condition grows worse progressively. The formation of a natural anus is indicated in such patients.

Stretching the anus with the fingers under anaesthesia improves the passage of the intestinal contents in considerably increased tonus of the sphincter *ani internus* muscle. The manipulation is repeated at intervals of 10–14 days.

Surgical treatment. Swenson–Hiatt's, Duhamel's, and Soave's operations are the radical surgical methods used most extensively in the management of Hirschsprung's disease.

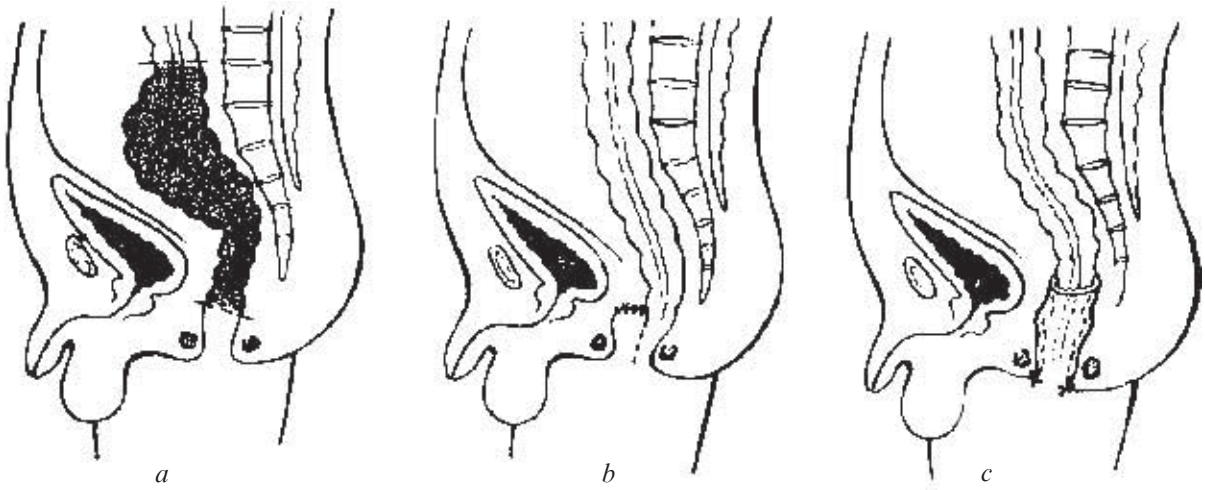


Fig. 3.3.19. Main operations in Hirschsprung's disease (represented schematically). *a* — Swenson's method (extraperitoneal resection with establishment of oblique anastomosis after Isakov); *b* — Duhamel's method; c — Soave's method

The principle of the Swenson–Hiatt's operation (Fig. 3.3.19 *a*) consists in the following. Before resection, the segment of the large intestine is mobilized distally, to a distance of 3–5 cm from the anus anteriorly and a little more (to a distance of 1.5–2 cm from the anal skin) on the posterolateral surfaces. An oblique anastomosis is then established extraperitoneally (Yu. F. Isakov) by two-stage evagination of the mobilized part of the intestine through the anal opening. Extraperitoneal resection of the aganglionic segment and the dilated part of the large intestine is performed.

Serious complications such as urinary and partial faecal incontinence may occur during the Swenson–Hiatt operation, which is linked with manipulations in the region of the nerve plexuses of the pelvic floor during exposure of the rectum.

With Duhamel's method (see Fig. 3.3.19, *b*) the rectum is divided above the dilatation, its lower end is closed with sutures, the upper (proximal) end is brought out through a canal formed between the sacrum and rectum to the sphincter oni externus muscle. At a distance of 0.5–1 cm from the junction of the mucosa and the skin on the posterior semicircumference of the anus, the mucous membrane is separated upward for 1.5–2 cm. A cut is then made through all the coats of the rectum and the sigmoid colon is brought downward onto the perineum through the "window" thus formed. G. A. Bairov introduced significant modifications into this method. After applying retractor sutures to the anterior wall of the transposed intestine and the posterior wall of the rectum, the aganglionic zone and part of the dilated intestine are resected. A special crushing clamp promoting spontaneous anastomosis is applied to the formed "spur".

The main stage of Soave's operation in the modification suggested by A. I. Lenyushkin consists in separation of the sero-muscular coat of the aganglionic zone from the mucous coat for almost the whole length (to a distance of 2–3 cm from the sphincter ani intemus muscle). The large intestine is evaginated through the anus onto the perineum by drawing it through the muscular cylinder of the rectum. The evaginated intestine is resected leaving a small freely hanging segment 5–7 cm long (Fig. 3.3.19 *c*). The redundant part of the intestine is cut off in a second stage 15–20 days after the sutureless anastomosis has formed.

The main advantage of this method is that the intestine is brought downward through the natural anorectal canal as a result of which the anatomical structures around the rectum are not damaged.

ANORECTAL MALFORMATIONS

Congenital and acquired diseases of the anorectal region are quite common among children (1 per 1500–5000 newborn babies). They are found more often in boys than in girls.

The aetiology of anal and rectal anomalies has not yet been studied sufficiently, however, the most widely accepted theory at present is that they are polyaetiological in nature, i. e. their origin is attributed to various harmful agents acting in the critical period of embryo development and in the period of organogenesis.

According to F. Stephens (1963), anorectal malformations occur at the stage of cloaca division and the formation of the perineum, and malformation options depend on the sex. This is connected with the interposition of genitals between the rectum and urinary tract in females. Various defects occur during violation of various stages of embryogenesis.

In males, these stages are conditionally divided into:

• Stage of cloaca division. At this stage the following defects can form: urinary rectal fistula (often in Lieutaud's triangle or prostatic part of urethra), and if the fistula obliterans, malformation appears as atresia of the rectum and anus.

• The stage of formation of the perineum: disclose of anal membranes with or without rectal atresia,

stenosis of the anus and rectum, ectopic of the anus, covered anus.

In female embryos occurrence of anorectal malformations has significant differences.

• Stage of cloaca division: during occurrence of rectovesical fistula doubling or bicornuate uterus forms, doubling of the vagina, the formation of the cloaca, or rectovaginal or rectovestibular fistula, which may be associated with atresia of the rectum and anus, rectal atresia without fistula.

• The stage of formation of the perineum: perineal fistula, and the rest are similar to those of the boys.

In embryos of both sexes during normal development of the anus insufficiency of posterior perineum is marked: clefts of perineum, adrectal congenital fistula, and rectovestibular fistula during normally located anus, a diverticulum of the distal rectum.

Closing apparatus at malformations of the rectum often has expressed morphological and mostly topographic anatomy deviations. During aresia often occurs inadequate (underdevelopment) of external sphincter in a lack of anterior muscle bundles, sometimes there is no sphincter at all. Muscle lifting the anus is less than others subjected to change, but in the absence of the rectum there is a reduction of the muscle, particularly its central portions (puborectal sling). Sling instead of the rectum covers the urethra like a ring in males or vagina in girls, i. e. strongly displaced anteriorly.

In deficient development of the cloacal membrane. Atresia of the rectum occurs when the blind end of the gut fails to reach the retracted ectodermal anal membrane and the cloacal membrane separating them does not resorb because the entodermal germ has not developed sufficiently. In deficient development of the ectodermal germ and ectodermal gut the distal part of the gut is developed and the ectoderm is not retracted as a result of which atresia of the anus and rectus occurs. Congenital stenosis of the anus is a consequence of an incomplete rupture of the cloacal membrane (Fig. 3.3.20).

Among the great number of existing classifications, the most rational is that compiled according to the embryogenesis of the malformation and the specific features of the surgical management in each concrete case.

Nowadays Melbourne classification of anorectal anomalies is common.

I. High (supralavatory)

1) Anorectal agenesis

Boys: a) without fistula;
b) with fistula: rectovesical, rectourethral;

Girls: a) without fistula;
b) with fistula: rectovesical, rectocloacal, rectovaginal;

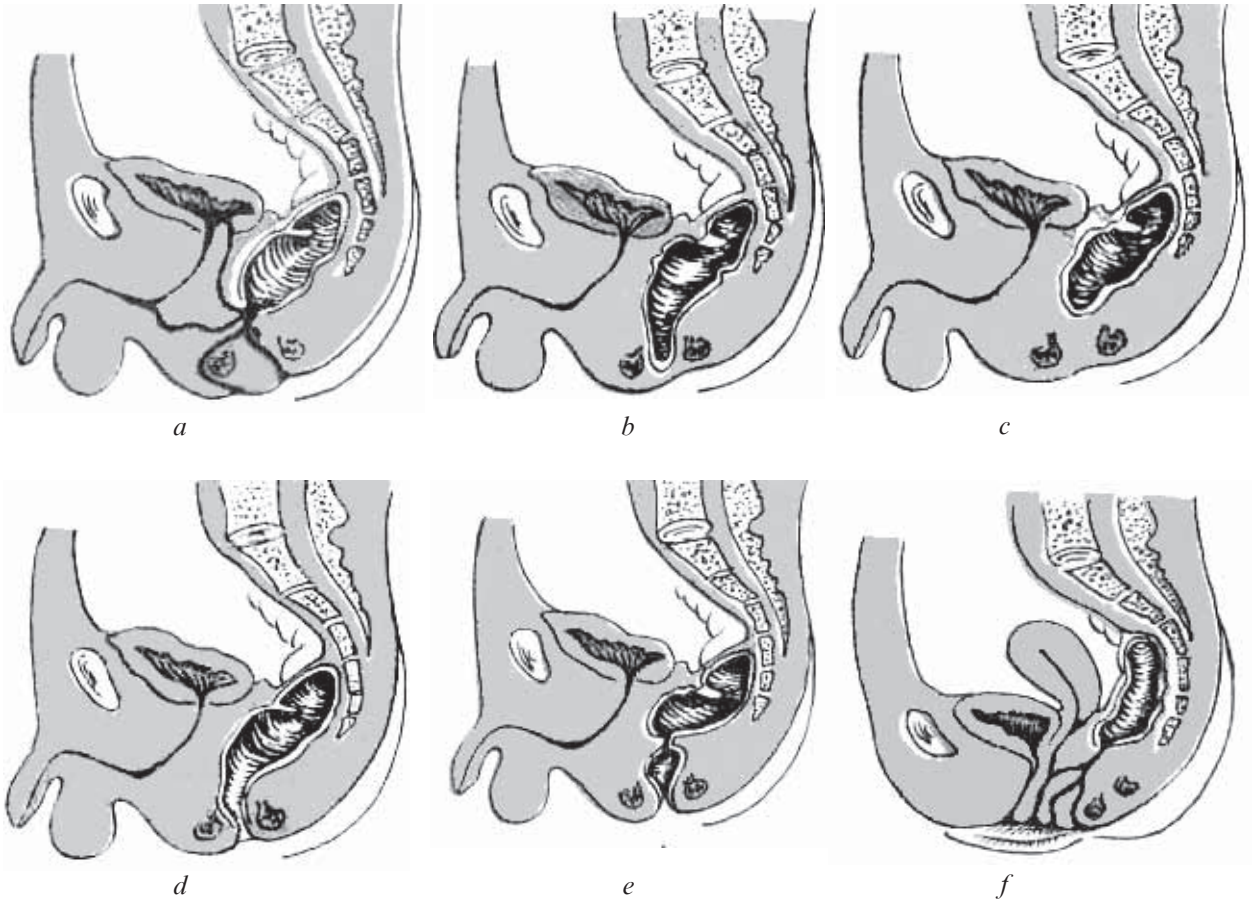


Fig. 3.3.20. The most commonly encountered forms of rectal malformations. Non-fistulous form: *a* — anal stenosis; *b* — anal atresia; *c* — atresia of the anus and rectum; *d* — rectal atresia; fistulous form (*e* — in boys; *f* — in girls)

2) Rectal agenesis (boys and girls);

II. Average (intra medial)

1) Anal agenesis;

Boys: a) without fistula;
b) with fistula: rectobulbar.

Girls: a) without fistula;
b) with rectovestibular fistula;

2) Anorectal stenosis (boys and girls);

III. Low (translavatory)

Boys and girls: covered anus — a simple, anal stenosis.

Boys: Front perineal anus, rectoperineal fistula.

Girls: Front perineal anus, rectovestibular fistula, vulvar anus, anovulvar fistula.

Atresia

Atresias are the most numerous group of anorectal malformations.

Anal ectopia. About ectopia should talk in those cases when the normal anus (well opened and shrinks, functions normally) is located in an unusual place — close to the vulva. In this case, all the anatomical structures of anal canal, including the external sphincter, are saved.

Clinical picture and diagnosis. Functional disturbances of the act of defecation are absent. One can discern the perineal and vestibular ectopia. At perineal ectopia anus is opened close to the root of the scrotum in boys or rear toward commissural of crotch in girls, and skin bridge remains. Sometimes in girls this bridge is missing and the mucous of vestibule goes into the anal canal mucosa (vestibular ectopia). A true ectopia must be differentiated from sinus forms of atresia. During atresia inevitably with time persistent constipation appears, during ectopia it is not observed. During atresia during the research of anal reflex, sphincter is reduced outside the anomalous openings.

So the absence or minimal expression of functional disorders and inside sphincter location of anus opening are the main distinctive features of ectopia of the anal opening from the sinus forms of atresia.

Treatment. Some authors do not recommend intervention in childhood and find it necessary to postpone it to adult age, so that one can decide for own the question about the operation. This view is reinforced by the fact that there are no functional impairment, and a good outcome of the operation cannot be guaranteed.

A. I. Lenyushkin considers that vestibular form must be surgically corrected. The most acceptable in these cases is the operation of Stone. The principle is to mark ectopic anus with the muscular system and transfer it to a new position on the perineum.

Congenital fistulae. In cases with a normally formed anus these fistulae are consequent upon incomplete closure of the vertical cloacal septum (formation of communications with the urogenital system) or disturbed formation of the perineum in later stages (relation with the vagina or its vestibule).

Rectovestibular and vaginal fistula

For rectovestibular and vaginal fistula spontaneous discharge of feces and gases through the genitalia gap in the background of regular, natural stool is typical. In newborns and infants, when there is a semi-liquid stool, constant release of stool through the fistula leads to a rather abrupt irritation of external genitals mucosa and perineum skin, vulvovaginitis. In older children, formed stool stand in smaller numbers, but there has been a steady involuntary flatus.

External opening of the full primary fistula is usually localized in the center at the bottom of the scaphoid fossa or on the back wall of the vagina immediately above the hymen. If the fistula is opened later after the inflammatory process, the external opening may be localized at the base of the *labia majora*. Sometimes there are two or more openings with skin bridges between them: in the genital lip and threshold of the vagina. Sinus diameter of the holes is 0.5–1.5 cm fistulous course can be cylindrical and conical. Depth of occurrence of inner opening of fistula is usually 1.5–2 cm from mucocutaneous transition of anal canal. To define it, one injects into the rectum a finger, and into the external opening of fistula the probe that is well felt by finger. The same probe easily goes through the anus. At external examination of the anus in most girls with congenital fistula is detected a peculiar skin appendage, which is a hypertrophied anal papilla. This symptom may facilitate the diagnosis of diverticular fistula forms when there is no external opening.

Treatment. A child with rectovestibular or vaginal fistula must be placed at the dispensary registration as soon as a diagnosis is set. It was revealed that fistula, even minor is not prone to self-closing. With the method of choice in the treatment of vestibular and rectovaginal fistula is congenital surgery. The operation can be postponed until the age of 3–4 years if the child is well cared.

It is necessary to examine the urine regularly and conduct a thorough perineum toilet several times a day. In some cases, with regular sharpening of pyelonephritis and vulvovaginitis operation is conducted before the deadline.

The operation consists in separating fistula from the vagina to the bowel wall. Through the inner hole of fistula, in fistula mosquito clamp is injected using it to turn inside all mobilized fistulous course in the intestine. Through the wound of perineum fistula base dual suture, intestine tissue is sutured in layers. In conclusion, from the side of intestine inverted fistula is stitched, tied and cut as a hernial sac.

Stump is treated with 3% iodine tincture. The sutures on the vaginal mucosa are not imposed; the wound is left to heal by secondary intention.

Operation during rectovaginal fistula differs from the previous one. The approach to fistula is carried from perineal access, then cross it in the vaginal wall, 2–3 catgut sutures are placed on the vaginal wall. The rest of the fistula is turned inside into the intestine lumen and process as was previously described. Perineum tissue of perineum is sutured tightly in layers.

Rectoperineal (pair rectal) fistula. There is reason to believe that this type of fistula is a "male type" of rectovestibular fistula. Clinically the presence in the newborn of pointlike orifice in couple rectal tissue is typical, from which in thick fat layer goes strand toward the anal canal. Typically, external opening of fistula is opened in front of the anus along the median suture, but in some cases fistula may be located in the side. Congenital adrectal fistula for some time may exist asymptomatic but sooner or later it becomes inflamed, there is a serous-purulent discharge, and these symptoms recur.

The inflammatory process is not transferred to adrectal fiber, as wall of fistula is a fairly reliable barrier. Peripheral hole of fistula sometimes self closes. Fistula is almost always within sphincter; its length is about 0.5–1.5 cm. The peculiarity of adrectal congenital fistula is their forward stroke.

Treatment is operative. Surgical intervention is indicated after the first year. Fistulous course dissect by a single block with the subsequent imposition of a blind stitch on the wound or its open charge.

Rectourethral and rectovesical fistulas

The clinical picture of anastomoses of the rectum with urinary tract is due to localization of fistula and the width. During a wide fistula in the bladder feces are constantly mixed with the urine, and it springs turbid during the whole urination. At the same time the gases leave through the urethra. During urethral fistula through the external urethral opening from time to time, regardless of the act of urination, gases and feces leave. Urine is turbid at first, then more transparent. Urinary tract infection causes relapsing urethritis, balanitis, and pyelonephritis. These symptoms are not always clearly marked, the most typical symptoms appear episodically and often parents do not pay attention to them.

Great help in establishing the correct diagnosis is rendered by radiography (urethrocystography) and sigmoidoscopy with the introduction into the urethra of coloring liquid.

Treatment. Rectourethral and vesical anastomoses needs surgery for diagnosis.

The management of the fistula depends on its location. Vesical fistula is eliminated from laparotomic access. Urethral fistula is eliminated by perineal fistula. Necessarily urethra is cathetered. Fistula is allocated from all sides and cross. Defects in the intestinal wall and urethra are sutured. For the prevention of recurrence of fistula, the seams on the intestine and the urethra are removed in different sides and fixed to the surrounding tissues.

Congenital stenosis of the anus and rectum results from incomplete destruction of the anal septum at the early stages of embryonal development.

Stenosis of the anus, of the rectum, and of both the anus and rectum is distinguished according to localization. The form of the stenosis varies from one caused by a fine mucosal membrane to stenosis due to a hard fibrous ring.

Congenital narrowing of the anus and rectum

Narrowing is located mostly at the junction of the endodermal part of intestine in the ectodermal, i. e. in the area of scalloping line of the anal ring. But sometimes narrowing can be placed on a few inches above the anus. In rare cases, stenosis captures the anus and part of rectum.

The shape and length of the stricture are variable. Sometimes it's membrane of thin skin or mucous membrane, in other cases — a dense fibrous ring. The length of constriction is from several millimeters up to 2–4 cm.

Clinical picture and diagnosis. Within the first days and months of life congenital stricture may not be shown clinically. However, during severe stenosis constipation is observed from the first day. Parents report that stool is released like a narrow belt or cylinder. With the introduction of complementary foods constipation become more severe, the act of defecation is accompanied by strong straining and screaming. The picture of secondary megacolon, which is the brighter pronounced, the older the child. On the severity of clinical signs of stenosis the degree and nature of constriction, as well as child's care affects significantly.

Diagnosis of congenital narrowing is based on complaints, inspection of the area of the perineum, digital rectal examination, sigmoidoscopy data, and radiopaque research. In some cases anus is abnormal: having a type of funnel. During the introduction of the finger into the anal canal space of narrowing is felt in some cases as a flexible ring: finger as though goes through the rubber plate, the edges of which are pulled behind a finger, while in other cases the stricture hardly passes fingertip. However, sometimes stenosing ring is high and it cannot be detected by a finger, in such cases the diagnosis is helped by sigmoidoscopy. Radiopaque examination of the colon helps to determine exactly the length of stricture and the state of the upstream sections of the colon.

During the low-lying strictures with leaky and rather narrow ring in some cases can be limited with a longitudinal incision the site of stenosis with subsequent suturing the wound in the transverse direction. Excision of the stenotic ring from the side of crotch is most appropriate; at the height of stenosis is not more than 2.5 cm.

At high constrictions, with irritated rectum, combined rectoplasty is performed.

Congenital anal stenosis is differentiated from a perineal fistula forming in atresia of the anus. Test of the anal reflex is of decisive importance: it is always extrasphincteral in atresia with a perineal fistula.

Atresia constitutes the largest group of anorectal malformations and accounts for 80–85 per cent of them (N. B. Sitkovsky).

Atresia of the anal canal and rectum is the most common and atresia of the anal canal less common forms in the group of simple atresias. A closed "imperforate" anus and isolated rectal atresia are rare malformations.

Atresia of the anus and rectum is easily recognized in most cases.

An imperforate anus is the most favourable form; the newborn liaby has no anus, in place of which there is a slight depression, sometimes a ridge of skin, which is a continuation of the median perineal suture. The skin is so thin in some cases that the meconium can be seen through it.

In rectal atresia, the anus is normally formed but the intestine ends blindly at a distance of 1 to 5 cm from the transitional fold, (lursory examination of the baby at birth may lead to a diagnostic error, and if no meconium is passed within the first 24 hours each newborn baby must be examined by means of a rubber catheter introduced through the anus. Normally the catheter can be advanced for a distance of 10 cm and more, in rectal atresia it meets an obstacle, bonds over and comes out of the anus. Meconium is not passed.

If emergency aid is not given, the baby becomes restless, sucks poorly, and regurgitates. Signs of low intestinal obstruction develop gradually: vomiting occurs, abdominal distension increases, the skin on the anterior abdominal wall is stretched. The distended intestinal loops are sometimes outlined. Respiration is difficult because of the high position of the diaphragm. Complications, such as perforation of the large intestine and meconium peritonitis, may dovelop in late hospitalization of the child.

The level of atresia is very important from the surgical standpoint. In low atresia, a jolt is felt in response to slight pressure of the finger in the region of the projection of the anus. In high atzzzzzresia the sign of the "jolt" is absent. The atresia is considered low if the blind end of the rectum is 2 cm from the anal skin.

Several auxiliary methods for determining the exact level of atresia have been suggested. Wangensteen's non-contrast X-ray examination is one of them. A marker (coin, clip, etc.) is fastened in the region of the projection of the anus and a X-ray is taken with the baby held head first (Fig. 3.3.21). The level of atresia is judged from the distance between the bubble of gas in the atretic intestine and the marker on the perineum. This method fails to yield convincing results within the first hours of the child's life because there is still very little gas in the intestine and no clear image is produced on the X-ray. Large accumulations of meconium in the blind end of the intestine, which prevents the gas from spreading, may also give the impression of a very high level of atresia. In view of this, some surgeons perform puncture of the perineum to a depth at which meconium is aspirated into the syringe and judge the height of atresia from the distance to which the needle was advanced. Complex appraisal of the results obtained allows the level of atresia to be determined more accurately and the rational method of surgery chosen.

Fistulous forms of atresia are encountered in approximately 50 per cent of cases.

Fistula into the genital system is only found in girls. The clinical picture is determined by the localization, width, and length of the fistula, and by the diameter of its opening.

In atresia with a rectovaginal fistula the opening is usually near the hymen, less frequently deep in the vagina. In atresia with a rectovestibular fistula the opening is in the vestibule, at the junction with the perineal skin at the posterior commissure of labia majus. The fistulous forms of rectal atresia in girls may be combined with malformations of the genitals (duplication of the uterus, etc.).

Atresia with a fistula into the genital system is usually marked by persistent constipation which in severe cases leads to the development of faecal toxicosis and secondary megacolon. Constipation causes loss of rectal sensitivity and is often combined with

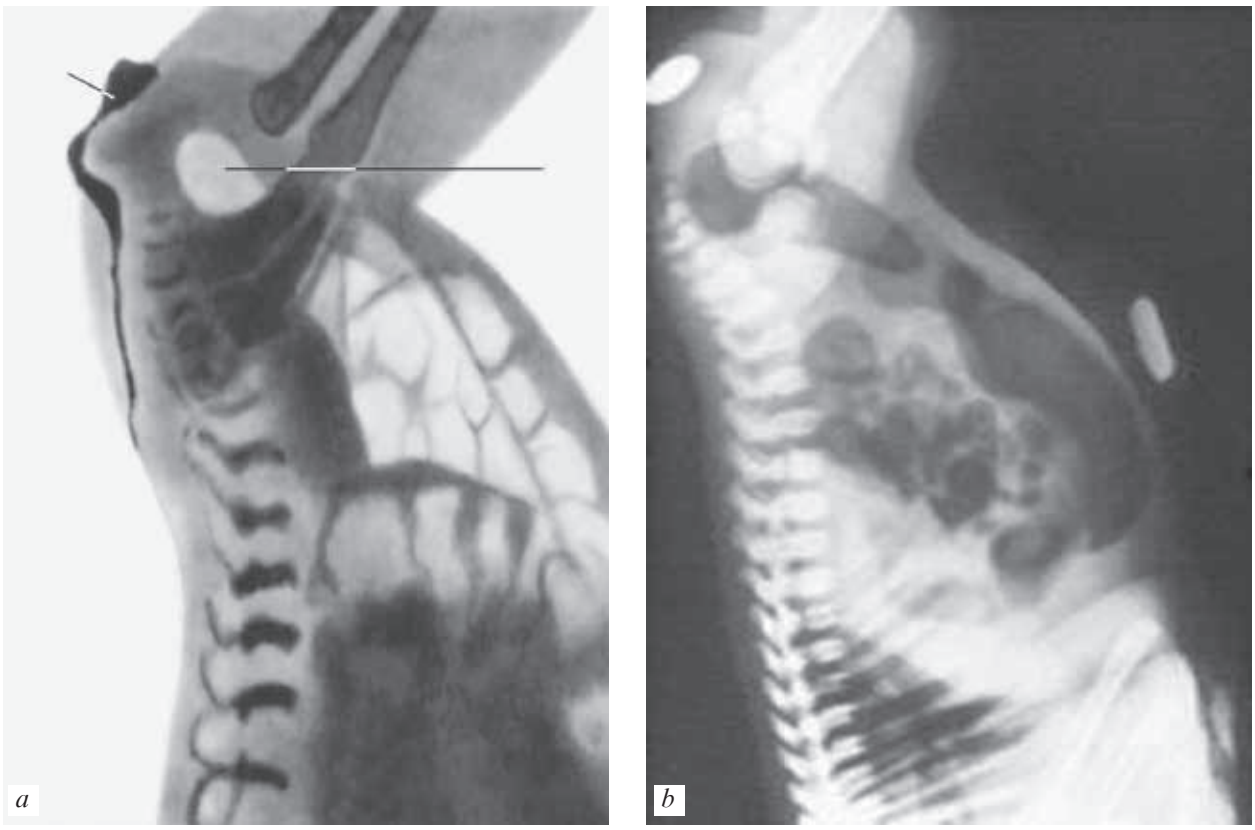


Fig. 3.3.21. X-ray examination in atresia of the rectum (after Wangensteen) *(a, b)*

continuous discharge of the faeces. The fistula may be plugged by the thick finical material in which case signs of acute intestinal obstruction may appear.

Fistula into the urinary system is encountered in boys as a rule. It opens in most cases into the urethra and rarely into the bladder lie urine of such children is clouded by faecal admixtures, but may be clear at times when the discharge of faeces ceases temporarily. Most authors believe that every baby boy born with an atretic rectum must be subjected to urological examination (cystourethrography) for a communication between the rectum and urinary system. Microscopy of the urine for detecting elements of meconium in it helps in making the diagnosis.

The group of atresia with external fistulae includes perineal fistula opening at the root of the scrotum or on the inferior surface of the penis. The fistula is usually small and the faecal material is discharged from it with great difficulty.

Treatment. Operations at low forms of atresia.

Patients with low anomalies have good prognosis, even without surgery. In these forms defect a simple bougienage is usually sufficient. Sometimes there is a need in minimum back mobilization of fistula and its moving to the center of the external sphincter.

During operations on the lower forms of atresia combined perineal access and posterior sagittal perineal access by Pena can be successfully used. Such access is recently favored by the fact that it is possible to visually monitor the position of intestine in the center of relegated muscle apparatus of the crotch.

Covered anus, when only a foil separates the lumen from external environment, just in a cross or oval way dissect this foil with hemostasis.

During atresia with fistula to the perineum blind end of the intestine is always low. Essentially anus is covered by hypertrophied middle seam of the perineum. If you cannot spend bougienage of fistula, Salomon surgery is conducted. To the fistula hole grooved director inject, pull intestine tissue and cut the wall of fistula of previously marked sphincter. Remove excess mucous membrane, sutured layers of tissue in the formed wound and shape anus.

Posterior sagittal section starts from the middle of the sacrum and continue to the center of external sphincter. During persistent cloaca cut is extended to the last opening at the crotch, separating the external sphincter. With malformations like vestibular fistula, it is enough to do a smaller incision (limited back sagittal anorectal plastic). Low anomalies require very little access (minimum posterior sagittal). The main factor justifying rationality of access is the fact that no important nerves and blood vessels do not pass through the middle line and do not cross it. During the intervention fibers of striated muscle in fact do not cut, but raised.

Such operations as Dieffenbach surgery, used during anal atresia without fistula should be noted. During atresia of anus with recto-vestibular fistula is used perineal rectoplasty by Stoun–Benson.

Operations during high forms of atresia

According to A. I. Lenyushkin in cases of atresia of the anal canal and rectum, regardless of the internal fistula or without it, to bring down the intestine through the perineal access is impossible, so the combined abdominoperineal or sacro-abdominoperineal access aare used. A so called intrarectal abdominoperineal plastic by Romuald Rebeyn is preferable.

Postoperative treatment and closure of the colostomy

During atresia of posterior sagittal procto plastic without abdominal access, during 3 days after surgery antibiotics are injected intravenously, in addition, topically antibiotic ointments for 8 to 10 days are applied.

In 2 weeks after the surgery bougienaging and spending it start twice a day. Each week, increase the size of the bougie on one number until they reach the size that meets the child's age. Once you've reached this size of bougie, the colostomy close. Further bougienage follows the scheme.

The hazard of ascending infection of the urinary tract is high in patients with a fistula into the urinary system, and surgery is therefore indicated within the first hours after birth. Abdomino-perineal proctoplasty after Romualdi is the most physiological intervention. Its peculiarity consists in removing the mucous coat from the distal atretic portion of the mobilized large intestine and bringing out the intestine onto the perineum through the natural anorectal canal. The normal intestinal wall overlaps the defect in the urethra or vaginal wall.

The method for treating atresia with a fistula into the genital system in girls is chosen individually. In atresia with a rectovestibular fistula, for instance, emergency operation is not needed because with good care the child develops normally and an operation performed at an older age (3–4 years) is much simpler technically. Perineal proctoplasty is conducted.

An earlier operation is indicated in cases with persistent constipation attended by Coprostasia and toxicosis and causing an effect on the child's development and leading to retardation of growth. Surgery in such cases is undertaken at the age of 6–8 months. Abdominoperineal proctoplasty is preferable in fistula into the vagina.

Stenosis diagnosed early can be corrected by bougienage. Hegar's dilator is introduced into the rectum for 10–15 minutes once or twice a day. The size of the dilator is gradually increased to that normal for the child's age. Positive results are produced in 2–4 months. Surgery is indicated if the manipulations prove ineffective. The stenosing ring is cut and then sutured transversely or the intestine is resected.

A fistula in a case with a normally functioning anus must be managed surgically. In rectovesical and retrourethral fistulae the communication is divided through a perineal or abdomino-perineal approach immediately after the diagnosis has been established. A fistula into the genital system in girls is corrected surgically from the age of 6 months.

The treatment of a perineal fistula is started by means of non-operative measures immediately after it is diagnosed. It is cauterized with 10 per cent silver nitrate solution or iodine tincture. After the age of 12 months the fistula is excised if it shows no tendency to close.

It is important to provide rest and the appropriate care of the perineal wound in the postoperative period. This is done by placing the newborn on a

bloster put under the pelvis and fastening with soft cuffs the abducted lower limbs suspended from a frame. A plaster cast suggested by I. K. Murashov is applied for the same purpose in older children.

Early bougienage must be undertaken to prevent cicatrization if the wound heals by second intention.

Cicatricial stenosis of the anus irresponsive to non-operative measures is corrected by a second operation: the cicatricial tissues are excised, the rectum is mobilized, brought out, and a new anus is formed.

The late operative results are determined to a great extent by the form of the malformation. Incontinence of faeces and urine is the most severe complication; it is encountered in severe malformations (agenesis of the rectum with deficiently developed muscular and nerve apparatus of the anorectal region) even if the operation had been technically performed properly. With gradual growth and development of consciousness, the deficiency of the sphincter ani muscles in some cases is compensated for by the activity of other muscles and the child is capable of holding the stool and gases.

In children with milder anorectal malformations the prognosis may be considered favourable if the operation is conducted in time and correctly.

3.4. MALFORMATIONS OF THE ANTERIOR ABDOMINAL WALL

Specific objectives of the lesson:

1. To master list of defects of the anterior abdominal wall and recognize the main clinical manifestations of hernia of umbilical cord, gastroschisis, umbilical and inguinal hernias.
2. Identify the main causes of hernia of the anterior abdominal wall.
3. Interpret auxiliary methods: ultrasound and X-ray, computed tomography, laboratory and biochemical parameters.
4. To analyze causal relationships occurrence of defects of the anterior abdominal wall.
5. Develop an algorithm of actions of doctor at hernia of umbilical cord, gastroschisis, umbilical and inguinal hernias. Summarize basic principles of treatment of the anterior abdominal wall defects.

Abdominal wall defects are always observed and descriptions of this disease reach to us from primitive times, but the last century had fatal consequences. First breakthrough in the treatment of children with large abdominal wall defects were made by Grosso, who proposed a phased closure technique for omphalocele patches of skin. Another breakthrough in the treatment of these patients was the invention of prosthetic materials that allowed gradually reset defective organs and perform a gradual closure of the defect. Both techniques have been used to treat bowel functioning of omphalocele, which was protected by intact amniotic membranes. Today survival with acceptable quality of life is expected for the majority of children with isolated anterior abdominal wall pathology and complications primarily relate to combined anomalies.

Formation of normal ventral wall requires the successful interaction of complex factors arising from the fourth to the twelfth week of fetal development. These factors include rotation, merger, growth and differentiation of embryonic tissue that later become muscle of the abdominal wall, and the navel tract.

Disruption of the normal course of events can lead to numerous congenital malformations, which include not only the absominal fissure, omphalocele as well as umbilical hernia, Meckel diverticulum, extrophy of bladder, cloaca and prune belly.

Congenital lesions that are caused by a defect of the anterior abdominal wall (hernia of embryonic umbilical cord, absominal fissure) are quite rare defects, their frequency is from 1: 1200 to 1: 21,000 newborns. Thus absominal fissure prevails in relation to omphalocele — 2:1 or 3:1.

EMBRYONIC HERNIAS OF UMBILICAL CORD

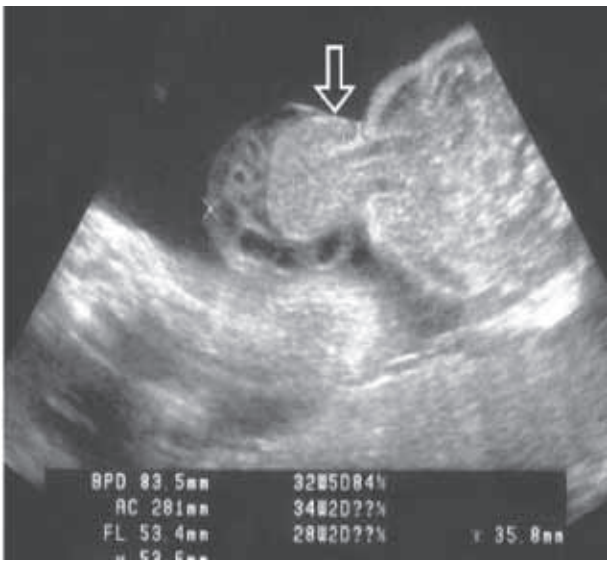
These hernias are also called omphalocele or embryonal hernia (not to be confused with umbilical). Omphalocele — a malformation, characterized with prolapse of the abdominal cavity through defect of umbilical cord in embryonal shells. Embryonic hernia of umbilical cord occur as a result of violation of the first period of rotation of the intestine at the stage of "physiological umbilical hernia" when there is a discrepancy rate of growth of the gut and increase of abdominal cavity. Normally up to 3-month gestation, this hernia should disappear. In addition violation of the abdominal wall closure simultaneously can happen due to muscle hypoplasia of its parts. At omphalocele muscles of the abdominal wall are intact, but dysplasia is marked.

These two aspects play a major role in the formation of hernia of umbilical cord. Usually there is midgut in hernial sac (intestines from the duodenum to the mid-lumbar colon) and partly liver, stomach, spleen and gonad. Abdomin is correspondingly small, at giant defects chest can also be deformed.

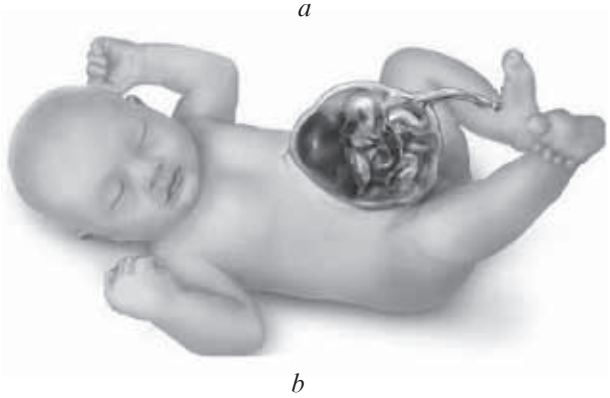
Sometimes omphalocele can be part of a complex defect of the abdominal wall that includes also extrophy of bladder and/or cloaca. In the presence of hernia of umbilical cord frequency of multiple malformations of other organs and systems (CNS, cardiovascular, urinary) and genetic diseases (trisomy 13-th and 18-th pairs of chromosomes) is about 54 % (Fig. 3.4.1).

Classification. Embryonic hernia of umbilical cord that arose before 3 months, hernia of fetus (fetal hernia), formed after 3 months, and mixed hernias.

At embryonic hernia of umbilical cord liver is part of the hernia and is covered with primitive peri-



a



b

Fig. 3.4.1. Omphalocele *(a, b)*

toneum, which is the inner layer of cord membranes. Fibrous membrane of liver (Glisson's capsule) is absent, so any attempt to separate the membranes of the liver is accompanied by bleeding.

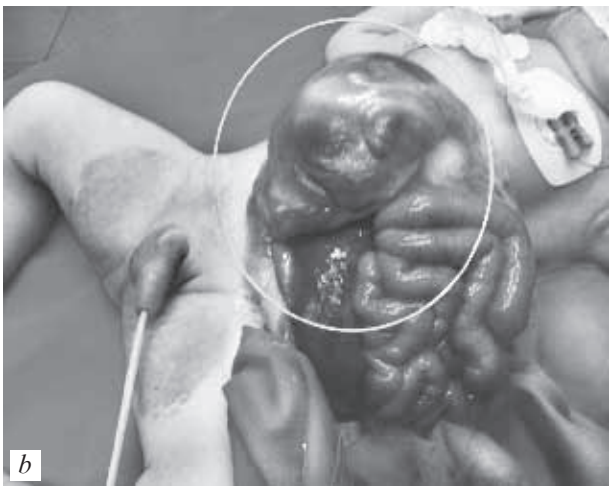
At hernia of fetus (fetal hernia) under transparent cord membranes can be seen the free abdominal cavity, and hernia membranes move freely over the liver, which, unlike embryonic hernias, after opening the shells are not bleeding. At mixed type of hernia umbilical cord upper part of the hernial sac is tightly soldered to the liver, and abdomen is not yet formed, while the lower part of the sac covers intestinal loops, which are free in the abdominal cavity, which is already formed (Fig. 3.4.2).

Thus formation is often covered with transparent membranes through which can be seen the contents of abdomen (loop of intestine, liver). Shells outwardly are composed of amnion, from the inside — of primitive peritoneum, and between them — Wharton jelly. Within the first hours after birth these membranes are shiny, transparent but at the end of the first day are borne by fibrinous layers. Generally, membranes are clearly separated from the skin of the abdominal wall by roller of bright red color (interruption zone of skin capillaries). In some cases hernial protrusion can be in any degree be covered with skin.

In order to manage the treatment, they are divided into small (diameter to 5 cm), medium (10 cm) and large (over 10 cm). For premature babies defect diameter of 5 cm is considered a large hernia. The dimensions of the defect of the hernial sac usually match, but the volume of the abdominal cavity is defined by volume of organs that remained in it. Depending on the progress of the shells and status they are classified as follows: uncomplicated, complicated (prenatal rupture of membranes, intestinal fistula, rupture and loss of internal organs, inflammation of the membranes hernia, ectopia heart related malformations). The clinical course of hernia of umbilical cord can be complicated with rupture of membranes, especially at large hernias and eventration of organs with subsequent infection and peritonitis. Infection of membranes can run without their damage, while also contact peritonitis and sepsis may develop.

At intrauterine eventration loops of guts are covered by a fibrinous layer, walls are swollen, peristalsis is absent, the skin from the edge goes into remains of amniotic membranes. Infection of shells usually happens after the first day of life, if they were not protected by a sterile dressing with special antiseptics. These children have hernial protrusion covered

a



b

Fig. 3.4.2. Patient with omphalocoele *(a, b)*

with a dirty gray layers with mucus, there are areas of necrosis. At progression of process, damage reaches deeper layers of membranes and umbilical cord, peritonitis develops (Fig 3.4.3).

Postnatal diagnosis is not much difficulty because the defect bulging of internal organs, transparent gray film, which consists of amniotic membrane, Wharton jelly, hernial sac (primary peritoneum) visible to the naked eye.

Differential diagnosis should be done with gastroschisis in which the contents of the abdominal cavity eventrate through defect of the abdominal wall to the right of the navel, manifested during ultrasound diagnostics.

Prenatal diagnosis is more significant lake in gastroschisis. Taking into account the fact that up to 13 weeks in a norm bowel must return to the abdominal cavity, during this period omphalocele may be diagnosed.

Treatment should begin in the hospital. We need to create conditions that prevent infection, rupture of membranes and hypothermia. For this membranes of hernia process with 70% alcohol or 1% iodine solution and cover with sterile dry cotton-gauze bandage.

At the same time antibiotic therapy should be initiated. The child should be delivered to a specialized hospital by ambulance with mobile team of neonatal resuscitation. The presence of intact amnion eliminates the need to operate a child in urgent procedure within the first hours of life. The child must do an ultrasound and X-ray picture to exclude accompanying diseases. In those patients who are determined anomalies incompatible with life or defects that are not able to give the child recovering from surgery, physical disease, significantly impair the child's condition, resorting to conservative treatment. In fairly rare cases omphalocele can be combined with Beckwith–Videmann syndrome, which needs immediate diagnosis, early treatment of hypoglycemia which can lead to lesions of the nervous system.

Even a slight gap of amnion should be sutured before conservative treatment. Also conservative intervention is conducted at large amounts of defect and infection of membranes.

At conservative treatment should handle everyday membranes with antiseptic solution and tannic fluids to form a protective layer that counteract bursting shells. From the first days of life every 2–3 hours membranes are processed with 70% alcohol, with silver sulfadiazine ointment or 1% iodine. Hernia protrusion should be placed under the frame because daily dressing can injure membranes and lead to their breaks. After the formation of thick crusts elastic bandage is imposed on hernia, to gently reset internal organs and gradually increase the size of the abdominal cavity. Over time, residual ventral hernia can be closed operatively. Surgery is usually performed for children with small and medium-sized hernias when it is possible to reset them to abdomen and conduct original plastic without the risk of respiratory failure and compartment syndrome. If the defect is large enough, it is best to leave the amnion or use a synthetic material. The best in these cases is to use smooth, soft and nonadhesive material such as Gore-Tex®.

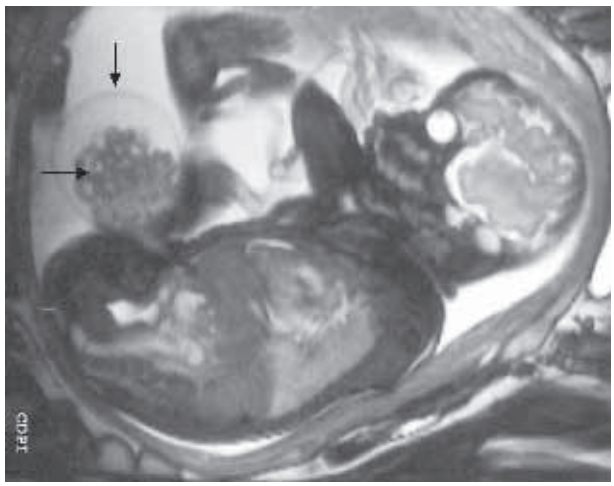


Fig. 3.4.3. Sagittal MR image fetus with omphalocoele

At one stage operation after incision of the skin, which borders skin transition into the amniotic membranes, it is necessary to carefully select up to aponeurosis and tie umbilical vessels and urahus. If you find obstructive bile duct it should be removed. If the child has an intestinal obstruction, reconstructive surgery is performed. Then, on the edge of hernia defect carefully mobilize the rectus abdominis muscle and aponeurosis, which should be easy to converge on the middle line. If the defect is of small size, it is necessary to cut the ring to reduce the trauma at reduction of organs into abdominal cavity. The defect of the abdominal wall is suturated with layers.

At large defects, when at closure of the abdominal cavity insufficient ventilation develops, as well as infection with artificial material, one can hold a two-stage transaction proposed by Gross (1948), in which only the skin is closed. Leather grafts can be mobilized so as to bring skin edges together. Sometimes to increase the elasticity of skin grafts they put a notch in a checkerboard pattern.

A year later the second phase of the operation — the removal of ventral hernias takes place (Fig. 3.4.2). However, at this operation removal of amnion because adhesions formed between the skin and intestines and liver is carried. Further treatment may be severe and accompanied by a large blood loss. Sometimes amnion at these operations is left untouched. However ventral hernia eventually increases in size and itself can lead to multistage treatments.

Today popular landmark operations using expanders of artificial materials. Operation of choice is considered to be syloplasty or formation of extra-abdominal bag.

GASTROSCHISIS

The term "gastroschisis" is translated from the Greek as disruption of abdomen. Gastroschisis (defined by T. C. Moore and G. E. Stokes) — extraumbilical malformation of the anterior abdominal wall with eventration of organs during the prenatal peri-

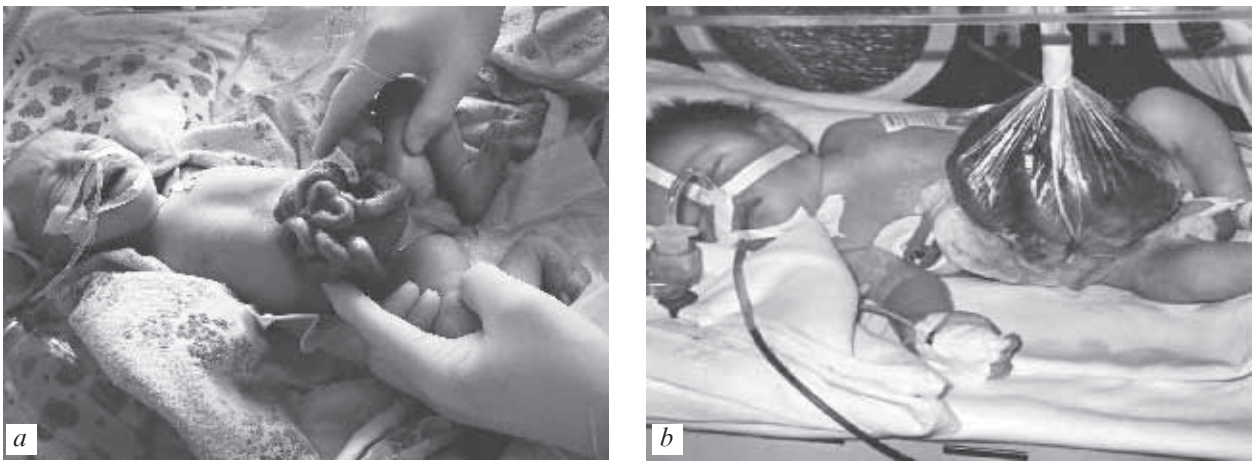


Fig. 3.4.4. Patient with gastroschisis *(a, b)*

od. It should be noted that in this pathology attachment cord remains normal.

Frequency is 1:6000 newborns. Defect in more than 90% of cases is situated in the right of the umbilical cord and looks like crack, but umbilical ring remains intact. Defect itself is usually smaller than the omphalocele. There are no parts of amnion left. At gastroschisis, unlike omphalocele, are no covering membranes. In the defects eventrated stomach, intestines with mesentery, bladder and sometimes appendages of girls. Poorly differentiated departments of bowel, mesentery is thickened and swollen. Pinching ripples in the defect are impossible, because their development takes place at the same time (Fig. 3.4.4).

Eventrated intestine is always shortened. As with omphalocele, available small abdominal cavity, which leads to visceroabdominal disparity — discrepancy between the volume of the abdominal cavity and eventrated authorities. Visceroabdominal imbalance affects the course and complications in the postoperative period. The development of cardiovascular and bronchopulmonary complications on the background of increased intra-abdominal pressure at submerged in reduced abdomen abdominal hernia content called compartment syndrome.

In gastroschisis unlike omphalocele, multiple malformations are significantly less (21%). Mostly these are intestinal abnormalities.

Pathogenesis. The reason for dilatation of gastroschisis is called intrauterine vascular accident in the distal part of the right omphalomesenteric artery, resulting in torn umbilical ring and hernial protrusion of abdominal contents.

Pairing of omphale mesenteric arteries occurs in early embryogenesis. Omphalomesenteric proximal part of the right upper artery becomes mesenteric artery. In turn, the distal portion of this blood collector accompanies omphalomesenteric passage through umbilical ring and ends in the gallbladder, situated to the right of the fetus. At infringement of distal part of right omphalomesenteric arteries heart attack and necrosis of umbilical cord arise, prolapse of the intestine through a place of healing and tissue resorption around the edges of defect before birth. This explains the typical localization of the defect in the anterior abdominal wall to the right of the navel. Interruption of the proximal part of the omphalomesenteric artery also may be the cause of atresia and stenosis of intestine. Gastroschisis appears on the early phase of embryogenesis between 5th and 8th weeks of gestation.

Gastroschisis reasons are still unknown, but risk factors are known, which include age of the mother. Thus, the incidence of fetal gastroschisis during mothers aged 15 to 19 is particularly high and reaches 1:400 newborns. Also an important role plays insufficient nutrients in the diet of pregnant women, especially alpha-carotene and amino acids. Risk factors are used within the first trimester gestation medcines (NSAIDs, pseudoephedrine, fenilpropanolamid), drugs.

Clinical picture. Gastroschisis is quite an obvious drawback because diagnosis does not cause complications. The defect of the anterior abdominal wall usually has a diameter of 2 to 5 cm. As mentioned above, due to this defect intestine eventrates, stomach, bladder, uterus, ovaries and others. These bodies are usually coated with fibrin, formed *in utero* in response to amniotic fluid, especially urine contained in the fluid from 30th week. This case can be quite strong, it is difficult to differentiate midgut under it, which loops are welded together, with violation of the outflow of venous blood and lymph. Peristalsis is violated or missing. Loop color from gray to bluish, therefore the surgeons who have experience in treating gastroschisis can interpret as a state of necrosis as a result of strangulation.

Gastroschisis diagnostics, as well as omphalocele, should begin in utero. In most cases, both of these defects are clearly visible in the ultrasound. Also for differentiation of gastroschisis from omphalocele the level of amniotic alpha-fetoprotein and acetylcholinesterase can be used, which will be higher at gastroschisis. If the ultrasound detected omphalocele, we need a more thorough investigation to rule out other related defects. You can even use karyotyping for planning postnatal patient management or to determine the issue of immediate abortion (Fig. 3.4.5).

Management of childbearing at gastroschisis and omphalocele is quite difficult question. Some authors believe it is expedient to have delivery through

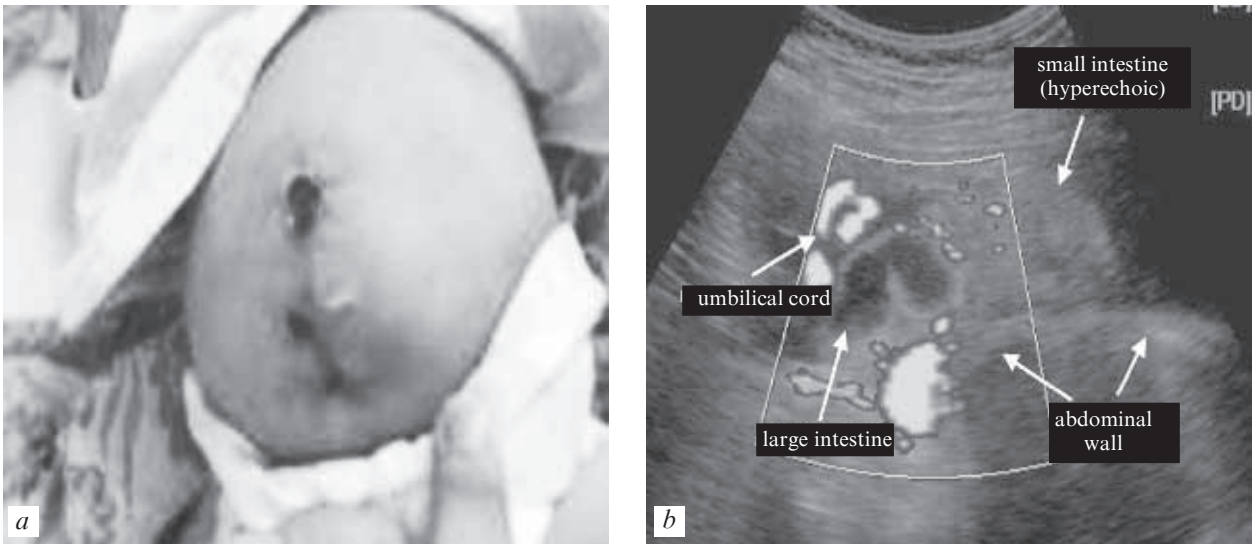


Fig. 3.4.5. Gastroschisis *(a, b)*

caesarean section to reduce the risk of damage to intestinal membranes and the passage of the baby through the vagina. However, R. A. Strauss, R. Balu deny the possibility of complications and advocate for natural childbirth. Indeed, today rather well documented in the meta-analysis and independent reports entirely equal number of complication at caesarean section and natural childbirth.

Another question — appointment of early labor at gastroschisis at 34–38 weeks of gestation, but here there is no substantiated evidence that early births by caesarean section or by natural means reduce the risk of damaging the intestine.

Differential diagnosis of gastroschisis is conducted with hernia of umbilical cord. In case of gastroschisis hernial sac is absent.

Treatment. And still clinicians debate about strategies of abdominal wall closure: what is best — primary closure or by stages.

Very important is maintenance of the child at the stage of pregnancy, and then transportation from maternity hospital to hospital plays an important role. At this stage it is important to maintain the body temperature of the child (eventrated organs should be placed in a sterile plastic bag and dry cotton gauze bandage, incubator with temperature of 38°C and humidity of 100%), decompression of the stomach (nasogastric tube installation) takes place, non-narcotic analgesics anesthesia (some authors suggest using broad-spectrum antibiotics at this stage). If in a child an isolated form of gastroschisis without visceroabdominal imbalances is marked, intestine in good condition, you can use nonanesthesia reduction by Bianchi. Thus must first be sure to empty the bowel in the intensive care ward under the control of heart rate, blood pressure and saturation, at sedation by any non-narcotic analgesic, nonanesthesia reduction of intestine and suture the skin is conducted. This manipulation takes about 12–15 minutes. After nonanesthesia reduction is better set passage through the intestines (4–6 day), child is released from hospital on the 14th day.

Help according to Bianchi can be used in about 21% of all cases of gastroschisis, all other children require surgery.

Approximately in 70% of patients with gastroschisis have conditions for primary closure of the defect, in addition, fibrin layer that covers the intestines, dissolves quickly if touches parietal peritoneum rather than the flap of skin. However, the primary plastic of defect is possible after immersion of eventrated organs into abdominal cavity, postoperative wound can be closed by layers, easily or with little tension.

At one stage operation it is necessary to use a nasogastric tube as well as a urinary catheter. You must empty the colon most possibly using the preevacuation of meconium by expanding the anus and administration of hypertonic enema solution.

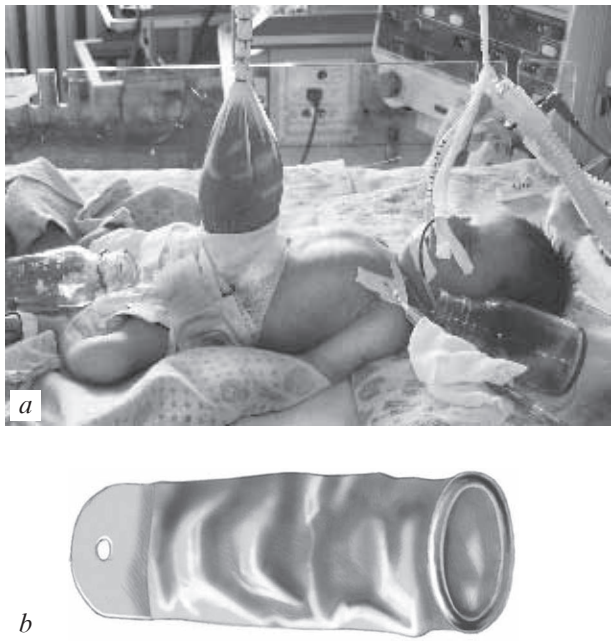


Fig. 3.4.6. Usage of preformed traction silo bag (Shuster) for repair of big abdominal wall defects (omphalocoele and gastroschisis) *(a, b)*

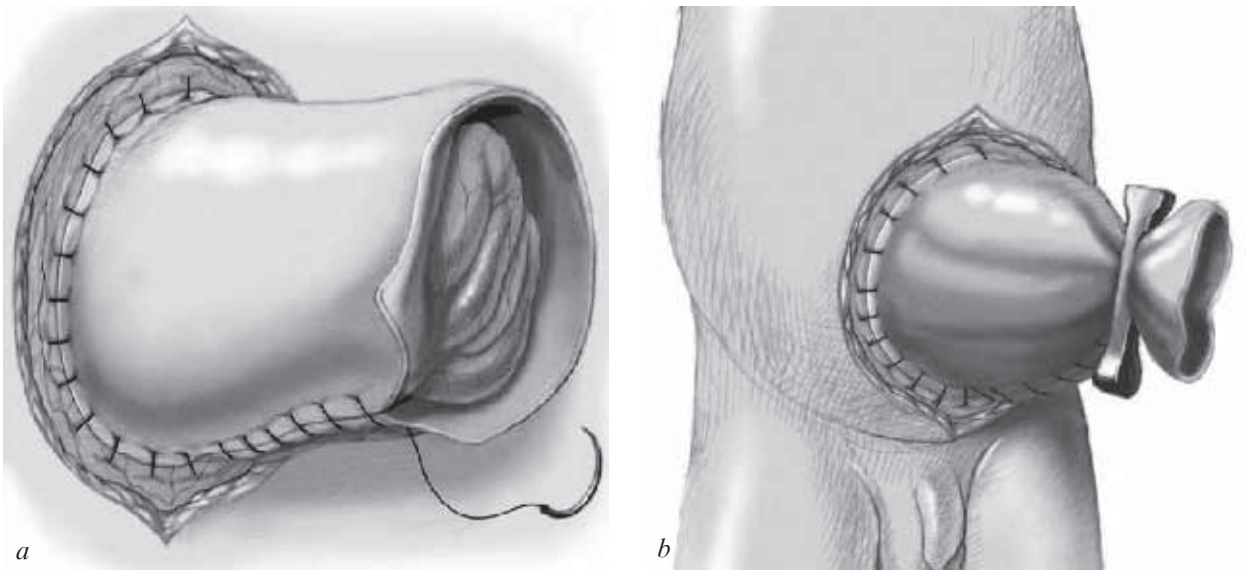


Fig. 3.4.7. Use of a silo for repair of big abdominal wall defects (omphalocoele and gastroschisis) *(a, b)*

If eventrated organs can not be reseted, the most efficient method is Silastic Silo or another method of forming ventral hernias. However, syloplasty with the use of a bag made of artificial material is a fairly safe alternative (Fig. 3.4.6).

This method makes it possible to cover aseptic environment in the gut for a long period. Typically, firstly is performed fascial defect traction vertically, and then to the fascia suture artificial bag. The basis of the bag should be slightly higher than the top. The choice of material from which the bag is made is very large.

Is often used reinforced silicone (sylastic), which suture to the borders of defects and sylastic boundaries are suturated so as to create extraabdominal bag (silo-tower). The bag gradually reduces in volume, daily immersing its contents into the abdominal cavity, followed by suturing of fascia. Usually during 2–4 days is possible to dip the contents of the bag into abdominal cavity. Only the last stage of the operation (closing skin defect) is performed in the operating room.

Approximately in 5–20% of children with gastroschisis is observed small intestine atresia. In such cases, where possible are trying to accomplish economical resection with anastomosis imposition of "end-to-end" or impose proximal enterostoma. Both can be used as the primary plastic, and at staged one. After 2–4 weeks. fibrin is resorbed and allows to distinguish anatomy easily, therefore, impose a more secure anastomosis. This approach is most appropriate for proximal atresia, in which you can make effective decompression quite a long time by a nasogastric tube. Atresia of distal part of iliac and large intestines can be effectively eliminated by using a nasogastric tube and often leads to perforation or segmental necrosis. Therefore, in this case it is necessary to impose stoma, but the presence of stoma and syloplastic sack significantly increases the risk of life-threatening sepsis (Fig. 3.4.7).

A very important point in the postoperative period is adequate providing the child's adaptation to high intra-abdominal pressure and compression of the vena cava. For this purpose a long mechanical ventilation (3–5 days) using muscle relaxants is conducted. Mandatory antibiotic therapy, physical therapy, which promotes fibrin resorption of the case, therefore, the restoration of peristalsis. Before the advent of peristalsis child is prescribed with total parenteral nutrition. Peristalsis may appear in a few days after surgery, but more typical is its start in 1–2 weeks. And normal peristalsis is established only after 3–6 weeks since the operation.

According to CIS clinical picture postoperative mortality is quite high, ranging from 4 to 8%.

UMBILICAL HERNIAS

Umbilical hernias account for 4% of all abdominal wall hernias. Umbilical hernia — a cleft of aponeurosis of umbilical ring, which passes through the peritoneum, which is the hernial sac itself. Often in the hernial sac can detect induration and loops of small intestine. There is relationship between umbilical hernia and low weight of newborn. According to Vohr et. al., in 75% of infants weighing less than 1500 g in the age of 3 months is diagnosed umbilical hernia. Also umbilical hernias are common at Down syndrome, a trisomy at 18 th and 13 th chromosome, mucopolysaccharidoses, congenital hipotyreoid and the presence of ascites.

Embryogenesis. By the 10th week of gestation midgut should return to the abdomen, and then the abdominal cavity is closed. Lateral wall of the body (somatopleura) is bent medially, narrowing yolk-intestinal isthmus, as a result forms yolk duct.

Somatopleura narrows around the yolk duct in the type of tightening pouch. Thus navel is formed. To it attaches amnion, body stalk and the remains of the yolk sac, allantois and umbilical vessels. Fi-

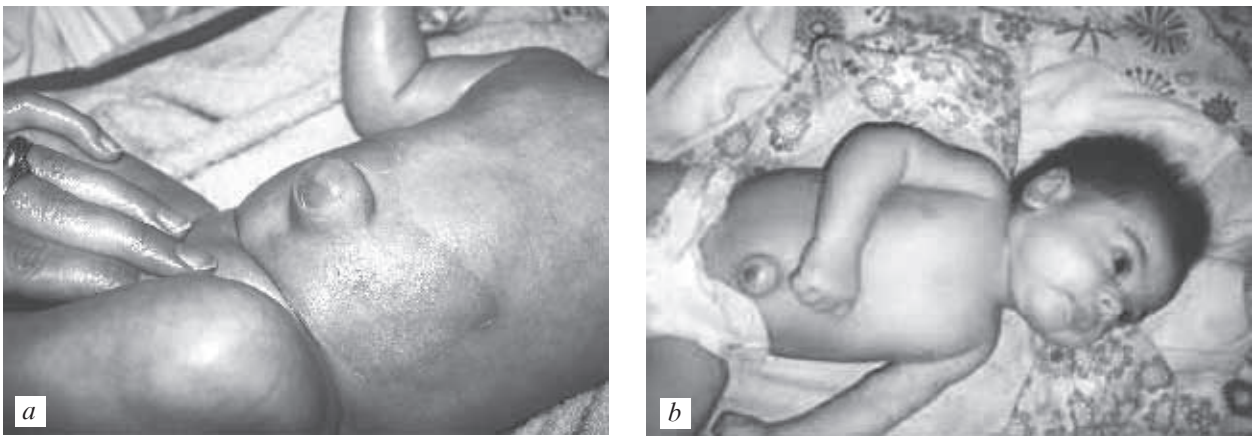


Fig. 3.4.8. Umbilical hernias in the neonatal period *(a, b)*

nally umbilical ring closes after defection of umbilical cord. The strength of the tissues that cover it are not the same. The strongestis the bottom half, where there are the remains of the umbilical artery, adventitia of which turnes into scar tissue. The strength of the lower half of the ring is caused by urinary ducts. The upper half of the ring is weaker, at this point passes only umbilical vein with thin walls. Between the wall of the vein and the upper limit of the umbilical ring freespace remains, covered with a thin layer of connective tissue and umbilical fascia. Most cases occur in umbilical ring in the area of the umbilical vein. Umbilical hernias are often already in the neonatal period, little bit more often in girls (Fig. 3.4.8).

Clinical picture. Within existing umbilical ring there is rounded protrusion which is covered with a thin skin. The contents of hernial sack and in most patients is easily reduced into abdominal cavity. After reduction are well palpated limits umbilical ring, which can be of any size — from that which appears difficult to see, to defect of 1.5–2 cm, and sometimes, in rare cases, even more. You can pay attention to the condition of the skin on the protrusion.

In very rare cases jamming is possible (less than 0.26%) and at about 7% can be observed incarcerated hernias. The relationship between umbilical ring defects in children and umbilical hernia in adults is unknown. It should be noted that in adolescence congenital umbilical hernias are fairly atypical.

Treatment. Tactical approach to the treatment of umbilical hernia is determined, first of all, by patient's age.

There are a few observations, which reported that umbilical hernia was closed between 5 and 11 years. But after 3 years there is no hope for self-closure of defect. Umbilical hernias that have a smaller diameter are more likely to disappear than those whose diameter exceeds 1.5 cm. Quite common method of treatment of umbilical hernia is the imposition of strips like tiles, but many authors point on the groundlessness of this treatment, believing that plaster strips in any case can not bring edges of aponeurosis closer, therefore somehow contribute to closing hernias.

Some authors suggest that conservative treatment measures under 3 years old have no justification and serve only for the psychological peace of parents who can not imagine that the only time plays a role in self-medication of patients (Fig. 3.4.9).

Incarcerated infringement or perforation are absolute indications for surgical intervention. Incarcerated hernia can be eliminated by careful manipulation under anesthesia, and surgery can be performed the next day. If hernia incarceration failed, it is necessary to perform urgent surgical intervention. Relative indications for surgery is the age of child older than 4 years, persistent occurrence of hernias as well as hernias of large size in infants. At surgery inci-

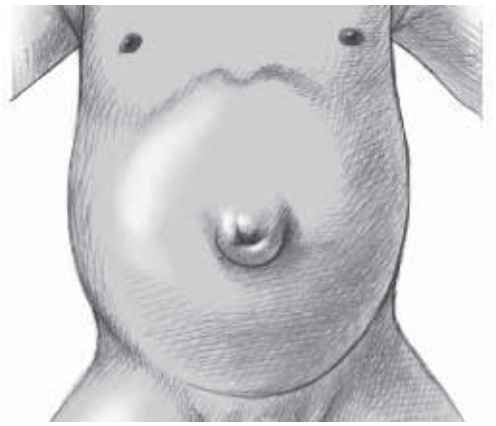


Fig. 3.4.9. Semicircular incision for repair of umbilical hernia

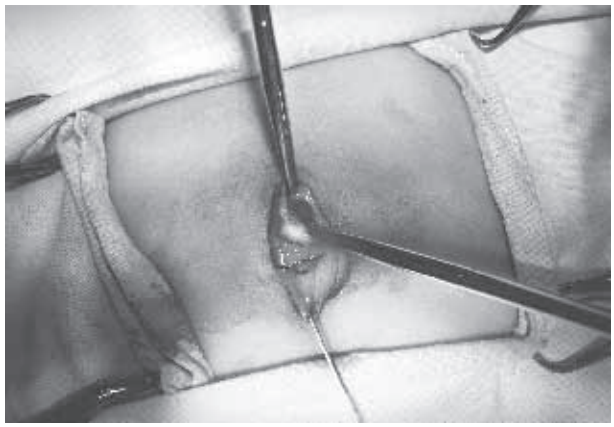


Fig. 3.4.10. Repiar of umbilical hernia

sion is performed over the course of lower umbical skin fold. The bag is cut off from the skin, the content is immersed in the abdomen, navel is fixed to subordinate fascia with a suture. Skin is suturated by intradermal seam (Fig. 3.4.10).

Postoperative complications at surgical treatment of umbilical hernia are unusual, but sometimes there are wound infections (0.8%) and hematomas (13%). Relapse of umbilical hernia is almost impossible. According to Lassaletta et al., there are no noticed cases of postoperative mortality.

INGUINAL HERNIA

Violation of obliteration of vaginal process of peritoneum is a common and basic element in the pathogenesis as congenital inguinal hernia as well as congenital hydrocephalus testicle (hydrocele). Wide process allows internal organs of the abdominal cavity penetrate into the hernial sac, narrow process facilitates penetration of only peritoneal fluid. By the time of birth vaginal process obliterates only in 75%, in all other cases is observed one of the two above-mentioned conditions.

Inguinal hernia in children is a protrusion of the abdominal cavity contents in the inguinal canal and on to the boys scrotum or labia lip — in girls. Inguinal hernia in children belongs to oblique because the inguinal canal is short, and the inner and outer ring are opposite to one another, protrusion along the spermatic cord takes place through both rings. Congenital inguinal hernia is the most common surgical pathology in children and is often diagnosed within the first 3 months of life. Among full-term infants inguinal hernia is observed in approximately 3.5–5% of cases, among premature can occur in about 30%.

Classification. In boys two types of hernias are distinguished: inguinal and inguinal-scrotal, among the latter funiculus and testicular hernias. At funiculus hernia inguinal process is open at the top and the middle parts, and in bottom is obliterated; at testicular vaginal process is open throughout and it may seem that the testicle is in the hernial sac. In girls analogue of vaginal process of the peritoneum is called Nuka channel, which preferably has to obliterate to the time of birth, so inguinal hernias in girls are observed in 8–12 times less frequently than in boys. But remember that girls who have inguinal hernia should be conducted genetics consultation. It should be noted that approximately in 1.6% of cases in girls with inguinal hernia *androgen insensitivity syndrome* occurs. In this case girls phenotypically normal with XY genotype but in the abdomen they have testes, which produce testosterone, which did not have masculinized genitalia. It is noted that girls with double-sided inguinal hernias are more likely to have the syndrome. It should also be noted that 75% of girls with this syndrome have bilateral inguinal hernia. Quite often children with inguinal hernias have connective tissue dysplasia syndromes, such as Ehlers–Danlos and Marfan syndromes, and hip dysplasia can also occur.

In newborns inguinal canal is short, relatively wide and goes back to front. Its length is usually 1–1.5 cm at a relatively large diameter of the external inguinal ring. But it is sufficient to have a small increase in abdominal pressure at shouting for that the abdominal organs, stretching hernia gate, dropped to a pre-formed hernial sac. Inguinal hernia in premature infants is always a surgical problem. Group of premature and low weight children — the most common group of patients among patients with inguinal hernias. The incidence of hernias varies depending on gestational age. In children with weight up to 1500 g hernia frequency reaches 11%, and in children with extremely low weight (1000 g) — up to 17%. Full testis omission and obliteration of the process in these patients is less likely if gestation was shorter or if development was not perfect. Thus, prematurity is among the most common risk factors for inguinal hernia. But irreducible risk in premature babies is very small and is approximately 13%, and cases of pinching or punching of the bag among these children are rare.

You should also distinguish resetable, nonresetable and strangulated hernias. Nonresetable hernias, unlike strangulated, do not cause compression of the bag, and thus have severe clinical manifestations. Content of the hernial sac is often the intestine or omentum. Girls may have ovary, sometimes with a tube. If the back wall of the bag is dome of the cecum or the bladder wall, these are called sliding hernias. This usually happens in large hernias, as along with their zoom protrusion captures a part of parietal peritoneum. A case when Meckel diverticulum gets into the bag is called Litre hernia.

Clinical picture. The child in the inguinal area has tumor formation, it may grunt when attempting to reposition. Often in the neonatal period in the child in the inguinal area protrusion appears, which increases during crying, anxiety. At rest and during sleep it disappears. At inguinal-scrotum hernia protrusion descends to the scrotum, causing its asymmetry. At palpation protrusion has a soft elastic consistency, in horizontal position resets into the abdominal cavity, sometimes with characteristic rumbling. After reduction for palpation is available extended external inguinal ring, is defined positive symptom of cough shock (Fig 3.4.11).

In girls hernial protrusion is usually located in the upper inguinal canal, but sometimes at large hernias can occupy the whole area of pudendum lips. Very rarely, the first symptom of hernia is its appearance together with symptoms of strangulation. Frequently parents themselves go to the doctor after finding hernial protrusion, but at the first examination the surgeon can not always detect hernia, even if he examines the child when crying or if the child "inflates" the stomach. In such cases, clear history together with palpated thickened formation, crossing the pubic tubercle (symptom of silk gloves), is enough for a diagnosis of hernia. You can also ask parents to admit clinic when protrusion reappears.

Differential diagnosis is conducted with dropsy of testicles and spermatic cord. It is characterized by

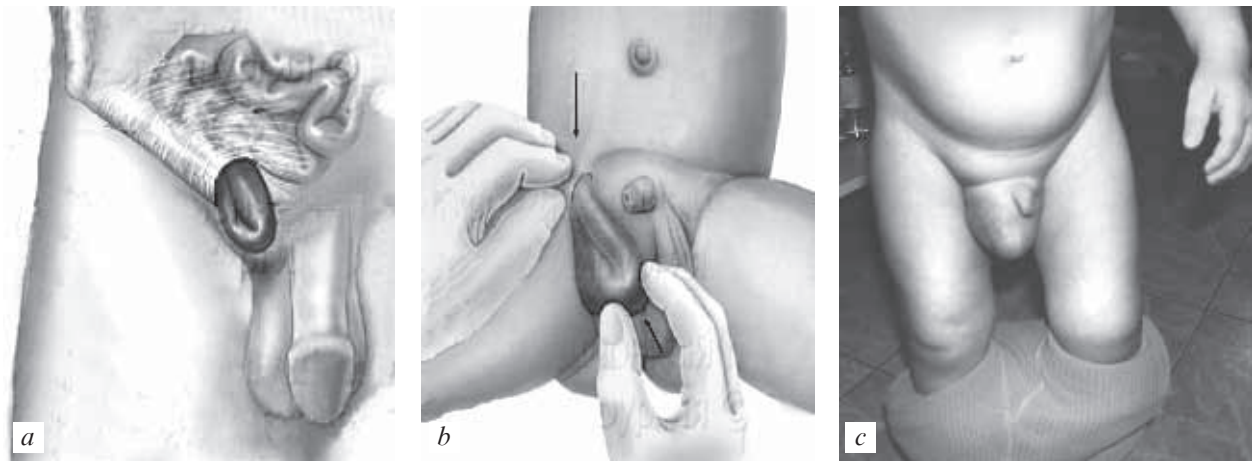


Fig. 3.4.11. Patient with inguinal hernia *(a–c)*

a gradual change in the formation: hydrops virtually disappears in the morning and increases in the evening. At palpation dropsical swelling is not elastic and can't be reseted into the abdominal cavity. At transillumination — a positive symptom of translucence. Isolated hydrops of testicle has an oval shape, more intense, with pressure does not change shape. Distinguishing the spermatic cord cyst is difficult but it is less painful, its borders are smooth, oval shaped, at pressure does not change, partial reduction into abdominal cavity enables the diagnosis.

At the differential diagnosis with cryptorchidism or testicular ectopias with hernial protrusion attention should be concentrated on the main syndrome — absence of testicle in the scrotum. Sometimes there is a need to distinguish inguinal hernia from increased lymph node of Pirogov–Rosenmüller, which is located below and under inguinal ligament, and sometimes diagnosis is carried out with tumors of inguinal area (chylangioma, dermoid cyst) and with inguinal lymphadenitis. At lymphadenitis usually on the lower limbs, in the groin or perineum one can detect inflammation, and all signs of inflammation will be available.

Complications. The most common complications — incarceration of inguinal hernia — require urgent surgery (Fig. 3.4.12).

In this case strangulation occurs within the first 3 months of life in 50% of cases. Among all cases of inguinal hernia complications are found in approximately 12% of patients, in preterm children they occur more frequently — up to 31%. Often there are no reasons for incarceration. Hernia that has been reseted earlier, stops reseting. In this case organs that were in the hernial sac, are compressed into aponeurotic ring. From the pathophysiological point of view, incarceration is developing due to the increasing swelling of organs, which are in a confined space of inguinal canal, which violates the lymphatic and venous drainage, leads to further increase in swelling. Finally, the pressure in the channel exceeds perfuse blood pressure, thus developing gangrene and necrosis of hernial content. The frequency of testicle atrophy after incarceration ranges from 1 to 11.7%, according to different authors, and during surgery testicles look cyanotic in 2.2–5% of cases. The frequency of bowel necrosis at incarceration is approximately 1.4%.

Basic clinical symptoms of strangulation are quite typical: restlessness, crying, refusal of food, vomit-

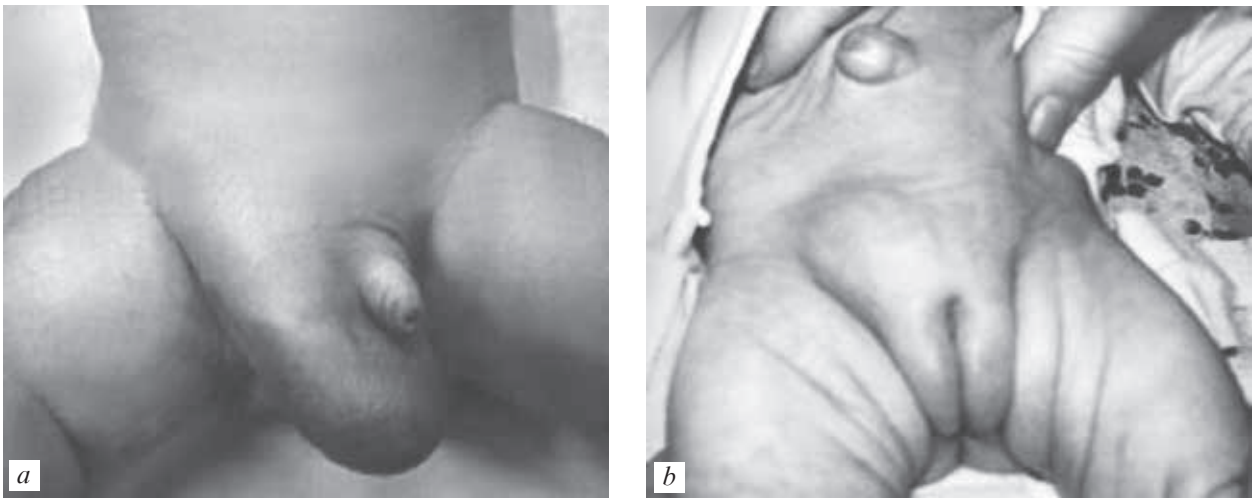


Fig. 3.4.12. Patient with inguinal hernia *(a, b)*

ing, time which parents usually indicate quite clearly. Hernia protrusion, which used to be fairly easy to reset into abdominal cavity became nonresetable, stressful and painful. Over time, the pain intensity decreases, the child becomes sluggish, increasing signs of intoxication. Swelling and reddness of the scrotum appear.

As mentioned above, inguinal hernias are the least frequent in girls than that of boys, but incarceration occurs frequently. In the hernia they are usually the uterus, which tend to rotation and necrotising quickly. In doubtful cases, the issue should be resolved in favor of surgery.

Treatment. Now the state of surgical and anesthesia services enables surgery to children of any age. In uncomplicated hernia surgery is performed after 6 months routinely after diagnosis.

In determining the indications for surgery the nature of the hernia protrusion should be taken into account. We should not allow the formation of large inguinal-scrotum hernias that violate the normal development of the child. Nonresectable hernia is an indication for surgery in the near future. The main purpose of the operation is to eliminate the connection between the abdomen and vaginal shoot. Narrowing of inguinal ring and consolidation of the anterior abdominal wall are only of secondary importance.

Today is quite common method of laparoscopic inguinal hernia correction. Multicentral studies have shown that laparoscopic incision of hernia is fairly safe at recurrence level within 3%. Note, however, that it is somewhat greater recurrence than is observed at open surgeries concerning the hernia, in which the recurrence rate is 1%. However, this level is lower than that marked among newborns (9%). The advantages of this method as compared to open herniotomy are as follows: 1) better visualization of blood vessels and reducing the risk of damage; 2) a revision of organs in case of strangulated hernia is easier to carry out; 3) giving access to the contralateral side, and at open vaginal process there is a chance to close it immediately; 4) making it possible to conduct a revision of the ovaries in girls. Disadvantages of this method are the need in endotracheal intubation and potential risk of trocar or instruments. The essence of laparoscopic correction is to ensure that from the abdominal cavity under conditions of pneumoperitoneum (10 mm Hg) with the help of laparoscope 3 mm in the inner ring of hernia Z-shaped suture with monofilament thread that does not resolve is imposed. Also examination of contralateral rings takes place and seam imposing if necessary also.

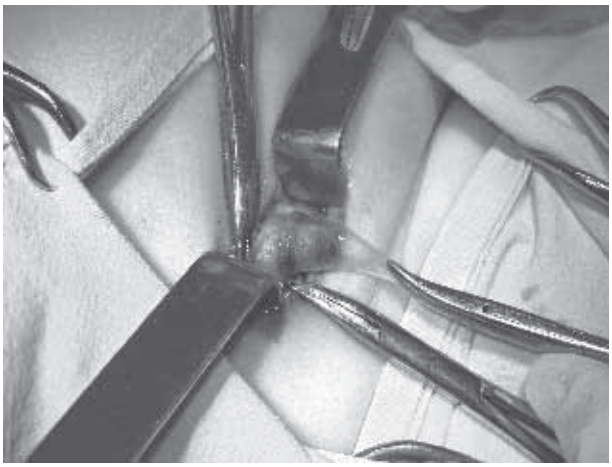


Fig. 3.4.13. Repiar of inguinal hernia

Open operations are conducted by Duhamel I and II. In infants and children up to 2 years operation is usually performed by J. Duhamel I. During the operation it is necessary to find hernial sac, allocating its cervix in the area of the external inguinal ring, followed by a blunt way separating it from the ejaculatory ducts and blood vessels. Hernial sac is open, revision of its content takes place, which must be returned to the abdominal cavity. Stitching and bandaging the cervix of the hernial sac as high as possible, then its cut. The external inguinal ring is not usually narrow. If necessary, on the legs of the inguinal rings one seam by Lambert type is imposed. At girls inguinal canal is closed tightly. In children after 2 years the principle of separation of the abdominal cavity by hernial sac can be saved, or can remove the sac completely. The most important thing is to comply certain characteristics, which are caused by a small operating field (Fig. 3.4.13).

Surgical technique in children older than 2 years is different. It is necessary making autopsy of aponeurosis of external oblique abdominal muscles. Most often the hernial sac is in the inguinal canal in the front or slightly medial to the spermatic cord. So for extracting the bag and the spermatic cord into the wound of tissue should capture them by soft clips. It is important to carefully separate the elements of the spermatic cord from the hernial sac.

After separation it is necessary to cut the bag and conduct its revision. If the content of the bag has no signs of incarceration and circulatory problems, it's returned to the abdominal cavity. Further bag in the proximal part can be sutured; binded and separates, and its distal part can be left in place. If during removal of neck of the bag stands out its bottom, in such cases it is necessary to remove the whole bag.

Usually strengthening of both the front and back walls of the inguinal canal is inexpedient even at expanding inguinal ring because the intersection of the vaginal process leads to the elimination of hernia protrusion and normalizes anatomical condition of the site.

At the incarceration of inguinal hernias it is permissible within the first 12 hours since the incarceration to conduct conservative measures aimed at creating conditions for self reduction.

With this purpose, 0.1% solution of atropine and 1% solution of promedola is introduced by counting 0.1 ml per year of life and a warm bath prescribed.

The child is placed with a raised pelvic end. Do not try to reset hernia by hands, as this may cause damage to the shells and bodies. In the absence of effect from conservative therapy for 1.5–2 hours emergency surgery is indicated. Principles of operation are

almost the same as in the planned opening of the hernia, except that the ring in which the infringement arose is opened only after revision of the hernial sac content.

One should conduct thorough revision of the contents of the hernial sac and in the absence of necrosis to dip it in the abdominal cavity. If there are doubts about the viability of the intestine, it should be warmed up using warm napkins and inject Novocain in the mesentery. In the absence of pulsation and peristalsis it is necessary to conduct resection with anastomosis imposition of "end to end". Also after insertion and separation of sac it is needed to inspect testicle to detect lesions inflicted by infringement.

Then, if aponeurosis is too weak, has slit-like defects plasty by Martynov is performed, which consists in the suturation of aponeurosis with separate seams to the inguinal ligament over spermatic cord, and the lower edge of the aponeurosis of external oblique muscle is imposed over the sutured aponeurosis, thus reinforcing the front wall of the inguinal canal.

You can also strengthen the front wall of the inguinal canal by Roux–Krasnobayev by suturing to the inguinal ligament uncut aponeurosis of external oblique abdominal muscle with underlying muscle tissues with several U-shaped sutures.

Jona showed that complications caused by inguinal hernias, both in the preoperative and in the postoperative periods are significantly reduced at earliest possible medical management.

MUSCLES' APLASIA OF ANTERIOR ABDOMINAL WALL

Prune belly, or aplasia of the muscles of anterior abdominal wall — a very rare defect that develops due to violation of lateral processes development of primary vertebrae. This defect is usually accompanied by bilateral cryptorchidism, atony of the urinary system, atresia of the rectum, incomplete reversal intestines, heart defects, poly- and syndactyly, arthrogryposis, and others Fig. 3.4.14.

Clinical picture. Abdomen is flaccid, wrinkled, 'frog' in a horizontal position and prune belly — vertically. Immediately after birth one can draw attention to a sharp increase in the abdomen and its prostration. Through sophisticated abdominal wall peristaltic loops of intestine can be seen. The testicles in the scrotum are absent. In most cases high, retroperitoneal standing of gonads is observed. The bladder is increased sharply, sometimes you can palpate stones in urethra.

Phimosis is often marked. That's why establishing the diagnosis of "prune belly" is revealed with a full urological esamination. Sometimes aplasia is marked only in the part of muscles whereas abdomen is asymmetrical.

There are five levels of muscles aplasia of the anterior abdominal wall.

1. Aplasia of the abdominal muscles without dilation of the urinary tract.

2. Megavesica, without the obstruction of the urethra; megaloureter, bladder-ureteric reflux.

3. Megavesica, megaloureter with proximal stenosis, dysplasia and reducing the urethra function.

4. Megavesica, massive bladder-ureteric reflux, pronounced ureterohydronephrosis, reduced renal function.

5. Megavesica, obstruction and stenosis of different areas.

Treatment is aimed at eliminating the malformations of the urinary system and plastic of anterior abdominal wall. There are several variants of plastic anterior abdominal wall. The most common is to create folds by suturing the inner surface of the abdominal wall, and then removal of surpluses of skin.

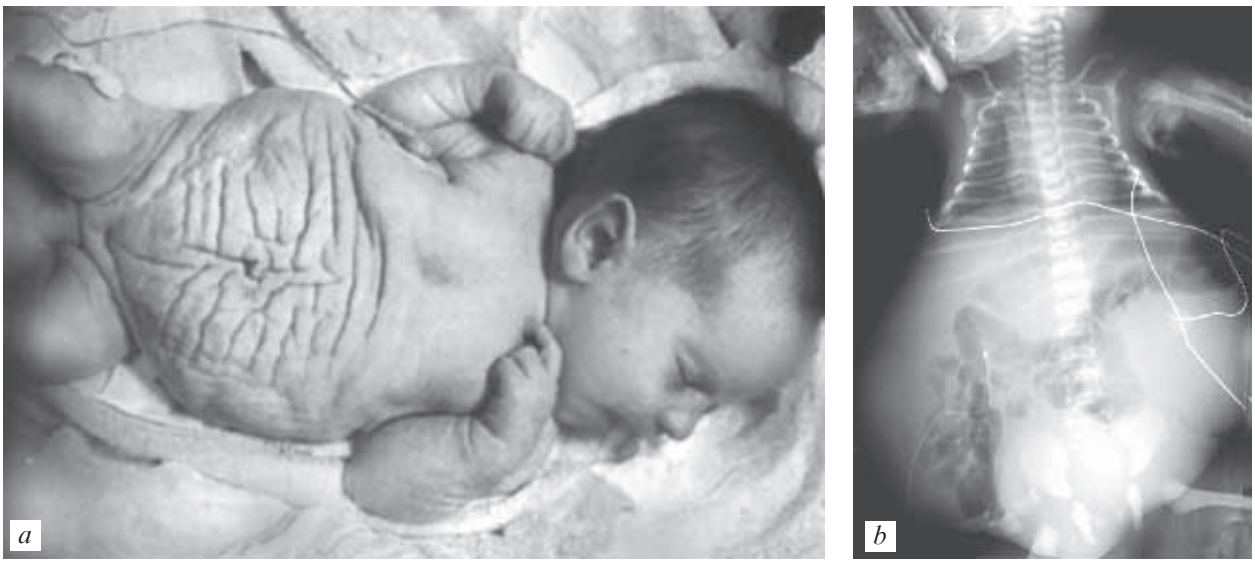


Fig. 3.4.14. Muscles' aplasia of anterior abdominal wall *(a, b)*

3.5. MALFORMATIONS OF THE URINOGENITAL SYSTEM

Anomalies of the urinary system form 33–40% of all congenital anomalies of development. Some anomalies don't threaten the child's life, course is asymptomatic and don't require treatment. Other defects lead to disrupt the flow of urine and the possibility of infection that requires immidiate diagnosis and treatment. Severe defects may be the cause of death of a child after birth or later in life.

Congenital urinary tracts are explained by complex ontogenetic development of the urogenital tract. During embryonic development of the child primary and secondary kidneys develop from meta nephrogenic tissue. Pronephrosis (Mullerian body) does not work, is reduced by the end of seven months of fetal life, male sex organs are developed from wolf's ducts and body. Secondary (metanephrosis) kidney — final kidney is formed of the caudal segments of meta nephrogenic tissue.

There are more than 100 different anomalies of urogenital system. They can be uni- and bilateral, and refer to amount, shape, location of the kidneys and urinary tract.

MALFORMATIONS OF THE KIDNEY

The anomalies of the amount, positions, relationships, values and structure of the kidneys are distinguished into several types:

— amount anomalies include agenesis and a third, added kidney;

— position anomalies are different types of dystopia of the kidneys;

— relationship anomalies are kidneys fusion. There are symmetrical and asymmetrical forms of unions. The first group includes horseshoe and galete kidneys, the second S-, L- and I-shaped kidneys;

— value and structure anomalies include aplasia, hypoplasia, a doubling of the kidney and cystic abnormalities.

AGENESIS

The absence of kidney origination occurs in 1 of 1,000 newborns. Bilateral renal agenesis occurs 4 times less than unilateral and mostly at male children (3:1). Children with agenesis of both kidneys (arenia) are unviable, born dead. However observations described fairly long survival. This feature can be explained by the child's body, when other organs function as the affected organ. The function of elimination is implemented by the liver, intestines, skin and lungs. Agenesis of the kidney is usually combined with the absence of the bladder, genital dysplasia, often with pulmonary hypoplasia, meningocele and other congenital malformations.

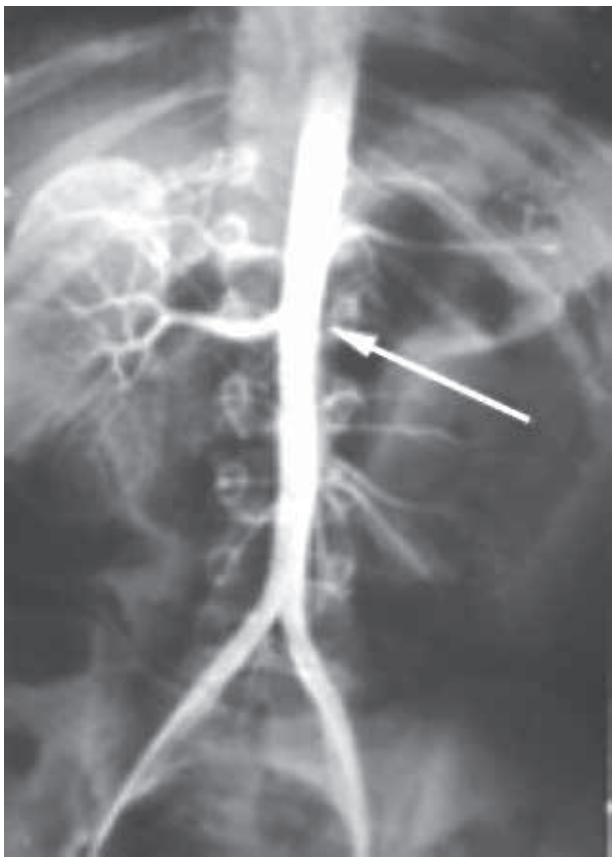


Fig. 3.5.1. Angiography: the absence of renal artery in the left

Clinical picture and diagnosis. On the one hand unilateral renal agenesis is associated with a lack of formation of nephroblastoma. Thus, as a rule, there is no corresponding ureter, hypoplasia of the half of bladder and often sexual apparatus are marked. Solitary kidney is usually hypertrophied and completely performs implementation function. In such cases, the anomaly occurs asymptomatic.

Suspection at the solitary kidney occurs during palpation of painless enlarged kidneys, but the diagnosis can be established only on the basis of X-ray urological research (excretory urography, cystoscopy, renal angiography. During the urography the contrast side of agenesis is absent. Cystoscopy reveals the absence of the ureter orifice and hemiatrophy of urocystic triangle. Angiography indicates the absence of renal artery (Fig. 3.5.1).

ADDITIONAL KIDNEY

Additional kidney is an extremely rare anomaly. Additional (third) kidney is much less that usual but has a normal anatomy. It is supplied separately with blood through arteries that branch from the aorta. The ureter falls into the bladder as an independent mouth, but can be ectopic or be connected

with the ureter of the main kidney. There are cases of the blind end of the ureter.

Additional kidney must be distinguished from the upper segment of the double kidney. The difference is that a doubling of the collecting system of the lower segment of the kidney is represented by two large renal calices, and of the top segment — by one. Segments of the doubled kidney form a indissoluble circuit of parenchyma. In case of additional kidney its parenchyma is separated from main kidneys and collector system contains three renal calices, like the main, only in miniature.

Clinical picture and diagnosis. Additional kidney has clinical significance only when ectopic ureter orifice (permanent incontinence) or as a result of an inflammatory lesion, tumor or other pathological process. Diagnosis can be made on the basis of excretory urography, retrograde pyelography, angiography. *Treatment* of diseases of the kidneys consists of nephrectomy because of low functionality of the organ.

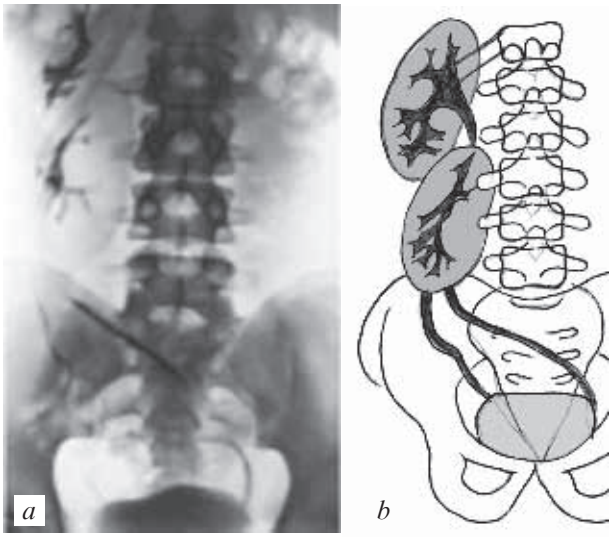


Fig. 3.5.2. Cross dystopia *(a, b)*

DYSTOPIA

Under this title understand the unusual location of the kidneys due to the disruption in the process of embryogenesis lowering. Frequency of anomalies is approximately 1:800. Dystopia of the kidney is more common in boys. Since the processes of subsidence and rotation are interdependent and dystopic kidney turned out, the lower the dystopia is, the ventrally pelvis is located. Dystopic kidney often has a loose type of blood supply, its blood vessels are short and limit kidney's displaceability. Functional conditions of dystopic kidneys are usually reduced. Typically kidney has a lobe-shape structure. Its shape can be various — oval, pear-like, flat and irregular.

There are distinguished three types: high, low and cross dystopia. Intrathoracic kidney refers to high dystopia. It is a very rare anomaly. The amount of its descriptions in the world literature do not exceed 90. During intrathoracic dystopia, kidney becomes a part of a diaphragmatic hernia. Ureter is lengthened, falls into the bladder. The types of low dystopia are lumbar, iliac and pelvic. During lumbar dystopia little interiorly rotated pelvis is at level of the IV lumbar vertebra. Renal artery usually deviates above the aortic bifurcation. Kidney moves are limited. Iliac dystopia is characterized by more severe rotation of the pelvis forward and its positions at level L_V–S_I.

Its displacement is medial in comparison with the lumbar dystopic kidney. Renal arteries are usually multiple, moving away from the common iliac artery or aorta at the site of bifurcation. There is almost no mobility of the kidney during changing the body position. Pelvic kidney is located on the center line at the bifurcation of the aorta, a bladder and a little above it. It can have various forms. It is usually hypoplastic in differend degree. Vessels of the kidneys which have loose types are branches of the common iliac or different pelvic arteries. A combination of pelvic dystopia with lumbar or iliac dystopia of contralateral kidney is possible.

Cross dystopia is characterized by the contralateral shift of kidneys (Fig. 3.5.2). As a rule two kidneys are fused and form S- or I-shaped kidney. Ureter of the dystopic kidney falls into bladder in the usual place. Vessels of the kidney deviate below normal with uni- or contralateral side.

The frequency of cross dystopia of kidney is 1:10,000 — 1:12,000.

Clinical picture and diagnosis. During dystopia of kidney clinical picture is caused by abnormal organ position. The main symptom is pain, which occurs while changing organ position, physical exertion, and flatulence. During cross dystopia pain is localized in the iliac region and radiating to the groin opposite side. As far as dystopic kidney is affected by pathological process (hydronephrosis transformation, pyelonephritis) more often than usually located kidney, often the symptoms of these diseases are joined. Hilar dystopia can simulate mediastinal tumor by clinical manifestations and data of X-ray. During the lumbar and iliac dystopia, kidney is palpated as slightly painful sedentary formation. Dystopia is revealed with excretory urography, and in case of a sharp decline of the kidney function — with retrograde pyelography. Characteristic sign of dystopia is the rotation and the unusual location of the kidney with limited iliac. Sometimes difficulties in the differential diagnosis of lumbar and iliac dystopia and nephroptosis arise, especially in the case of fixed nephroptosis, which is characterized by low localization and small displaceability of kidneys. However, at a fixed urogram with nephroptosis the location of the medial pelvis and long tortuous ureter can be noted. Sometimes only renal angiography helps to distinguish this condition, which reveals short vascular pedicle during dystopia and elongated — during nephroptosis.

Treatment. Management during kidney dystopia is maximum conservative. The operation is carried out during dystopia, complicated by hydronephrosis or calculus. In case of death of dystopic kidney, nephrectomy is performed. Operative movement of

the kidney is extremely difficult because of the loose type of blood supply and small caliber of vessels.

FUSION OF THE KIDNEYS

Fusion of the kidneys is about 13% of all renal anomalies. Symmetrical and asymmetrical forms are distinguished. Horseshoes and galete-shape belong to the first type, S-, L-and I-shaped kidney — to the second.

With **horseshoe abnormality** kidneys fuse with the same ends, renal parenchyma has the form a horseshoe. The anomaly is associated with impaired development of lowering and rotation of kidneys. Horseshoe kidney is situated below normal, renal pelvis are directed anteriorly or laterally. Usually blood supply is accomplished by multiple arteries that branch from the abdominal aorta and its branches. More often (98% of cases), the lower ends of the kidneys are fused. At the junction of the renal isthmus connective tissue or complete renal parenchyma is presented, which may have a separate circulation. The isthmus is located in front of the abdominal aorta and inferior vena cava, but can be placed between them or behind them.

Anomaly occurs in infants with a frequency of 1:400 — 1: 500, and boys 2.5 times more often than girls. Horseshoe kidney is often combined with other anomalies and malformations. Dystopic location, poor motility, abnormal discharge of the ureters and other factors contribute to the fact that the horseshoe kidney traumatizes easily.

Clinical picture and diagnosis. The main clinical sign of a horseshoe kidney is a symptom of Martynov–Rovsing, whish is causing pain when bending of the body. The appearance of pain attack is associated with compression of blood vessels and the aortic plexus by renal isthmus. Often the pain is vague and accompanied by dyspepsia. Horseshoe kidney can be determined by deep palpation of the abdomen as a dense sedentary formation. With good preparation intestine kidney is visualized as a horseshoe by X-ray. Most clearly contours of the kidney are defined during angiography in a phase of renogram.

On excretory urograms horseshoe kidney is characterized by rotation of pyelocaliceal system and changing of the angle, formed by the longitudinal axes of the kidneys. If this angle is normally open from to bottom, then during the horseshoe kidney — upwards. Shadows of the ureters depict "a flower vase": moving away from the pelvis, ureter go to the sides, and then on the way to the bladder gradually converge.

Treatment. During horseshoe kidney operation is performed only with the development of complications (hydronephrosis, stones, tumors, etc.). To determine the nature of the blood supply before surgery renal angiography is advisable. **Galete** kidney is a flat formation, located at promontory or below, formed by fusion of the two kidneys with both ends to the beginning of their rotation. Blood supply of galete kidney is implemented by multiple vessels that branch out from the bifurcation aorta. Pelvis is located anteriorly, ureters are shortened. Anomaly occurs with a frequency of 1:26 000.

Diagnosis is based on data from abdominal palpation and rectal digital examination, as well as on the results of excretory urography and renal angiography.

In the case of **S- and I-shaped** kidney longitudinal axis parallel to the fused kidneys, and the axes of the kidneys, that form an L-shaped kidney, perpendicular to each other. Pelvis of S-shaped kidneys are looking in opposite directions.

I-shaped kidney arises from the dystopia of one kidney, often rights to the opposite side. During this kidneys fuse, forming a single column of the renal parenchyma with pelvis, located medially. Fused ectopic kidneys may compress surrounding organs and great vessels, causing ischemia and leading to disease.

Diagnosis. Anomalies are identified with excretory urography and renal scan. If you need surgery (removal of stones, plastic), a renal angiography is conducted. Operations on the fused kidneys are technically difficult because of the blood supply problems.

RENAL APLASIA

Renal aplasia is a severe degree of underdevelopment of kidney's parenchyma, which can be combined with the absence of ureter. The defect is formed in the early embryonic period, before the formation of nephrons. There are two forms of renal aplasia — great and little. During the first form, kidney is presented by the lump of fibrolipomatosis tissue and small cysts. Nephrons are not defined, ureter is absent. The second form of aplasia is characterized by fibrocystic mass with a small number of functioning nephrons. Ureter is refined, has the opening, but can not reach the renal parenchyma, resulting blind. Such kidney has no pelvis and formed renal pedicle. Frequency of renal anomalies ranges from 1:700 to 1:500. In boys it is more common than in girls.

Clinical picture and diagnosis. Kidney aplasia doesn't reveal clinically and diagnosed during contralateral renal disease. Some patients complain of pain at the side or stomach, which is associated with compression of the nerve by fibrous tissue. Detection of renal aplasia is based on data from X-ray and imaging studies. On X-ray in a few cases on the spot of the renal aplasia, cysts with calcareous walls are determined. Against the background of the air, injected retroperitoneally, with good intestinal preparation kidney is visible in the tomograms as a small lump. During aortography arteries are not defined.

Aplasia needs to be differentiated from non-functioning kidney, renal agenesis and hypoplasia. Retrograde pyelography and aortography helps to distinguish kidney that lost function in result of pyelonephritis, tuberculosis or any other process.

Agenesis is characterized by the absence of marker of renal parenchyma. Herewith urogenital apparatus does not develop: ureter is absent, represented by band of fibrous tissue or ends blind, there is hemiatrophy of vesical triangle, testicle is absent or not descended. Cystoscopy helps differential diagnosis, which reveals the opening of the corresponding ureter during kidneys in aplasia in the half of the cases.

Hypoplastic kidney differs from aplasia by a functioning parenchyma, the ureter, which is defined throughout, and is visualization of vascular pedicle during aortography.

Treatment. The need of therapeutic measures in renal aplasia occurs in three cases:

1) a pronounced pain in the kidney area,
2) the development of renal hypertension,
3) at reflux to hypoplastic ureter.

Treatment consists of performing the removal of the kidney and ureter.

KIDNEY HYPOPLASIA

Hypoplastic kidney macroscopically is a normally formed organ in a miniature. On a section cortical and medullary layers are well-defined. Histologically, there are three forms of hypoplasia:

1) simple hypoplasia;
2) hypoplasia with oligophronia;
3) hypoplasia with dysplasia.

A simple form of hypoplasia is characterized only by reducing the number of renal calices and nephrons. During the second form reducing the number of glomerules is coupled with the increase of their diameter, fibrosis of interstitial tissue, dilatation of tubules. Hypoplasia with dysplasia is conducted by the development of connective tissue or muscle sleeves around the primary canals. This form of hypoplasia, in contrast to the first two, is often accompanied by abnormalities of the urinary tract.

Clinical picture and diagnosis. Unilateral hypoplasia may not show all its life but it is noted that hypoplastic kidney often manifested as pyelonephritis and is a source of development of renal hypertension.

Bilateral renal hypoplasia is detected early — in the early years and weeks of life. Such children retard in growth and development. Often there is pallor, vomiting, diarrhea, fever, signs of rickets. A decrease in the concentration of renal function is marked. However, the results of biochemical studies of blood for a long time still remain normal. Blood pressure rises only with the development of uremia. The disease is often complicated with pyelonephritis. Most children with severe bilateral renal hypoplasia die of uremia within the first years of life.

Unilateral hypoplasia is detected by X-ray examining pyelonephritis. Reduction in the size of kidneys with good contrast to the system is marked on excretory urograms. The contours of the kidney can be different, pelvis moderately dilated (Fig. 3.5.3).

During hypoplastic kidney calyx is not deformed as during pyelonephritis, but only reduced their number and volume. Compensatory hypertrophy of the contralateral kidney is marked on urograms. Renal angiography makes a great help in the differential diagnosis. During hypoplasia arteries and veins are uniformly refined, while in the secondary contracted kidney angiogram looks like a picture of charred wood. Kidney biopsy may also be useful, but in practice its value in diagnostics during hypoplasia is limited.

Treatment. In the case of unilateral hypoplasia complicated pyelonephritis and hypertension, treatment is reduced to nephrectomy. In bilateral hypoplastic kidney which is complicated with severe renal insufficiency, the patient may be saved only by bilateral nephrectomy with subsequent kidney transplantation.

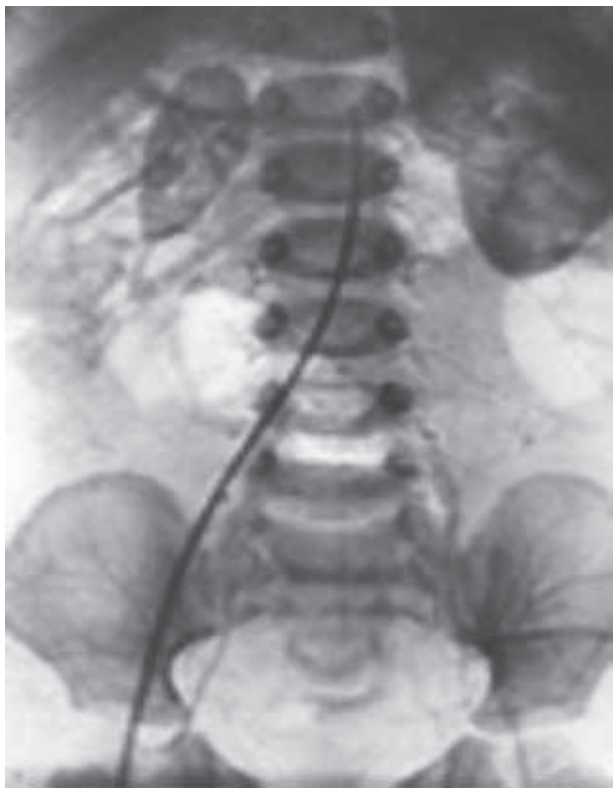


Fig. 3.5.3. Renal hypoplasia on the right

CYSTIC ABNORMALITIES

Cystic kidney abnormalities are observed at 1:250 but more often diagnosed in adulthood. The most common of cystic lessions is polycystic renal disease. Polycystic renal disease (polycystic degeneration, cystic disease) — genetic abnormality, which affects both kidneys (Fig. 3.5.4).

Cystic disease, which occurs in adulthood, is transmitted with autosomal dominant and monomeric gene and the so-called children malignant polycystic renal disease — by recessive type.

Development of polycystic renal disease is associated with the violation within the first weeks of embryogenesis, leading to non-union metanephrosis tubules with collecting duct of ureter anlage. An important role is played by insufficient blood supply to the renal parenchyma. Cysts are divided into glomerular, tubular and excretory. Glomerular cysts have no connection with the tubular system and therefore do not increase. They are found in newborns. Early development of renal failure is typical, which leads to rapid death of the child. Tubular cysts are formed of meandering canals and excretory — from collecting ducts. These cysts are uneven, constantly increasing due to the difficulty of discharge.

Among children with polycystic renal disease 5% of the common cystic changes occur in the liver, 4% — spleen, rarely in lungs, pancreas, and ovaries.

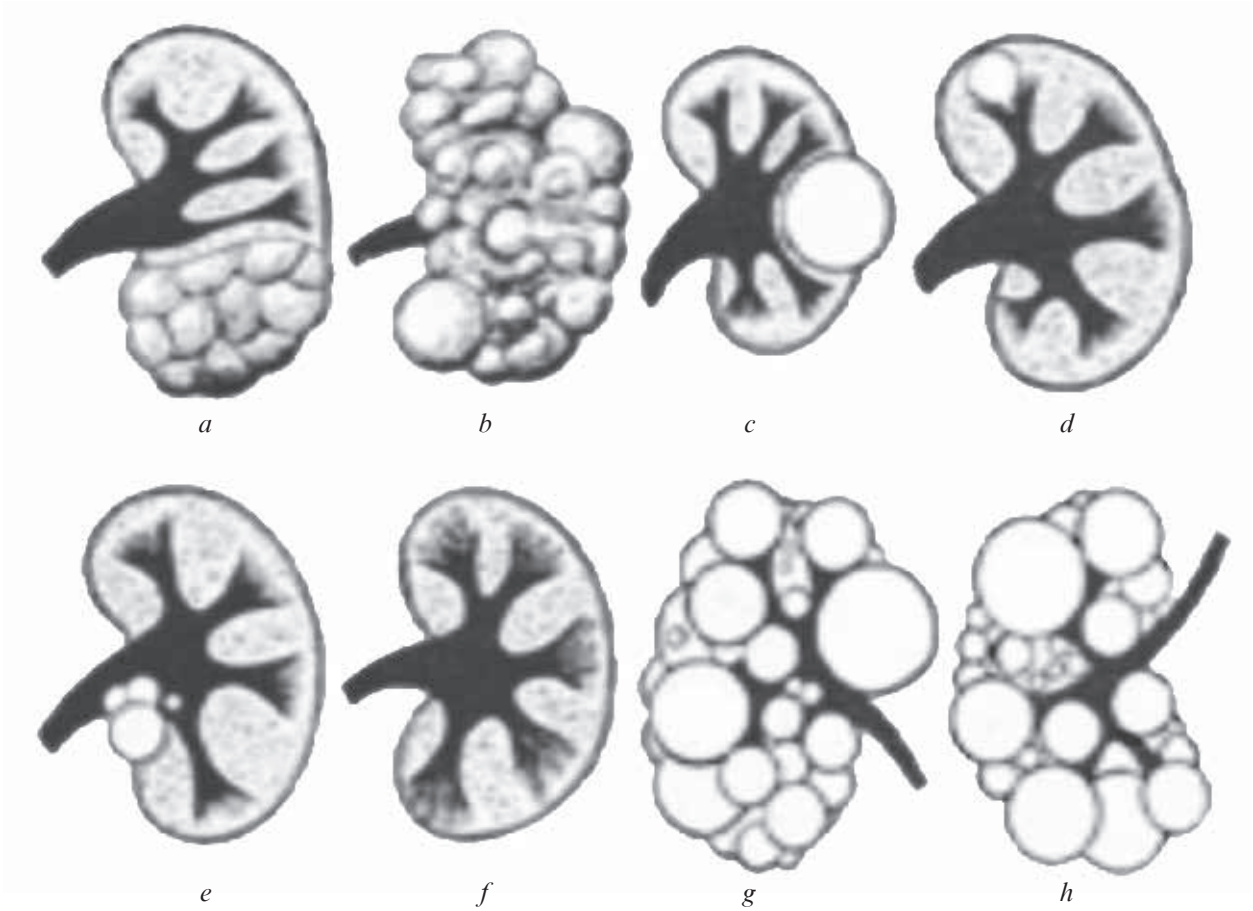


Fig. 3.5.4. Cystic renal disease *(a–h)*

Clinical picture and diagnosis. Polycystic renal disease in different age groups is displayed in different ways. In young children polycystic renal disease is often complicated by severe pyelonephritis, which quickly leads to anuria and uremia. In elder children the disease is accompanied by a dull ache in the lower back, periodic hematuria and hypertension (70%). Polyuria and nocturia are marked. Enlarged hummocky kidneys are determined by palpation. In such cases it is important to find a family anamnesis that will facilitate diagnosis. The addition of pyelonephritis is characterized by corresponding changes in the urine, which leads to progressive kidney failure.

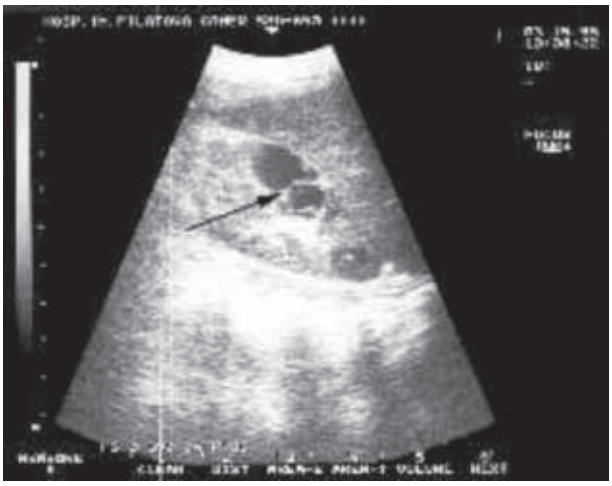


Fig. 3.5.5. Renogram

Polycystic renal disease is detected by excretory urography, renal scan and angiography.

Enlarged kidneys, elongation and spreading of the cervix of the renal calices with bulb-shaped deformations are defined on urograms during stored function. Scanning detects defects and increase of the kidney and parenchyma due to uneven uptake. Angiography determines depletion of vascular net, spreading of the arterial branches. Renogram has a spotted view (Fig. 3.5.5).

The differential diagnosis spends with other cystic lesions of the kidney and William's tumor. It should be remembered that polycystic disease is always bilateral, unlike the above-mentioned diseases, which often affect one kidney. Angiography allows distinguishing polycystic disease from William's tumors, which reveals the increase of only one kidney area and its increased vascularization with the presence of a tumor.

Treatment. Aim of the treatment is struggle with pyelonephritis, hypertension, and correction of water and electrolyte balance. Surgical intervention is necessary in renal bleeding profusely, obturate stone, pyonephrosis or development of malignant kidney tumor.

With terminal stage of renal failure chronic homotransplantations and kidney transplantation are using.

The prognosis of cystic disease is unfavorable. Patients rarely live more than 10–12 years after the onset of clinical symptoms, although there are cases of prolonged (up to 70 years) survival in the benign course of the disease.

Sponge kidney (sponge kidney with pyramids) — a rare hereditary congenital anomaly in which collecting tubes pyramids are cystic dilated. Anomaly is clinically manifested if complications of nephrocalcinosis or pyelonephritis are not developing. However, half of the carriers of the anomaly and in uncomplicated cases have constant moderate proteinuria and microhematuria.

The diagnosis is established on the basis of excretory urography for typical features ("bouquet" in the area of the pyramids).

Treatment. In the absence of complications the problem does not require treatment.

Multicystic dysplasia. Anomaly, in which one or rarely both kidneys are replaced by cystic cavities and have no parenchyma, is called multicystic dysplasia. Ureter is absent or rudimentary. Sometimes the testicle or an appendage of the appropriate side joins the kidney. Bilateral anomaly is incompatible with life. During a unilateral lesion complains arise only if the cyst grows with compression of surrounding organs, causing nephrectomy. A solitary cyst is a solitary cystic formation of round or oval shape, which comes from the renal parenchyma and rises above its surface. Dermoid cyst is very rare and contains derivatives of the ectoderm: the hair, sebaceous mass, teeth.

Clinical picture and diagnosis. Main signs of solitary cyst are a dull pain in the kidney area and transient hematuria. The temperature rises and pain increases with festering cyst. In some cases, the disease is complicated with pyelonephritis and hypertension.

The diagnosis is established by urography: crescentic defect of pelvis or calyx and apart of the cervix of the calyx is detected. Treatment consists of husking cysts if localization permits or in the detection and plugging its cavity perirenal fat.

Prognosis at the long-term term after surgery is favorable.

DOUBLING OF KIDNEYS

It is the most common abnormality of the kidneys that occurs in 1:150 newborns, with girls twice as often as boys. It can be single or double-sided. The anomaly is associated with splitting ureteric anlage early or on the way to the growing into a nephrogenic blastema.

The upper end of the kidney is about 1/3 of the renal parenchyma, drained by upper group of renal calices, which flow into a separate pelvis. Lower and average, and lower groups of renal calices flow in the lower segment of the pelvis. In about half the cases, each segment has an isolated double kidney blood flow from the aorta. Ureters, which set out from the double kidney pelvis, pass by and flow into the bladder or separately, or merge into one barrel at one or another level. During the confluence the deal is about their not full doubling. This condition can lead to ureteroureteral reflux associated with no synchronous contraction and relaxation of the branches of the ureter (Fig. 3.5.6).

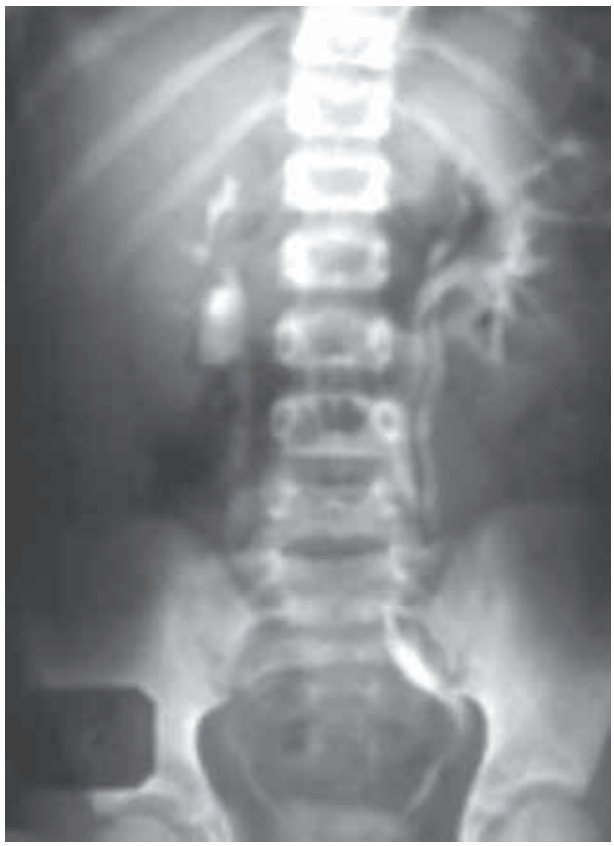


Fig. 3.5.6. Doubling of kidneys

Uretroureteral reflux is a functional barrier that contributes to stagnation of urine, the development of pyelonephritis.

At full doubling of the ureter the main trunk, which takes out from the lower segment of the double kidneys, opens at the mouth of vesical triangle, and the second — near or distal (law of Weigert — Meyer).

Clinical picture and diagnosis. Doubling the ureters in some cases becomes the cause of vesicoureteral reflux because of inferiority of locking mechanism of the mouth. Reflux occurs more frequently in the lower (main) segment of the doubled kidney. The anomalous structure of the kidney and ureter promotes various acquired diseases (approximately 30%), among which pyelonephritis is at the first place. The corresponding ureter is significantly extended, enlarged, and tortuous (megaureter).

The man with the double kidney can live a long life without any complaints and clinical manifestations. Anomaly usually occurs in the examination about pyelonephritis.

Treatment. Operative treatment of renal and ureteric doubling is conducted in the following cases:

— with full anatomical and functional degradation of one or both segments of the kidney (nephrectomy or heminephrectomy is performed);

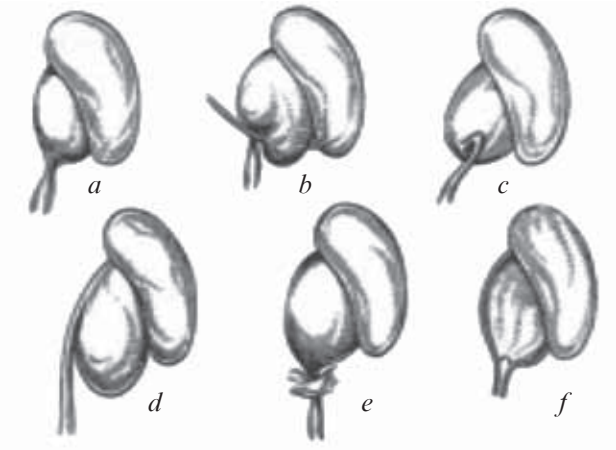


Fig. 3.5.7. Causes of hydronephrosis *(a–f)*

— during reflux in one of the ureters (ureterouretero- or pyeloanastomosis is imposed, if there is reflux, antireflux surgery is performed);

— in the presence of ureterocele its excision is performed with ureter neoimplantation into the bladder.

HYDRONEPHROSIS

The progressive extension of the pelvis and calyces — hydronephrosis transformation is resulting from the breach outflow of urine at the site of ureter pelvic segment.

Reasons that cause hydronephrosis may be anatomical, which include:

— stenosis of ureter pelvic junction,

— embryonic tissue bands and adhesions,

— fixed inflection of the ureter,

— high-discharge of the ureter,

— aberrant vessel, which moves junction — ureteric segment,

— valve ureter,

and functional caused by:

— dysplasia, muscle and nerve elements of the wall segment,

— impaired patency of peristaltic waves in it (Fig. 3.5.7).

The most common cause of hydronephrosis in childhood is stenosis of ureter pelvic junction (Fig. 3.5.7, *a*). Its occurrence is associated with impaired development of recanalization of the ureter in embryogenesis.

Consequence of antenatal fetal inflammation are bands and adhesions (Fig. 3.5.7, *b*), which compress it from the outside or are the cause of a fixed inflection. For some patients difficulty of emptying the pelvis is associated with additional aberrant vessels (Fig. 3.5.7, *e*), which constant pulsation can lead to sclerotic changes in the wall of the ureter and a violation of its patency. The high discharge of the ureter (Fig. 3.5.7, *d*) is the result of congenital anomalies, resulting in the dilatation of the lower part of calix.

One of the causes of hydronephrosis are valves of the ureter (Fig. 3.5.7, *f*), which are located at the site of ureter pelvic junction and are fold mucosa (mucous valves), or they contain all the layers of ureter.

Whatever the reasons, which cause the development of hydronephrosis, its pathogenesis is the same in all cases. The delay of urine in the pelvis due to the complicated outflow causes ischemia and gradual atrophy of the renal parenchyma. The speed of this process depends on the degree of obstruction and the type of pelvis. During intrarenal pelvis this process occurs more rapidly. In the presence of an obstacle to the outflow, pelvis copes with the function of the output of urine through the working muscle hypertrophy, then its atony comes, pelvis is very extended, renal calices increase and get coin-shaped forms, their walls are greatly enhanced.

It's interesting that even with severe block the kidney remains employable for a long time. Increased pressure in the pelvis leads to the return of urine from the renal pelvis into the tubules (tubular reflux). Fornical zones might be broken during severe ureteric obstruction, while urine is introduced into the interstitial space, where are spread by venous and lymphatic vessels. At the same time, pielorenal reflux leads to poor blood supply to the parenchyma and its replacement by scar tissue. Stasis of urine and organ ischemia promotes the accession of pyelonephritis, which occurs in 87% of patients.

Clinical picture and diagnosis. The main clinical manifestations of hydronephrosis are pain, changes in the urine and symptom of palpable tumor in the abdomen. Pain syndrome is seen in 80% of patients. Pain has different nature, from the dull aching to attack of colic. The frequency and intensity of pain is associated with the accession of pyelonephritis and/or distortion of the renal capsule on the background of the severe disruption of the flow of urine. As usual, pain is localized in the umbilical region, while elder children complain of pain in the lumbar region. The changes of urinalysis are characterized by pyuria and bacteriuria (during accession of pyelonephritis) or hematuria.

The symptom of palpable tumor is a common clinical manifestation of hydronephrosis, especially of small children with poorly developed abdominal wall. Formation is usually determined by a doctor occasionally during the palpation of abdominal wall. Sometimes it is determined by the parents, which is the reason for seeking medical attention. Tumor formation has clear contours, fluid, supple texture, located at the navel or above on the right or left side of the abdomen.

The main methods of diagnosis of hydronephrosis in children are ultrasound, excretory urography, radionuclide study and renal angiography. Renal enlargement is determined by the ultrasound due to the dilatation of its collector system and the induration of parenchyma. Ureter is visualized. The next step is to carry out studies of excretory urography. Typical X-ray picture of hydronephrosis: the dilatation and deformation of coin-shaped renal calices, their isthmus, expanding pelvis. It should be noted that while doing the excretory urography in pa-

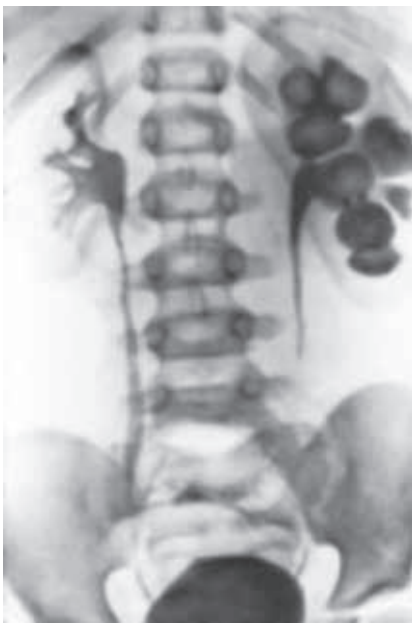


Fig. 3.5.8. Hydronephrosis transformation of the left kidney

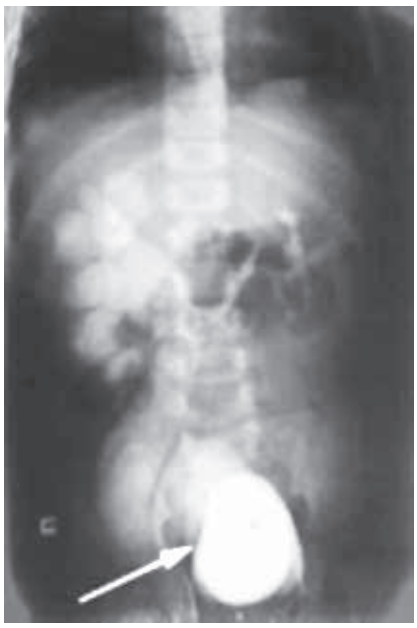


Fig. 3.5.9. Megaureter

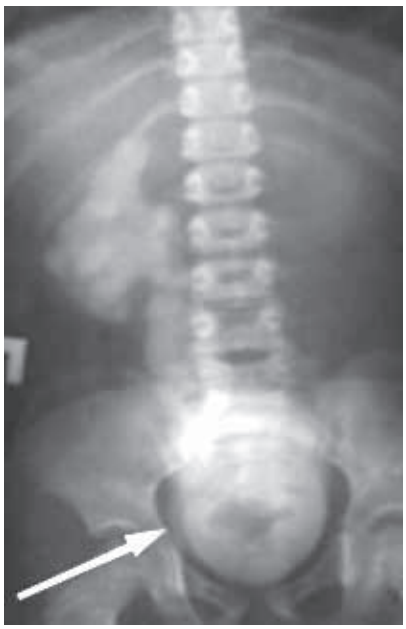


Fig. 3.5.10. Reflux megaureter

tients with suspected hydronephrosis needs implementation of delayed X-rays to get a clear picture on the background of decreased kidney function, and a large volume of the collector system of the kidney. Ureter with hydronephrosis is rarely determined. It is narrow, contrasted by cystoid type, its position is normal (Fig. 3.5.8).

Radionuclide study helps to assess the degree of preservation of renal function and to determine the treatment strategy.

Treatment of hydronephrosis is only surgical. Indications for surgery are determined after confirmation of the diagnosis. Volume of surgery depends on the preservation of renal function. If kidney function is reduced slightly, reconstructive plastic surgery is performed — resection of modified ureter pelvic junction with further pyelitis ureterostomy (operation by Hynes–Anderson–Kucera).

If the changes of renal function are irreversible, there is the question of nephrectomy. Postoperative prognosis mostly depends on the degree of preservation of renal function and activity of pyelitis-nephrotic process.

Clinical supervision of children after surgery for hydronephrosis is jointly urology and nephrology. Control X-ray is performed at intervals of 6–12 months. Good cross ureteropelvic junction and no exacerbation of pyelonephritis for 5 years can remove the child from the hospital registration.

MEGAURETER

Megaureter is a significant dilatation of the ureter, caused mechanically. Depending on the cause of development there are such types of megaureter: obstructive, reflux and achalasia ureter (Fig. 3.5.9).

Obstructive megaureter develops in the presence of stenosis at the site of the ureter or ureterocele. Violation leads to emptying its substantial dilatation and megadolichoureter, dilatation of the collector system of kidneys, the rapid emergence of urethritis and pyelonephritis. Chronic renal failure develops early during bilateral process.

Reflux megaureter proceeds not so hard, but over time reflux causes the development of reflux nephropathy, growth of the kidney, sclerotic changes of renal scarring of parenchyma. Pyelonephritis accelerates scarring process of the kidney.

Achalasia of ureter is characterized by its local dilatation, limited by the distal or middle cystoids without dilatation of the pelvis and calyces. Its reason is the immaturity of neuromuscular ureteric wall structures that tend to mature, which can lead to self-healing.

Clinical picture and diagnosis. The display of megaureter is due the course of chronic pyelonephritis. Parents report weakness, pallor, retarded growth of the child, unexplained lift of temperature. Urine is sometimes muddy; there are pyurias, and bacteriuria, cystitis and sometimes red blood cell in the analysis. During exacerbations frequent and painful urination appear. On excretory urograms there is a delay of allocation of contrast material by the kidney, the deformation of the collector system, dilatation and sinuaty of ureter. Evacuation of contrast matter is slow (Fig. 3.5.10).

However, one third of patients, especially during bilateral megaureter, can not establish a satisfactory passage of urine, and many of them are candidates for kidney transplantation.

MALFORMATIONS OF THE URINARY DUCTS

Urachus is a tubular formation, which comes from the top of the bladder and is sent to the umbili-

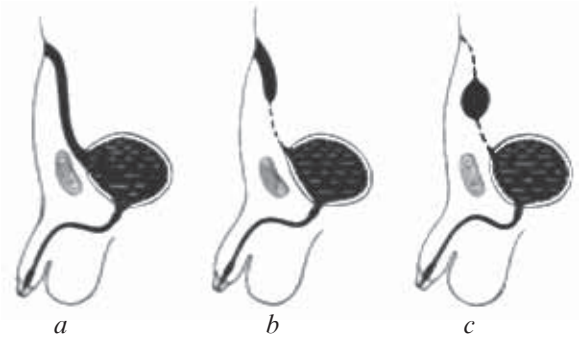


Fig. 3.5.11. Malformations of urahus

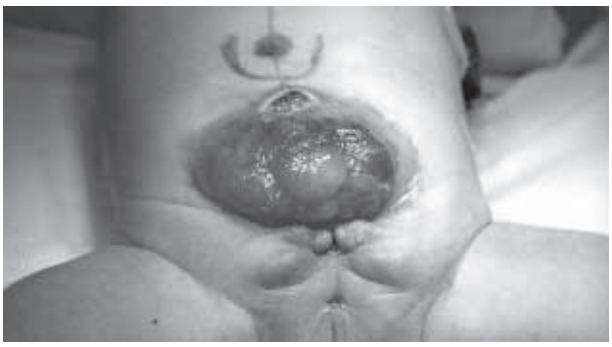


Fig. 3.5.12. Bladder extrophy

cus between the peritoneum and the transverse fascia of the abdomen. In an embryo, it serves to drain the primary urine in the amniotic fluid (Fig. 3.5.11).

On the 4–5 months of fetal development, urachus obliterates, turning into the median umbilical ligament. However, in some cases, it can be left open at the time of birth, and obliteration occurs within the first year of life. At a delay of obliteration urachus may remain open throughout (vesicoumbilical fistula) or in some areas.

Vesicoumbilical fistula is the most common abnormality of the urinary duct. Clinically, it appears by the release of urine from the navel, cystitis. The continued existence of the fistula may be complicated with pyelonephritis and lithogenesis in the bladder.

The diagnosis is confirmed with a test of indigo carmine. The dye of the fluid is injected along the urethra into the bladder or intravenously: urine, released from the fistula, is colored. Indigo carmine test can be introduced into the fistula, then colored (blue toned) urine comes from the urethra. Diagnosis is confirmed by X-ray at cystography or fistulography.

Treatment consists of excision of the urinary duct.

Urachus cyst is the second abnormality by frequence. The content of the cyst is mucus and serous fluid. For a long time the cyst remains very small and not clinically manifested. Sometimes it can be palpated by suprapubic midline. In the case of a sharp increase, the cyst may compress the bladder, causing dysuric phenomenon. It tends to fester, manifested by fever, pain, abdominal wall tension, redness and swelling of the tissues below the navel.

Differential diagnosis of cysts of the urinary duct is carried out with a diverticulum of the bladder, abdominal wall hernia, umbilical cyst (omphalocele). Cystography, ultrasonography and CT scan help to clarify the diagnosis.

Treatment of the cyst, that causes dysuric phenomenon, is operative excision. During the abscess it is opened and drained.

Umbilical fistula (incomplete) appears with soak of the navel, signs of inflammation (omphalitis) and pus from the umbilical ring. During poor draining of the fistula fever and intoxication appear. Gradually, granulation tissue is growing in the navel area.

For solving the question about connecting of obliterated part of urinary duct with the bladder, after decrease of inflammation, fistulography is made.

Treatment consists of daily baths with potassium permanganate, processing navel with 1% solution of brilliant green and cauterization of granulation 2–10% silver nitrate. If the conservative measures are ineffective, radical removal of the urinary duct is made.

MALFORMATIONS OF THE BLADDER

Diverticulum of the bladder is formed due to incomplete obliteration of the urinary duct. It is asymptomatic for a long time and there is a chance to find it accidently on the cystography, which is done during dysuria and pyuria. There are front pouch mainly in boys and often combined with bladder infravesical obstruction.

Treatment involves removing the diverticulum. At the same time the bladder infravesical obstruction is eliminated.

Bladder extrophy is a severe malformation, which is conducted as a congenital absence of the anterior wall of the bladder and the corresponding section of the anterior abdominal wall. Bladder extrophy is always accompanied by a total epispadias and divergence of bones of the pubic symphysis. Anomaly occurs in 1 by 40,000–50,000 newborn boys, more often than girls (Fig. 3.5.12).

Formation of bladder extrophy occurs on 4–7 weeks of intrauterine development.

Permanent incontinence, severe deformity of external genitalia, absence of abdominal wall above the split-bladder causes severe physical and mental suffering for both patients and their parents, and are the main complaints to see a doctor.

Clinical picture and diagnosis. The clinical picture of bladder extrophy is quite specific: through a rounded abdominal wall defect falls bright red mucous membrane of the posterior wall of the bladder. Umbilicus is located above the top edge of the defect. The mucous membrane of the bladder is very vulnerable, often covered with papillomas growths and bleeds easily. Diameter of vesical plate is 3–7 cm. Mucosa scars over time. The isthmus of the ureter is opened in the lower plate of bladder or hidden between the coarse folds of mucous membrane. Urine flows continually, causing maceration of the skin of abdominal wall, the inner thighs and peri-

neum. In boys the penis is shortened, pulled up to the anterior abdominal wall, split urethra contacts with the mucous membrane of the urinary bladder. Scrotum is underdeveloped, cryptorchidism is often observed. Girls with splitting of the urethra have clitoral splitting, adhesions of large and small labia. Anus is anterior ectopic.

Often bladder extrophy is combined with inguinal hernia, fallout of the rectum, malformations of the upper urinary tract. Direct contact with the external environment ureter leads to rising pyelonephritis. For patients with bladder extrophy "duck" gait is typical because of instability of pelvic ring.

Treatment of bladder extrophy is only surgical. To avoid joining the ascending pyelonephritis, surgery, if the child's condition permits, must be made within the first 3 months of life. In the future, it makes social adaptation of the child easier, because it saves from urinary incontinence.

Patients, operated for bladder extrophy, are in need of constant supervision of nephrologists and urologists. Under medical check-up main attention should focus on the assessment of the upper urinary tract, preventing the formation of kidney stones, treatment of pyelonephritis.

Neuropsychiatrist participation in patient's treatment with bladder extrophy is necessary and allows, especially in adolescence, to avoid severe reactive states and the development of neurosis.

HYPOSPADIA

This malformation is characterized by the absence of the lower wall of the urethra in the distal sections, and the external opening of the urethra is displaced proximally and is located on the ventral surface of the penis, scrotum or perineum. The penis is deformed, bent down as a "hook". Formation of anomalies is associated with the infringement of embryogenesis on the 7–14th weeks of pregnancy.

Hypospadia takes the first place among the anomalies and malformations of the urethra: it occurs in 1/400–1200 births. The anomaly is the "privilege" of boys, although extremely rare, it occurs in girls.

According to the degree of underdevelopment of urethra, the forms of hypospadia are distinguished:

— capitate,
— stem,
— scrotal,
— perineum,
— chord type, "hypospadias without hypospadias" (Fig. 3.5.13).

Capitate form. It is the most common and easiest type malformation during which the urethral opening reopens on the site of frenulum of the penis. Foreskin on the ventral side of the penis is absent, and the dorsal, looming in the form of an apron, not completely covers the head. Penis is straight; sometimes deviation of the head downwards is marked.

In this form of hypospadias narrowing of the external opening of the urethra or its shield by thin skin is often observed, which can significantly hamper urination and lead to dilatation and atony of the urinary system, which are located above.

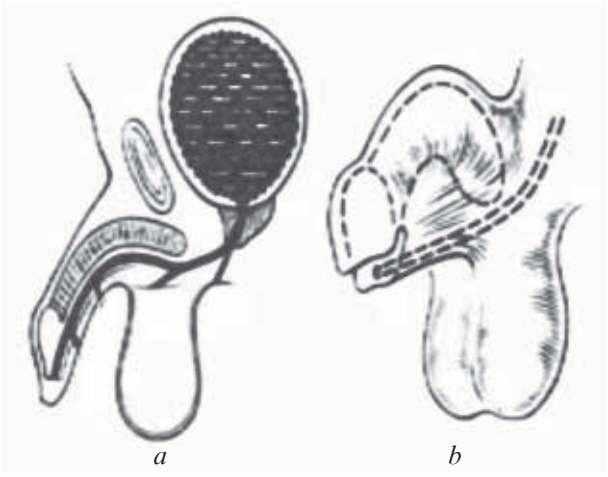


Fig. 3.5.13. Hypospadias *(a, b)*

Stem form. In this form the urethral opens on the ventral surface of the slightly opened trunk of the penis. Penis is deformed by fibrous strands that branch out from head to hypospadia opening of the urethra, is hook shaped, especially when erect. Urination is male typed, but the patient has to tighten his penis to the stomach for the foreskin. The growth of the cavernous bodies is difficult, their deformation increases with age, erection is painful. Sexual intercourse with stem hypospadias is possible, but if the opening of the urethra is located at the base of the penis sperm doesn't enter the vagina.

Scrotal form. This form is accompanied by an even more marked hypoplasia and deformity of the penis. The external opening of the urethra reopens at scrotum, which is split and resembles large labia. Urination is conducted by a female typed. Sexual intercourse is usually not possible because of the sharp deformation of the penis.

Perineum form. View of genital organs is very changed, which causes difficulty in determining the sex of the patient. Penis for its size and shape is similar to the hypertrophied clitoris, scrotum is split in the form of the labia. Urethral opening reopens at perineum, often there is rudimentary vagina. In this form of hypospadias a single or bilateral cryptorchidism is seen more often, than in other forms.

In addition to the above forms hypospadias occurs, with the absence of dystopia of the urethra opening, but there is a distinct strain of the cavernous body of penis. This is called "hypospadias without hypospadias". Synonyms: congenital short urethra, hypospadias by chord type. During this anomaly urethra can be 1.5–2 times shorter than cavernous body. Painful erections, coitus is not possible.

Treatment. Capitate hypospadias doesn't need treatment, unless it's accompanied by narrowing of the external opening of the urethra or the presence of urethra with covered opening — a membrane. This needs porotomy (opening the external opening of the urethra) or removing of the membrane.

The main efforts of the physician during the treatment of other forms of hypospadias should be direct-

ed to the rectification of the cavernous body, the creation of the missing part of the urethra, and in severe cases an additional problem — correction of gender may occur.

The first stage of surgery is performed at the age of 1.5–2 years old. The operation consists of the careful dissection of fibrous tissue and displacement of hypospadias hole proximal, which achieves maximum smoothing of cavernous body. The second stage of treatment — urethroplasty — is performed at the age of 5–7 years. Recently, one-step procedure is widely used — smoothing of the penis and urethroplasty from the leaves of the foreskin or the skin of dorsal surface of the penis on the vascular pedicle. This operation can be performed in children from 2–3 years old.

HERMAPHRODITISM

Intersexuality (hermaphroditism) belongs to the severest genital malformations. True and false hermaphroditism are distinguished. False hermaphroditism is divided into male and female.

True hermaphroditism is characterized by the presence in the body at the same time male and female sex organs. This type is rare, detected in 10–12% of patients with severe forms of hypospadias. It has been associated with chromosomal aberrations in the embryonic period.

There are organs of one sex during false hermaphroditism and the external ones are so underdeveloped that their appearance resembles the other sex. Appearance of false female hermaphroditism is associated with congenital hyperplasia or tumor of adrenal glands (adrenal form of pseudohermaphroditism — adrenogenital syndrome).

The external manifestations of different types of hermaphroditism are quite similar, so the establishment of a real sex is most often associated with great difficulty and needs for special studies.

Treatment. The vast majority of the patients underwent surgical correction. In the case of true hermaphroditism during correcting sexual psychoaffiliation of the patient should be considered, which begins to manifest itself at the age of 2 years. During sex reassignment of female type hypertrophied clitoris and testicles are removed. Correction by the male type is more complex and involves the removal of the uterus and ovaries, straightening of the cavernous body of the penis, urethroplasty and orchiopexy (bringing down the testis).

EPISPADIAS

It is a congenital splitting of the upper wall of the urethra in the distal part or throughout (Fig. 3.5.14). Anomaly occurs in 1 in 50,000 of newborns, boys are affected five times more often than girls. According to the degree of splitting the urethra of boys distinguish epispadias of head, of the penis and full epispadias, of girls — clitoral and complete. Complete (total) epispadias is noted three times more often than other forms.

PHIMOSIS

Cicatricial narrowing of the openings of the foreskin — phimosis — prevents exposing penis head (Fig. 3.5.15).

Boys (younger than 3–5 years) have foreskin, as usual, completely covering the head of the penis and hangs as a skin trunk with a narrow opening.

During the growth of the penis, head pushes preputial sac, and its exit becomes free. That is why for children within the first year of life phimosis is physiological and no treatment is required.

Clinical picture and diagnosis. Often the inner layer of the foreskin is fused with the head of the penis by soft embryonic adhesions (synechiae). This

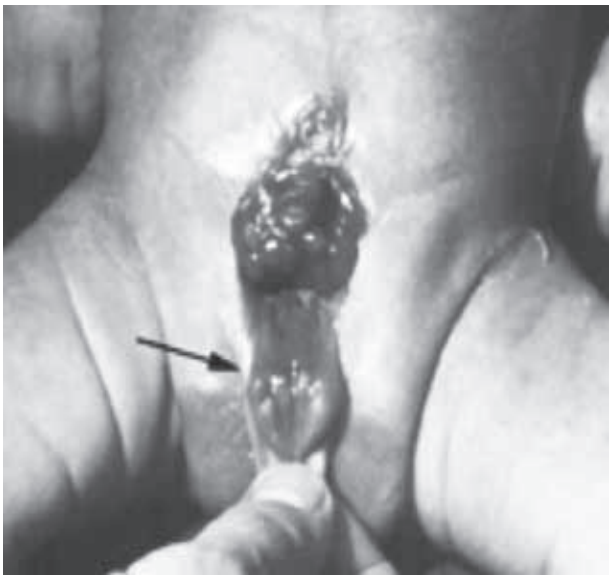


Fig. 3.5.14. Epispadias

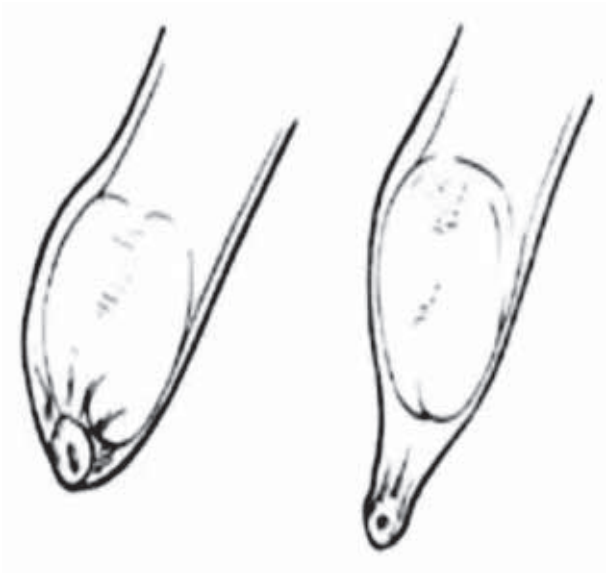


Fig. 3.5.15. Phimosis

may facilitate the accumulation of smegma, which is a good breeding ground for microorganisms.

Therefore not careful toilet of external genitalia in boys first years of life can lead to balanoposthitis — inflammation of the head of the penis and genital circumcision. During inflammation subsided, at the site of the opening of the foreskin adhesions often form, which prevents not only the output of the head, but also lead to a violation of urination. Difficulty of urine flow, in turn, can be the cause of an ascending infection and lead to cystitis and pyelonephritis.

The main complaints during cicatricial phimosis are disorders of urination and inability to display the head of the penis. During urination child worries and is straining. Urine, getting in preputial sac inflates it and through the narrowed opening follows by a thin stream or drops. In the event of the accession of inflammation there is pain at the site of the head and foreskin, edema and hyperemia of the prepuce, pus discharging.

During a careful examination, diagnosis is not difficult, but it should warn against forcible withdrawal of the head, in order to avoid injury to the foreskin. Hypertrophic and atrophic phimosis are distinguished. The first one is characterized by excessive development of the foreskin, the second one — it fits closely to the head and has a pin hole.

Treatment. During cicatricial phimosis surgery is necessary — circular excision of the leaves of the foreskin.

In the case of balanoposthitis, treatment begins with conservative measures: warm bath with a fluid of potassium permanganate or furadilin 5–6 times a day for 4–5 days, injection of antiseptic ointments to the preputial sac. Observation of a child should be placed after the liquidation of the inflammatory process. Development of scar changes in the area of the external opening of the foreskin is an indication for surgery. Recurrent balanoposthitis, which are not amenable to conservative treatment, are an indication for excision of the prepuce, even without it marked scarring.

Paraphimosis is a limitation of the head of the penis by a tapered ring of the foreskin. It is caused by violent pulling of the foreskin over the penis' head, which is often observed in children during masturbation. If the skin of the prepucethe skin of the prepuce is not returned to its original position in time, tissue edema is developing, which impairs blood flow to the prepuce and the head of the penis, which can end up with necrosis of the infringed ring.

The clinical picture of paraphimosis manifests itself in acute pain at the site of the head of the penis, swelling of the foreskin, which eventually increases, swelling of the head itself. Because of the significant pain syndrome and because of the increased swelling of the child, urination is infracted.

Treatment in the early stages consists of the immediate reduction of the head under anesthesia. In the later periods during severe swelling, perform excision of infringed ring and trimmed flesh.

With modern diagnosis and early treatment, prognosis is favorable.

MALFORMATIONS OF THE TESTIS

Testicular hypoplasia is caused by the loss of blood supply and is most common in cryptorchidism. Bilateral hypoplasia is accompanied by endocrine disorders. As a rule, children have adipose-genital obesity, puberty is late.

In some cases, the anomaly is combined with microfoam, or "hidden penis". Treatment is carried out by endocrinologist.

Monorchism is a congenital anomaly, which is characterized by the presence of only one testicle. The anomaly is associated with the violation of embryogenesis before placing the final kidney and sexual gland. That is why combination of monorhizm and solitary kidney is not rare.

During monorhizm besides the lack of the testicle, testicular appendage and deferent alveus are not developed. Corresponding half of the scrotum is applized.

Diagnosis of the monorhizm is final only after fruitless searches of the testicle with broad audit of retroperitoneal space. Congenital lack of one testicle at a normal second usually does not manifest in endocrine disorders or results in infertility. But in some cases, the only testicle is cryptorhired. Then hypogonadism can exist in one or another degree.

Treatment. During "clear" monorhizm treatment consists of implantation of testicle prosthesis, made of silicone, into the scrotum. Surgery is performed for cosmetic reasons in adolescents of 12-14 years. During hypoplasia of the only testicle do hormonal replacement therapy.

Anorchism is a congenital absence of both testes, due to no tab of fetal gonads. As usually vice is combined with agenesis or aplasia of the kidneys, but can occur as a distinct anomaly. During agenesis and renal aplasia children are unviable. In extremely rare cases anorhizm, as an independent anomaly, is characterized by pronounced eunuchidizm, hypoplasia of the external genitalia, and absence of the prostate and seminal bubbles. Secondary sexual characteristics do not develop.

Treatment consists in the appointment of hormones.

Polyorchism is the anomaly, which is characterized by the presence of an additional (third) testicle. Usually it is reduced, hypoplastic, devoid of the epididymis and is located in the scrotum above the main testicle. Extremely rare testicular ectopia of additional testicle under the skin of the thigh, back, and neck are described.

Treatment involves removing the extra testicle, because it can be a source of malignant tumors.

Cryptorchidism is the anomaly of the testicular position in which one or both testicles during intrauteral development did not drop into the scrotum. They are located on the ground of lower pole of the archinephron, in the abdomen or in the inguinal canal. More often most the latter localization is met. Predominant form is the right-sided cryptorchidism. Bilateral is noted in 20% of cases. There are true and

false cryptorchidism. The latter one is characterized by the opportunity of relegation testicle into the scrotum, but it can return to its original place.

Cryptorchidism has a high frequency of infertility, which reaches 15–60%. According to different authors, cryptorchidism in term children occurs in 3%, in premature — at 30%. There are abdominal and inguinal retention testicle.

The reasons that determine cryptorchidism occur during intrauterine development. There are five stages of testicular migration:

1) laying of gonads,

2) migration of the testis from a anlage to the entrance of the inguinal canal,

3) formation of the inguinal canal openings,

4) the passage of the testes through the inguinal canal into the scrotum,

5) obliteration of the vaginal process of the peritoneum.

The migration of the testes from the abdomen into the scrotum begins with 6 weeks of fetal development. Testicle reaches inner ring approximately to 18 to 20 weeks, and by the time of birth, the gonads are located on the bottom of the scrotum. And if the transabdominal phase doesn't depend on the level of androgens and may pass through the intra-abdominal pressure, the passage of the testis through the inguinal canal is very dependent on the concentration of androgens, which are produced by embryonic testicle. But leading role belongs to androgens, which are produced by the pituitary gland of the fetus at the last trimester of pregnancy.

The causes of cryptorchidism may be mechanical factors, such as lack of development and the wrong position of vaginal process, fibrous adhesions and others. Cryptorchidism also often accompanies abdominal wall defects, the syndrome of a "plum" belly, *spina bifida* and chromosomal defects.

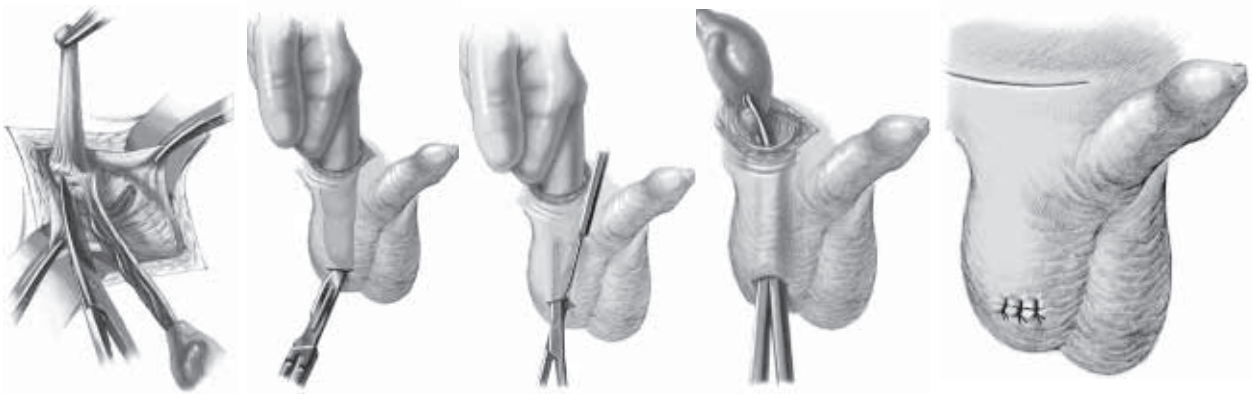
Clinical picture and diagnosis. On examination of the patient with cryptorchidism the doctor should remember that it is possible to identify children with a false cryptorchidism or with increased cremaster reflex. Scrotum is usually well developed at such children. During palpation in the groin, in the direction from the inner to the outer ring, gonad can be brought down to the scrotum. Parents of a child say that while swimming testis descends into the scrotum itself. In children with true cryptorchidism testicle can not bring down into the scrotum. In this case one or both halves of the scrotum are hypoplastic, gonad is palpable in the groin, pubic, perinea areas or in the other half of the scrotum. On palpation of the testis in the groin area it is necessary to make a *differential diagnosis* of inguinal retention and groin ectopic gonads, because with any form of ectopia hormone pre-operative preparation is not necessary. The elements of the spermatic cord are well developed, are long enough for downgrading the testis in the scrotum with the operational way. At the same time during inguinal retention gonad is in the inguinal canal, and the vessels of the testicle do not have sufficient length for free relegation. Therefore, patients with inguinal retention need additional preoperative hormonal therapy.

Differentiation of inguinal ectopia from inguinal retention is possible after palpation of the study. In cases when the palpable gonad is shifted exclusively along the channel, repeating its anatomical way with high probability we can say about the retention of the testicle. Displacement of gonads in all directions may indicate ectopic groin.

The most complicated group of patients is with abdominal retention (both in terms of diagnosis and treatment of the position). First of all, a patient with the syndrome of non-palpable testis should be tested to determine the sex and eliminate chromosomal abnormalities. The algorithm study of children with non-palpable testis includes an ultrasound scan of the abdomen, karyotyping if necessary, angiography, computed tomography, etc. Laparoscopic research is quite effective, which allows us to estimate the condition of vessels gonads, testicles and determine the location of the state of the gonads by external signs. Biopsy is performed if necessary.

Treatment. For the treatment of cryptorchidism drugs of chorionic gonadotropin are used. In the treatment of true cryptorchidism efficiency reaches 10.5%.

Surgery. In cases when the operation is performed at children under 2 years old, normal spermogram was estimated in about 80%, in the group operated at the ages of 2–4 years old, the proportion of patients with normal spermogram is approximately



a *b* *c* *d* *e*

Fig. 3.5.16. Stages of Petriwalsky surgery *(a–e)*

58%. During operation at the age of 5–8 years old — 36%, and those who were operated at older age, normal spermogram is registered in less than 30% of cases. So the best time for the operation is under the age of 2 years old. This is due to the influence of hyperthermia and ischemic testicular parenchyma, as well as increasing of the diameter of the seminiferous tubules till 9 years old.

A typical example of a one-step operation with using the principle of permanent fixation is an operation, which suggested Petriwalsky. This operation is quite common around the world because it provides an opportunity for careful traction of gonads (Fig. 3.5.16).

There have been recent reports of laparoscopic treatment of cryptorchidism with abdominal location of the testicle. Very common is Fowler–Stephens surgery, which is performed in two phases: in the first phase of laparoscopy the location of the testicle and place hemostatic clips is identifyed and established, and after 6 months after laparoscopy a second step follows, in which we define a broad cuff from peritoneum, testicle and deferent duct, and this complex relegates to the scrotum.

Differential diagnosis must be done with inguinal lymphadenitis and tumor formations, ectopic testis.

Complication of cryptorchidism: volvulus, strangulation, malignant degeneration syndrome, swollen scrotum.

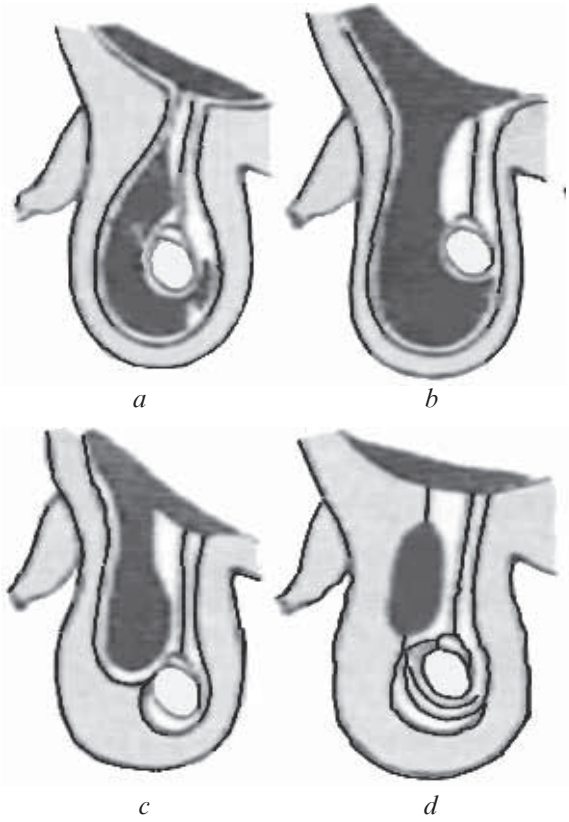


Fig. 3.5.17. Malformations of vaginal rame *(a–d)*

HYDROCELE OF TESTIS MEMBRANES AND SPERMATIC CORD

Hydrocele of testis membranes and spermatic cord are very frequent abnormalities in children. Their development is associated with cleft vaginal process of the peritoneum and its accumulation in the cavity of serous fluid (Fig. 3.5.17). During the absence of obliteration of vaginal process in the distal edema, hydrocele of testis membranes forms (Fig. 3.5.17, *a*).

In the case of no obliteration of all inguinal process combined hydrocele of testis membranes and spermatic cord occurs (Fig. 3.5.17, *b*). If the process obliterans in the distal part and proximal remains open and has a connection with the abdominal cavity, it is a concomited spermatic cord hydrocele (Fig. 3.5.17, *c*).

If there is a bone in the obliteration of the distal and proximal parts, and fluid accumulates in the middle of its department, it is about a mismatched hydrocele of the shells of spermatic cord or cyst of the spermatic cord — funiculitis (Fig. 3.5.17, *d*).

Formation of hydrocele is associated with low absorptive ability of vaginal wall and imperfection of lymph apparatus in the groin. With age of the child gradual reduction and disappearance of edema may occur (Fig. 3.5.18).

In older children and adults the causes of funiculitis are injury and inflammation. When you hit in the groin, fluid in the shells of the spermatic cord may accumulate in the shells of the spermatic cord, which does not resolve long time. In these cases we deals with acute spermatic cord cyst.

Clinical picture and diagnosis. Hydrocele is characterized by an increase of the half, and during bilateral disease — of the entire scrotum. During isolated hydrocele of the testicle swelling is round, near its lower pole the testicle is determined. Combined hydrocele appears in soft elastic oblong formation, which upper edge is palpated near external inguinal ring. With straining that formation increases and becomes denser. Palpation of the swelling is painless. Transillumination reveals a characteristic symptom of knockouts. During the valve character the connec-

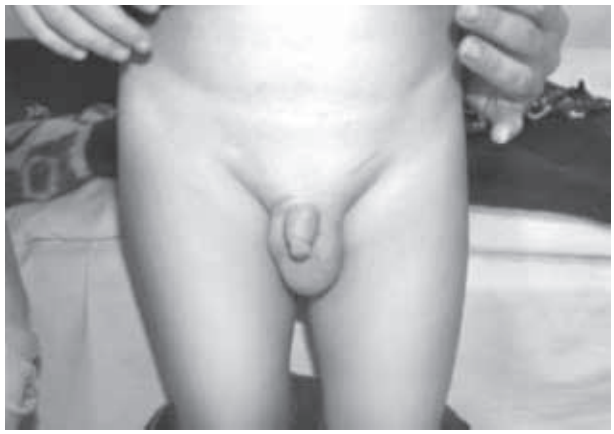


Fig. 3.5.18. A child with hydrocele of testis

tion with the abdominal cavity hydrocele is tense, may cause anxiety of the child.

Spermatic cord cyst has a round or oval shape and sharp contours. Its upper and lower poles are well defined. Hydrocele often has to be differentiated with inguinal hernia. With reduction of hernia contents, characteristic is audible rumble, immediately after the reduction swelling in the groin disappears. During incongruous hydrocele attempt of reposition fails. Dimensions of formation in a horizontal position decreases, but more slowly than in the reduction of hernia, and without the characteristic sound. Great difficulties arise in the differential diagnosis of occurred cyst with strangulated inguinal hernia. In such cases, often resort to surgery with a preliminary diagnosis "strangulated inguinal hernia."

Treatment. Because for the first 2 years of life self-healing is possible due to the finishing of the process of obliteration of the processus vaginalis, surgery is performed in children over this age. During isolated and acquired hydrocele of testicular membranes common is Winkelmann surgery, which consists of dissection of shells of hydropic cavity and stapling them in twisted position around the testicle and epididumis.

With combined hydrocele Ross surgery is used, the purpose of which is termination of connections with the abdominal cavity and the creation of an outflow for hydropic fluid. Vaginal process is tied around the internal inguinal ring and partially removed with the leave of the testicle in its own shells, through which the fluid exits and dissolves into the surrounding tissues. This operation is easier than Winckelmann surgery, not accompanied by traumatization of the testicle and gives a good effect.

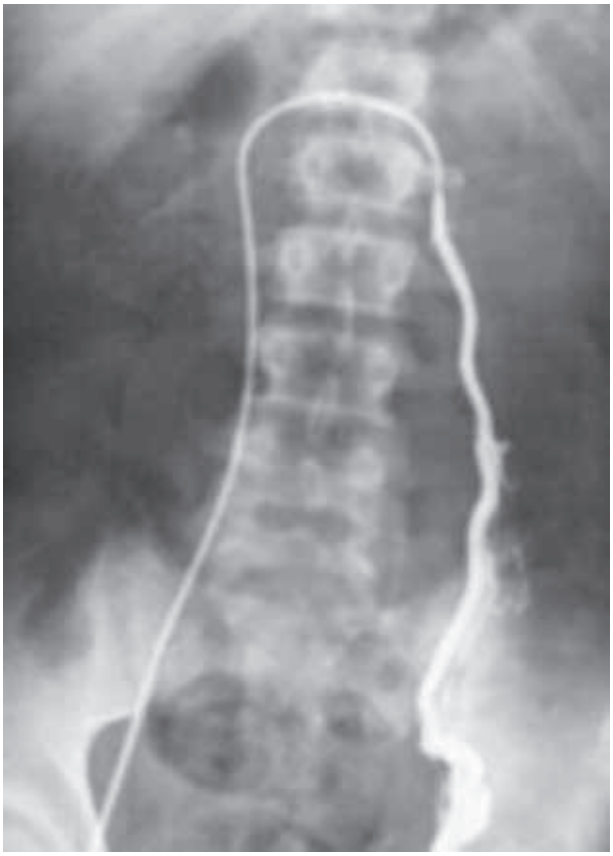


Fig. 3.5.19. Varicocele, angiography

In children younger than 2 years old in the case of complicated hydrocele of testicle shells, which causes anxiety, the use of the puncture method of treatment is indicated. After the evacuation of hydropic fluid suspensory is applied. Repeated puncture is performed with the accumulation of fluid. Suctioning reduces the compression of the testicle and allows delaying the surgery.

VARICOCELE

Varicose veins of acinar (withe-type) plexus is common in boys mostly after the age of 9–10 years with frequency up to 15% (Fig. 3.5.19).

Idiopathic (primary) and symptomatic (secondary) varicocele are distinguished. Development of a secondary varicocele is due to compression of the outflow tract of blood with some testicular volume retroperitoneal formation (tumor, enlarged lymph nodes, cyst).

Usually primary varicocele is formed on the left and has a rather complicated genesis. As known, the blood from the testicle flows in three veins: testicular, cremast and vein of the deferens. Right testicular vein falls to the bottom hollow, and the left — in the renal vein. The left renal vein, going to the bottom of a hollow, is in the so-called aorta mesenteric tweezer (between the aorta and superior mesenteric artery) and can be compressed, resulting in renal hypertension and venous outflow of blood through the spermatic vein. Sometimes renal vein is compressed by abnormal testicular artery, which is thrown over it.

In pre-pubertal and early pubertal boys are growing rapidly, which is manifested in further increased pressure in the acinar plexus due to the increase of orthostatic pressure. At the same period there is 4 times increase of the arterial blood flow to the testis. Increased blood flow re-stretches testicular vein, pushing the valves and thus opening the way to the path retrograde flow of blood from the renal vein in a testicular. Significantly influenced by the increasing pressure, varicose testicular vein wall deformation and acinar plexus develops. In the horizontal position of the patient, aorta mesenteric "tweezers" opens, the pressure in the renal vein goes down and the blood on the spermatic vein is flowing freely from the acinar plexus to the kidney.

Prolonged venous stasis leads to ischemia, the development of sclerotic changes in the testis and differentiation of spermatogenic epithelium. During this damages blood-testis barrier, this performs the function of a protein shell of the testicle, basement membrane and Sertoli cells. Autoimmune aggression is developing. In general lines antibodies appear that due to various reasons can overcome testis barrier of the right testicle and cause violations of its morphology and function. In future, this may manifest decline in total spermatogenesis and development of infertility.

Clinical picture and diagnosis. Very rarely varicocele is diagnosed at young (2–5 years) age. In the

history of these patients can be determined the factor that caused prolong damage to outflow of blood from the testicle (trauma, inflammation, surgery).

Sometimes varicosis is marked on the right side or on both sides. Varicocele only on the right side is connected with the abnormal falling of the right testicular vein into the renal. Bilateral varicocele is due to the presence of between testicular anastomoses in which high blood pressure in the left testicle is transmitted to the right side. After treatment of the left-sided varicocele delatation of acinar plexus disappears.

Children with varicocele usually do not make complaints, and varicose veins found in checkups at school. Only the older kids sometimes have feeling of heaviness and pain in the left side of the scrotum.

Clinically three degrees of varicocele are divided:

— veins above the testicle are determined only by palpation in the vertical position of the patient during the tension of the abdominal muscles,

— extended and tortuous veins clearly visible through the skin of the scrotum (a symptom of worms in a bag), in horizontal position veins are fallen down,

— on the background of fixed veins reduced testis and dough form is determined by palpation.

During varicocele, which does not fall down in a horizontal position, a study is conducted to identify volumetric retroperitoneal formation (excretory urography, ultrasonography, CT).

Treatment. During idiopathic varicocele the operation is performed — ligation of spermatic veins in the retroperitoneal space or thrombosis at angiography. It stops the flow of blood from the kidney to the testis and contributes recession of varicose veins.

SYNDROME OF SWOLLEN SCROTUM

This condition occurs as a result of trauma, testicular torsion or epididymis, Morgagni necrosis, orchiepididymitis.

Clinical picture and diagnosis. The main symptom of syndrome is pain, swelling and redness of the scrotum half. With similar clinical manifestations, each of these disorders is characterized by certain features. Closed trauma of the testicle is described by the rapid emergence of these symptoms. Depending on the degree of injury (bruise, break of the testicle, compression), the severity of the pain varies from mild to the development of traumatic shock. During the localization of pain, mainly along the spermatic cord, testicular torsion or epididymis can be suspected. Twisted testicle is usually lifted up and sharply painful. Morgagni necrosis is accompanied by appearance of liquid in the testicle shells, but you can find a place of the greatest tenderness or palpate gidatide itself. During the break of the testicular parenchyma pain is diffused, half of the scrotum is greatly increased, bluish; swelling often extends throughout the scrotum.

In order to identify the nature of the content of testicular membranes (blood, exudate) perform transillumination and diagnostic puncture.

The differential diagnosis is spent with the orchitis, which is complicated by mumps, and angioedema. The latter, as a rule, increase whole scrotum, fluid permeates all its layers, forming water bubble under a delicate skin. Palpation of the scrotum is not painful.

Treatment. The syndrome of swollen scrotum needs emergency surgery because the testicle is very sensitive to ischemia and can quickly be lost. After opening the shells of the testicle clarify the diagnosis. If there are breaks of the testicle, evacuate the hematoma, unsustainable part is removed and protein coat is sutured. During identifying twisted testicle it is eliminated and the testicle is fixed by the protein shell to the total tunica vaginalis. Necrotic gidatide is removed after ligation of the legs. During purulent orchoepididimic cavity of own testicle shell is drained. Testicle is removed only during explicitly necrosis (dark color, which after eliminating of the twist, warming, and novocaine blockade of the spermatic cord does not change). After sparing surgery aspirin in half dose is prescribed to remove the autoimmune response. Delaying of the surgery during a swollen scrotum threatens with testicular atrophy.

3.6. MALFORMATIONS OF THE MUSCULOSKELETAL SYSTEM

TORTICOLLIS

Specific objectives of the lesson:

1. Master classifications of congenital hip dislocation, congenital clubfoot, congenital muscular torticollis, congenital extremities, congenital spine.

2. Recognize main clinical symptoms at malformations of the musculoskeletal system in children of all ages.

3. Interpret auxiliary methods at malformations of the musculoskeletal system.

Fig. 3.6.1. View of a child with torticollis

4. Demonstrate examination of the child with defect of the musculoskeletal system.

5. Master algorithm of actions in identifying malformations of the musculoskeletal system.

6. Interpret main principles of malformations treatment of the musculoskeletal system in children.

Contracture of the neck is called torticollis due to the shortening of sternoclavicular-liners muscles (m. sterno-cleido-mastoideus), sometimes accompanied by the primary (congenital) or secondary (caused by trauma) change of the trapezius muscle, fascia of the neck. Rare "bilateral congenital muscular torticollis" occurs as a result of shortening of both sternoclavicular-nipple muscles.

Formation of congenital muscular torticollis can cause:

— Incorrect forced position of the fetal head at unilateral excessive pressure on it in the area of uterus, which approached the point of attachment of sternoclavicular-liners muscles, which leads to its shortening with fibrous degeneration (Fig. 3.6.1);

— Intrauterine ischemia of muscle under pressure of the cord, twined around neck of the fetus or at birth;

— Intrauterine inflammation of sternoclavicular-liners muscle with the transition into chronic interstitial myositis;

— Hematoma and rupture with severe birth of sternoclavicular-liners muscles in the lower part, in the transition area of muscle fibers into the tendon with subsequent scarring and delay muscle growth in length;

— Malformation of the sternoclavicular-liners muscle;

— Hyperextension or microtrauma of young immature muscle during birth with further overproduction of connective tissue.

Clinical picture and diagnosis. Congenital torticollis is detected from the first days of life. Head tilted to one side, the chin is turned to the other, m. sterno-cleido-mastoideus on the side of the bending tense, shortened and contoured under the skin. Motion in the cervical region is limited, passive correction of lateral curvature is possible.

In children, during first 8–12 days of life signs of congenital muscular torticollis subtle and identified at a small minority of patients. Early symptoms of the disease appear at the end of the second and beginning of the third week by spindle-shaped thickening of the middle or lower third of the sternoclavicular-liners muscle that is a consequence of intrapartum injury with hemorrhage and edema of dysplasia muscle.

This thickening has a dense texture, easily shifted along with the muscle, with no signs of inflammation. Clearly contoured muscle thickening maximum increases to 5 to 6 weeks (up to 2–2.5 cm in diameter), and then gradually decreases and disappears to 4–8th month of life. In the area of missing thickening stays indurateion of the muscle, its elasticity decreases like tendon bundle, lagging behind in growth compared with the same named muscle on the opposite side. Convergence of fixed points of attachment of sternoclavicular-liners muscle forms the head tilt to the affected side, and at the same time its turn in the opposite, that is, forced incorrect position of the head and neck, or torticollis. The advantage of the slope of the head points to the overwhelming defeat of clavicular leg, the advantage of turning — to the defeat of the sternum.

In children younger than 1 year of age, deformation is expressed slightly. Not timely diagnosed torticollis, left untreated progresses, especially during the rapid growth of the baby, after 3–6 years. Along with the increase of fixed tilt and rotation of the head, limitation mobility of the neck appear secondary compensatory adaptive changes, which depend on the severity of lesions of sternoclavicular-liners muscle. Asymmetry and hemihypoplasia of facial skeleton is seen. The size of a face of the affected size reduces vertical and increases horizontal. As a result, eye gap is narrower and places a little lower, cheek contour is smoothed, raises angle of the mouth. Nose, mouth, chin are located on a curve, concaved from the affected part. The quest to the vertical position of the head is compensated by high standing of shoulder girdle and scapula, scoliosis in the cervical and thoracic sections, and in older children — S-shaped scoliosis of the cervical, thoracic sections and lumbar spine.

During bilateral congenital muscular torticollis equivalent shortening of sternoclavicular-nipple muscle appears by a slope — the nomination of the head forward with significant cervical lordosis, limitation of motion of the head, especially in the sagittal plane, high standing of the clavicles. Different degree of muscle lesion is often diagnosed as unilateral congenital torticollis.

External identity of secondary changes that develop in congenital muscular torticollis, requires differentiation with other:

— congenital (syndrome of Klippel — Feil, congenital additional tapered neck hemivertebrae, fusion of the cervical vertebrae, additional cervical ribs; webbed neck);

— and acquired forms of torticollis (Grisel disease, spasmodic torticollis due to suffered encephalitis, birth brain injury, injury and destruction of the cervical vertebrae, post-burn scars).

Conservative treatment. Conservative treatment should be started from a 2-week age of child's life, that is from the time the symptoms of the disease occur. Corrective exercises are performed for up to 5 minutes 3–4 times a day: with both hands taking the child's head, who lies on his back, and without effort, gently tilt it to a healthy side during turning to the affected side. Exercises are completed with the massage of muscles of the healthy half of the neck, and the affected muscle — at the level of compaction only with stroking.

For keeping baby's head in the position of overcorrection cotton-gauze collar by Shanz or a large cotton-gauze roll by K. A. Krumin is imposed with latching through the armpit of the unaffected side (Fig. 3.6.2–3.6.5). During putting a healthy baby in the bed, the healthy side of the neck should be turned away from the wall; as a result, watching the room, the child unconsciously stretches affected sternoclavicular-liners muscle. However, UHF

therapy is recommended to the site of indurated-muscle, and from 8 weeks of age — physiotherapy courses, a combination of electrophoresis of potassium iodide, ronidase (lidase) with paraffin bath. Treatment is continued until 1–1.5-year age of a child. Only some children with severe underdevelopment of sternoclavicular-liners muscle, started promptly and carefully conducted conservative treatment does not lead to a complete recovery.

Surgical treatment is carried out after the age of 2 years. Depending on the severity of the changes of sternoclavicular-liners muscle, surrounding tissues, strain, age, basically there are two methods of surgery: myotomia of sternoclavicular-liners muscle with its partial excision and opening the fascia of the neck and plastic elongation of the muscle. After the operation, cotton-plaster collar for 4 weeks is used, and then for several months physical therapy, massage and physiotherapy are conducted.

Favorable results of conservative and operative treatment does not exclude the need for follow-up, including during puberty. During the absence of treatment of congenital muscular torticollis, severe irreparable deformation forms: head with rotation firmly tilted to the upper arm, pronounced asymmetry of the face and scoliosis of the spine.

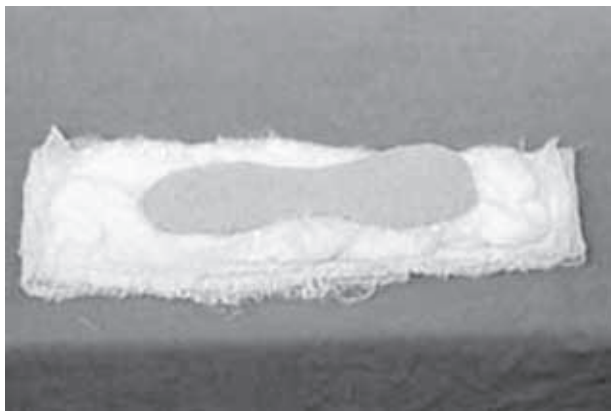


Fig. 3.6.2. Cotton-gauze Shanz collar

Fig. 3.6.3. A child with Shanz collar

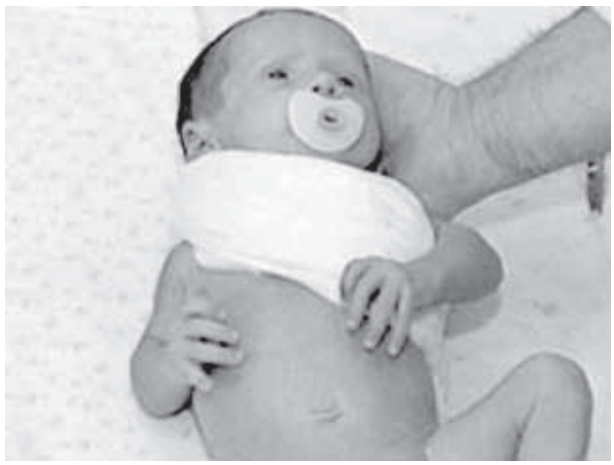


Fig. 3.6.4. Cotton-gauze roll by Krumin

DEFORMATION OF THE SPINE

Deformation or deviation from the normal development of the spine in children may be congenital or acquired, isolated and combined; in all cases, the deformation is accompanied by a violation of posture. To identify spinal deformity, normal development and the formation of its natural curves should be remembered. Spinal deformation in children is identified from 7.7 to 29% among children. First spinal deformation was described by Galen (II century BC) and introduced the following definitions:

1. Kyphosis — the deviation of the spine in the sagittal plane to the back.
2. Lordosis — the deviation of the spine in the sagittal plane forward.
3. Scoliosis — curvature of the spine in the frontal plane.

Newborn's spine has a shape of flat curved back arc, which is a uniform *kyphosis*: in the supine position on a flat surface spine becomes straight. On the 2–3 month of life the child picks up and holds the head, reclining it back; as a result of balancing the neck and scalene muscle physiological *cervical lordo-*

Fig. 3.6.5. Treatment of torticollis

sis develops. At 5–6 months age, baby is sitting well, spine is easily movable, and under the force of the weight of the head, shoulders, interiors, durable lordosis and balance by back muscles forms *thoracic kyphosis*. At the age of 8–9 months baby starts standing, 10–12 months — walking, due to the muscles that flex the hip (mainly m. psoas major), the pelvis tilts forward, grabbing the lumbar spine. In vertical position the body is balanced by sciatic muscles and back muscles — physiologic *lordosis of the lumbar spine* forms.

Until the end of the first year of life physiological curvature of the spine in the sagittal plane arise, inhered for the adult spine, which continue developing and individually form during the growth of the child, which completes by 17–22 years of age. Orthostatic or vertical position of human determines *posture*. Great influence on the posture have external conditions, daily routine, diet, physical overload in sports, diseases, and everything that makes imbalance on the balanced state of musculo-ligamentous skeleton and spine. This results in a *normal or abnormal posture.*

Posture is the vertical free position of the body.

To determine the posture, detection of spinal deformations one should examine the child in an upright position in the front, posteriorly and from the sides. At the same time attention is payed to the position of the head, shoulder girdle, shape and symmetry of the chest, protruding ribs, position of the blades (the level of the lower corners, distance of vertebral line from the center line and chest); a symmetry of triangles, formed by hollow of waist and arm; bends of spine, their severity, the presence of lateral curvature vertical and during the slope of the body forward; the position of the pelvis, the ischial fold symmetry, size and shape of the limbs.

It's necessary to study child's gait and range of motion in the large joints of the limbs. Volume of the spine motion and soreness is tested by bending body forward, backward, sideways and rotation about an axis. Local pain is determined by palpating the spinous processes and a moderate load on the shoulder girdle.

During normal posture, vertical axis in the frontal plane of the body passes the line from the middle of the parietal part on the back of the line, connecting the corners of the lower jaw, down through the line, which joins two hip joints and the middle of the foot. The angle of inclination of the pelvis in the 4th year of life equals 22°, on the 7th — 25°, in men — 31°, in women — about 28° (4° deviation is considered normal). In normal posture vertical axis in the sagittal plane, which is dropped from the mount of the occipital bone, passes through the spinous process of VII cervical vertebra, the crease between the buttock muscles and the middle of the quadrangle (the site formed by the surface of the feet and between them). As a result, the power of the body weight is distributed evenly on both feet, which is optimal. Vertical is deflected to the left or right during the uneven distribution of weight force. This occurs during the development asymmetry of certain groups of muscles of the trunk, especially during a time of rapid growth, and is determined during the inspection in an inclined position. During unequal length of limbs curved spine is removed by underlaying of special insoles under the foot of shorted limb in a vertical position.

Increase or decrease of physiological convexity of the spine in the sagittal plane with the increase or decrease in pelvic tilt creates pathological types of posture (F. Staffel):

— hunched back (increased thoracic kyphosis),

— flat back (flattening of the physiological curves),

— plano concave back (thoracic kyphosis decrease with an increase in lumbar lordosis),

— kyphosis (increase of all physiological curves).

The degree of fixation of spine curvature is tested by holding the baby in a vertical position with both hands for armpits or lying on stomach. During such method of research non-fixed curvature straightens. Floating curvatures of the spine in the frontal plane, which disappear during sloping of the body and lying, refer to the scoliotic posture. If the curvature of the spine combines with the rotation around the longitudinal axis of the body, and at an inclination of the body is found even a slight protrusion of the ribs backwards on one side or protrusion of lumbar muscles, or both, should think of scoliosis, not of pathological posture. Pathological posture precedes the formation of fixed spinal curvature: pathological kyphosis, lordosis and scoliosis. Fixed curvature of the spine can be either congenital (malformations of the spine), and acquired.

KYPHOSIS

Kyphosis — curvature of the spine in the sagittal plane by the concave back. Thr upper chest, lower chest, lumbar and total kyphosis are distinguished.

Congenital kyphosis is rare and is the result of additional wedge-shaped vertebra, synostosis of two vertebras, and hypoplasia of the anterior body of vertebra in the thoracic or upper lumbar section of the spine, which determines by X-ray. The deformation is detected early — within the first six months of life, as soon as the baby starts to sit. As the child grows, deformation increases significantly, is painless and without neurological symptoms, reaching pronounced sizes to the period of puberty. Child's growth is delayed.

Treatment. During identifying the deformation plaster bed is used. Massage of back muscles, prolonged exposure on stomach, crawling is indicated. Corrective exercises are carried out by the developed system under the supervision of an orthopedist. Swimming, in winter — skating and skiing, spa treatment is recommended. During progressive forms of kyphosis surgical spinal fixation is conducted.

Acquired kyphosis can result from rachitis, tuberculous spondylitis, osteochondropathy, compression fractures of the vertebral bodies. Rachitic kyphosis is a consequence of the general muscular hypotonia

during severe rachitis. It develops quickly, once the baby starts to sit.

Clinical picture and diagnosis. Rachitic kyphosis is characterized by uniform curvature of lower thoracic and lumbar spine posterior, but may be formation of more acutely salient angle. Deformation decreases significantly if the child is put on the abdomen, or is eliminated entirely, if the child's legs and pelvis are raised up. During this study, deformations during congenital kyphosis, tuberculous spondylitis don't disappear. For the correct orientation in the diagnosis it is sufficient to evaluate other changes in the skeleton: the presence of craniotabes, rachitic rosary, a thickening of the epiphyses of the limbs and typical clinical signs of rachitis.

Treatment. There is a need in complex treatment of rachitis. Unfixed rachitic kyphosis is eliminated if the child is laid on a flat hard mattress; do not allow him to sit. Supine position is alternated with the position on the stomach. Child is fixed with a special bra to bed to keep in horizontal position. During fixed pronounced kyphosis it is recommended to lay the baby in plaster bed with underlying cruciformly stacked rolls for eliminating distortion. Plaster bed is changed every 1.5–2 months depending on the elimination of kyphosis. Simultaneously using massage to strengthen the back muscles, abdomen, and extremities. In most cases with the treatment of rachitis kyphosis disappears. Extremely rare, despite the complex of antirachitic treatment, deformation as kyphoscoliosis remains for whole life.

Abnormal kyphosis may be the result of infraction of chondrogenesis, namely spondee dysplasia: Calve disease (damage to the vertebral body, or *vertebra plana*) and Sheuermann disease (defeat of vertebral apophyses, juvenile kyphosis). The disease occurs in adolescence and is characterized by progressive kyphosis of thoracic, back pain. X-ray detect changes in the lower thoracic spine, depending on the stage of the process. The principles of treatment are described above.

LORDOSIS

Lordosis is curvature of the spine in the sagittal plane, concaved front. Physiological lordosis of the cervical and lumbar regions of the spine is the result of the formation of the human orthostatic condition. Abnormal or excessive, lordosis is localized mainly in the lumbar spine, may be due to pathological changes in the lumbar spine and surrounding tissues of congenital (*spina bifida occulta et aperta*, spondylolysis and spondylolisthesis, wedge-shaped vertebrae) and acquired origin. Most often strain develops as a compensatory curvature during localization of the main process in another region of the spine (kyphosis of thoracic section, round back), during deformation of the lower limbs, impaired function of muscles of the trunk and limbs (*coxa vara*, ancylosis of the hip in a vicious position, congenital hip dislocation, weakness of greater sciatic muscle and hyperfunction of hip flexor), etc. A child with hyperlordosis in the supine position can easily be passed the brush under the loin. While in this position the child's legs are bent at the hip and knee joints and hyperlordosis persists, it is fixed.

Congenital hyperlordosis in older children with a hidden passage may be shown in fatigue, soreness in the lower back. Compensatory lordosis of children most often occur asymptomatically.

Treatment. It is necessary to eliminate the main cause, which caused the development of pathological lordosis. A special set of therapeutic exercises, massage, heat physiotherapy are recommended.

SCOLIOSIS

Scoliosis — is a persistent lateral curvature of the spine, which is coupled with its torso, due to pathological changes in the spine and paravertebral tissues, prone to progressive deformation with static-

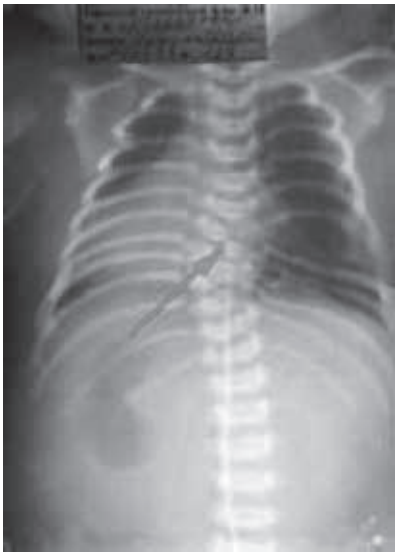


Fig. 3.6.6. X-ray during scoliosis

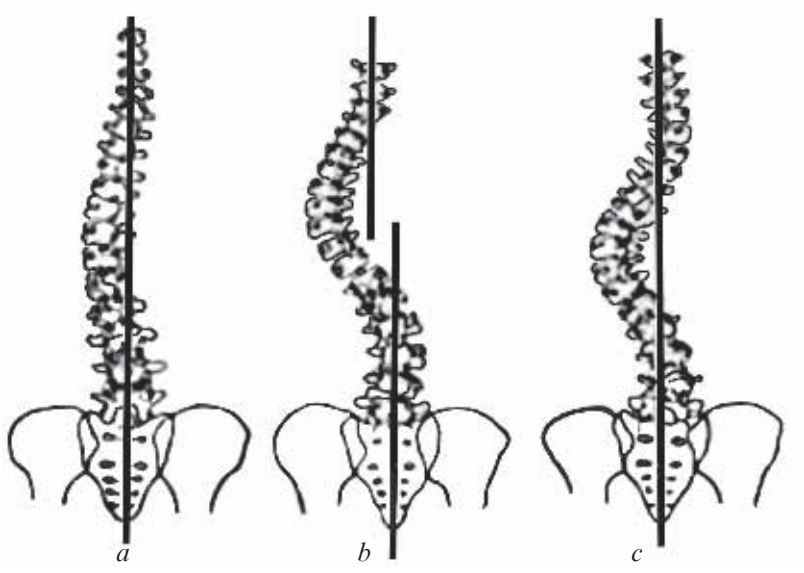


Fig. 3.6.7. Development scheme of scoliosis *(a–c)*

dynamic disorders and functional changes in the internal organs. During etiology congenital and acquired scoliosis are distinguished (Fig. 3.6.6, 3.6.7).

Congenital scoliosis. Congenital scoliosis is formed in the presence of defects of spine and ribs development (extra wedge vertebra and hemivertebra, synostosis of spinous processes, extra ribs, synostosis of ribs on one side, etc.), dysplasia of the lumbosacral section of spine, which defines as "dysplastic scoliosis" (spondylolysis, lumbalization, sacralization, *spina bifida*).

V. A. Dyachenko divides malformations of the spine on ontogenetic and phylogenetic. Ontogenetic defects occur within the first years of life:

— Anomalies of the vertebral bodies (cracks, defects, wedge vertebrae, platispondylitis, brachyspondylitis, microspondylitis, ace);

— Anomalies of bifida (cracks, hypoplasia of the half, anomaly processes, spondylolysis);

— Mixed anomalies (Klippel–Feil syndrome, blocking, vertebral hypoplasia, underdevelopment of vertebra).

Phylogenetic defects occur in puberty and are defined by location:

— Traumatic cervical (atlanta assimilation, atlanta demonstration);

— Cervicothoracic (dorsalization, cervicalization);

— Lumbar-sacral (sacralization, lumbalization);

— Sacrococcygeal (assimilation).

Vertebrae cracks — spina bifida — can be the front (of bodies) and back (of arches); they are caused by connection problems of one somite halves of vertebrae. Rachischisis — the presence of cleft of the body and arches of vertebra.

Wedge-shaped vertebra (hemivertebra), depending on the degree of underdevelopment can be lateral, back, single, multiple, alternating (two hemivertebra on opposite sides and different levels), "butterfly-view", active in the progression of scoliosis and blocked if scoliosis is not progressing.

During violation of the vertebra forming depending on the type of hypoplasia and ace-aplasia occur: platispondylitis — flattening of vertebra, brachyspondylitis — reduced height, microspondylitis — reducing of the size. Spondylolysis — a violation of the connection of arch and vertebral body.

During Klippel–Feil syndrome, which clinically appears as short neck, low hairline, limited mobility of the head, identify and reduce brachyspondylitis, reducing quantity of cervical vertebra, their fusion.

Assimilation and atlant manifestation can lead to limitation of the mobility of the head. Dorsalization — increasing of the number of vertebra due to VII of the cervical and the appearance of the cervical vertebra due to ribs. Cervicalization — increasing of the number of cervical vertebrae due to agenesis of the first rib. Sacralization — increasing of the number of sacral vertebrae due to lumbar. Lumbarization — increasing of the number of lumbar vertebra. Bilateral defects alter posture, unilateral lead to scoliosis.

Anomalies of the spine in 76% of cases may be accompanied by malformations of other organs and systems (according to V. E. Ulrich, A. Y. Mushkin): defects are localized in the organs that develop from a single somite with abnormal vertebra, or these organs have a segmental innervation with abnormal vertebra. This is embryopathy of skin and soft tissues (hemangioma, lymphangioma, spots, and hypertrichosis), urinary tract, spinal cord.

Acquired scoliosis is a sign of other diseases and differentiates due to the causes. Static scoliosis is observed during shortening of the lower limb, unilateral congenital hip dislocation, ancylosis in a vicious position and contractures of hip and knee joints. Neurogenic and myopathic scoliosis are caused by an imbalance of the back muscles, obliques after polio disease, Little disease, during neurofibromatosis, syringomyelia, family Friedreich's ataxia, muscular dystrophy type Erb–Roth, rachitis. Scoliosis is known because of the large post-burn scarring of the body, diseases and operations of the chest and rib cage. The cause of scoliosis can be spinal tumors of paravertebral localization. Metabolic disorders such as cystinosis, mucopolysaccharidosis, Marfan syndrome, Chernogubova–Ehlers–Danlos syndrome may be accompanied by scoliosis.

Idiopathic (dysplastic) scoliosis is the most common form of independent disease, manifest from 5 to 6 years of age. According to systematic concept of the pathogenesis (V. V. Butukhanov), scoliosis is a display of dysplastic-dystrophic syndrome that is combined gradual display of systemic disease areas of bone growth. It is proved that the root cause of dysplastic scoliosis is dysplasia of the vertebral disc, dismetabolism in it. Metabolic contravention in nucleus pulposus leads to decentration and abnormally located annulus fibrosus, appearance of irregular (wedge) shape of the disc and secondary deformation of vertebra with torsia due to uneven load and changes of the axis.

Congenital scoliosis gene anomalies of the spine and dysplastic scoliosis, which are characterized by a progressive course, lead to the scoliosis development. Scoliosis disease has etiopathogenesis, dynamics, clinical picture and stages, progresses during the "growth" pushes. During scoliosis disease after primary arc curving one or two arcs of anticurving appears. If the angle of the anticurving arc equals cor-

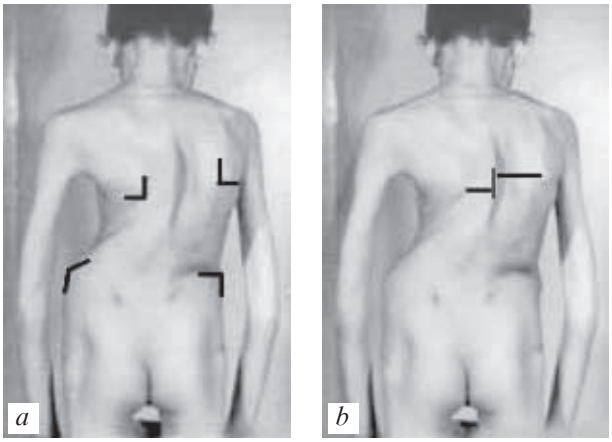


Fig. 3.6.8. Scoliosis diagnostics *(a, b)*

ner of the main distortions, such scoliosis is compensated; if the sum of anticurving angles is less, the scoliosis is decompensated.

Clinical picture. A careful examination of the patient is conducted in vertical position in front, back, laterally, sagging while standing, as well as in sitting and lying position. During anterior examination attention is payed to the posture; can be identified forced position of the head and facial asymmetry, different levels of shoulder girdle location, chest deformity, deviation of the umbilicus from the midline and a different level of the iliac crest. During back examination assessing deformity of the spine due to the spinous processes (for objectifying label projection of peaks of spinous process) (Fig. 3.6.8).

During scoliosis one girdle is higher than the second, blade on the concave side of the curvature of the spine is close to the spinous processes, and is located below the other, deformation of the chest and rib hump, asymmetry triangles waist are defined (Fig. 3.6.9).

During examination from the side, severity of kyphosis, size of rib hump are examined. In the slanted trunk position vertebra torsia on the asymmetry of paravertebral muscle rollers are detected, on height and location of the top of the rib hump, on volume of the trunk movement forward, sideways, backwards, and in the degree of elongation (stretching of the head or armpit), the mobility of the spine is determined. During the examination of the patient with scoliosis "balanced child" is always determined. Hypermobility of the spine, combined with increased mobility and recurvation in elbow, knee joints is a bad prognostic sign. Resilience of the spine describes an ability of back and stomach muscles to a prolonged power tension.

In addition, during clinical examination of the child disembryogenesis stigma, symptoms of connective tissue dysplasia (CTD), the pathology of other organs and systems are revealed. During dysplastic scoliosis of I–II degree, signs of dysplasia of connective tissue are detected in 100% of cases. To main CTD — syndrome signs include scoliosis, kyphosis, chest deformity, joint hypermobility, high-arched palate, angiodysplasia, blurred vision, arachnodactyly, pes plana, and cyanosis bulbi, mitral defects, antimongoloid eye shape, skin hyperelasticity, and dolichostenomelia.

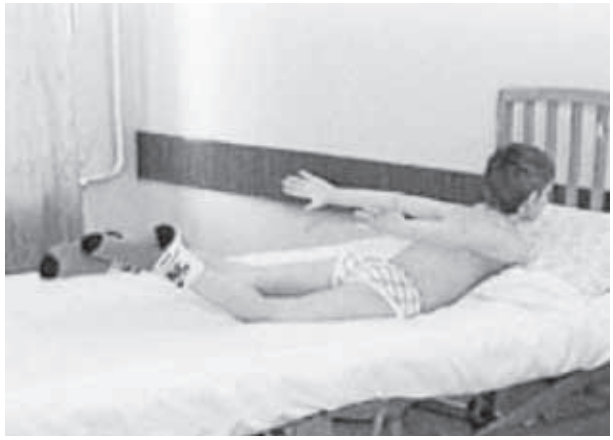


Fig. 3.6.9. LFC during scoliosis

Due to the shape scoliosis the C-and S-shaped are observed. By Poncetti–Fridman upper chest, thoracic, thoracolumbar, lumbar and combined scoliosis are distinguished.

During scoliosis of the I degree are determined asymmetry of back relief, one arc of curvature (doesn't disappear during sloping) and lumbar muscle spindle (Chaklin symptom). During II degree 2–3 cur-

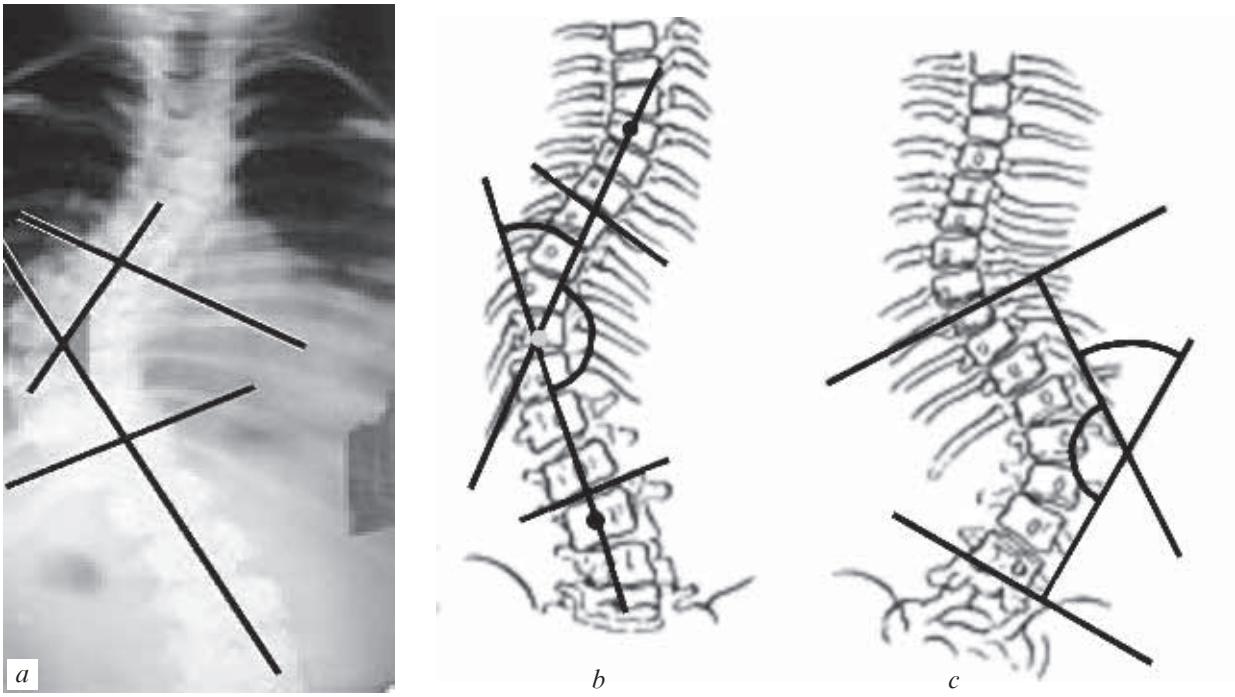


Fig. 3.6.10. Scoliosis diagnostics on Fergusson or Cobb techniques *(a–c)*

vatures of the spine, asymmetry of the chest, moderate rib hump are defined. During scoliosis of III–IV degree deformation of torso in front and back, large rib hump are defined.

Instrumental diagnosis. Radiography of the spine allows specifying the type of deformation (congenital, acquired), location and characteristics of the bending, making spondilometry. In order to perform diagnosis of scoliosis X-ray of the spine including pelvis in the capture of direct projection of the patient in an upright position and lying, and profile picture in the prone position are conducted. According to X-ray value of the angular deformity in the coronal plane on Fergusson or Cobb techniques are calculated; ray morphological changes of vertebra are clarifyed (Fig. 3.6.10).

By Fergusson the centre of the vertebral body at the top of the curvature and center of the neutral vertebrae above and below the arc of the curve are marked. These points are connected by straight lines; the angle of intersection of which matches the value of curvature. According to Cobb method lines on X-ray are performed which are parallel to the upper and lower surfaces of the neutral vertebra above and below the arc of the curvature. The intersection of perpendiculars of these lines an angle forms, which is equal to the value of curvature (see Fig. 3.6.10)

Determination of the angles of the deformation helps to determine the nature of curve arches — primary and secondary. Typically, the primary arc curving is bigger than secondary, is fixed, there is a torsion of interested vertebra. Above and below the primary arc secondary compensatory curvatures of the spine are defined.

According to localization these types of scoliosis are distinguished:

— Cervical thoracic (upper chest) with the localization of the primary vertex of the arc of curvature at the level on Th_{IV}–Th_{V} vertebra;

— Thoracic scoliosis with the top level on Th_{VIII}–Th_{IX} vertebrae (can rapidly progress to severe functional disturbances of breathing and cardiovascular system);

— Thoracolumbar with top level distortion on Th_{X}–Th_{XI} vertebra (prone to progression with impaired respiratory function and cardiovascular system, accompanied by pain syndrome);

— Lumbar with the top of curvature on the level of L_{I}–L_{II} vertebra (differs by mild course, rarely has severe degree of deformation);

— Lumbosacral, during which pelvis is included to an arc curving, creating a bias with an elongation of one foot (may be accompanied by pain);

— Combined (S-shaped) type of scoliosis is characterized by two primary curve arches at Th_{VIII}–Th_{IX} and L_{I}–L_{II} vertebra (is characterized by stability; formation of deformation at the level of L_{III} indicates on compensatory curvature during thoracic scoliosis).

On X-ray vertebra torsion is determined due to displacement of the shadow of the spinous processes from the center line in side of concavity of the curvature. During increasing of the torsion around the longitudinal axis kyphoscoliosis is formed, that is a combination of curvature in the frontal and sagittal planes.

To assess the severity of scoliosis classification system proposed by V. D. Chaklin is used:

1. I degree — angle of curvature on X-ray is up to 10° (170°) during the vertical position, during the horizontal — it is reduced or eliminated, moderate vertebra torsion is defined.

2. II degree — angle of curvature is up to 10°–30° (155°), severe torsion, there is a compensatory arc.

3. III degree — angle of curvature is up to 30°–50°(140°), there are chest deformity and rib hump.

4. IV degree — angle of curvature is bigger than 50°, severe fixed deformity with the presence of anterior and posterior rib humps, stiffness in the spine.

Progression of scoliosis depends on the patient's age, the type and degree of deformation. Intensive increase of deformation is observed during the rapid growth of the child, reaching a maximum at puberty, girls at 11–13 years, boys at 14–16 years and usually ends after growth stop.

Adverse course scoliosis, which are formed to 6 years of age, more favorable — after 10, especially after 12 years.

CT, MRI, which allows making exact spondilometry, to measure bone mineral density of vertebra, inter vertebral cartilage status, the presence and size of the protrusion and drive's extrusion, etc. belong to additional tests. Electromyography (EMG) helps to determine the bioelectric activity of the muscle fibers, electrogenesis of paravertebral muscles at rest and stress, qualitative changes in the EMG-signal, the imbalance of the regulation of muscle tone and posture, and objectively evaluate the effectiveness of the treatment of neuromuscular spinal deformities.

For determination of the treatment strategy it is important to predict scoliosis course. Risk factors of the progression of deformation are:

— The age of the child (the younger the child, the worse the prognosis)

— The child's sex (girls have harder disease course)

— Syndromal of pathology

— Chest and combined type of curvature

— Decompensated forms of scoliosis

— Instability of deformation (difference between the bending angle standing and lying is more than 10°),

— Connective tissue dysplasia of the II–III degree, etc.

During congenital spine deformities progression of deformation occurs in more than 80% cases (by E. V. Ulrikh).

Treatment. Children with compensated scoliosis of initial degree should be treated in a clinic and at home, during slow progressive course — better in specialized boarding schools, during vigorous progression — in an orthopedic hospital. Conservative treatment of children with non-progressive scoliosis of I–II degree consists of a set of measures aimed at reducing the static load on the spine and correct motoring organization in preschool, school and home. Thus it is necessary to eliminate the causes that contribute spinal deformity: prolonged sitting,

wrong position in bed, poor posture while standing, carrying heavy loads in one hand, the shortening of legs, etc. Therapeutic exercises aimed for stabilizing the existing distortions, preventing its progression, combining massage, especially of back muscles, recommended sports, ball games, swimming, skiing.

Much more complex is method of treatment of scoliosis of I–II degree with progressive course. Combined treatment is aimed for arresting the progression of scoliosis and the stabilization of the pathological process. First of all, there is a need in a strict reduction of load on the spine, reducing the time of the patient sitting or standing.

Child staying for a long time in a horizontal position needs to be combined with corrective exercises, massage and orthopedic activities. Therapeutic exercises with tonic effect on the body should be aimed primarily at arresting the progression of scoliosis and compensate scoliosis by increasing the stability of the spine and developing the orthostatic position with the restoration of the balance between the curvatures of scoliosis. During this exercises for spine stretching and intense deformation correction are excluded. A set of physiotherapy exercises is defined by the physical therapy specialist for each patient or group of patients individually. The procedure of physiotherapy is performed twice a day for 45 minutes and ends with position correction — briefly putting the patient for 15–20 minutes to the side on curvature of the convex part on the soft cushion. To restore the functional capacity of back muscles and abdominal, gymnastics should be united with massage. Electrical apparatus using AFM-2 "Amplipuls" for 10–15 minutes, up to 30 treatments helps for strengthening and enhancing muscle tone on the convex side of the curvature.

Prosthetic-orthopedic measures in the complex of conservative treatment influence on correction of scoliosis. For preschool-aged children with unstable scoliosis during daytime rest and at night a plaster bed, made at the maximum correction of the spine is used. To prevent an increase of vertebral torsion during kyphoscoliosis in bed on a projection of deformation cotton-gauze pad is used. For children older than 3 years, with the unbalanced, unstable, progressive scoliosis are prescribed corset. At the I degree of scoliosis brace usually is not using. To create favorable conditions for stabilizing during hypermobility of the spine and deviation of the body from the midline corsets of fixed type is used; 7 year old children are recommended corsets with lightweight constructions. For older children during localization of the main curvature above VI thoracic vertebra tire-leather corset with the holder of the head is used; during thoracic, thoracolumbar and lumbar scoliosis fixed frame corset is widely used. Elimination of body's deflection with process stabilization, but possible progression of scoliosis is an indication of a change of this corset into functional. Functional corset, which restricts lateral movements, support spine without loss of motion in the sagittal plane.

All types of corsets first are recommended for continuous wearing, in the future — for the daytime period, which involve loading on the spine (school studies, tired muscles in the second half of the day, etc.).

The definition of the indications for surgical treatment of scoliosis in children is quite responsible. Believe that with rapidly progressing scoliosis of II degree surgery is performed. Progression of deformation on 20–30° in a relatively short time (1–1.5 years) is a poor prediction test and there is a need in stabilization of the spine. However, this depends on the type and etiology of scoliosis. Thus, early surgery leads to better results during cervical-thoracic scoliosis, which responds poorly to conservative treatment with severe cosmetic consequences, and during thoracic scoliosis, which is progressing rapidly in 70% cases of patients and is accompanied by severe disorders of the respiratory and circulatory system.

However, during dysplastic lumbar scoliosis, which has a relatively benign course, as well as scoliosis of syndromic forms of metabolic disorders (Marfan syndrome, Chernogubova–Ehlers–Danlos syndrome, and cystinosis) surgical treatment must be treated cautiously. It is used during III–IV de-

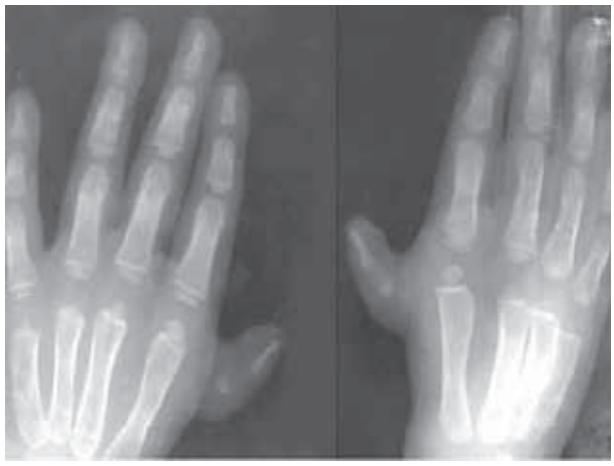


Fig. 3.6.11. Aplasia of fist thumb

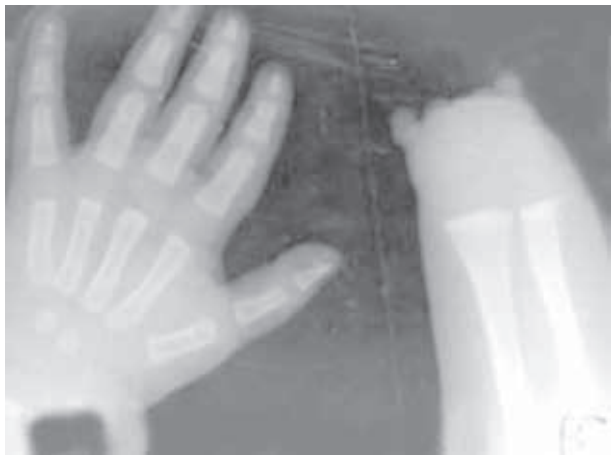


Fig. 3.6.12. Ectrodactylia

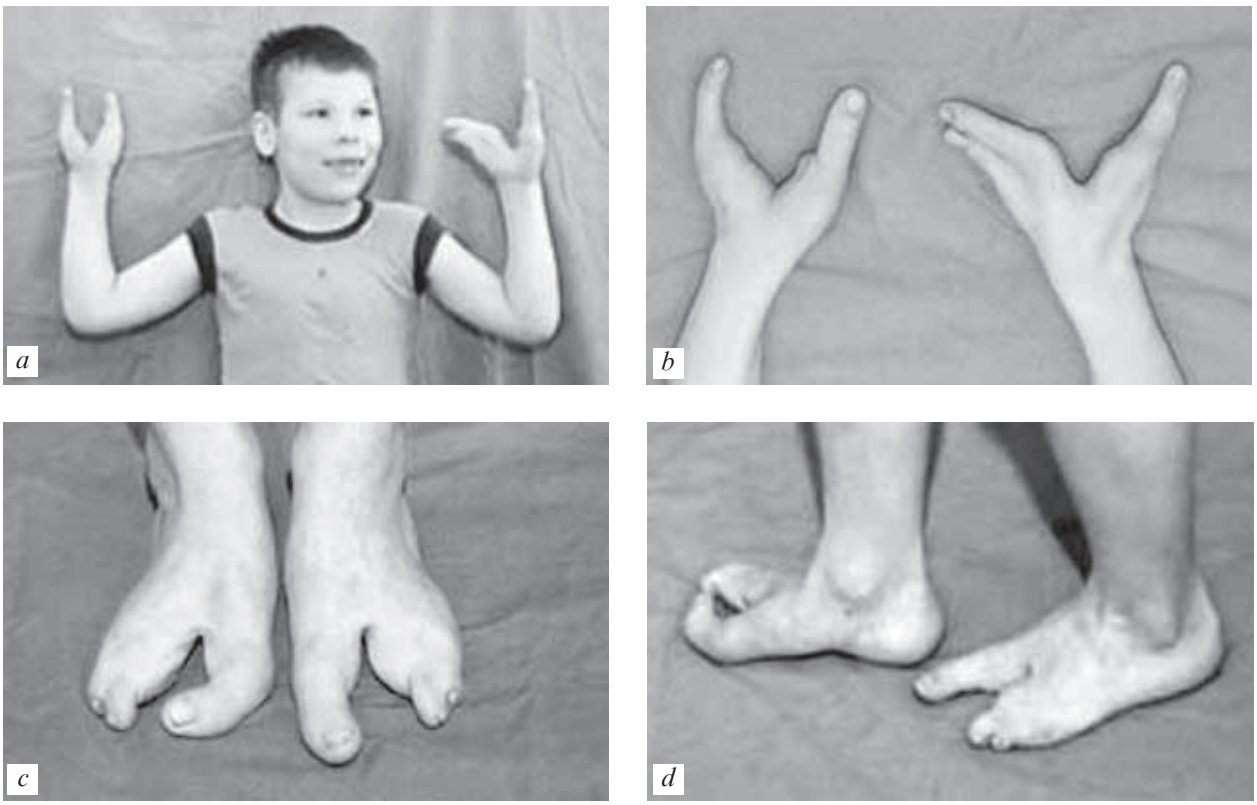


Fig. 3.6.13. Malformations of the upper and lower limbs *(a–d)*

gree scoliosis, but with more labor-intensive intervention and less effectiveness. Age of the patient for surgery is usually defined as 10–14 years; in patients aged 17–18 years, indications are very limited.

The goal of surgery is to correct the deformation and fixation of the spine. The imposition of different distractors (Kazmin, Harrington distractors), Rodniansky and Gupalov endocorrector, Shulutko tenoligamentocapsulotomy, and wedge resection of vertebra, Tsivyan vertebratomy, and enucleation of the disco belong to these operations. After the operation long resuscitation treatment is needed.

CONGENITAL DEFORMATIONS OF THE UPPER LIMB

Among the diseases of the upper limb malformations make up 7.4%. They are characterized not only by cosmetic defects, but also by severe dysfunction.

It is settled that congenital limb deformities can be both genetically caused, and as a result of the pathological action of exogenous factors on the embryo (embryopathy) or fetus (fetopathy). This leads to the possibility of the development malformations display as an independent nosological form, with aplasia or deformation of some segment of the limb

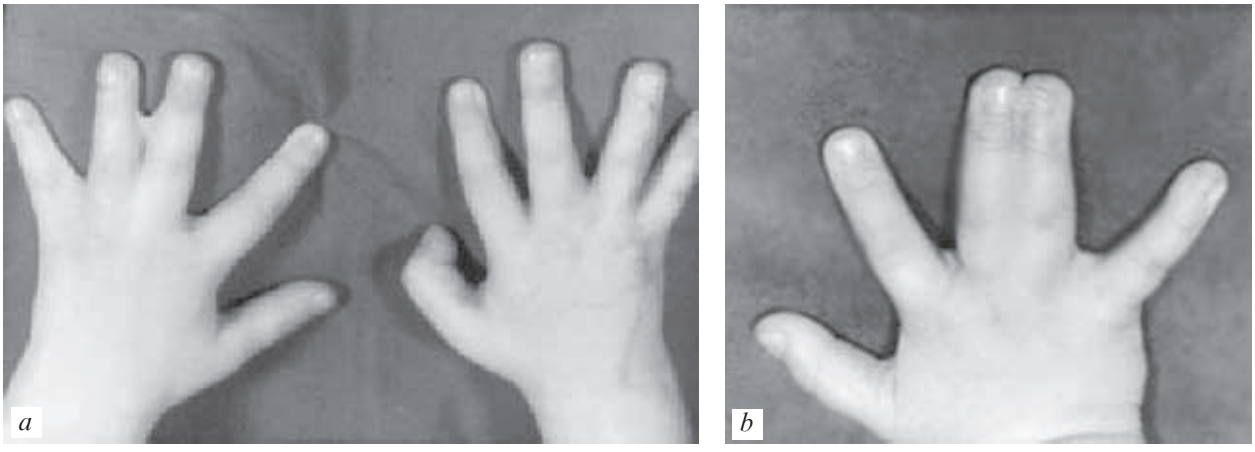


Fig. 3.6.14. Syndactyly *(a–b)*

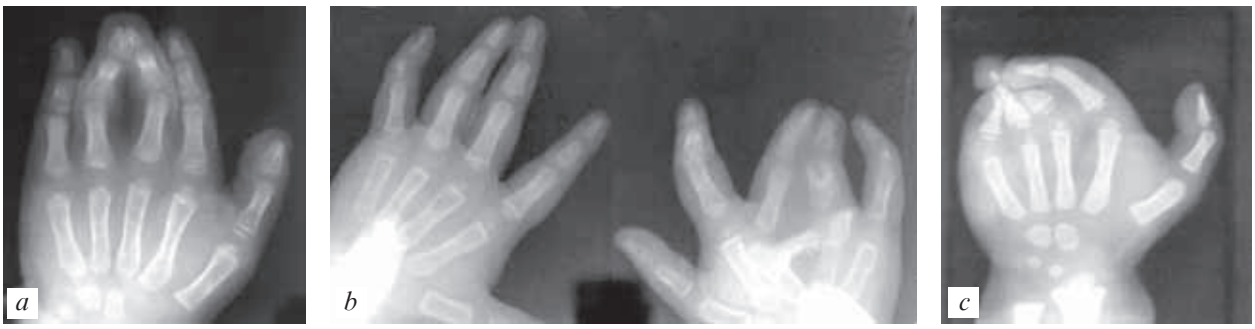


Fig. 3.6.15. X-ray during syndactyly *(a–c)*

(ectrodactyly, aplasia of phalanges, etc.), and as a complex with other diseases of syndromic pathology (Fig. 3.6.11–3.6.13).

SYNDACTYLY

Syndactyly — a partial or complete fusion of two or more fingers, the anomaly of the fingers development due to violations of their division in the embryonic period, when on the 3rd–9th week the formation and differentiation of the limbs takes place. In most cases III–IV fingers are not separated, rarely I and V with others, which can obviously be explained by the dependence of the period of differentiation violations (Fig. 3.6.14).

Clinical picture and diagnosis. During a simple form of syndactyly membranous, skin, bone and end options can be observed. During webbed syndactyly, fingers are joined together with a fold in the form of membrane, which consists of two sheets of skin; movement in the fingers is preserved in full volume, their development is not affected. Skin syndactyly — is more intimate fusion of finger without bone changes; only a joint movement of the fingers is possible (see Fig. 3.6.14).

Bone syndactyly, except fusion of soft tissue adhesions, is characterized by fusion of bonfires phalanges. During the end syndactyly skin or bone fusion of terminal phalanges is observed, often with their deformation. The complex shape of syndactyly occurs in conjunction with other malformations of the limb: poly- and oligodactyly, ectrodactyly, aphalangia with amniotic bands, brachidactyly, etc. (Fig. 3.6.15).

Treatment. Syndactyly can be eliminated only by surgery. The goal of surgery is not only to eliminate the distortion, but also to restore full function of the hand and fingers as it possible. With the growth of the child syndactyly increases along with the lack of differentiated movements, gross deformation of fingers, which increase the functional deficiency of the hand, making it training hard, limiting the choice of profession.

Operative treatment during syndactyly of fingers is advisable from 2 years of age of the child. The exception is the end syndactyly, when early surgery in the second half of first year of life prevents uneven growth of individual segments with secondary deformation of fingers.

All proposed types of surgery can be divided into four groups:

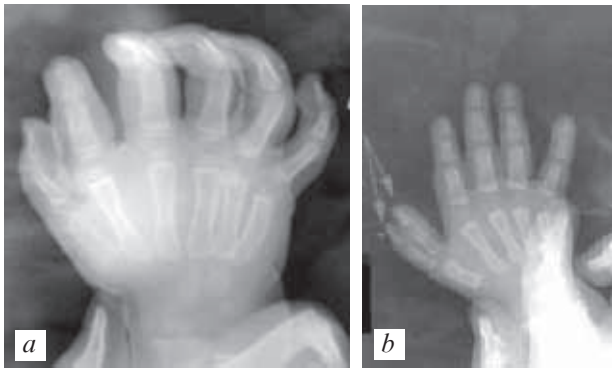


Fig. 3.6.16. Polydactyly *(a, b)*

— Simple dissection septum of interdigital septum without plasty;

— Dermepenthesis by local tissues after dissection of fused fingers;

— Free dermepenthesis by split or fully layered skin graft;

— A combination of skin plasty by local tissues with free transplantation of skin autograft.

Surgery during syndactyly can help to achieve normal anatomic structure of fingers in development and restore full function of the hand. Nowadays these problems are being solved through the use of distraction techniques in the external fixation sets. It does not require more gentle free skin plasty, provides good anatomical and functional results.

POLYDACTYLY

During polydactyly deformation is determined by additional fingers and toes, it is often hereditary, can be combined with other abnormalities — syndactyly, brachidactyly, congenital hip dysplasia, clubfoot (Fig. 3.6.16).

Clinical pricture. Extrafingers are often arranged in radial or ulnar edge of the hand, at least in general row. However, they may approach to normal structure of the size and the number of phalanges or be vestigial appendages. Most often, the deformation is observed in the form of additional little finger, which does not work, hangs on the thin skin stem, or in the form of doubling of the thumb, sometimes with a split of phalanx.

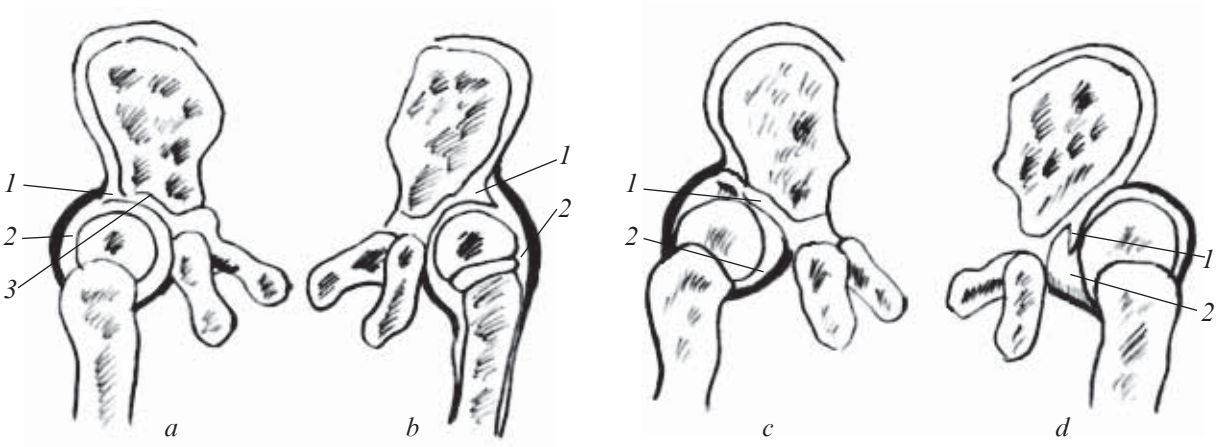


Fig. 3.6.17. The scheme of the hip: *1* — limbus, *2* — joint capsule, *3* — acetabulum *(a–d)*

Treatment is only surgical. If extra finger is hanging on a thin stalk, it is advisable to remove it within the first weeks of child's life. During normally formed additional finger, phalanges separation leads to a difficulty in deciding about the anatomical and functional disability of finger or segment. It is rational to perform surgery at the end of the first year of life, which creates the possibility of normal growth and development of other fingers.

CONGENITAL HIP DISLOCATION

Congenital hip dislocation — it's a severe degree of hip dysplasia, which is one of the most common deformations of the musculoskeletal system, which leads to disruption of statics and earning capacity of the patient. Hip dysplasia means developmental disorder of bone cartilage basis, ligament-capsular and muscular system of the joint.

Etiology and pathogenesis. Today, in the occurrence of congenital hip dislocation the fact of inheritance is undisputed. Mutagenic ability of teratogenics factors (ionizing radiation, some biologically active chemical compounds) often shows itself in one generation, rarely mutations become inherited.

Violation process of normal development of the hip joint is due to the influence of several harmful factors in embryonic and early fetal period of fetus revolution. If earlier the emphasis was made on mechanical factors (narrowness of the uterus, hypamnion, position particularity of the fetus), now the main focus is on toxic influences, effects of biological agents (viruses, protozoa, etc.), ionizing radiation, hormonal deficiency of the parent body, toxemia of pregnancy; does not exclude the value of vitamin deficiency. Disruption of the normal development of the hip joint is expressed in its flattening and irregular shape with changes in cartilage structures; slowed process of ossification and the small size of the femoral head; in turning of the proximal femur anteriorly (antetorsion); over-stretched capsule — ligament and changes of the muscles that surround the joint.

All components of underdevelopment, displayed in differend degree, affect the ratio of femoral head and the acetabulum, the formation of the type of deformation (Fig. 3.6.17).

Before-dislocation, subluxation and dislocation of the hip are distinguished. In newborn before-dislocation of the hip is characterized by maintaining the relationships in the hip joint, but stretched, relaxed ligament-capsular apparatus can cause lung dislocation and subsequent reposition of the head into acetabulum (hip dysplasia). During subluxation of the hip, head shifts upward and to the side, but does not go beyond the limbus of the acetabulum; the latter is moderately flattened and elongated. During hip dislocation joint capsule remains stretched, acetabulum flattened, the femoral head is out of the acetabulum; during the displacement of elastic limbus into the joint creates interposition of capsule and limbus. Changes in the hip joint progress as the child grows. In 15–25% of cases before-dislocation is transformed into subluxation and dislocation of the hip.

Because of the lack of lower limb abduction and the right balance in the hip joint with the time the child develops muscle contractures, deformity progresses. Flattening of acetabulum increases on account of underdevelopment of upper-back edge, thickening of the cartilage layer and growth of connective-scar tissue.

The head and neck of the hip deforms, development of head's ossification core is delayed, the neck-shaft angle is obtuse (135–145°), antetorsion increases. With the start of the static load displacement of the femoral head up and back, reaching the level of the body becomes pronounced, the middle of the ilium wing, approaching to sacro-lumbar junction (I–IV degree dislocation). Dislocation of the femoral head is accompanied by stretching the joint capsule, its restriction in the form of "hourglass"; up to 5 years isthmus is formed, which prevents reduction of femoral head's dislocation together with the deformed limbus. Together with the increase of dislocation degree, the center of weight is moved back, static suffers considerably, pathological compensatory lordosis of the lumbar spine is formed.

The frequency of congenital dislocation ranges from 3 to 8 per 1,000 births. In girls, congenital hip dislocation occurs 4–7 times more often than in boys.

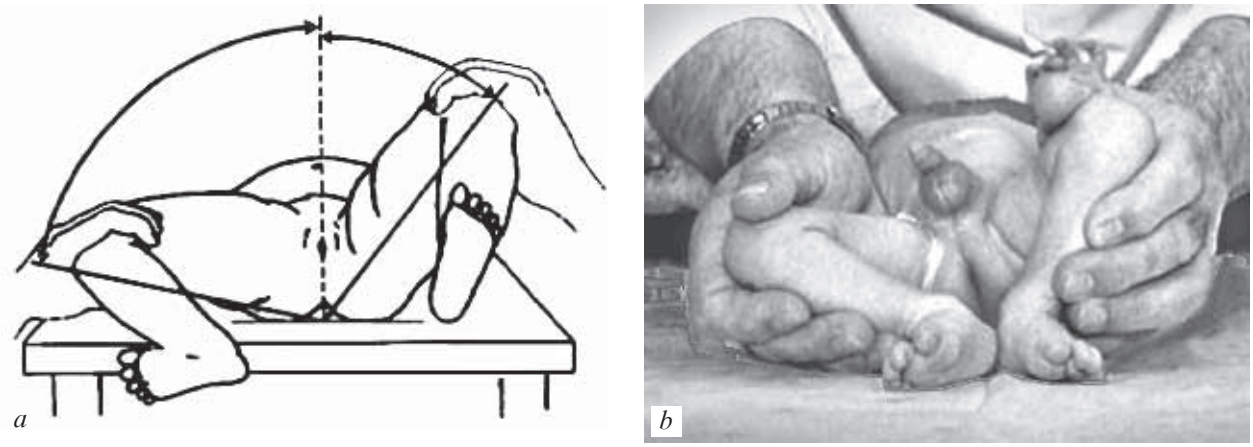


Fig. 3.6.18. Limited hip abduction symptom *(a, b)*

The frequency of unilateral dislocation dominates over bilateral. Among the infants with hip joint pathology in 74% of cases dysplasia is diagnosed, in 15% — subluxation, 11% — dislocation.

Clinical picture. Clinical displays are pretty miserable at the time of the static load. The earliest clinical sign of the disease of newborns is a symptom of *slippage, or reduction and dislocation* of the femoral head. In order to identify this trait in the child in the spine position bent legs at the knee and hip joints at an angle 90°. The first fingers doctor places on the inside and the others on the outside part of the hip, pushing the II–III fingers in the greater trochanter. During careful abduction with simultaneous traction on the hip axis of the distal course and light pressure on the greater trochanter, femoral head with a click sound reduces in a hip socket, during casting dislocating head dislocates with the same characteristic sound. Slip symptom — a sign of instability of the hip. It is typical for infants and often disappears on the 10th day, that is the time of muscle tone increasing, rarely preserves in children up to 2 months of age.

Next clinical sign of hip joint pathology is a symptom of *limited hip abduction* (Fig. 3.6.18).

For its identification in a supine position with no efforts legs raise to the sides, bent at the hip and knee joints, at 90°. Normally legs can be pulled to horizontal plane (85–90°), during dislocation abduction is limited. During unilateral congenital dislocation difference in the degree of foot abduction is marked, that is, at the same physiological muscle rigidity restriction of abduction is sharper expressed on the side of dislocation.

Adduction in young children is so expressed that the hip of dislocated limb can be freely placed on the groin of opposite leg. Rotating in and out in the hip joint can be doubled. In healthy leg rotation in and out can be up to 45°, during dislocation — up to 90–100°, the total rotation during dislocations can be 180–200°.

The sign of *skin folds asymmetry* of hips is determined in position of the child on his back, legs in hip and knee joints are fully straightened up and stretched. The front internal hip side of the healthy child is usually expressed by three deep wrinkles: groin, adductor, wheel, they are symmetrical and regular.

During unilateral dislocation, a shift of folds on the side of dislocation is proximally observed, sometimes their number increases. During the prone position attention is drown to the level of sciatic folds location, on the affected side sciatic fold is situated above. This characteristic must be taken into account only in combination with other symptoms, because sometimes in healthy children asymmetry and number increases of the skin folds on the hips are observed. During unilateral hip dislocation limb shortening is marked. In infants and children of first months of life is almost impossible to determine the leg length with measuring tape, so the difference in leg length is judged in terms of the knees location. For this purpose the child is placed on his back, legs bent at the hip and knee joints strictly in the sagittal plane and in the stop position on the changing table determine the level of knee location. On the side of hip dislocation, level of the knee joint is lower (Fig. 3.6.19).

A child who is calm, during full muscle relaxation can be observed the external rotation position of feet as a result of dislocation and pathological antetorsion. Earlier mentioned clinical signs are low fidelity, except for the symptom of slipping. Only a com-

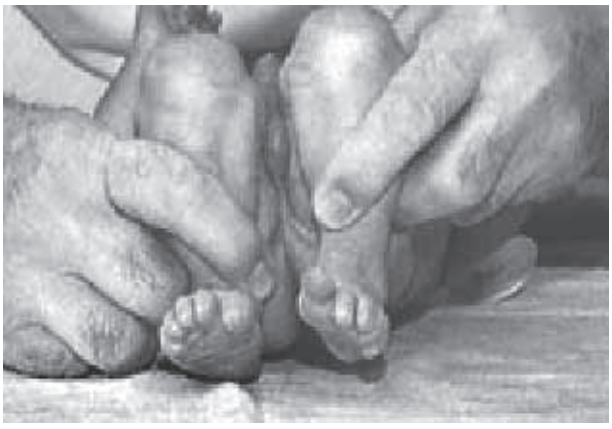


Fig. 3.6.19. Limb shortening during unilateral hip dislocation

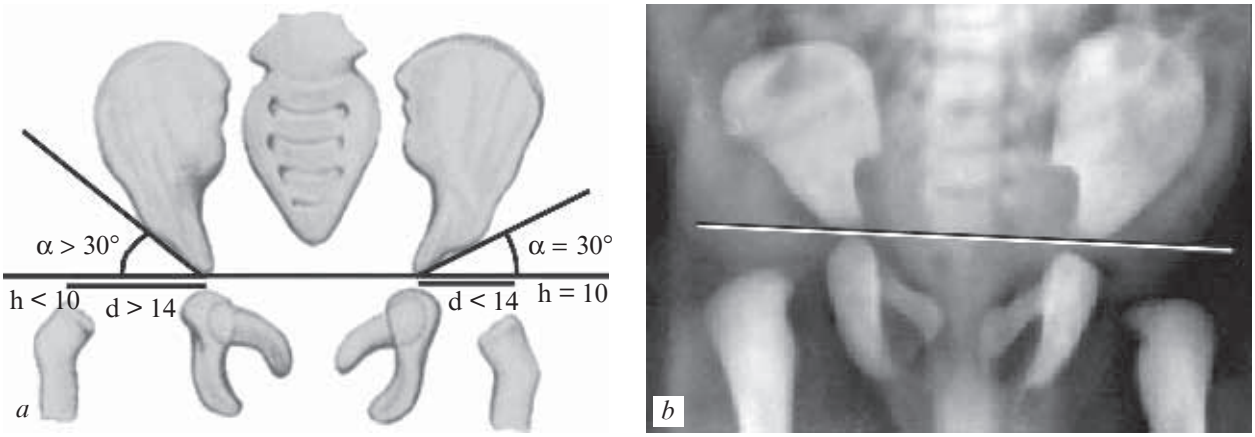


Fig. 3.6.20. Hilgenrayner–Erlaher scheme: angle α — acetabular index, formed by a horizontal Kehler line, carried out through the center of acetabular cartilage and Y shaped tangent to the roof of the acetabulum (normal up to 30°) *(a, b)*

prehensive interpretation allows to suspect the presence of congenital subluxation and dislocation of the hip.

Picture of congenital hip dislocation becomes pronounced after the static load. Children begin walking late, at the age of 1.3 years up to 1.5 years and even later during bilateral dislocation. Gait is not stable: during unilateral dislocation — lameness, during bilateral — "duck" gait. Gait disturbance in children over 5 years old is accompanied by a rapid fatigue and intermittent pain in the hip joint. Gradually compensatory lumbar lordosis forms. During unilateral dislocation relative limb shortening is observed, and during flexion contracture — functional. Absolute length of limbs is the same.

The amplitude of rotational motions of the hip is mainly increased due to external rotation. With age range of rotational motions during the hip subluxation is reduced. Maximum bent leg at the hip and knee joints during congenital dislocation reaches stomach in oblique direction, during normal knee joint does not go beyond the abdominal midline (Erlaher symptom). Duchenne–Trendelenburg syndrome is constantly defined: if a child is standing on a dislocated leg, bending the healthy leg in hip and knee joints at 90°, the tilt of the pelvis into the healthy side occurs and drooping of the sciatic folds of this side — a positive symptom; if at standing on a healthy leg pelvis does not drop and sciatic folds are at the same level, a symptom is negative. This symptom reflects the state of sciatic muscle: during the dislocation the sciatic muscle tone is reduced by the points convergence of attachment. Often reveal Dupuytren's symptoms: during the pressure on the heel area in the situation of the child on the back, mobility by axis of the limb is expressed.

Diagnosis. Early detection of hip joint pathology requires a special examination of a newborn during first 4–7 days after birth by a pediatrician and obstetrician in maternity hospital, then by a pediatrician and an orthopedist in pediatric clinic at the age of 3–4 weeks and in the future in accordance with the terms of follow-up — at the age of 3, 6 and 12 months. Diagnostic capabilities during the inspection of children on the 7th day after birth reveal brighter than in first months of life. In general, a period of timely diagnosis of children with before dislocation, subluxation and dislocation of the hip are considered the first 3 months; later periods are late.

X-ray examination has a decisive value at the early diagnosis of hip joint pathology. X-ray is performed in the direct projection in the position of the child on the back with a symmetrical position of the pelvis, with outstretched legs, straighten up in the hip and knee joints, in the middle position between the external and internal rotation. The central beam is directed through the pubic joint. During X-ray interpreting, infants and children up to 3 month age have some difficulties because skeleton part is represented by cartilage non X-ray contrast tissue. Spine bone contours provide only indirect evidence for the presence of disease. Therefore, X-ray study of hip joints is preferably performed after 3 months from birth. For the early diagnosis of hip dysplasia ultrasound perform, which allows us to estimate the state of the joint, their deviation from the norm. For the analysis of the X-ray picture Hilgenreiner–Erlacher scheme should be used, which main reference points are (Fig. 3.6.20, 3.6.21):

1) the height ***h*** — perpendicular, drawn from the femoral neck to the Kehler line (at newborns not less than 10 mm), determines the degree of proximal femur displacement up;

2) the value ***d*** — from the angular point to the perpendicular ***h*** (newborn no more than 13–14 mm);

3) the value ***d*** — Erlaher line; the distance from the medial-proximal contour of the femoral neck to the ischium (at newborn no more than 5 mm). The values ***c*** and ***d*** specify lateroposition of proximal femur;

4) the core of femoral head ossification appears at the age of 3–6 months, so only one-sided significant delay of its appearance for 1.5–2 months should be defined as abnormal.

Identifying acetabular roof bevel, the delay of nucleus of femoral head ossification appearance, lat-

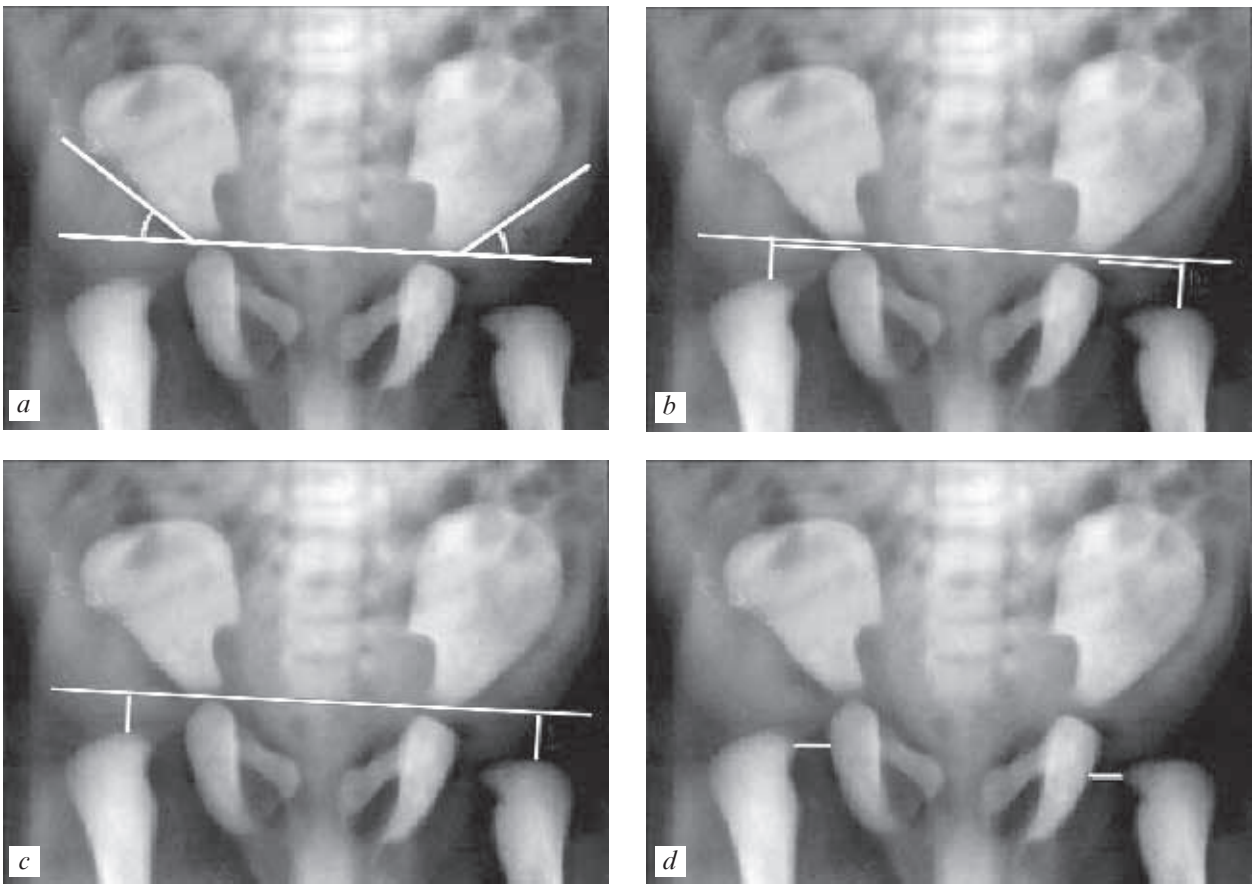


Fig. 3.6.21. X-ray. Hilgenreiner–Erlacher scheme *(a–d)*

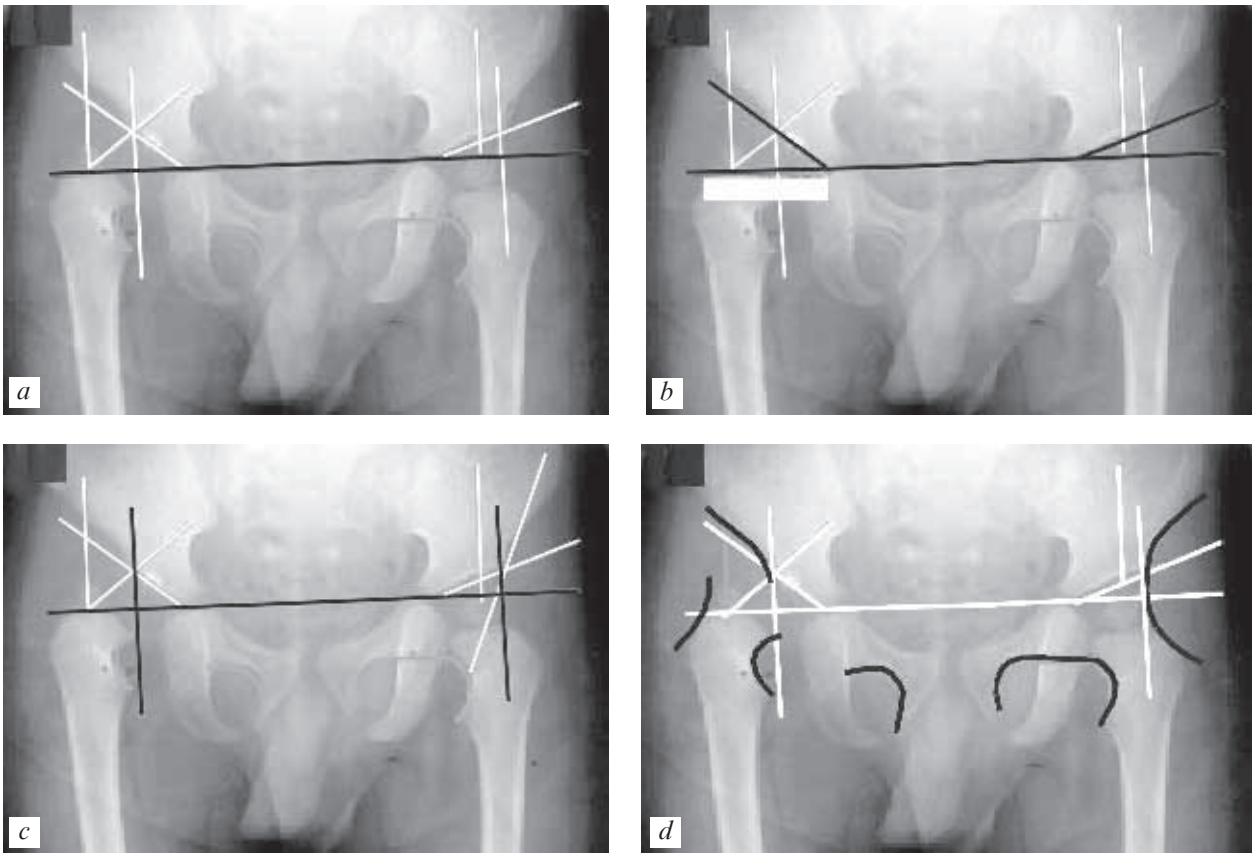


Fig. 3.6.22. Roentgen study *(a–d)*

ero- and supraposition of the hip (Putti triad) are sufficient for X-ray diagnosis of dysplasia and dislocation of the hip.

Deviation from the normal indicators on X-ray provides a basis for the accurate diagnosis of congenital chest before deviation, subluxation or dislocation of the hip. During before dislocation acetabular index is increased; during subluxation the acetabular index is increased, ***h*** value is decreased, ***c*** and ***d*** values are increased; during dislocation acetabular index is significantly increased, proximal femoral neck section is on Kehler line and over it.

For the interpretation of X-rays at older children Reinberg scheme is rational to use: the balance of femoral head and the acetabulum is evaluated based on Ombredan and Kehler lines, congruence of the articular surfaces, their deformation, structure, size of the head and neck of the femur and their relation to the diaphysis (Fig. 3.6.22).

During congenital dislocation the femoral head is above Kehler line and out of the Ombredan line, acetabular index is increased (rate at 6 months is 20–21°), Shenton and Calvet curving arcs are marked.

On the basis of X-rays the degree of deformation is determined: during I grade the femoral head is above Kehler line near the top of the sloping acetabulum (subluxation); during II degree it is located above the top edge of the basin at the level of the ilium body; during III degree — in the projection of the iliac bone wing; during IV degree — at the top of the iliac bone wing.

The differential diagnosis of congenital hip dislocation of older children is conducted with congenital and acquired varus deformation of femoral neck, pathological dislocation, epiphysis dysplasia, Legg–Calve–Perthes disease, juvenile epiphysiolysis of femoral head.

Treatment. The key to success consists in the early use of the functional method of treatment, the basic principle of which is to achieve centration of the femoral head in the acetabulum by gradual abduction and flexion of the hips, holding this position while maintaining mobility in the joint to full anatomical reconstruction of the hip. This is achieved through the use of special tools: wide swaddling, preventive pants, Frejk pads, Pavlik stirrups, abductor tires. Selecting devices and duration of treatment depends on the child's age and degree of pathology. For babies wide swaddling, preventive panties, Frejk pillow are used. After 3 months — Pavlik stirrups, later — diverting tires. Treatment includes physical therapy, massage and physiotherapy. For eliminating dysplasia swaddling with folded in several layers diapers, fixed in another diaper crotch in a triangle are enough. This allows you to keep your child's feet in the position of abduction and flexion of the hip joint. If at the age of 1 month slipping symptom remains, and the X-ray determines partial concentration of the femoral head in the acetabulum, it is necessary to start treatment in Pavlik stirrups. In addition to wide swaddling massage, therapeutic exercises are recommended.

First exercise: child's legs are bent at the hip and knee joints, divert to the changing table without effort, then in reverse position and straighten; *second exercise*: in position of bent at the hip and knee joints of the feet with the breeding rotational movements with easy force on hip axis are performed. Exercises are performed 4–5 times a day before feeding up to 8–12 exercises per session.

For hip subluxation treatment of children after the age of 1 month Pavlik stirrups are used, then after 6 months to 1 year has worked well constantly fixing tapping bus-strut (Fig. 3.6.23).

The method is simple: in the lead and legs bending position at the knee and hip joints in the lower leg above the ankle joints is applied cuff of soft tissue, they are fixed with stick-strut (25–30–35 cm), wrapped in the same cloth. During 3–4 weeks centration of the head in the acetabulum occurs. Term fixation is 3–5 months. No complications are observed. For the treatment of subluxation other devices are also used: Vilenskiy splint, splint of vinyl plastic, the splint CITO.

Treatment of congenital dislocation of the hip is a more difficult task. From the neonatal period up to 6 months treatment is carried out by the functional method with permanently fixing tire-strut for 4–6 months. It is possible to above-mentioned devices. Usually re-

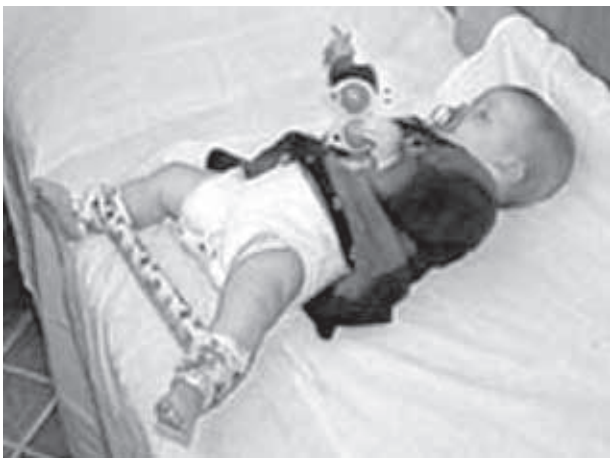


Fig. 3.6.23. Bus-strut

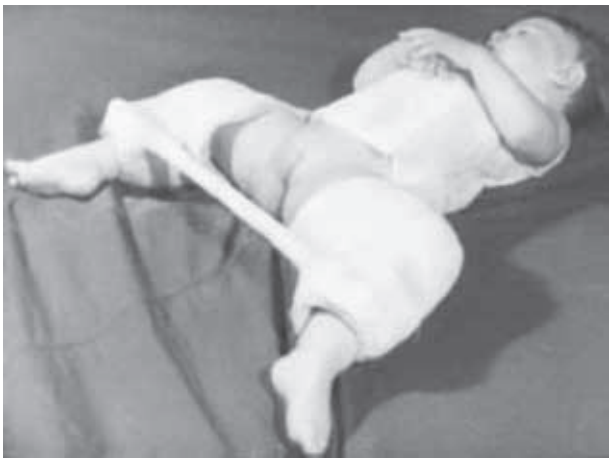
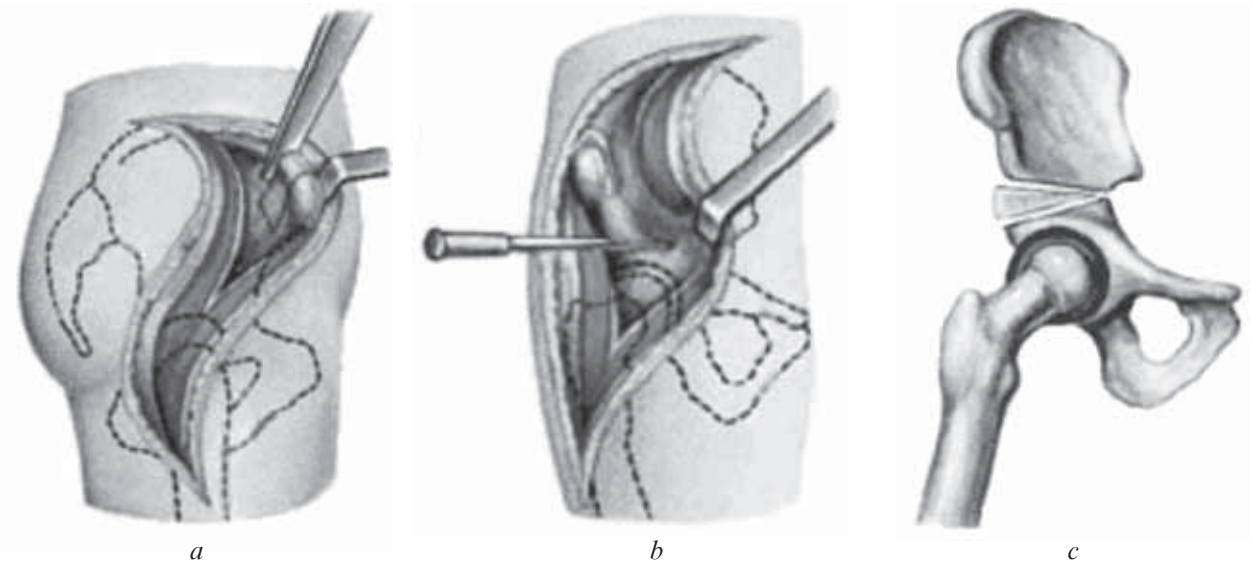


Fig. 3.6.24. Plaster cast in Lorenz position



a *b* *c*

Fig. 3.6.25. Surgery scheme *(a–c)*

duction — centration of the femoral head is produced by 3–4 weeks, which is controlled by full abduction (90°) of hips and X-rays. After centration of the femoral head in the hollow after 1–1.5 months massage of sciatic muscles, gymnastics are used. If within 2 months dislocation on diverting tires can not be straightened, it is advisable to go to a gradual reduction through functional lightweight cast. In case of delayed diagnosis of hip dislocation from 6 to 12 months a combination treatment is recommended: during a month of using outlet tire-strut for gradual overcoming retraction of adductor-muscles, then the use of a lightweight functional plaster cast for reposition of dislocation and retention of the femoral head centered in the cavity.

Methods of applying a lightweight plaster cast: on the baby's legs, bent at the hip and knee joints up to 90°, from the upper thigh to the lower third of the leg cotton-gauze pads and plaster bandages are applied, baby's legs with no effort are diverted and a plaster cast is stuck to record-strut (during high dislocation acsilar position is attached).

After 5–7 days allocation is increased by the change of stick-strut. After 3–4 stages a femoral head is typically reduced in acetabulum, which is confirmed clinically (pronounced and symmetrical femoral-sciatic folds) and X-ray. After 5–6 months bandage is removed, but for 2–3 months child should be in a tire-strut or Vilenskiy tire. Massage and physiotherapy are needed.

In children of 1–2 years old are very hard to achieve self-reposition of congenital dislocation of the hip on the tires. Most orthopedists recommend temporarily closed reduction by Lorentz, Zatsepin, Shantz, Schneider methods. Manipulation is performed under anesthesia. After the reduction of dislocation circular plaster cast is imposed in the "Lorenz I" position (bends of the legs at an angle of 90° in the hip and knee joints during full abduction of the hip to the frontal plane), in the treatment process is created "Lorenz II" position — leg's extension at an obtuse angle; "Lorenz III" — legs fully straightened up and settled aside a bit (Fig. 3.6.24).

Fixation period lasts for 8–12 months. By S. D. Ternovskiy and M. V. Volkov method, in order to prevent avascular necrosis of the femoral head and persistent contractions, after the reduction of dislocation in a month the front part of the pelvis of plaster cast is removed. The child is able to sit through the movement of the acetabular head around the fixed hips. The bandage is removed after 5–6 months; after the massage and physical therapy during a month, a child is allowed to walk.

Conservative treatment (if necessary) provides the best results. Complete anatomical and functional recovery of the hip can be reached during the treatment within the first month of life in 100% of cases, at the age of 2 to 6 months — in 89.2%, from 6 to 12 months — 78%, 1–2 years — in 57.2%. Surgical intervention is a very traumatic event; during congenital dislocation of the hip, it does not fully eliminate the anatomical and functional changes.

Methods of surgical treatment of congenital hip dislocation are divided into two main groups: intra- and extraarticular (Fig. 3.6.25).

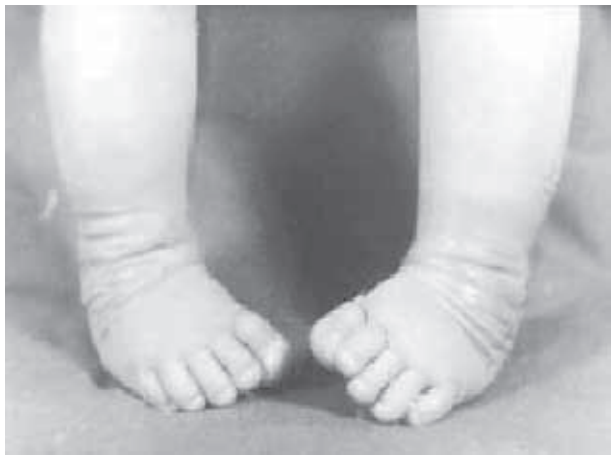


Fig. 3.6.26. Congenital clubfoot

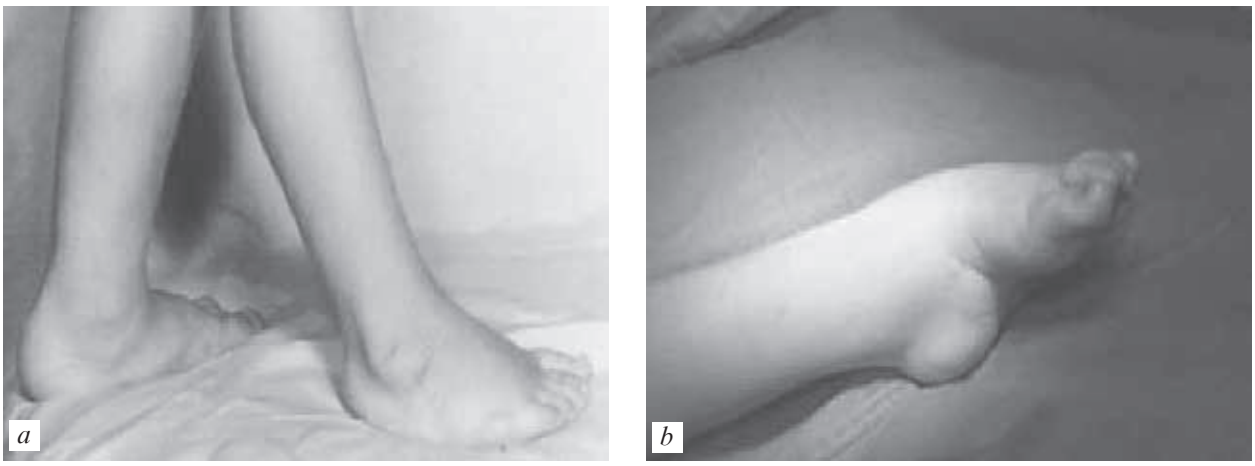


Fig. 3.6.27. Clubfoot *(a, b)*

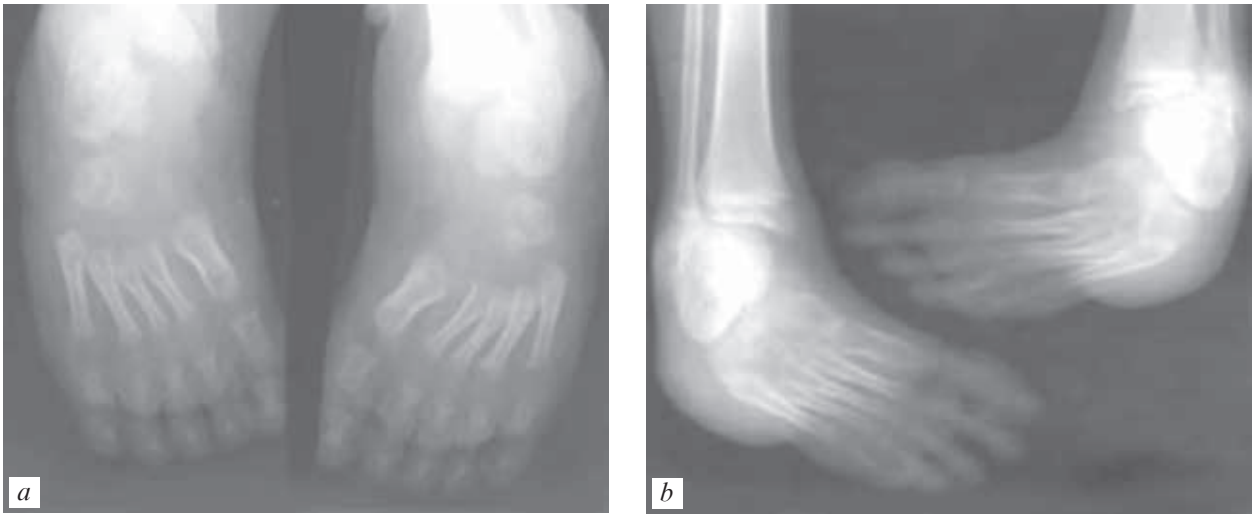


Fig. 3.6.28. Clubfoot on the X-ray *(a, b)*

CLUBFOOT

Clubfoot is a stable deformity of the foot. It can be congenital or acquired.

Congenital clubfoot

Most researchers associate occurrence of deformations with the influence of endogenous and exogenous pathological factors (amnion fusion with the surface of the embryo and pressure of amniotic banners, umbilical cord, uterine muscles, toxemia of pregnancy, viral infection, toxoplasmosis, toxic effects, vitamin deficiency, etc.) on embryogenesis and early fetal period of fetal development. Hereditary character of disease is established.

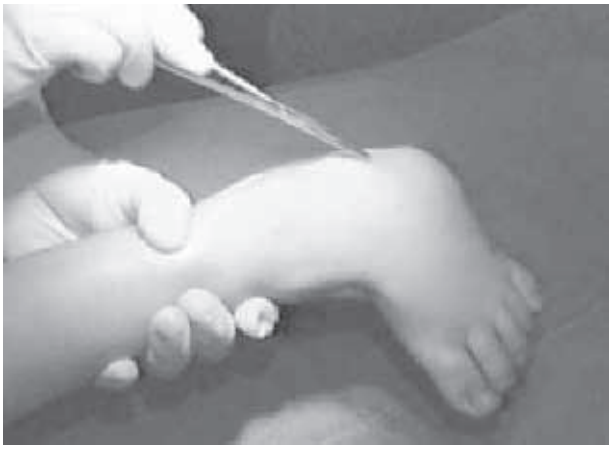


Fig. 3.6.29. Clubfoot

Congenital clubfoot is shown by dysplasia of bone ankle articular surfaces, especially captains, the joint capsule and ligaments, tendons and muscles — with their shortening, underdevelopment, and offset of attachment points (Fig. 3.6.26).

The frequency of children's birth with congenital clubfoot is 0.1–0.4%, 10–30% of this pathology is combined with a congenital hip dislocation, stiff neck, syndactyly, etc.

Clinical picture and diagnosis. Wrong position of child's foot is determined from birth. Deformation during congenital clubfoot consists of such components (Fig. 3.6.27–3.6.29): plantar flexion of the foot (*pes equinus*), supination — rotation of plantar surface medially with lowering the outer edge (*pes varus*), bringing the anterior section (*pes adductus*) with the increase of the longitudinal arch of the foot (*pes excavatus*).

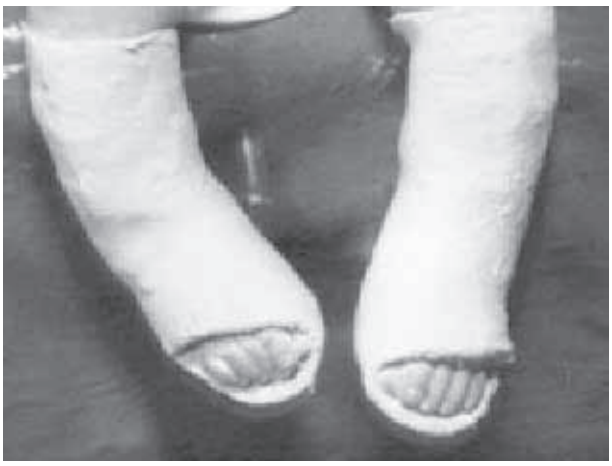


Fig. 3.6.30. Plastering during clubfoot

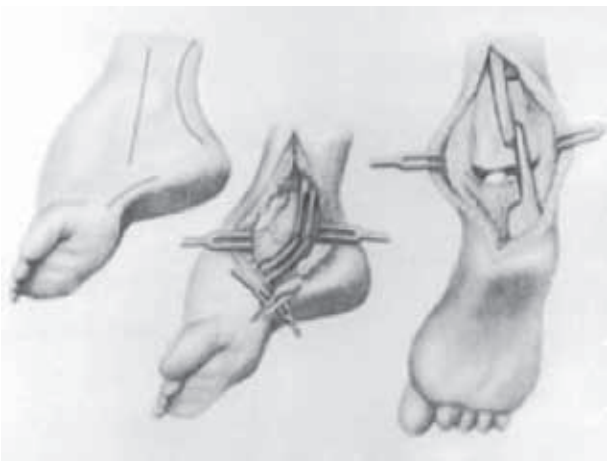


Fig. 3.6.31. Scheme of operative treatment

With the age of the child deformation increases, strain malnutrition of leg muscles appears, internal tibial torsion, hypertrophy of the outer ankle, placing the head of the talus from the outer back side of the foot, the sharp decrease of the inner ankle, varus deviation of the toes.

Due to the foot deformation, children begin to walk later. Gait with a support on the back-outer surface of the foot is typical, during single-sided deformations — lameness, during two-sided — walking with small steps in children of 1.5–2 years age, in older children — with stepping through the opposite deformed foot.

In 7–9 years, children begin to complain of fatigue and pain during walking. Depending on the opportunity to make a passive correction of foot deformation the following degrees of congenital clubfoot are distinguished:

I degree (mild) — deformity components are easily removed and can be eliminated without much effort;

II degree (moderate) — a movement in the ankle joint is limited, determined at correction springy resistance mainly from the soft tissue, which prevents the removal of some components of the deformation;

III degree (severe) — movements in the ankle and foot are severely limited, deformity correction with hands is not possible.

Provided typical form of congenital clubfoot should be differentiated from atypical during arthrogryposis, amniotic shin band, *spina bifida aperta* during myelodysplasia. During arthrogryposis together with the deformation of the foot by type of clubfoot from birth are observed contraction and deformations the knee, hip joints, often with a dislocated hip, flexion contractures of the upper limb, usually the wrist joint. Amniotic bands are formed by adhesion of the amnion with different parts of the fetus, often causing spontaneous amputation or deep circular retraction and deformations of the distal section (on lower leg on the type of deformity) with functional and trophic disorders.

Treatment. Elimination of deformation of congenital clubfoot starts from first days of life by conservative methods. The basis of conservative treatment is manual correction of deformations and retention of achieved correction. Manual correction of deformation consists in redressing gymnastics, consistent correction of foot deformation components: adduction, supination and equinus. During mild degree of deformation corrective exercises is carried out before serving for 3–5 minutes, finishing it with leg and foot massage, and repeat 3–4 times a day. Foot after gymnastics is held in corrected position by a soft flannel cloth bandage or fustian (length of bandage is 1.5–2 m, width 5.6 cm) by the Finc–Etting method.

In moderate and severe deformations the above mentioned method should be used as a preparatory stage for the treatment by milestone corrective plaster casts. Treatment is carried out by orthopedist of the clinic from two weeks of age. The first plaster cast-bootee is imposed from the toes to the knee joint, without correction of the deformity (Fig. 3.6.30).

In future, with each change of the plaster bandage in 7–10 days, successively eliminate varus and adduction, then the plantar flexion of the foot.

To correct the deformation, the child is placed on the abdomen, legs flex at the knee and by hand fix for the heel and lower third of the leg, by second hand with a slight movement without effort, slowly, gradually stretching the soft tissues and ligaments of the foot, performing the correction. Plaster cast is put on the leg with cotton-gauze pad. Layers of plaster bandage are applied loosely, by a circular motion against the direction of the deformation, from the outside of the foot on the back surface inside, carefully modeling bandage. It is important to monitor the toes. Deformation is eliminated in 10–15 stages depending on the degree of clubfoot. Then in the overcorrection position of a foot a plaster bootee is superimposed for 3–4 months, changing it every month. After removing the plaster boots recommend massage, therapeutic exercises, physical therapy (warm baths, paraffin or ozokerite applications). It is allowed to walk in the shoes with a padded on the entire surface of the sole by the pronator. To hold the foot in the correct position for the night splint put in plaster or polymer materials (polivik). During fail-

ure of conservative treatment in children older than 6 months as well as in late diagnosis conduct surgical treatment — tenoligamentcapsulotomy by T. S. Zatsepin (Fig. 3.6.31).

After surgery fixing with a plaster cast for at least 6 months is needed, then massage, physiotherapy therapeutic exercises and wearing orthopedic shoes for 2–3 years. Older children (12–14 years) may require surgery osteoarticular apparatus: the crescent and wedge corrective osteotomy, arthrodesis of the small joints of the foot.

Earlier conservative treatment can get about 90% of the favorable results. To judge about the total correction of congenital clubfoot can be no earlier than after 5 years. Dispensary supervision of children up to 7–14 years of age is necessary.

Acquired clubfoot

It occurs less often than congenital. Its causes may be damage to the bones of the foot and distal metaepiphysis shin bones, burns, acute and chronic inflammatory processes sluggish and spastic paralysis, etc. It is therefore necessary not only to determine the type of deformation, but also to clarify the anamnesis and carefully examine the nature of previous disease. Choice of medical management in each case is individual. Acquired clubfoot is not treated by plaster bandages and redressation.

During paralytic clubfoot due to damage to peroneal nerve and muscle polio, deformations are eliminated by restoring tendon and the damaged nerve or tendon transplanting anterior or posterior tibial muscle on the outside of the foot, in older children, a favorable result gives a subtalar and heel-cube-shaped arthrodesis.

During cicatricial post-burn deformities it is necessary to remove the cicatrical, restoring the tendons and skin autotransplantation. Post-traumatic and postosteomielitic deformation persists corrective osteotomy with bone-plastic operations, often using individual design of the Yelizarov device.

American professor Ignacio Ponseti in 1948 has developed a unique method of conservative treatment of this disease, and has used it for more than 50 years. Ponseti method is a special treatment technique based on specific manipulations of the foot, followed by the imposition of plaster casts and subcutaneous intersection of Achilles tendon. This method is the gold standard for the treatment of clubfoot in many countries. Treatment can be started from a 2–3 weeks aged baby. 5–7 plaster bandages are superimposed with a week long intervals.

Bandages are superimposed from the toes to the upper thigh in flexion in the knee joint on 90°. Applying a series of plaster casts gradually manage to remove all the elements of clubfoot except for equinus (plantar flexion of the foot), which is eliminated by simple surgical procedure. The foot is removed from the cycle position, and the last plaster bandage is imposed for 3 weeks, during this time there is a restoration of Achilles tendon.

Subsequently, after removal of the dressing, to prevent recurrence, the child needs to wear a special abduction brace (orthopedic shoes). Thanks to method of professor Ignacio Ponseti child is fully cured from clubfoot during 2–3 months. There is a special technique for the treatment of clubfoot in children older than three years.

3.7. MALFORMATIONS ACCOMPANIED BY CARDIOVASCULAR INSUFFICIENCY

The specific objectives

1. To study the features of heart embryogenesis.
2. To learn the features of circulation in the fetus.
3. To study normal anatomy of the heart, normal intracardiac hemodynamic parameters.
4. To learn classification of congenital heart defects (CHD) and MS.
5. To learn tetralogy of Fallot (TOF) clinical picture.
6. To learn clinical picture of interventricular septum defect (IVSD).
7. To learn clinical picture of interatrial septum defect (IASD).
8. To learn clinical picture of coarctation of the aorta (CoA).
9. To learn clinical picture of patent ductus arteriosus (PDA).

FETUS BLOOD CIRCULATION

During pregnancy, fetal blood circulation with primitive form undergoes significant changes (Fig. 3.7.1). Blood from placenta flows through the umbilical vein and enters the venous (Arantsiyev) duct and liver vasculature. From several branches of Arantsiyev duct arterial blood goes to fetus liver. Arantsiyev duct flows into inferior vena cava (IVC), in which venous blood from the lower body of fetus is mixed and already circulates arteriovenous on a grosser circle of circulation. On admission to the right atrium blood is divided into two streams: main amount enters the left atrium through the oval hole, and its smaller part plus superior vena cava blood and coronary sinus enters the right ventricle. Antenatal flow through the pulmonary artery makes up only 12% of blood flow. The main volume through the patent ductus arteriosus (PDA) flows to descending part of the aorta.

Radioisotope method research found that blood from hollow veins still mixes in the right atrium. Thus, none of fetal tissues except the liver is not supplied with blood, saturated on more than 60–65%.

Blood from the right ventricle is also divided into two streams: a large amount of blood comes into arterial duct and then into aorta, and smaller amount passes through lungs. Lung function in the fetus performs placenta, from which umbilical vein delivers blood to the fetal liver, on 80% saturated with oxy-

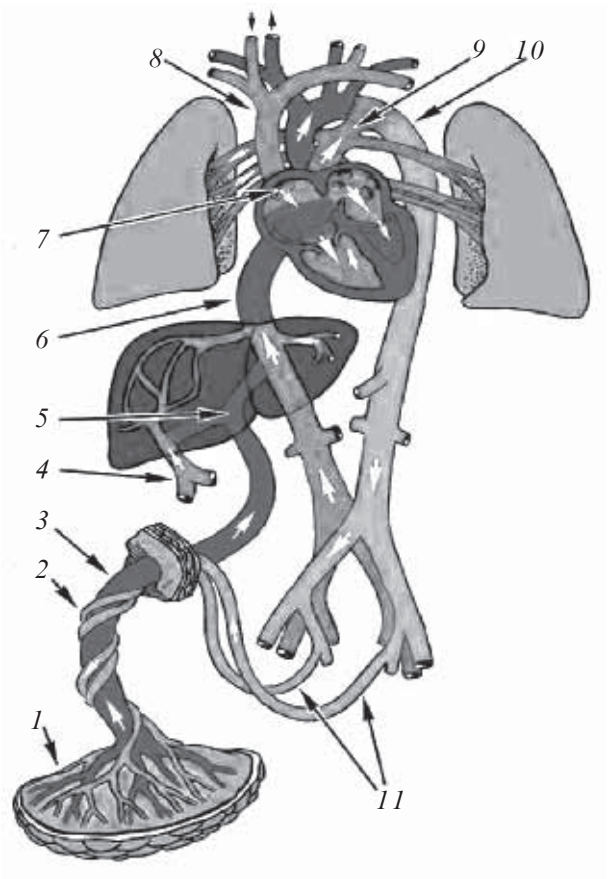


Fig. 3.7.1. Scheme of fetus blood circulation: *1* — Placenta; *2* — umbilical artery; *3* — umbilical vein; *4* — Hepatic portal vein; *5* — venous (Arantsiyev) duct; *6* — inferior vena cava; *7* — oval hole; *8* — superior vena cava; *9* — arterial duct; *10* — aorta; *11* — hypogastric artery.

gen. Oval window and Botalli duct act as bypass grafts, allowing blood from hollow veins, bypassing lungs, to flow into the system of systemic circulation. The peculiarities of fetal circulation are the conditions under which, despite the presence of significantly reduced pulmonary blood flow, the work of two ventricles is kept the same. Thus, the balance of right and left heart segments is maintained during fetal life. With the development of lungs — the period lasts from 24 to 38 weeks of pregnancy — the amount of blood through Botalli duct decreases.

None of the pump works so long as the heart. This "small organ sized as clenched fist" (quoted by William Harvey) supplies blood throughout the entire body during man's life.

It should be noted that right and left halves of the heart are connected in parallel, not sequentially, as in adults, and therefore the same amount of blood in both halves of the heart is not a big deal. It has been proven that the left ventricle of fetal pumps blood by 20% less than right.

The main feature of fetal heart is that both ventricles pump blood into aorta, into systemic circulation. Small heart of fetus allows providing tissues and organs with such amount of blood that excess in two, three times the bloodstream of adults.

Arterial blood is marked with red color; blue — venous; red with blue dots — mixed blood, similar in composition to the arterial; blue with red dots and purple — mixed blood, similar in composition to the venous (level of carbon dioxide is slightly less in blood, marked with purple color).

CONGENITAL ANOMALIES OF HEART AND GREAT VESSELS

Congenital heart defect can lead to different degrees of dysfunction of blood circulation and have a wide range of clinical manifestations. CHD vary widely and have few consistently recognizable groups of signs or symptoms. Many of these defects vary with age: pulmonary stenosis at *Fallot's tetrad* becomes more pronounced, defect in muscular part of the interventricular septum becomes lower, vascular resistance in lungs decreases.

Many newborns have obvious symptoms and signs of heart defects, while others may have very complex defects, but not detected during the first days and months of life. The most important features in diagnosis is to identify: noise in the projection of the heart, shortness of breath during feeding, poor weight gain, frequent acute respiratory infections, increased fatiguability, irritability and cyanosis of the skin. Once clinically heart defect is suspected, screening should be continued until an accurate diagnosis will be installed. Chest X-ray and electrocardiogram should be made in all cases, as a part of the primary diagnosis. For additional diagnostic screening and anatomical definition, you need more specific noninvasive studies such as echocardiography. Bidirectional color Doppler echocardiography eliminates the need to perform cardiac cavities catheterization for each child with suspected CHD. And especially in newborns, cardiac cavities catheterization can be difficult in execution and has some risk of fatal complications.

Echocardiography provides sufficient diagnostic information to decide to have the operation or not. However, in many cases with CHD, cardiac cavities catheterization and cineangiocardiography give the most accurate picture of the anatomical and physiological condition of the heart and great vessels. The procedures must be carried out meticulously accurate, as the type (palliative against radical) and consistency of operation (early vs. late) is determined by a survey.

COMMON APPROACHES IN SURGERY OF CONGENITAL HEART DEFECTS

Almost always treatment of CHD consists of their surgical correction. Medication, diet and physical activity regulation are important in many forms of CHD, but the pediatrician should recognize the severity and type of defects, recommend a treatment program leading to surgical correction in optimal time. Thorough, dynamic monitoring should prevent the development of irreversible complications in type of lung vessels sclerosis, cerebral thrombosis or development of infective endocarditis.

SURGICAL TREATMENT

CHD is fully corrected surgically. Suture ligation of PDA and closure with small patch of atrial or interventricular defects are seen as radical surgery. However, surgery of CHD, although physiologically corrective, in some cases, can result in complete recovery and return to normal status. The next occurrence of arrhythmias, ventricles dysfunction, and valve failure prevents understanding of these operations from considering their complete correction. Palliative operations are intended to reduce or increase pulmonary bloodstream; thus narrowing of the pulmonary artery in infants with interventricular septum defect may decrease pulmonary blood flow and allow the child to grow so that the defect could be closed with a lower risk to life when the child becomes greater. Shunt procedure — to send more blood to the lungs, can be used in a child with obstruction of the pulmonary artery and some form of interventricular connection in anticipation of later radical correction with less risk to life. Most cardiac operations require artificial circulation (AC), which can be used in children with the same risk as in adults. So many corrective operations are performed in childhood.

In general, artificial blood circulation is usually done by draining of blood via catheters that are placed in VPV and lower PV. Blood that is drained through a catheter from hollow veins (venous blood) gets into heart-lung machine (HLM), with help of oxygenator oxygen-rich blood (arterial blood) is pumped back to the patient through a small cannula into aorta. Amount of blood fully bypasses the heart, allowing heart not to act, so that surgical correction could be made within the heart chambers. Hypothermia during EC reduces blood flow level, which returns to the patient because metabolism of body tissues is reduced when the body temperature is reduced. Thus, to perform more accurate surgical correction, the amount of blood in heart can be reduced by adding hypothermia to EC system. In some infants with CHD type aortic arch rapture, complete correction is performed with full cardiac flow stop and maintained without EC during 30–40 minutes.

In most of these cases, babies are cooled, generally to 16–18°C, using HLM to a complete stop of EC. The head is also covered with ice to increase brain cooling. Neurological complications through a complete stop of EC are rare, and occur much less after adequate surgical correction. Protection of heart during correction is performed by various methods, including a stop with fibrillation, local or systemic hypothermia and cold cardioplegic solution (crystalloid or blood), which provide protection and stop of myocardium usually at temperatures 4–6°C.

POSTOPERATIVE TREATMENT

After open heart surgery, most patients are at endotracheal intubation and are controlled by mechanical ventilation during 4–24 hours. Mechanical ventilation reduces the need for oxygen because efforts to implement breathing are medically eliminated by using muscle relaxants, although myocardial function can be improved with early extubation. Drainage catheters in mediastinum to control postoperative bleeding are usually removed the next morning. Arterial blood gases and pH are measured by using intra-arterial monitoring.

The most common complication after open heart operation — postoperative bleeding. This is due to the patient's blood heparinization during SHIK and its neutralization with protamine but efficiency is not always complete. Approximately 2–3% of these patients should be returned to the operating room because of uncontrolled bleeding. Postoperative bleeding most often occurs at cyanotic cardiac cavities defects because of relative lack of coagulation factors in connection with polycythemia.

Problems with the presence of arrhythmias and poor cardiac output — not so frequent after correction of congenital heart defects, as they occur at more severe acquired myocardial diseases. Most patients receive antibiotics intravenously because venous lines are saved, as bare areas of intracardiac structures have prosthetic materials that impose major risk of bacterial endocarditis or sepsis.

OBSTRUCTIVE CONGENITAL HEART DEFECTS

Obstructive CHD impede blood flow and increase ventricular afterload. A good representative of this group of defects is coarctation of the aorta is one of the most frequent CHD. In the absence of interventricular defect, obstructive CHD with the damage to aortic or pulmonary valve contribute to the occurrence of ventricular hypertrophy. Sudden death is not an exception in patients with aortic or pulmonary stenosis and ventricular hypertrophy degree causes concern of cardiologists because of high frequency of arrhythmias, ischemic changes in the myocardium

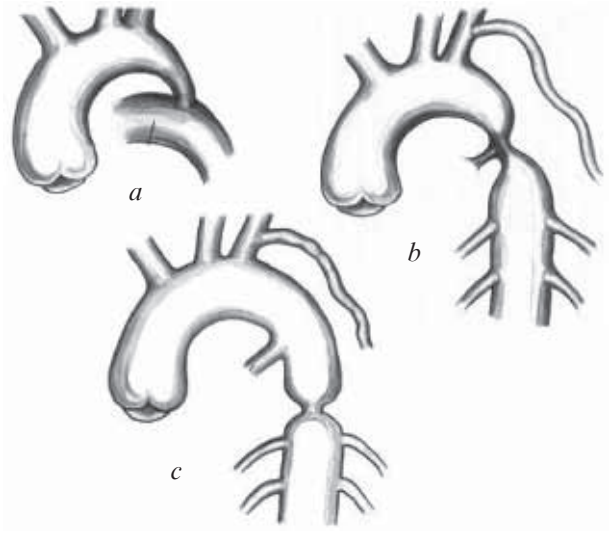


Fig. 3.7.2. Coarctation of the aorta *(a–c)*

and permanent myocardial damage or its replacement with fibrosis.

Coarctation of the aorta (CoA)

Criteria for the diagnosis:

— Newborns and infants may have severe heart failure;

— Lack or weak pulse in the femoral artery;

— Systolic pressure is higher on upper limbs than on lower extremities;

— Rough systolic murmur is heard on the back.

Coarctation of the aorta is a congenital heart defect that occurs twice as often in men than in women. 98% of all aortic coarctation is located close to aorta isthmus (Fig. 3.7.2) — aorta segment is adjacent to patent ductus arteriosus or ligament. Approximately 40% of these patients, aortic valve — bicuspid, aortic narrowing is usually well localized with external narrowing and inner luminal membrane. At coarctation systolic and diastolic hypertension in the proximal aorta and upper limbs occurs, large collateral blood vessels develop that connect branches of proximal aorta and subclavian artery to artery originating from the aorta below the coarctation. In most patients, blood flow to the lower body is not reduced, but the pulsating pressure in the distal aorta is significantly reduced, and the work of the left ventricle is increased.

Clinical manifestations. Hemodynamic effects of aorta coarctation depend on the level of open Ductus arteriosus closure, severity of obstruction, collaterals development, and combination with other cardiac defects. There are two groups of patients with these diseases: patients who had disease at early childhood and those at whom it appeared later in childhood. Infants with coarctation may have severe heart failure. More than half of these infants have combined cardiac pathology in the form of Ductus arteriosus, ventricular septal defect of interventricular septum or atrial septum defect. Some babies may actually be ductus-dependent for blood flow to the lower parts of the body, so VAP should remain open via infusion of prostaglandin E1, until the correction is performed. Rare forms of coarctation of hypoplasia type or the ascending aorta or can also be observed in this group.

Infants with severe coarctation require immediate diagnosis and surgical correction. The mortality rate within the first year of life without surgery — about 75%.

Much older children with coarctation — asymptomatic and well developed. There are complaints of headache and frequent nose bleeding. Most of these children have hypertension in the upper extremities. Many have left ventricular hypertrophy on electrocardiography.

Treatment. Operative correction of coarctation of the aorta is recommended for all patients. It is believed that in a younger age correction is made, the less severe degree of hypertension. Types of surgical correction of aorta coarctation: (1) resection with anastomosis end to end, (2) aortoplasty using flap of the left subclavian artery, (3) aortoplasty with polytetraphtoretilen (Gore-Tex) or Dacron, (4) percutaneous endovascular balloon dilatation of coarctation places.

Contradictions continue regarding the optimal surgical approach, but any method may be better depending on the anatomy and stability condition of the patient. Resection of coarctation segment with anastomosis "end to end" is recommended at newborns in the presence of severe heart failure, reduced left ventricular ejection fraction, but is commonly used in pre-school children at its isolated forms. Some forms of coarctation are performed using padge plasty, sacrificing left subclavian artery in infants. Another technique is based on correcting of aorta with synthetic material like polytetrafluorethylen or dacron. With this technique, left subclavian artery is saved and orthopedic patch can be made slightly larger than the lumen of the aorta increases with waiting for the posterior wall of the aorta enlargment. Aortic aneurysm in such plastic is possible in a remote period so you need dynamic monitoring of these patients.

Results of coarctation correction are good. Residual hypertension remains a problem, although operative correction makes hypertension a more easily controlled medication. Blood pressure may remain elevated for long time, and the question is that this state should be controlled by cardiac surgeon or cardiologist, using antihypertensives. Performance indicators of mortality are low (5–15%) even in infants, and lethal cases due to the presence of concomitant severe heart defects. Paraplegia or paraparesis is a rare but catastrophic complication, probably due to inadequate blood flow in the descending aorta and depends on the length of the aorta squizing at anastomosis formation. Mesenteric vascular spasm may occur in the days following aortoplasty. It manifests itself by increasing pressure in descending aorta after restoration of blood flow. Hypotensive medications that lower blood pressure, are shown for patients with permanent hypertension in early post-operative period or those who developed pain in the abdomen.

CONGENITAL HEART DEFECTS WITH INCREASED PULMONARY BLOOD FLOW

Approximately in 50% of all CHD blood is shunted from the systemic blood circulation to the pulmonary (reset from left to right). Frequently flaws in this group — open ductus arteriosus, atrial septum defect and defect of interventricular septum. As pliability of thick left ventricular is less than of the right ventricle, due to the fact that systemic vascular resistance is usually about ten times higher than pulmonary vascular resistance, the pressure in the left chambers of the heart and arteries in the system is higher than the corresponding pressure in the right sections of heart and pulmonary arteries. Higher pressure makes part of oxygenated blood in the left heart and aorta shunt through pathological anatomical communications and re-circulates through the lungs. Excessive pulmonary blood flow causes the

increase of pulmonary vascular resistance, ending with frequent respiratory infectious diseases and additionally loads involved ventricle (right ventricle at atrial septal defect, left ventricle at arterial and both ventricular at interventricular defects). The increased ammount of "afterload" or "preload" leads to the growth of the involved ventricle. As the ventricle expands, end-diastolic pressure increases.

Increased pulmonary blood flow gradually increases pulmonary arterial blood pressure. Although the lungs are very stretching (and therefore pliable), pulmonary arterial pressure in normal lung approximately doubles when pulmonary blood flow triples. Increased left atrial pressure because of excessive blood flow or increased left ventricular end diastolic pressure, increases pulmonary arterial pressure and pulmonary vascular resistance. If pulmonary arterioles also reduce in response to increased blood flow, vascular resistance increases (hyperkinetic pulmonary hypertension) and pulmonary arterial pressure may increase further if blood flow is not reduced. Pulmonary arteriolar vasoconstriction may be completely changed by inhalation of oxygen or intravenous tolazolin — this test is used to differentiate hyperkinetic pulmonary hypertension of sclerotic forms of pulmonary vascular disease.

In some patients, increased pulmonary blood flow and increased pulmonary arterial pressure, eventually generates a muscular hypertrophy of pulmonary arterioles media (stage 1), intimal proliferation (stage 2) and eventually hyalinization and media, adventitia fibrosis (stage 3). These morphological changes, called pulmonary vascular disease are acquired but they occur at congenital heart defects which induce high pulmonary arterial pressure and large artery-blood flow (ventricular septal defect, full atrioventricular canal, total arterial trunk) than in those defects, which generate only increased pulmonary blood flow (atrial septal defect, partial anomalous pulmonary venous drainage). Pulmonary venous hypertension and chronic hypoxemia from residence in the high mountains also promotes the development of pulmonary vascular disease. Since the area of pulmonary vascular channel is reduced as a result of morphological changes, pulmonary vascular resistance increases and the ratio of pulmonary vascular resistance to systemic vascular resistance increases. The amount of blood, which is shunted from left to right, reduces. When pulmonary vascular resistance equals or exceeds systemic vascular resistance, blood flow from left to right through the defect is terminated or becomes crossed. At Eisenmenger's syndrome, pulmonary vascular obstruction reduces pulmonary blood flow and forces blood to shunt from the right to left. Patients who developed pulmonary vascular disease (stage 3) with reverse shunting (Eisenmenger syndrome) are doomed, the only surgical care is lung transplantation or organ complex "heart — lung."

The narrowing of the pulmonary artery trunk — palliative operation designed to reduce pulmonary arterial blood flow, creating an artificial anatomic barrier for increased with high blood pressure, blood flow to the lungs. The cuff compresses the trunk of the pulmonary artery over the valve and adds resistance to pulmonary vascular resistance. Good bandage can reduce pulmonary arterial blood flow to relieve left ventricular failure and prevent rapid progression of pulmonary vascular disease. The narrowing of the pulmonary artery trunk just palliative and usually only used in very young infants, when the risk of radical correction is very high, or condition of the patient cannot allow radical surgery.

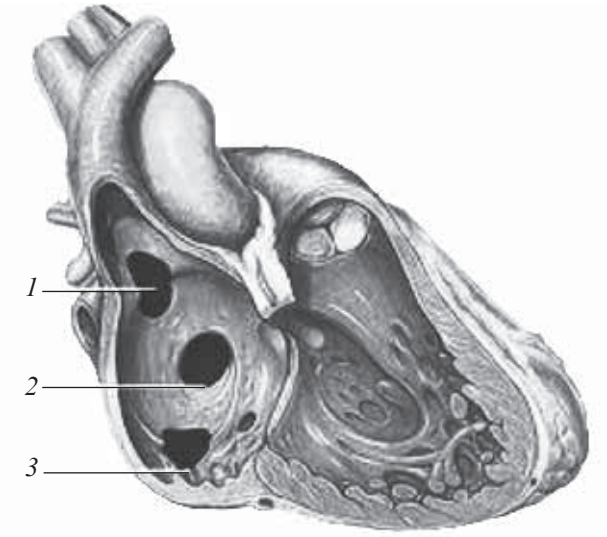


Fig. 3.7.3. Variations of secondary IASD: *1* — defect at superior vena cava; *2* — defect at inferior vena cava; *3* — secondary ACD (central)

SECONDARY INTERATRIAL SEPTUM DEFECT (IASD: II)

Interatrial septum defect (IASD) occurs as an isolated defect in 5–10% of CHD cases (Fig. 3.7.3).

In females it occurs 2 times more often than in men. Anatomically right atrium is separated from the left atrial membrane, which consists of three different parts of different anatomy and embryonic origin. From top to bottom secondary membrane, then primary and membrane of atrial channel are located. Atrial septum defects are classified on localization relative to the oval window, the embryonic origin and size. There are three types of IASD: secondary (50–70%), primary (15%) and defect of venous sinus (10%).

Criteria for the diagnosis:

— Acyanotic, asymptomatic;

— Right ventricular hypertrophy;

— Second tone is widely split and fixed.

Secondary intra-atrial septum defect is in the open oval window and can be single or multiple. The secondary defect is the easiest and certainly the largest of atrial defects. High secondary defects are often associated with partial anomalous drainage of right pulmonary veins, the so-called "venous sinus defect". Partial anomalous drainage, associated with atrial defect is considered mostly by physiological point of view, than anatomical during surgical correction. The lateral wall of interatrial septum is often absent and it is difficult to say whether the

right pulmonary veins actually enter anatomically to the right or left atrium. Patch of autopericardium is used for the correction of most of these atrial defects.

Heart failure of secondary interatrial simple defect may occur in young children, but more common in adults. Pulmonary-vascular disease is also rare but interatrial arrhythmias are marked with increasing age.

The average life expectancy in patients with interatrial septal persistent defect is reduced from right heart failure, arrhythmia and sometimes pulmonary vascular disease.

Treatment. Surgical closure is conducted in all patients with secondary interatrial defects. The mortality rate is very low, even small defects have danger of paradoxical embolism or infective endocarditis. Patch is usually used to close the defect, made of autopericardium, polytetrafluorethylen or dakron. Prediction after surgery is excellent.

PRIMARY INTERATRIAL SEPTUM DEFECT (IASD: I)

Criteria for the diagnosis:

— Acynaotic;

— Asymptomatic or shortness of breath during physical exercise;

— Widely split tone II;

— Apical systolic murmur;

— ECG shows left axis deviation.

Primary interatrial septum defect develops duringanlage of atrioventricular canal. Partial atrioventricular canal, the defect is low in interatrial septum, adjacent to the coronary sinus and the hole of tricuspid valve. Aortic (front) mitral valve is usually split and in some cases septal tricuspid valve cusp — also split. Most patients are asymptomatic, although mitral insufficiency may develop signs of heart failure more familiar at primary atrial defects than at secondary atrial defects. We recommend surgical closure of the defect. When there is mitral insufficiency, mitral valve splitting should be removed to reduce the degree of failure. Sometimes anuloplasty is achieved by decrease in the size of the valve orifice. If there is no mitral insufficiency, plastic of the valve is not carried out, and defect is closed by pericardium or polytetrafluorethylen dacron.

Prognosis. The most typical complication is damage to the conduction system during surgical correction. Long-term prognosis depends on the growth and development of the mitral valve and mitral insufficiency or increase later in the late period.

INTERVENTRICULAR SEPTUM DEFECT (IVSD)

Criteria for the diagnosis:

— Asymptomatic if the defect is small;

— Severe heart failure with shortness of breath, frequent respiratory diseases and poor growth if the defect is great;

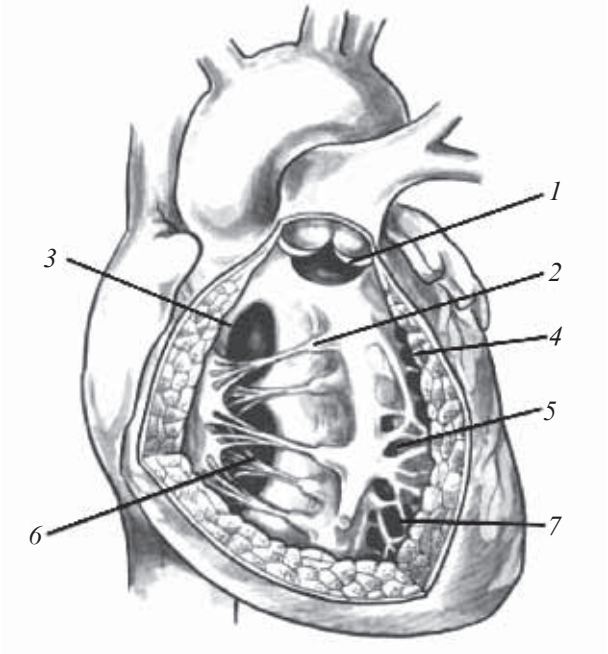


Fig. 3.7.4. Types of IVSD: *1* — under pulmonary defect; *2* — papillary cone; *3* — perimembranous defect; *4* — regional muscular defects; *5* — inferior muscular defects; *6* — flow defect; *7* — apical defect

— Pansystolic noise, maximum on the left edge of the sternum, second tone is loud with apical diastolic noise and bi-ventricular enlargement on plain chest X-ray if the defect is large.

Interventricular septum defect (IVSD) occurs in four anatomical positions of interventricular septum. Approximately 85% of IVSD occur in the membranous part of the membrane. Some IVSD are above supraventricular crest, the other below septal tricuspid valve or in muscular part of the interventricular septum. IVSD is often one of the most complex components of other CHD on type of general arterial trunk, complete atrio-ventricular communication, *Fallot's tetrad* or transposition of the great vessels with IVSD (Fig. 3.7.4).

Patients with isolated IVSD may have other abnormalities such as open ductus arteriosus and coarctation of the aorta. Patients with IVSD above supraventricular crest sometimes have aortic valve insufficiency.

Clinical signs associated with IVSD are usually like in children with increased pulmonary blood flow, ie, dyspnea during physical exercise, poor weight gain, light fatigue. Infants with major defects of the interventricular septum, usually lag behind in physical development of their peers and have chronic respiratory disease. Generally, on X-ray heart is increased and vascular pattern of lung fields is enhanced. Pulmonary vascular disease may develop as a result of high pressure and large blood flow in the lungs. Approximately one third of all patients with isolated interventricular defects has small defects and is generally asymptomatic. Many of these defects

are closed spontaneously in muscle membrane to 7–8 years.

Approximately one third of patients with large or multiple IVSD have symptoms of heart failure in infancy and require aggressive medical therapy, early topical diagnosis and surgical correction.

Optimal treatment consists of closing defect. Postoperative mortality is up to 3%. In infants weighing <3 kg, lower risk with performance of palliative operation by narrowing of the pulmonary artery trunk. In patients with multiple defects, complicated by high pulmonary hypertension in childhood is also possible narrowing of the pulmonary artery trunk, leaving radical surgery for a later time. Almost all IVSD are closed using synthetic prosthetic materials: dacron or teflon. In most cases access through the right atriotomy (perimembranous defects, *Fallot's tetrad*, transposition of great vessels) or ventrykulotomy (common arterial trunk), left ventriculotomy (with defects in muscular part of the membrane) or via pulmonary artery (defects above ventricular ridge). Diastolic murmur on the aorta is indication for early closure of septal defects located above the ventricular ridge, closure will in some cases stabilize the foundation of the aortic valve and can reduce or, at least, does not promote the growth of aortic insufficiency.

Prognosis. In general, after the operation, normalization of hemodynamic and reduce of cardiac shadows, but in patients with high pulmonary hypertension, pulmonary vascular resistance decreases slowly since the closure of defect. Thus, earlier surgery usually reduces the likelihood of lung vascular sclerosis and pulmonary vascular disease.

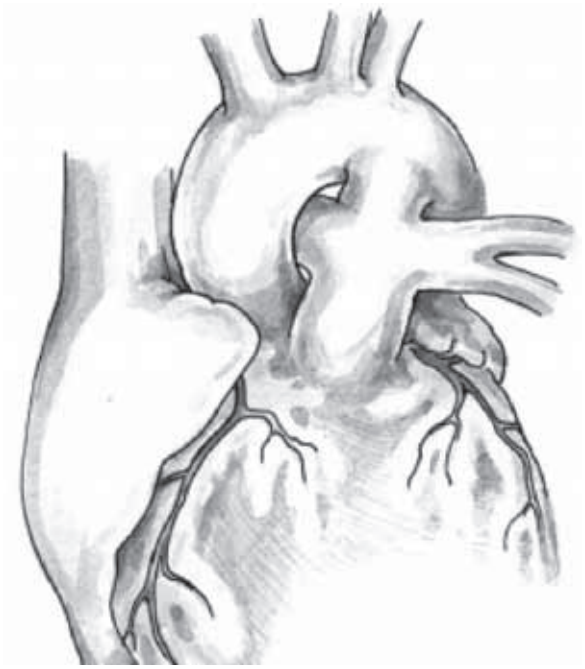


Fig. 3.7.5. PDA combines aorta with pulmonary trunk in place of bifurcation

OPEN DUCTUS ARTERIOSUS (ODA)

Criteria for the diagnosis:

— Older patients with small or moderately large ODA are asymptomatic and have continuous noise in the projection of trunk of pulmonary artery, another loud tone and limited peripheral pulse;

— Lag in physical development and frequent colds respiratory infections in infants with heart failure (wide ODA);

— Noise is usually systolic-diastolic;

— Increase in pulse pressure.

In most babies, born in time, open ductus arteriosus closes within the first days of life. The muscular component of the vascular wall PDA reduces when subjected to higher levels of oxygen in the blood at birth. For unknown reasons, the closure does not always happen. If ductus remains open and pulmonary vascular resistance decreases, pulmonary blood flow increases, leading to heart failure and stagnation in the pulmonary circulation. In most babies ductus manifested in classical "machine" systolic-diastolic noise in the second intercostal space to the left of the sternum. Functioning open ductus arteriosus is much more typical for premature babies and most of them are with symptoms of heart failure and demand its closure (Fig. 3.7.5).

Clinical manifestations. Diagnosis can be made clinically and confirmed by echocardiography study. Cardiac catheterization is sometimes used to prove the presence of PDA, but most patients will not probably require this extensive research, if not suspected of other linked CHD. Approximately 5% of full-term infants with open ductus arteriosus die of heart failure and pulmonary complications within the first year of life. The other 5% has great shunting of blood, eventually developing pulmonary vascular disease. The rest ones usually asymptomatic and ductus appears during a routine auscultation of the heart.

Treatment. Surgical closure of ductus (Fig. 3.7.6) by methods of ligation, clipping or dissection is the most effective methods of correction. Indomethacin, an inhibitor Prostoglandin E, can sometimes close PDA in full-term and preterm infants. Its effectiveness is usually observed within a week and surgical correction must be made if PDA is not closed during this time. Postoperative mortality rate is less than 1% and slightly higher in preterm babies, many of which weigh less than 1 kg. Video-endoscopic surgical techniques have recent use and successfully used at this defect, but it requires clarification. Interventional cardiology could provide new therapeutic methods.

Prognosis. In non-complicated cases long-term results are excellent.

CONGENITAL HEART DEFECTS WITH REDUCED PULMONARY BLOOD FLOW

The combination of the right heart obstruction defects and the presence of septal defect reduce pul-

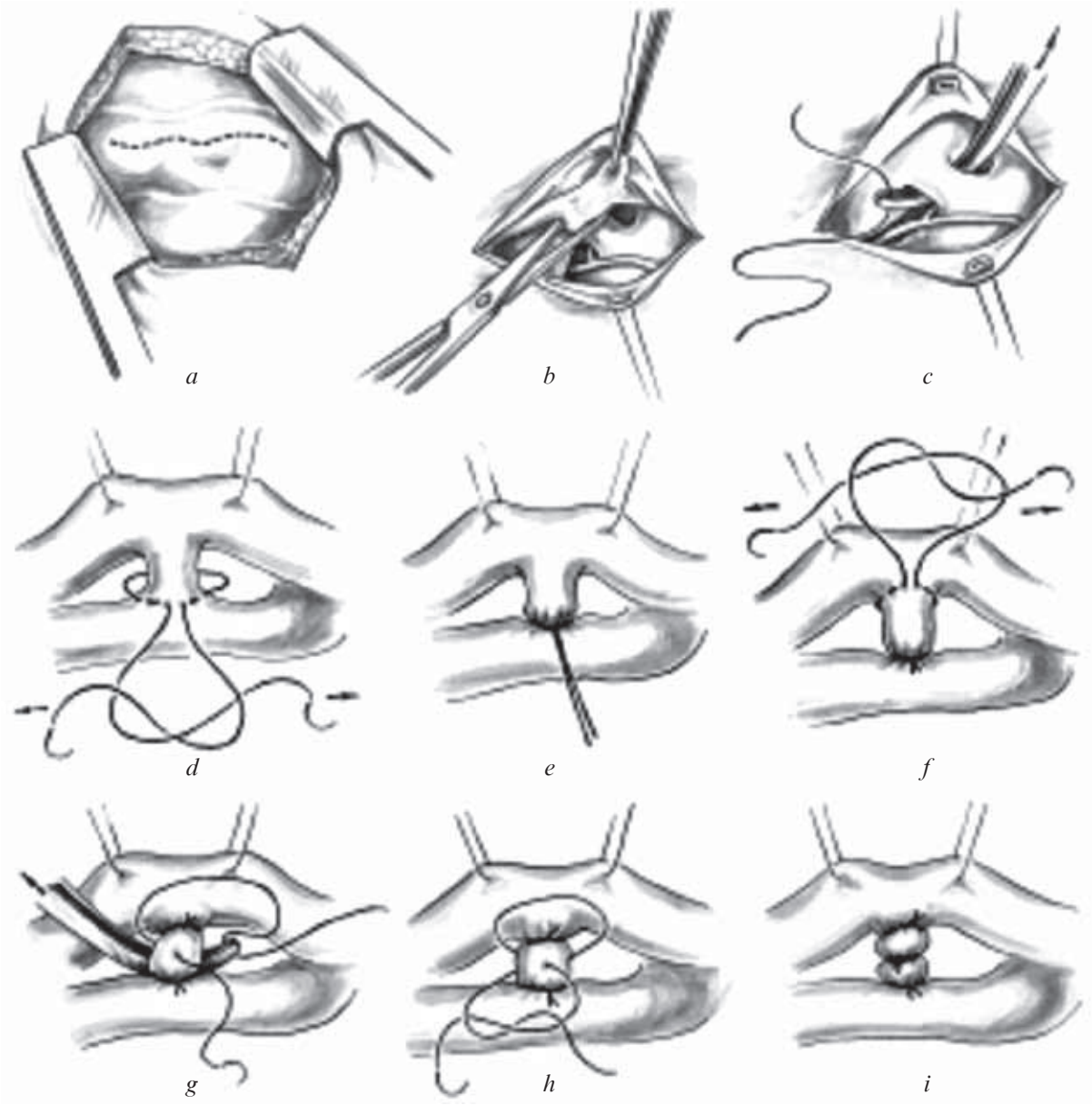


Fig. 3.7.6. Technology of PDA closure *(a–i)*

monary blood flow and cause a discharge of systemic venous blood into the systemic circulation directly, so shunting "right to left" takes place. The degree of cyanosis is directly proportional to the volume of shunt "right to left" and back, with the volume of pulmonary blood flow. Fallot's tetrad is the most severe anomaly in this group, which also includes pulmonary atresia, tricuspid atresia valve, Ebstein anomaly and other complex CHD. Severe cyanosis stimulates the production of red blood cells, which increase hematocrit and hemoglobin concentration. It improves oxygen transport, so that the blood that reaches the lungs, bind more oxygen. Raised hematocrit that can reach 80% or more, increases blood viscosity and may reduce some coagulation factors, particularly platelet count and fibrinogen. Dehydration in patients with very high hematocrit may cause systemic and pulmonary venous thrombosis, despite the reduced concentration of clotting factors.

Changing cyanosis, hypoxic seizures, squat and "drumsticks" are often connected with CHD with reduced pulmonary blood flow. Several factors can alter the degree of cyanosis, changing the ratio of pulmonary and systemic resistance. Physical activity reduces systemic vascular resistance, increases the systemic circulation and at Fallot's tetrad, reduces pulmonary blood flow and arterial blood oxygen saturation. Increased catecholamines or acidosis may also reduce pulmonary blood flow in patients with Fallot's tetrad.

Hypoxic seizures indicate a severe brain hypoxia because of the sharp reduction in pulmonary blood flow. Spasm of the output of the right ventricle is the most probable cause of hypoxia periods, which may occur without precursors. Infants and young children

lose consciousness for variable periods of time and sometimes die. The most effective treatment should include oxygen and small doses of morphine, placing the patient in position with knees bent, pressed to the abdomen and correction metabolic acidosis.

Children with a decrease in pulmonary blood flow, often sit on their heels. In this position the systemic resistance increases. Increased systemic vascular resistance reduces right-left blood discharge temporarily and thus increases pulmonary blood flow.

Fingers as "drumsticks" develop in early childhood due to proliferation of capillaries and small arteriovenous fistula creation in the distal phalanges. Reduced pulmonary blood flow stimulates the dilatation of mediastinal and bronchial arteries. These vessels are connected to the pulmonary artery and in some cases can provide an increase in pulmonary blood flow. At birth, open ductus arteriosus provides blood flow to the pulmonary arteries of patients with obstruction of outflow tract of the right ventricle. Unfortunately, PDA is almost always closed during the first few hours or days after birth. Intravenous prostaglandin E supports PDA for hours or days in some infants, thus allowing you to reduce acidosis, cyanosis and stabilize the infant for surgery. Several palliative operations that shunt blood from the systemic to pulmonary blood flow were invented for infants and young children who have inadequate pulmonary blood flow.

Blalock–Taussig surgery connects subclavian artery with the pulmonary artery branch with anastomosis "toward the end". Modified Blalock–Taussig shunt inserted into 4–6-mm, vascular prosthesis, usually polytetrafluorethylen (Gore–Tex) between the subclavian and pulmonary arteries. Operation by Waterston: anastomosis between the rear of the ascending aorta and right pulmonary artery branch. Potts operation: anastomosis "side to side" between the descending aorta and the left branch of the pulmonary artery. Waterston and Potts operation has mainly historical interest because of the difficulty of regulation of blood flow during creation and closure of the shunt during radical surgery. All these operations increase pulmonary blood flow, due to differences in pressure between the systemic and pulmonary circulation. Classic Glenn surgery connects the upper vena cava with the right branch of the pulmonary artery so that blood from the superior vena cava (SVC) should fall into the right pulmonary artery. Aa more modern bidirectional Glenn operation connects SVC "toward the end" with right branch of the pulmonary artery, providing blood flow to both lungs. Connection of right atrium or ventricle with pulmonary artery or directly by intracardiac tunneling, or using conduit is now commonly used for hemodynamic correction (Fontan).

Connection of right ventricle with pulmonary artery via conduit can be used for CHD with reduced pulmonary blood flow where the outflow tract of the right ventricle cannot be restored (pulmonary atresia, Fallot's tetrad) or more complex CHD with increased pulmonary blood flow (total arterial trunk, transposition of great vessels with interventricular defect and stenosis of the pulmonary artery). Conduits are usually made of dacron or polytetrafluorethylen may have included biological or artificial valve prosthesis. Human aortic or pulmonary homografts are also used.

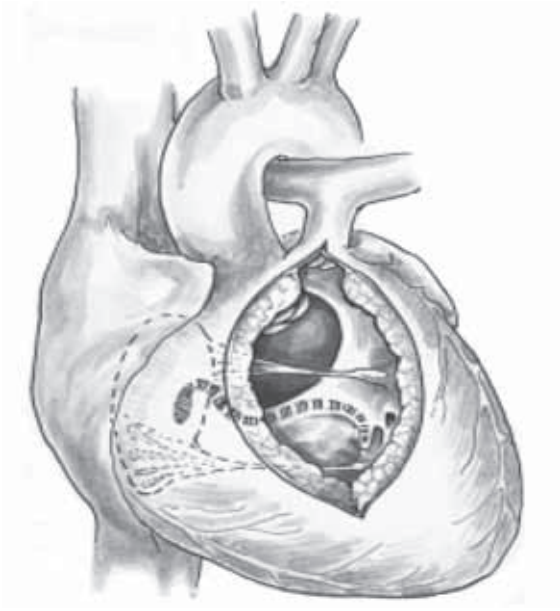


Fig. 3.7.7. Fallot's tetrad, pulmonary atresia with interventricular septum defect

FALLOT'S TETRAD (FT), PULMONARY ATRESIA WITH INTERVENTRICULAR SEPTUM DEFECT (PA WITH IVSD)

Criteria for the diagnosis:

— In anamnesis hypoxic seizures and sitting on heels;

— Cyanosis and "Drumsticks";

— A prominent right ventricular impulse, single second tone;

— Rough systolic murmur in the third intercostal space;

— Systolic murmur softens or disappears during hypoxic attack.

Fallot's tetrad has four main anatomical anomalies: interventricular septum defect, pulmonary stenosis, aortic dextraposition and right ventricular hypertrophy. If still attached pentalogy of Fallot. Pulmonary stenosis may be a valve or infundibular, but in most cases there is some degree of hypertrophy of muscle mass output of the right ventricle. In more severe forms, pulmonary valve can be complete-

ly atresia. Local distal narrowing of branches of the pulmonary arteries is also sometimes observed (Fig. 3.7.7).

Clinical signs are directly related with pronounced obstruction output of the right ventricle and the pulmonary blood flow number. PDA can mask defects of the heart and its symptoms within the first days of life. Systolic noise due to pulmonary stenosis is heard to the left edge of the sternum. Because of dextraposition of aorta and large ventricular septal defect, blood flow through the septal defect is a rare cause of noise. Hypoxic attack is the most severe sign, because it may end with cerebral hypoxia, brain damage and even death. The diagnosis can be usually made under cyanosis, the small size of the heart and reduced pulmonary blood flow seen on chest X-ray. ECG shows right ventricular hypertrophy. Cardiac catheterization is required and the degree of success of surgical treatment correlates with the size of the pulmonary arteries, palliative procedure can be performed in young children. Successful correction is determined on attitude of the right branch of the pulmonary artery to the ascending aorta size as 1: 3. More low ratio may indicate the need for initial palliative surgery.

Treatment. The choice of surgery for Fallot's tetrad is debatable. If the main pulmonary artery and distal pulmonary vessels have acceptable size correction procedure,probably, need, regardless of age. Way of outflow of the right ventricle and the pulmonary artery ring often require transannular plastic or plastic of output channel of pancreas by patch. Patch should not be so large, should be implanted with autopericardium so there are no pulmonary valve incompetence. Infundibular area increases by resection of muscular legs in this area. Interventricular septal defect is closed by patch made of prosthetic synthetic material. Correction is performed during artificial circulation. Complete correction is also performed through the right atrium, including closing of IVSD and infundibular resection. This operating approach avoids ventriculotomy. After surgery, patients with Fallot's tetrad, usually have increase in heart rate and a moderate degree of right ventricular failure. With all Fallot's tetrad one should perform left ventricle after surgery than before because pulmonary flow is restored.

If pulmonary arteries are narrow, the initial palliative procedure can be better. Outflow of output channel or valvular ring can be increased by patch without IVSD closing. An alternative approach — creating systemic-pulmonary shunt: Blalock–Taussig anastomosis of the subclavian-pulmonary is probably the best. At patients with artesia of pulmonary artery valve containing conduit for radical correction Fallot's tetrad, but the initial palliative operation is usually required at childhood.

Prognosis. Full success of surgical correction of can be used — 90–95%. Currently, many patients are exposed to full correction at an early age. Approximately 50% of patients who survived have no limitations in physical activities. Long term perspective, however, for patients with the residual insufficiency of the valve of pulmonary artery is hesitant. Sometimes some of them will demand further valve replacement because the pulmonary valve incompetence cannot be allowed for such a long period. Some patients have residual pulmonary stenosis, residual reshunting at patch in IVSD. These defects can usually be corrected easily.

QUESTIONS FOR THE FINAL CONTROL OF THE CHAPTER 3

1. Embrigenesis of congenital cysts of lungs.
2. Clinical course of congenital cysts of lungs.
3. Clinical symptoms of cystic diseases of lungs.
4. Medical treatment of congenital cysts of lungs.
5. Etiology of the congenital lobar emphysema of lungs.
6. Clinical symptoms and forms of the congenital lobar emphysema.
7. Medical treatment of the congenital lobar emphysema of lungs.
8. Atresia of esophagus and "VACTERL-association".
9. Forms of atresia of esophagus and clinical symptoms.
10. Diagnosis of atresia of esophagus.
11. Medical treatment of atresia of esophagus.
12. Classification of diaphragmatic hernia.
13. Clinical symptoms of diaphragmatic hernia. "Asphictic incarceration".
14. Differential diagnosis of diaphragmatic hernia.
15. Operative treatment of diaphragmatic hernia.
16. Congenital hypertrophic pyloric stenosis. Clinical course. Diagnosis. Medical treatment.
17. Classification of congenital intestinal obstruction.
18. Clinical picture of high intestinal obstruction.
19. Clinical picture of low intestinal obstruction.
20. Diagnosis of the congenital intestinal obstruction.
21. Volume of operation in case of congenital intestinal obstruction.
22. Omphalocele. Classification. Clinical picture. Medical treatment.
23. Gastroschisis. Clinical picture. Differential diagnosis. Medical treatment.
24. Umbilical thernia. Clinical picture. Medical treatment.

25. Inguinal hernia. Clinical picture. Differential diagnosis. Medical treatment. Complication.

26. Congenital dislocation of hip. Early diagnosis.

27. Congenital dislocation of hip. Methods of examination.

28. Medical treatment of congenital dislocation of hip in the children of the first year of life.

29. Medical treatment of congenital dislocation of hip in the children after 1 year age.

30. Congenital clubfoot. Clinical picture.

31. Medical treatment of congenital clubfoot.

32. Congenital torticollis. Clinical picture. Diagnosis.

33. Medical treatments of torticollis.

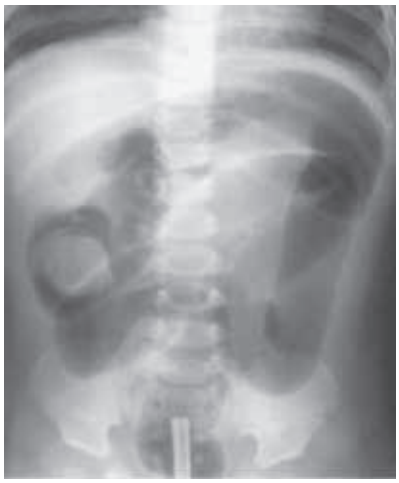
34. Hydronephrosis. Reasons. Clinical picture. Diagnosis. Medical treatment.

35. Vesiculo-uretheral reflux. Classification. Clinical picture. Diagnosis. Medical treatment.

36. Anomalies of development of urethra. Methods of diagnosis, complications. Medical treatment.

TASKS

TASK N 1



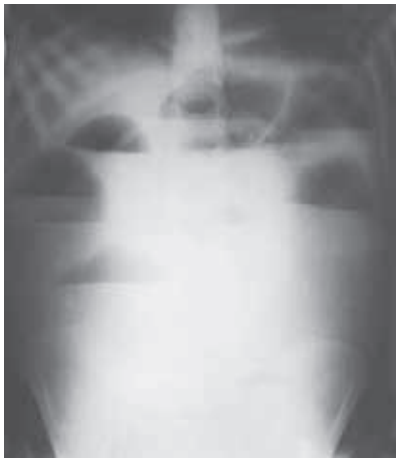
Examining a 6-months infant a physician has revealed that the infant is restless during the last 8 hours. He is screaming and drowing up legs. His anxiety is intermittent. Repeated vomiting and bloody stool passage are observed. The infant has eaten the vegetable puree this day first time. Abdominal palpation is painless, muscle guarding is absent, a solid oval-formed mass is found in the right upper quadrant.

1. Make a diagnosis.
2. Choose a treatment management.

Example of answers:

1. Due to the typical anamnesis and clinical symptoms (new food insertion, intermittent anxiety, repeated vomiting, bloody stool, palpated mass) the intussusception must be suspected.
2. Emergency admission for pneumodesinvagination is indicated.

TASK N 2



A 8-year old child complains of intermittent abdominal pain, multiple bilious vomiting, no stool passage. The child was operated 6 months ago because of acute appendicitis. The childs holds knee-chest posotion, his tongue is dry and coated, his abdomen is distended in the epigastric region, asymmetric and painful near the postoperative scar. Increased peristaltis is auscultated. X-ray shows a few Kloiber's cups of different sizes.

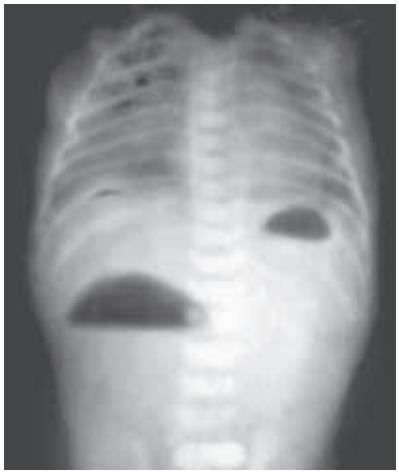
1. Make a diagnosis.
2. Choose a treatment management.

Example of answers:

1. The hystory, clinical and X-ray symptoms allow to make diagnosis of the late adhesive intestinal obstruction.

Conservative treatment: nasogastric tube, infusion therapy with potassium level correction, proserinum for peristalsis stimulation, hypertonic enema, epidural anesthesia. If effect is absent during 6-hour treatment, the surgery is indicated.

TASK N 3

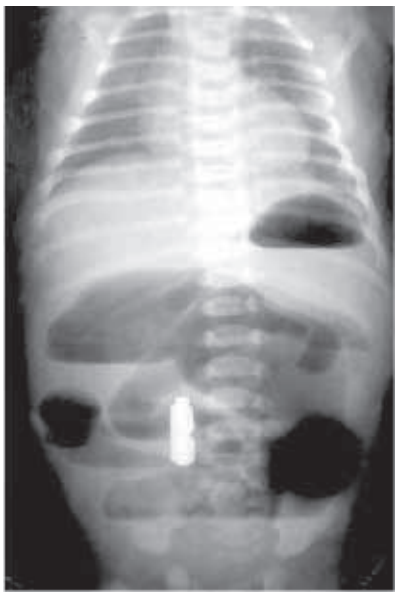


A 2.8-kg newborn boy developed bilious vomiting in 2 hours after delivery. The epigastric region is distended, muscle guarding is absent. Light-colored stool passed after the enema.

1. Make a diagnosis.
2. Administer necessary examinations.
3. Choose treatment management.

Example of answers:

1. High congenital intestinal obstruction.
2. A plain X-ray of the abdominal cavity is indicated. The typical symptom is "double bubble".
3. Insertion of the nasogastric tube, total parenteral nutrition, temperature maintenance and transportation to the surgical departement for operative treatment are indicated.



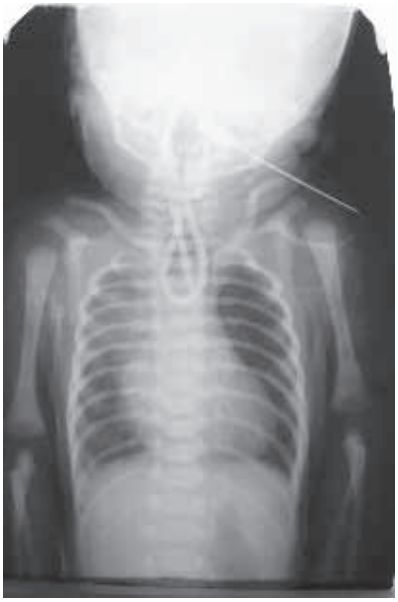
TASK N 4

A 2-day old newborn, who was full-term (birth weight is 3.1 kg), developed vomiting with bile and intestinal contents. Meconium has not been passed. Newborn's abdomen is distended with visible peristaltis.

1. Make a diagnosis.
2. Administer necessary examinatios.
3. Choose a treatment management.

Example of answers:

1. Low congenital intestinal obstruction.
2. Plain X-ray of the abdominal cavity is indicated. The typical symptom is multiple horizontal liquid levels.
3. Insertion of the nasogastric tube, total parenteral nutrition, temperature maintenance and transportation to the surgical departement for operative treatment are indicated.



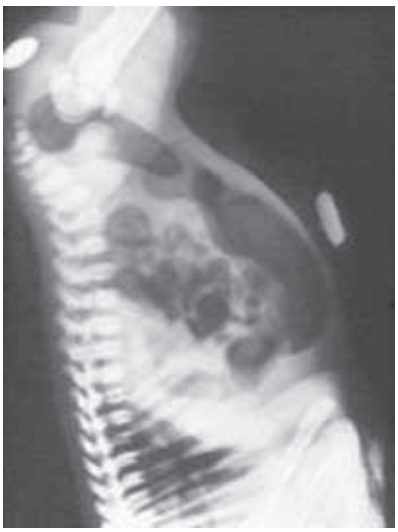
TASK N 5

A newborn had excessive foamy salivation from mouth and nose and cyanosis. After suctioning the salivation increased again.

1. Make a diagnosis.
2. Administer necessary examinatios.
3. Describe the rules of transportation.

Example of answers:

1. Esophageal atresia.
2. Nasogastric tubing, Elaphant's probe, X-ray of thorax and abdominal cavity with a contrast catheter must be done.
3. A special transport cradle for temperature maintenance, permanent oropharyngeal suctioning, tracheal intubation, infusion therapy and antibiotics are required for newborn transportation to a special surgical department.



TASK N 6

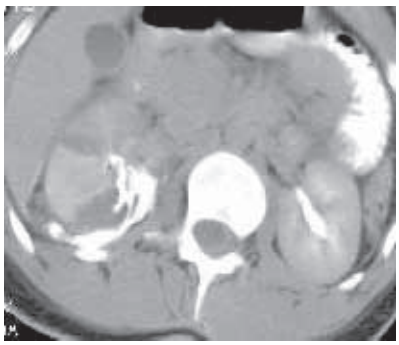
Examining a 3-days newborn a pediatrician noted newborn's restlessness, vomiting, abdominal distention, absence of the anus.

1. Make a diagnosis.
2. Administer necessary examinatios.
3. Choose a treatment management.

Example of answers:

1. Atresia of anus and rectum (inperforated anus).
2. X-ray examination (invertogramm by Vangestin) must be done in 12–16 hours after delivery.
3. Surgical treatment — perineum or abdominoperineum plasty.

TASK N 7



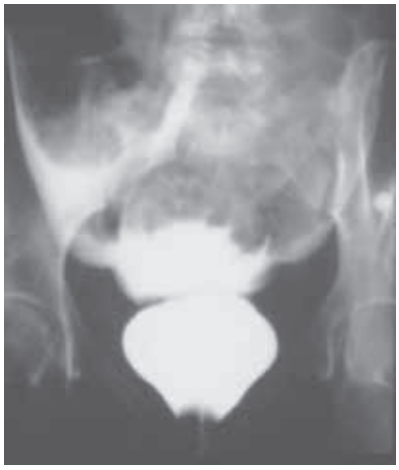
A 10-year-old boy is brought to the emergency room in 40 min after he has fallen from a tree. The child complains of pain in the right lumbar region. Lacerations in the right lumbar region, gross hematuria, pulse — 88/min, BP — 110/70 mm Hg.

1. Make a diagnosis.
2. Make a plan of examination.

Example of answers:

1. Blunt trauma of the right kidney.
2. Complete blood count, urinalysis, ultrasonography, excretory urography, computer tomography.

TASK N 8



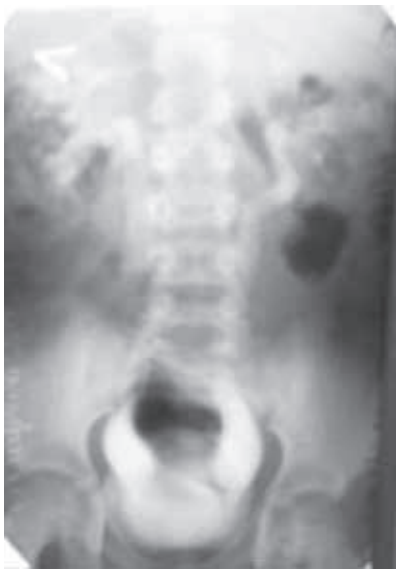
An 11-year girl was struck by an automobile. The child sustains a fracture of pelvis and complains of abdominal pain, painful and effectless urges to void urine. Tendereness and muscle guarding over the pubis are found.

1. Make a diagnosis.
2. Make a plan of examination.
3. Describe the X-ray.
4. Choose a treatment management.

Example of answers:

1. Bladder rupture.
2. Ascending cystography.
3. X-ray shows the intraabdominal rupture of bladder.
4. Operative treatment is indicated.

TASK N 9

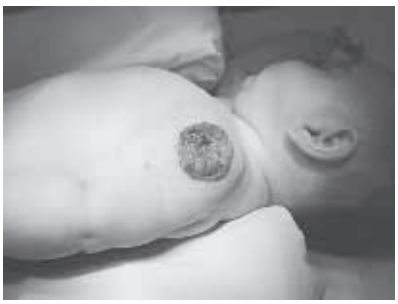


A 2-year old child presents with fatigue, vomiting, temperature 39°C, pale skin and cloudy urine. Complete blood count shows leukocytosis and shift to the left, urinalysis — leukocyturia and bacteriuria. Bilateral vesicouthetreal reflux I–II stage was diagnosed at 1 year old age.

1. Make a diagnosis.
2. Make a plan of examination and treatment.

Example of answers:

1. Bilateral vesicouthetreal reflux, complicated with acute pyelonephritis.
2. Urinalysis by Nechiporenko, urine culture, biochemical blood analysis (protein, creatinine, nitrogen), infusion and antibacterial therapy.



TASK N 10

A 3.5-month infant presents with a bright-red tumorous mass on the skin of the right shoulder. The tumor has been growing slowly and rising over the skin.

1. Make a diagnosis.
2. Choose a treatment management.

Example of answers:

1. Ulcered cavernous hemangioma of the right shoulder.
2. Applying of a compression bandage on the tumor and hospitalization for surgical treatment are indicated.

TASK N 11

A child of 2 years has played with LEGO-toy without supervision of adults. During game the child has had a fit of coughing, than has drawn attention of adults. It is delivered in clinic by parents in a severe condition, with the pronounced respiratory insufficiency. At examination the sonorous voice, retraction compliant places of a thorax is marked, at auscultation the symptom of "clap" is periodically audible.

1. Define the preliminary diagnosis.
2. Define management at a stage of transportation.

Example of answers:

1. The balloting foreign body of a trachea.
2. The patient should be delivered in a hospital by the car of first aid. In a way the child should be in a sitting position, in order to prevent moving an foreign body and asphyxia. If necessary hardware artificial breath is conducted.

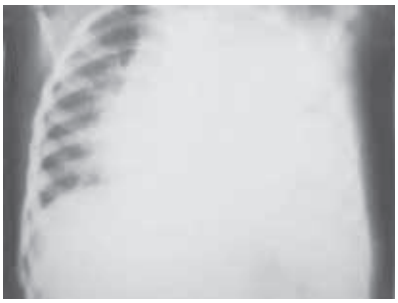
TASK N 12

At the child of 3 years old an acute pain during defecation is marked, last portion stool is painted by scarlet blood. In the anamnesis constant constisipations.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. The crack of a mucous direct gut.
2. Removal of pain and spasm of anal sphincter, normalization of a stool, use of the medicines to heal a crack.



TASK N 13

A child of 8 years within a week was observed by the pediatrist in-home in occasion of a flu. For last 2 days a condition has sharply worsened.

There is cough, temperature 39.0–39.5°C, a short wind (42 in mines). The condition of the child severe, integuments pale — grey, cyanosis, the child languid, temperature 39.6°C, frequency of breath 42/min, pulse 144/min. Left half of thorax lags behind in breath, during the percussion the sound at the left is truncated, during auscultation breath is sharply weakened. On the X-ray total blackout left hemithorax, mediastinum is driven to the right.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Considering data of the anamnesis (in current of week flu), objective examination (a grave condition, integuments pale-grey, cyanosis, the child languid, temperature 39,6°C, frequency of breath 42/min, pulse 144/min; the left half of thorax lags behind in breath, during the percutoion the sound at the left is truncated, during the auscultation breath is sharply weakened), and also data of X-ray, it is possible to put a diagnose: destructive pneumonia, the lung-pleural form, total piothorax.
2. Management: a puncture of a pleural cavity, and evacuation of pus. Transportation to the hospital by car of first aid. Treatment of destructive pneumonia includes: cure which is directed on macroorganism (desintoxication, symptomatic therapy, immune therapy); cure which is directed microorganism (rational antibiotic treatment with change of a medicine each 5–7 days, bacteriophage); cure which is directed on inflammation process (drainage of a pleural cavity, evacuation of contents and sanitation of a pleural cavity).

TASK N 14

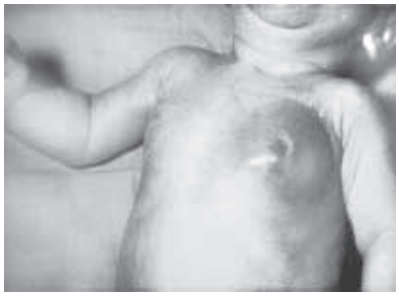
A child of 6 years has allocation of stool with an impurity of scarlet blood and slime, blood in the form of a spittle atop of last portion of stool is periodically marked. The general condition of the child is satisfactory. During rectal manual research on depth of 4 cm it is defined tumorous formation, 1.5×1.5 cm, which has a round form on a narrow thin leg.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Thepolyp of the rectum.
2. Polypectomy in the scheduled order.

TASK N 15



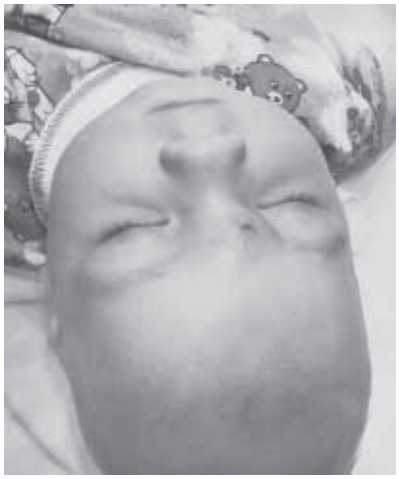
A newborn of 3 weeks becames restless, the temperature has increased up to 38°C, there was a hypostasis in the field of right mammary gland, the skin with hyperemia, during the palpation morbidity is marked.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. The rightsided mastitis of the newborn.
2. Management — conservative (antibacterial, symptomatic therapy, local — compresses, physiotherapy) in case of cataral form, in case of flegmonose — the child needs operation by opening by radial cuts.

TASK N 16



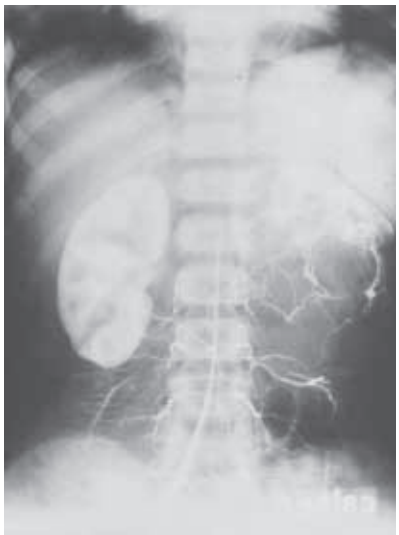
A mother with the child of 2 months, came to the doctor with complaints to of quickly growing red spot at the field of the nose bridge, towering above a level of a skin. The baby is ill from the date of a birth. At pressing the spot turns pale, the condition of the child does not suffer.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Simple hemangiome.
2. Treatment conservative — sclerous therapy or cryodestruktion.

TASK N 17



A mother with the child of 1.5 years has addressed in clinic with complaints of decreasein appetite, the child has lost weight, deterioration of the general condition, pallor, increase in a stomach. In left hypochondrium it is palpated tumorous formation 8×9 cm, inactive, painless. At carrying out of ultrasonic and excretor urography the increase in the left kidney and reduction in its function is revealed.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Embriosarcoma of the left kidney (Wilms tumour).
2. Carrying out of the combined therapy (beam therapy + operative treatment).

TASK N 18

A girl of 13 years has been ill for 4 day, has complains of intensive pains in the bottom third of the left femur. Rise in temperature up to 39°C is marked. The swelling, infringement of function is marked. The girl marks that it was a trauma before the beginning of disease. X-ray revealed no pathologies.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Acute hematogenous osteomyelitis of the left femur.
2. For specification of the diagnosis: a puncture of bottom third of the left femur, definition of intrabone pressure, bacterioscopy. Crop of contents.

TASK N 19

Parents of a child of 3 years old has addressed to the children's hospital. Parents complained that the behavior of the child was unnormal: child is languid, restless, unsleeping, rise in temperature up to 38°C, frequent vomiting, a liquid stool. At examination grave condition, temperature 38.3°C, tongue is dry, during palpation of abdomen the child cries, pushes away a hand of the doctor. At examination in an hour during a dream it is revealed a muscles defance of forward abdominal wall in all departments.

1. Define the preliminary diagnosis.
2. Define management.

Example of answers:

1. Acute appendicitis, a peritonitis.
2. Operative treatment after preliminary preparation

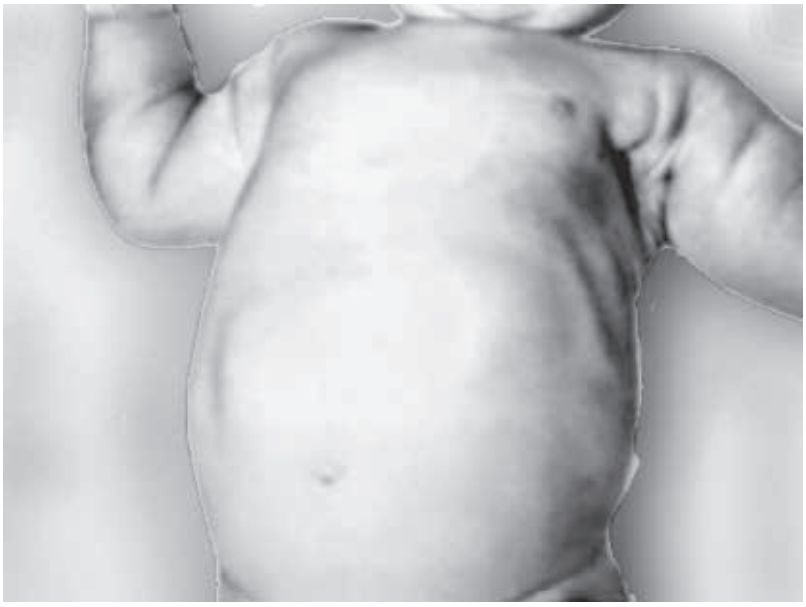
TASK N 20

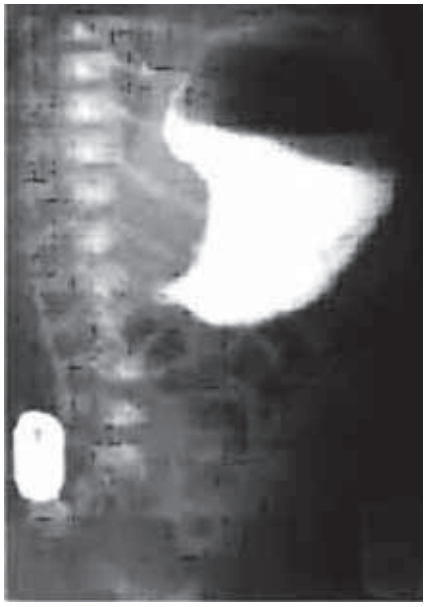
A child of 1 month, is il for 3 weeks. Disease has begun with regurgitation, then there was a vomiting by "fountain" milk without an impurity of bile 4–5 times in day. The weight of a body at a birth 3400, at survey body weight is 3600, the child is restless, shout weak, in epigastral area peristaltics of stomach as "sand-glass".

1. Your presumable diagnosis.
2. Define further management.

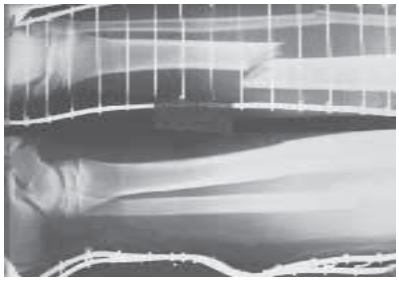
Example of answers:

1. Congenital hypertrophic pylorostenosis.
2. Fibrogastroscopy. Surgery by Frede–Ramshted–Weber





TASK N 21



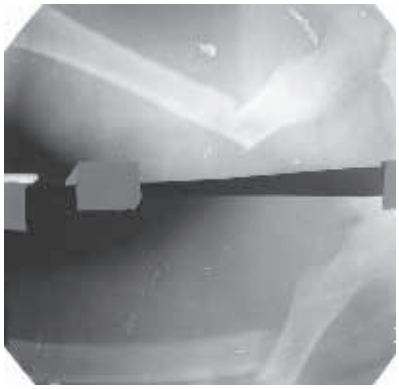
Closed oblique fracture of right tibia in middle third with dislocation by width on 1/3 was found in an 7-year-old child. The axis of extremity is correct.

Make the correct management.

Example of answers:

1. Applying skeletal traction.

TASK N 22



A 9 year old child was delivered to the clinic in bad condition with bloody vomiting, paleness, and black color stool. Palpation of the front abdominal wall reveals increasing of the spleen. The pulse is frequent, weak. The arterial blood pressure is 80/50 mm Hg. Distended veins were found under the fibrogastroscopy.

1. What is your tentative diagnosis?
2. What is your management?

Example of answers:

1. Extrahepatic form of portal hypertension.
2. The management is: sclerotising therapy, if there will be no effect, esophageal cirsodesis should be applied.

TASK N 23

A physician of ambulance was called to the 10-year-old child, victim of an accident. Examination revealed lost of conciousness, absence of the pupil reaction, pulse rate is 60/min, weakness, arterial blood pressure 80/10 mm Hg. There are bleeding from the nose and liquorrhea. Spontaneous breathing is absent.

1. What is your tentative diagnosis?
2. What is your management?

Example of answers:

Based on the objective data (bleeding from the nose and liquorrhea) there is possible to thinking about the fracture of the base of skull, complicated with traumatic coma.

The list of further measures:

— Cleaning of respiratory tract from mechanic obstacles;

— Artificial breathing;

— Introduction of 10% solution of glucose or reopolyglukin for increasing BP;

— Introduction of diuretics: 2% lasiks or 25% solution of mannitol;

— Hospitalizing to the neurosurgical department for specialized help.

TASK N 24

A14 years old boy has got a cutting wound of the right forearm as result of a fight in the school. The school physician found continues bleeding with the cherry-colored blood from the wound. Blood flow with the constant stream that become worth in down position of extremity.

1. What is your initial diagnosis?
2. What is your further management?

Example of answers:

1. Cutting wound of right forearm. External venous bleeding.
2. There is necessary to stop the bleeding timely. The simple and effective method of timely hemostasis is applying of the compressive bandage.

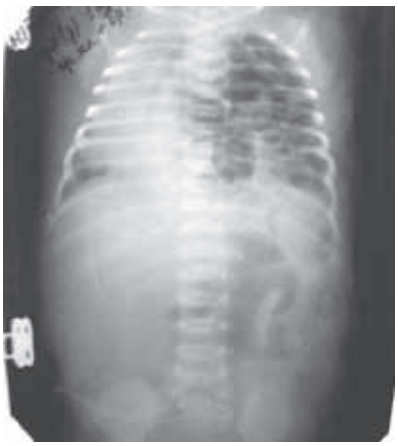
TASK N 25

A 8-year-old child was delivered to the reception department by the ambulance for 20 min after falling down from 3 meter height. Loss of consciousness, absence of reaction on surrounding, didn't answers on the questions was found under examination. Mimic reaction appears under the pain irritation (pricking by the needle). Reaction of pupil is weak, corneal reflexes are saved. Swallowing also is saved. There are abrasions in the left temple region and visible deformation of the left femur. Pulse rate is 75/min.

1. What is your initial diagnosis?
2. What is your further management?

Example of answers:

1. Combined cranial-cerebral trauma and skeletal injury complicated with the sopor are in the child.
2. The list of measures is following: performing X-ray investigation of the skull in 2 positions and X-ray of the left femur in two position; consultation of neurosurgeon and traumatologist; hospitalizing to the intensive care department.



TASK N 26

Signs of respiratory insufficiency were found in newborn on 2nd day of life. Baby becomes restless. Asymmetry of the chest in breathing movement with lagging behind of the left part was found under examination. Breathing in the left is absent instead of breathing peristalsis is there. Tympanitis is under the percussion in the left. In plane X-ray of thoracic cavity instead of lung picture there are cellular formations, shifts of the mediastinum to the right. Condition of the baby becomes worse in dynamics.

1. What is your initial diagnosis?
2. What is your management?

Example of answers:

1. The left sided false diaphragmatic hernia;
2. Intubation of trachea and oscillatory artificial breathing are indicated with hospitalizing the patient to the specialized hospital.

TASK N 27

A 12-year-old boy far from home after the fight with parents in the winter. He was all the night in the broken house, where he was found in the morning. In reception department there was found that child with blocked mind, answering the question monosyllabically and late, trembles all over, lying in the bad with the flexed legs and arms, body temperature is 34.5°C. Skin is pale, cyanotic, and cold. Foot fingers are white, without change of color under pressing; movement and pain sensory are absent.

1. What is your initial diagnosis?
2. What is your further management?

Example of answers:

1. Based on anamnesis (being all the night in a cold broken house in the winter), on data of examination (general overcooling, disturbance of movement, absence of pain and tactile sensory of the feet fingers, there is possible to suppose general shivering, frostbitten of the feet fingers.

2. Plan of first aid:

— Hospitalizing;

— General treatment — hot drink, improvement of microcirculation (i. v. introduction of reopolyglukin, trental, heparin, 0.25% solution of novocain), warming up to 38–39°C Ringer's solution, 5% solution of glucose;

— Local treatment — isolating dressing of the both feet and low parts of calf.

TASK N 28

A 12-year-old boy hit against the bottom by the head when diving. The boy complains of the pain in the neck region that increasing in attempt of movement.

1. What is your initial diagnosis?
2. What is your further management?

Example of answers:

1. The most typical is the fracture of the cervical part of spine in the diving on a shallow place.

2. Immobilizing of the neck part of spine with the cervical collar and delivering in to the hospital in horizontal position to prophylaxis of spinal cord injuries.

TASK N 29

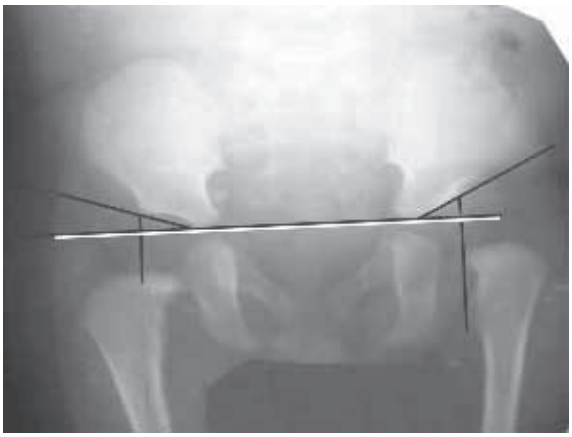
Pediatric doctor revealed asymmetry of skin folds, limitation of adduction of the left hip, relative shortening of the left leg and "klick" sign in the left in a baby girl under examination.

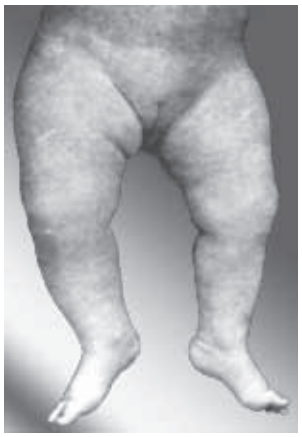
1. What is your initial diagnosis?
2. What is your further management?

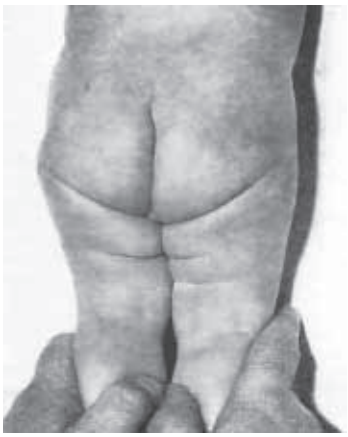
Example of answers:

1. Dysplasia of the left hip joint.

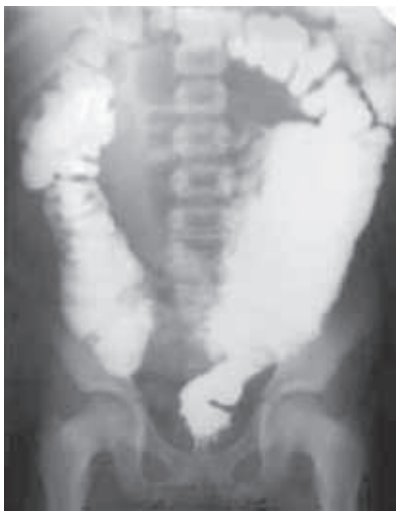
2. Management of treatment: physical training, massage, wide swaddle. After discharge from maternity home — ultrosonography of hip joints, Pavlick's stapes's.







TASK N 30



A mother with a 3-week old baby applied to the doctor with complains of the acute distension of abdomen, absence of stool from the time of discharge from maternity home. Poor liquid stool with bad smell appear last two days. Mather told about losing of baby's weight, vomiting, refusing of eat.

1. What is your initial diagnosis?
2. What is your further management?

Example of answers:

Based on anamnesis and complains there is possible to suppose acute form of Hirschprung's disease in a baby.

Hospitalizing to the surgical department for irrigography and surgical treatment is indicated in this case.

KROK-2 "PEDIATRIC SURGERY"

Topic 1
GASTROINTESTINAL BLEEDING

Portal Hypertension

1. A nine-year-old girl is admitted to surgery. Suddenly, among a full health there has appeared vomiting with a large number of impurities of the liquid blood in the form of clots. The patient is complaining on general weakness, drowsiness. The skin is pale; the veins of the anterior abdominal wall are enhanced. Tension of the abdominal muscles is not observed, the liver is not palpable, the spleen is 10 cm, firm, painless.

Free fluid in the abdominal cavity is detected. What is the probable cause of the bleeding?

A. +Portal hypertension.
B. Gastric ulcer.
C. Trombocytopenic purpura.
D. Diaphragmatic hernias of intestinal hole.
E. Mallory–Weiss syndrome.

Acute Inflammatory Diseases of the Abdominal Cavity

1. A newborn child with ulcerative necrotizing enterocolitis has got fecal vomiting, generated intestinal gas and zero stools. The abdomen is swollen; there is muscle tension of the abdominal wall, no peristalsis, no rebound tenderness (Schotkin–Blumberg symptom) and no hepatic dullness when percussing the abdomen. What is the cause of these symptoms?

A. +Perforation of intestine.
B. Sepsis.
C. Pneumothorax.
D. Pneumomediastinum.
E. Hepatic coma.

2. A boy of 2.5 years old was admitted to the emergency department. His parents report that within the day the child became capricious, weak, had slept poorly the night before, vomited twice, in the morning there was loose stool. On examination the boy has got severe condition, the body temperature is 39.8°C, there is tachycardia. The child is lying on his right side with his legs pulled up to the abdomen. On abdominal palpation pain is determined in the right iliac region, as well as muscle tension, symptoms of peritoneal irritation are positive. Rectal examination is normal. What kind of disease is it?

A. +Acute appendicitis.
B. Food poisoning.
C. Bowel intussusception.
D. Enterocolitis.
E. Coprostasia.

3. A three-year-old girl got sick 3 days ago: frequent vomiting, loose stool with mucus, fever more than 38.5°C. On the 4th day of disease abdominal pain intensified, there was some discomfort when changing the body position. Severe intoxication: exacerbation of facial features, xerostomia. The pulse was 120 beats/min. On palpation the abdomen is tense, painful in the area of the navel and the lower parts. Which pathology of acute abdominal organs can be assumed?

A. +Peritonitis of appendiceal origin.
B. Influenza, toxic form.
C. Acute dysentery, severe course.
D. Intestinal intussusception.
E. Acute pancreatitis.

4. A patient of 8 years old on the 10th day of follicular tonsillitis had signs of acute appendicitis. During the operation, appendicitis abscess was confirmed. There was found streptococcus in crops of purulent exudate. Which of the ways of infection in the appendix mentioned below is most likely in this case?

A. +Hematogenous.

B. Enterogenic.
C. Lymphogenous.
D. Mixed.
E. —

5. A child of 1.5 years got ill 8 hours ago, when there were pains in the abdomen, and then vomiting and loose stools added. The condition was satisfactory. He was sluggish. The body temperature was 38°C. In blood there were leukocytes $18–10^9$/l. On abdominal palpation during sleep study the muscle tension of the abdominal wall was determined in the right iliac region. While hand pressing on this area, the child woke up and started screaming. What is the diagnosis?
A. +Acute appendicitis.
B. Intestinal infection.
C. Renal colic.
D. Acute mesadenitis.
E. Acute enterovirus infection.

Acquired Intestinal Obstruction

1. A sick 15 year old patient suddenly felt severe abdominal pain that enhanced gradually, nausea. The abdomen was swollen moderately, generated no intestinal gas. No stool. Pasternatsky's symptom is negative. Plain abdomen examination revealed horizontal fluids, some loops formed an arcade. The clinical and X-ray diagnosis is?
A. +Intestinal obstruction.
B. Renal colic.
C. Perforation of a hollow organ.
D. Ulcerative colitis.
E. Bleeding into the abdominal cavity.

2. An 18-year-old sick patient has complained of abdominal pain on the right, which periodically has nature of cramping, nausea, repeated vomiting of food and bile, retention of stool and gas during the day. When he was 7 he had appendectomy. The pulse is 96 beats/min. The tongue is moderately moist. The abdomen is slightly swollen with some small asymmetry due to increasing of the right half. Some noise is detected. Peristaltic sounds are periodically amplified. Single Kloiber's cups are seen on plain radiograph of the abdomen. The most obvious diagnosis is?
A. +Adhesive intestinal obstruction.
B. Volvulus of the sigmoid colon.
C. Thrombosis of mesenteric vessels.
D. Ulcerative colitis.
E. Obstructive intestinal obstruction/tumor genesis.

3. A patient with dysentery suspected was hospitalized in the Department of Infectious Diseases (frequent stools mixed with blood like raspberry jelly). After one day the final diagnosis was determined — intussusception, the patient was transferred to a specialized department. What treatments should be used?
A. +Operative desintussusception.
B. Conservative desintussusception.
C. Infusion treatment with antispasmodics.
D. Manual desintussusception.
E. Perirenal blockade.

4. A baby of 6 months was admitted to surgery after 16 hours of a disease that began suddenly. The baby became restless, rubbing legs, refused any food. The panic attack was short-period. The boy calmed down and fell asleep. He woke up in 20–25 min, had repeated vomiting and sudden anxiety. The child is pale, adynamic. His diaper is in dark red formations. The preliminary diagnosis was?
A. +Intestinal invagination.
B. Enterocolitis.
C. Meckel diverticulitis.
D. Worm intestinal obstruction.
E. Tumor of the abdominal cavity.

5. A baby of 5 months is delivered to hospital within 6 hours from onset of disease, he is pale, cramping excited, with repeated vomiting. Last stool was 4 hours ago. From medical history it was found that the child first time got semolina. On examination the child is pale, watchful, there is tachycardia, his forehead is covered with cold sweat. The abdomen is not swollen, soft, on rectal examination the cecum could not be found in a typical sites — blood in the form of raspberry jelly. What disease are you dealing with?
A. +Invagination.
B. Dysentery.
C. Gastric ulcers.
D. Mucous anal fissure.
E. Rectal polyp.

6. A boy of 12 years was hospitalized with complaints of recurrent vomiting, abdominal pain, gas retention. His medical history had appendectomy. Objectively: the skin is pale, the pulse is 90 beats/min, ACT is 110/80 mmHg, the body temperature is 37.2°C. The abdomen is moderately swollen, asymmetric, somewhat resistant in the lower regions. Bowel sounds are overly tightened. Rebound tenderness (Schotkin–Blumberg symptom is negative). On examination the rectal ampulla is somewhat inflated, there is empty anal resting tone. What is the most probable diagnosis?
A. +Acute adhesive intestinal obstruction.
B. Food poisoning.
C. Renal colic.
D. Biliary colic.
E. Acute pancreatitis.

7. A child of 5 months had sudden paroxysmal abdominal pain, frequent vomiting, retention of stool and gas. On examination: mild stomach, some tumor formation is diagnosed in the right upper quadrant, moving, is slightly painful. On examination through the rectum: the fingers are stained with blood. What is the most probable diagnosis?
A. +Invagination.
B. Tumor abdomen.
C. Polyposis.
D. Peritonitis.
E. Intestinal tumor.

8. A girl of 12 years fell ill acutely 6 hours ago, when there cramping abdominal pain, nausea, repeated vomiting of food eaten the day before started. Stools and gas did not pass. Two months ago the patient had appendectomy. Some tumor formation contoured with high sound of bloat over it on the anterior abdominal wall to the right of the navel. What is the most probable diagnosis?

A. Adhesive intestinal obstruction.
B. Inflammatory abdominal infiltrate.
C. Intussusception.
D. Coelioncus.
E. Acute suppurative right-paranephritis.

Features of Fractures in Children

1. A boy of 2 years old was admitted to traumatology department with complaints of pain and deformity of the right hip. X-ray reveales a hip displaced fracture at length. What method of treatment is to be assigned to the patient?

A. +Skeletal traction by Schede.
B. Closed reposition.
C. Intramedullary osteosynthesis.
D. Overlay of Ilizarov apparatus.
E. On-bone fixation.

2. A boy of 12 years is admitted to the emergency unit with a fracture of the middle third of the right femur with displacement of bone fragments at length. Your management?

A. +Skeletal traction.
B. Open reposition.
C. Closed reposition.
D. Stretching by Schede.
E. Ilizarov apparatus.

3. Taking a sweater off the three-year-old girl's mother gave her a pluck at the wrist, and then the child began crying in pain. Pain is indicated in the elbow joint. When attempting active and passive movements the girl is showing discomfort. What damage to the child is most likely?

A. +Subluxation of the radial head.
B. Dislocation of forearm.
C. Traumatic neuritis of the radial nerve.
D. Stretching of bag — ligament apparatus of the elbow.
E. Epiphysiolisis of the distal humerus.

Injury of Internal Organs

1. After falling from the second floor height on admission a patient has got closed diaphysis fracture of the left femur, diaphysis of the right shoulder, III–IV left rib, pain and abdominal tension on his left, the blood pressure is 40/0 mm, the skin is pale, covered with cold clammy sweat. What additional tests should be immediately assigned?

A. +Laparoscopy or celiocentesis.
B. Diagnostic laparotomy.
C. X-ray of the abdomen inspection, radiography of injured limbs.
D. Ultrasound of the abdomen.
E. Excretory urography.

2. Patient K. was taken to surgery after a traffic accident with closed chest trauma and rib fractures on the right. The patient was diagnosed with intense right side pneumothorax. The patient was immediately assigned:

A. +Thoracostomy.
B. Tracheostomy.
C. Artificial ventilation.
D. Tracheal intubation.
E. Thoracotomy.

3. A child of 8 years was taken to the hospital 1 hour after abdominal trauma. There was a total severe condition. He was pale. The abdomen was bloated in a volume. Percussion all over — thympanitis, hepatic dullness was not detected. Tenderness diffused throughout the abdomen, there was distinct tension of the abdominal muscles. What is the most probable diagnosis?

A. +Hollow organ damage, peritonitis.
B. Subcapsular hematoma of the liver.
C. Rupture of pancreas, peritonitis.
D. Rupture of liver, intra-abdominal bleeding.
E. Extraperitoneal rupture of bladder.

4. A child of 12 years was hit in the stomach an hour ago. Moderate severity condition, forced position in bed. Pale skin. The pulse is 122 beats/min. The pressing on the left costal arch is painful. Weinert–Kulenkampf's symptoms are positive. Macroscopically urine is not changed. The most probable diagnosis?

A. +Rupture of the spleen, abdominal bleeding.
B. Rupture of the left kidney, retroperitoneal hematoma.
C. Rupture of pancreas.
D. Rupture of liver, intra-abdominal bleeding.
E. Rupture of a hollow organ, peritonitis.

Bacterial Destruction of Lungs

1. On the 10th day a child of 1 year with bacterial pneumonia had subcortical abscess up to 4 cm in diameter with perifocal infiltration at the bottom lobe of the right lung. Choose a method of lung abscess treatment without draining it into the bronchus.

A. +Puncture and catheterization of the abscess cavity.
B. Bronchoscopic lavage.
C. Radical surgery.
D. Medical treatment.
E. Lobe bronchus occlusion.

2. A child of 3.5 years was diagnosed bilateral viral bacterial pneumonia. During the last 4 days his condition changed for the worse. Increased breathlessness and pale, fever temperature. The child refuses eating. Objectively: the right half of the chest is

bulging, intercostal spaces are smoothed. When percussing right there is a dull sound, breathing is not ausculted. Shifted to the left border of the heart. In blood there are hyperskeocytosis, neutrophilic shift, toxic granularity of leukocytes. What is the most probable diagnosis?

+A. Two-way viral and bacterial pneumonia, right pyothorax.

B. Relaxation of the right dome of the diaphragm.

C. Pulmonary tuberculosis, pleuritis of right side.

D. Atelectasis of the right lung.

E. Right lung tumor.

3. A child of 5 years receives a comprehensive therapy for bilateral viral and bacterial pneumonia. The presence of small cavities containing liquid was marked at the last radiograph along with decrease in infiltration of the lung fields. Having a meal the child is coughing; he has become sharply restless; dyspnea has increased. Objectively: mucosal cyanosis, bulging of the left chest cells. When percussing on the left lung apex there is thympanitis, from the III rib down there is blunting, the breathing is not auscultated. The borders of the heart are shifted to the right. What is the most probable diagnosis?

A. +Two-way viral and bacterial pneumonia, pneumoempyema on the left.

B. Foreign body of the left bronchus.

C. Strangulated diaphragmatic hernia on the left.

D. Strenuous cyst of the left lung.

E. Lobar emphysema on the left.

Hematogenous Osteomyelitis

1. A five-year-old child got sick 7 days ago. There are is complaining on constant throbbing pain in the upper third of the tibia. There is follicular tonsillitis in the anamnesis. Sleeping is poor, appetite is absent, the body temperature is 38.7°C. The upper third of the tibia is greatly swollen, soft tissue is strained, he has got flushed skin, positive syndrome of fluctuations, leukocytosis 12×10^9/l. ESR 31 mm/h. Your diagnosis?

A. +Acute hematogenous osteomyelitis.

B. Tuberculosis of tibia bones.

C. Fractures of tibia.

D. Erysipelas of tibia.

E. Phlegmon of tibia.

2. A baby of 26 days was diagnosed with umbilical sepsis when the doctor drew attention to edema of the right shoulder, the lack of active movements of the shoulder joint, dangling of the right hand. Mother noted the child's deterioration during the last day, which revealed itself in denying the breast, anxiety, crying, fever up to 38.8°C. What is the most probable previous diagnosis?

A. +Epiphyseal osteomyelitis of the humerus.

B. Fracture of the humerus.

C. Traumatic brachioplexsitis.

D. Phlegmon of shoulder.

E. Fracture of the clavicle.

3. A child of 9 years has got pain in the upper third of the right tibia, high temperature of 39°C, cannot stand on the leg. There is an injured leg and angina in the anamnesis. Your diagnosis?

A. +Acute hematogenous osteomyelitis.

B. Bone fracture.

C. Acute rheumatism.

D. Tuberculous osteomyelitis.

E. Malignant tumor.

4. A boy of 12 years is admitted to pediatric surgery clinic. He is complaining on two fistulas in the lower third of the left thigh, fever, general weakness. He underwent acute hematogenous osteomyelitis of the left femur 6 months ago. On radiographs of the left hip there is femoral total sequestration of 12× ×3 cm size. The preliminary diagnosis?

A. +Secondary chronic osteomyelitis.

B. Pathological fracture of femur.

C. Tuberculosis.

D. Ewing's sarcoma.

E. Osteoid osteoma.

5. A patient of 18 years has been severe ill for 3 weeks. There is sharply increased temperature, severe pain appeared in the left tibia. X-rays of the area are showing multiply foci of destruction of irregular geometric shapes with irregular contours. In the areas of destruction sequestration shedows are determined. Around the affected parts of bone calcification peeled off periosteum strips are detected. What is the most probable diagnosis?

A. +Acute hematogenous osteomyelitis of the left tibia bone.

B. Tuberculous lesion of the left tibia bone.

C. Sarcoma of the left tibia.

D. Fibrous osteodystrophy of the left libia.

E. Metastatic bone lesions of the left tibia.

6. A boy of 13 years began feeling pain in the upper third of the left thigh, the body temperature increased up to 39°C. Swelling was noted in the upper third of the thigh, as well as flatness of the groin. There was finiteness in the bent position. Active and passive movements were impossible because of acute pain. The most actual diagnosis?

A. +Acute hematogenous osteomyelitis.

B. Acute coxitis.

C. Intramuscular abscess.

D. Osteosarcoma.

E. Brodie abscess.

Inflammatory Diseases of Soft Tissues

1. A child of 10 years old, who was on the street in the windy and cold weather, felt moderate pain and tingling in the fingers and toes. The boy returned home, parents discovered whitened fingertips and toes, loss of sensation. Started warming of affected areas, pain reappeared and tingling in the fingers remained. Pale color of the skin turned into crimson, tingling disappeared, there was mild itching and

slight swelling of the fingers. Determine the degree of frostbite in the child.

A. +I degree frostbite.
B. Chilblains.
C. II degree frostbite.
D. III degree frostbite.
E. IV degree frostbite.

2. A newborn child of 8 days was discharged from the hospital. The child has got a weeping umbilicus. The day before redness of the right breast appeared, it was hard and painful when pressed. The child is not calm, is eating badly; the body temperature is 38°C. Your diagnosis?

A. +Mastitis of a newborn.
B. Chylangioma.
C. Mastopathy.
D. Erysipelas.
E. Pemphigus.

Features of Childhood Cancer

1. A baby of 1.5 months has got a tumor formation of dark purple color in the parietal section in the right since birth, which is rising above the skin surface, persisting during pushing; the size is 3×2 cm. The diagnosis is?

A. +Cavernous hemangioma.
B. Chylangioma.
C. Pigment spots.
D. Angiofibroma.
E. Congenital vascular stain.

Malformations and Diseases Accompanied by Respiratory Failure

1. A newborn child is observed that the tube doesn't enter the stomach during the initial examination in the delivery room. When viewing X-ray with contrast probe in the vertical position it is observed that the tube is lifted in the blind end of esophagus, there is no gas in stomach and bowel. What diagnosis must be determined?

A. +Form of esophageal atresia.
B. Esophageal atresia with lower fistula.
C. Congenital stenosis.
D. Esophageal atresia with upper and lower tracheo-oesophageal fistula.
E. Isolated tracheo-oesophageal fistula.

2. A child of 3 years with manifestations of respiratory failure on auscultation. Moist rales are auscultated in the lower right. On radiographs it is marked the mediastinum shift to the right and availability of triangular shadow in the lower right. What is the possible diagnosis of the child?

A. +Cystic hypoplasia of the right lower zone of lung.
B. Lobar congenital emphysema of upper left lung.
C. Abscess of the right lower zone of lung.
D. Atelectasis of the upper zone of right lung.
E. Congenital emphysema of upper zone of right lung.

3. A child was admitted to hospital with strange radiocontrast body of the right intermediate bronchus. In the evening, during terrible coughing a child had cyanosis, bradypnoe, motor restlessness. On the left breathing was not auscultated. There is suspicion on the left bronchial obturation due to migratory foreign body. What kind of the first aid measures could be recommended?

A. +Emergency sanation bronchoscopy.
B. Pleural puncture.
C. Thoracostomy.
D. Postural drainage.
E. Micro tracheostomy.

4. Which localization of foreign body of the respiratory tract has the following features: reduction of percussion sound, relaxed breathing on the right, lung atelectasis on the right?

A. +Foreign body of the right bronchus.
B. Pharyngeal foreign body.
C. Foreign body of trachea.
D. Foreign body of larynx.
E. Foreign body of the left bronchus.

5. A boy of 5 years old was eating water melon and suddenly started coughing, became pale, began choking. Confused parents started shaking the child, knocking on the back, then breath shortness passed, but at night a cough appeared, and the body temperature rose up to 37.8°C the next morning. The district pediatrician discovered a clap symptom during respiration and sent the child to otolaryngology department immediately. What is the diagnosis?

A. +Foreign body of trachea.
B. Pharyngeal foreign body.
C. Larynx foreign body.
D. Foreign body of the bronchi.
E. Acute stenosing laryngotracheitis.

6. A newborn baby after 1 hour after birth had gradually increasing signs of respiratory failure. During re-examination it is marked a gradual shift of the heart to the right. The left half of the chest is lagging at breathing, percussing the normal lung sounds on the right, tympanitis is periodically determined on the left, and gurgling noises are auscultated during auscultation. On plain radiograph of the chest there is the mediastinum shifted to the right, there are air pockets of different sizes detected on the left at the 2 rib level. Which pathology in the newborn baby are you dealing with?

A. +Diaphragmatic hernia.
B. Pneumonia.
C. Aspiration.
D. Esophageal atresia.
E. Defect of the heart.

7. A child had abundantly secreted saliva foamed of the mouth and nose, increasing dyspnea, cyanosis from the first minutes after birth. The belly was sunken but full at the epigastric region. The tube

stopped when probing the stomach. What defect are you dealing with?

A. +Atresia of esophagus with fistula to the respiratory tract.
B. Atresia of choanae.
C. Atresia of the small intestine.
D. Esophageal atresia.
E. Atresia of the colon.

8. A newborn baby had a significant amount of mucus in the mouth immediately after birth. Atonic vomiting with unchanged milk was observed during the first feeding. An attempt to insert a tube into the stomach was unsuccessful. What kind of diagnosis was possible?

A. +Esophageal atresia.
B. Birth trauma.
C. Achalasia of the esophagus.
D. Hirschsprung disease.
E. Pilorospasm.

9. A newborn with prolonged rupture of membranes is observed with shortness of breath in the first hours of life, which is amplified in a horizontal position. On examination: the left half of the chest is bulging, the heart is shifted to the right, his breathing is puerile on the right, the left bowel sounds are heard, there is shortness of breath. The abdomen is sunken. What is the diagnosis?

A. +Diaphragmatic hernia on the left.
B. Left-sided pneumothorax.
C. Tensed lobar emphysema.
D. Bilateral aspiration pneumonia.
E. Tensed cyst of the left lung.

Malformations of the Digestive Tract. Malformations of the Anterior Abdominal Wall

1. A child of 2 months has vomited milk since the 3–4th week, stool retention, oliguria, visible peristalsis of the stomach. What is the cause of these symptoms?

A. +Congenital malformation — hypertrophic pyloric stenosis.
B. Encephalopathy.
C. Food poisoning.
D. Overeating.
E. Pyelonephritis.

2. A newborn child was diagnosed with the colon perforation during ulcer-necrotic enterocolitis. What management is rational?

A. +Altered bowel resection, anastomosis.
B. Conservative treatment.
C. Laparotomy, stitching of perforated ulcers and colostomy.
D. Stitching of perforation of ulcers without colostomy.
E. Dynamic observation.

3. A newborn is without departing meconium within 24 hours. The child is restless, refusing the feeding. The anus is normal. The abdomen is uniformly full, the baby is crying during palpation. On plain radiograph of the abdomen numerous air bubbles are variegated with horizontal level liquid (Kloiber's cup). Your diagnosis is?

A. +Congenital low intestinal obstruction.
B. Congenital esophageal atresia.
C. Congenital high intestinal obst.ruction
D. Congenital pyloric stenosis.
E. Congenital pilorospasm.

4. A child of the 1st month is observed with jaundice, which appeared on the first day and gradually became intense, getting green. The stool is colorless from birth, the urine has got the color of dark beer. There is increase in the liver. There is bilirubin is 10–15 mg/% in blood. Van den Berg reaction is straight. The presence of urobilin bile pigment is absent in the urine. Your diagnosis is?

A. +Atresia of biliary tract.
B. Physiological jaundice.
C. Hemolytic disease of newborn.
D. Septic jaundice.
E. Mechanical jaundice.

5. An infant is diagnosed with Hirschsprung's disease along with persistent constipation, flatulence of the belly increased in size on contrasting ergography. Specify the pathogenetic cause of the child's disease.

A. +Congenital agangliosis of the colon section.
B. Congenital narrowing (stenosis) of the rectosigmoid colon section.
C. Colonostasis.
D. Hypertrophy of muscular layer of the colon.
E. Congenital dilatation of colon.

6. During hernia excision of a patient (12 years) testes have been found in the hernial sac. What type of hernia does a patient haves?

A. +Birth braid inguinal hernia.
B. Acquired braid inguinal hernia.
C. Direct inguinal hernia.
D. Femoral hernia.
E. Obturator hernia.

7. A boy of 1.5 months has got vomiting after each feeding. He is not gaining in weight. There is reduction of subcutaneous fat layer, hour-glass deformity sign revealed on examination. What is the most probable diagnosis?

A. +Pyloric stenosis.
B. Pilorospasm.
C. Anorexia nervosa.
D. Food poisoning.
E. Adrenogenital syndrome.

8. There is a newborn in the delivery room. On examination having weight status of 2800 g, the child had the swollen abdomen in the epigastric region. Having sounded the stomach, 30 ml of the received contents were of yellowish color. Meconium did not leave. On plain x-ray there were two levels of gas and liquid found in the epigastrium with no gut pneumatization. What kind of the defect are you dealing with?

A. +Atresia of duodenum.
B. Atresia of the esophagus.
C. Ileum atresia.
D. Atresia of the colon.
E. Atresia choanae.

9. On examination of a newborn in the delivery room it was found that the newborn had weight status of 2600 g, the abdomen was normal all over. Having sounded the stomach, it was received 10 ml of transparent content. Meconium did not leave. The catheter, entered through the anus, was 2 cm. On plain X-ray there was pneumatisation of intestine throughout, gas and fluid levels. What kind of the defect are you dealing with?
A. +Atresia of rectum.
B. Atresia of 12 duodenal ulcer.
C. Ileum atresia.
D. Atresia of the anus and rectum.
E. Esophageal atresia.

10. In irriogramm a child of 2 years has got some narrowing in the area of the rectum and sigmoid colon with the colon dilatation over the place. Your diagnosis is?
A. +Hirschsprung's disease.
B. Dolichosigma.
C. Esophageal atresia.
D. Rectal atresia
E. Atresia of small intestine.

11. A child of 1.5 years has got constipation, bloating, malnutrition, choking since birth. Irriogramm shows narrowing in the area of the rectum with the colon dilatation over the place. The anorectal reflex is negative. Your diagnosis?
A. +Hirschsprung's disease.
B. Diaphragmatic hernia.
C. Dolichosigma.
D. Payr's disease.
E. Atresia of the colon.

12. After a while a newborn child has got vomiting of stagnant intestinal contents with impurities. Meconium is absent. Stool is in a form of thin mucous streaks. What kind of the diagnosis was possible?
A. +Low intestinal obstruction.
B. Pilorospasm.
C. Hirschsprung's disease.
D. Sepsis.
E. Cystic fibrosis.

13. There is a boy of 6 weeks in the children's unit of the district hospital, who has clinically and X-ray diagnosed congenital hypertrophic pyloric stenosis. Within 10 days he has been given antispasmodic, infusion and generally reinforcing therapy. According to the pediatrician, the child's condition has improved slightly. What are further treatment management?
A. +Surgical intervention.
B. Outpatient observation.
C. Purpose of antibiotics.
D. Intraorganic electrophoresis of antispasmodics.
E. Mechanical pilorodilatation.

14. A child of 3 years has stool retention from the date of birth; the mother makes enema every 3–4 days. The boy has failure to thrive. It is marked skin pallor, bloating. What disease should be assumed?
A. +Hirschsprung's disease.
B. Coprostasia.
C. Worm infestation.
D. Peritonitis.
E. Intestinal tumor.

15. An infant is 1.5 months old. The mother is complaining on daily infrequent vomiting, like a fountain, immediately after feeding. The volume of these masses is larger than consumed food. The baby has been sick since the 2nd week of life. The child is flabby, with pale skin, the folds slowly straighten. Weight deficit of the body is 24 %. Hour-glass deformity sign is clear. Constipation. Little ammounts of urine. What is the diagnosis?
A. +Pilorostenosis.
B. Pilorospasm.
C. Hipervitaminosis D.
D. Adrenogenital syndrome.
E. Cardiostenosis.

Malformations of the Musculoskeletal System

1. A student of a vocational school has blades asymmetry, the waist triangle is less prominent on the left; there is a muscle spindle on the back to the left; the arc angle of curvature of the spine is 20°; the bending partially disappears at active straightening of the back. The diagnosis is?
A. +Scoliosis grade I–II.
B. Stooped posture.
C. Kyphotic posture.
D. Lordotic posture.
E. Scoliosis grade III–IV.

2. On examination of a 10-year-old girl the doctor determines that all the physiological curves of her spine are barely marked. The head is tilted anteriorly due to low-grade cervical lordosis. What type of the back has the child got?
A. +Flat.
B. Slouch.
C. Round.
D. Normal.
E. Undifferentiated.

3. On examination of a newborn born in breech presentation, the pediatrician located passive abduction of the right thigh and click in abduction, asymmetry of the inguinal and femoral gluteal folds. What sign does the child's congenital hip dislocation reliably indicate?
A. +Clicking during hip abduction.
B. Birth in breech presentation.

C. Limitation of hip passive.
D. Asymmetry of inguinal and femoral skin folds.
E. No reliable signs.

TEST TASKS

1. A child had foamy nose and mouth discharges, attacks of cyanosis immediately after birth. What disease should you think about?
A. +Achalasia of oesophagus.
B. Congenital defects of development of lungs.
C. Obstetric craniocerebral trauma.
D. Stenosis of oesophagus.
E Atresia of oesophagus.

2. You are called to consult a three-day-old boy. The child is restless, has got repeated vomiting, the abdomen is bloated in a volume. Meconium doesn't evacuate. Your preliminary diagnosis?
A. +Pylorospasm.
B. High congenital intestinal obstruction.
C. Pylorostenosis.
D. Obstetric craniocerebral trauma.
E. Low congenital intestinal obstruction.

3. A newborn has got progressing cyanosis, harsh shallow breathing with participation of additional muscles. The abdomen is small in size; intestinal peristalsis is auscultated in the left part of the chest. The suggested diagnosis is diaphragmatic hernia. Which examination would be decisive?
A. +An X-ray of the gastrointestinal tract.
B. An abdominal X-ray.
C. An abdominal ultrasound.
D. Endoscopy.
E. Punction pleural cavity.

4. A six-month-old baby has been suffering for 8 hours. According to the mother' words among a full health there were panic attacks, some vomiting; a dark bloodstain has appeared on the napkin. Objectively: the abdomen is soft, a tumour is palpable on the right. The diagnosis is invagination. Your managements includes:
A. +Pneumodesinvagination.
B. Surgical intervention.
C. Laparoscopy.
D. Enema.
E. Ultrasound.

5. A child, 2 years old, was hospitalised into surgery department with complaints of the body temperature rise up to 38.3°C, repeated vomiting, abdominal pain. Objectively: the patient's state is moderate severity, the body temperature is 38.4°C, the tongue is coated, the abdomen is uniformly inflated, and muscle tension of the abdominal wall is determined on palpation. Your diagnosis is?
A. +Primary peritonitis.
B. Acute enterocolitis.
C. Invagination.
D. Coprostasis.
E. Acute appendicitis.

6. A newdorn of 2 days old was admitted with anal atresia from maternity hospital. What examination is necessary to determine the atresia type?
A. +An X-ray by Wangensteen.
B. An abdominal ultrasound.
C. An abdominal X-ray.
D. Pneumoirrigography.
E. Contrast X-ray investigation.

7. A newdorn had the body temperature rise up to 38,3°C, anxiety at the end of the second week of life. The right hip joint is increased in a volume, painful on palpation. The right lower limb is immobile. An increased articular slot of the hip joint was revealed on the ultrasound. What kind of disease could it be possible?
A. +Transitional coxitis.
B. Phlegmon of newborns.
C. Hemarthrosis of the right hip joint.
D. Metaepiphyseal osteomyelitis.
E. Trauma of a hip joint.

8. A newdorn had anxiety on the second week of life. The body temperature increased up to 38°C. On examination there was skin hyperaemia, oedema; tenderness is detected in the lumbar region on palpation. Your diagnosis is?
A. +Necrotic phlegmon of newborns.
B. Adiponecrosis.
C. Erysipilatous inflammation.
D. Sclerema.
E. Calcification.

9. Deformation of the right foot (supination, reduction of the forward part, equines) was detected on examination of a newborn. What defect of development of the child's foot should you think about?
A. +Arthrogryposis.
B. Congenital talipes.
C. Reduced stop.
D. Plainly-valgus foot.
E. Heel's foot.

10. Oedema and pain were revealed in a newborn on palpation of the right clavicle postpartum. The right upper limb was immobile. What pathology could be suspected?
A. +Fracture of clavicle.
B. Metaepiphyseal osteomyelitis.
C. Duchenne–Erb paralysis.
D. Right-side torticolis.
E. Children's cerebral paralysis.

11. Limitation of driving back in the left hip joint, linear shortness of the limb, asymmetry of dermal tucks were revealed on examination of a newborn girl. Other pathologies were not revealed. Your suggested diagnosis is?
A. +Metaepiphyseal osteomyelitis.
B. Congenital linear shortness of the limb.
C. Children's cerebral paralysis.

D. Birth trauma.
E. A congenital dysplastic dislocation of the left femur.

12. A newborn has got a fracture of the left-hand femur in the mean third aborning. Objectively: there is soft tissues oedema, deformation and linear shortness of the femur. What medical managements would you choose?
A. +Plaster extension by Schede.
B. Plaster bandage.
C. Skeletal traction.
D. Crede bandage.
E. Opened reposition.

13. What primary X-ray sign is the characteristic for acute hematogenous osteomyelitis in children?
A. +Bulbar periostitis.
B. Needle-like periostitis.
C. Periostitis for the "visor" type.
D. Osteosclerosis.
E. Linear periostitis.

14. A child, 7-year-old, is complaining on deformation of the thorax. It is known from the anamnesis, that the deformation was detected immediately after birth, then there were phenomena of a paradox inhalation. Objectively: there is recess of the sternum and ribs up to 2–2.5 cm. Your diagnosis is?
A. +Congenital funnel-shaped deformation of thorax.
B. Rachitic deformation of thorax.
C. Congenital defect of heart.
D. Marfan's Disease.
E. Version of the constitution.

15. A girl of 12 years is complaining on the body temperature rise up to 37.4°C, abdominal pain. On examination the girl is acyanotic, weak; the tongue is coated, the abdomen is inflated. The painful fixed tumorous formation was determined in the right ileac area on palpation. The sign of peritoneum irritation is absent. Your diagnosis is?
A. +Coprostasia.
B. Intestinal obstruction.
C. Ascariasis.
D. Swelling ileocecal angle.
E. Appendicular infiltrate.

16. You were invited to consult in a maternity hospital. There was a newborn boy, who had had vomiting during a day. It was marked abdominal distension, swelling of the anterior abdominal wall and genitals. Your suggested diagnosis is?
A. +Necrotic enterocolitis.
B. Meconium ileum.
C. Peritonitis of newborns.
D. Low congenital intestinal obstruction.
E. High congenital intestinal obstruction.

17. A child, 12 years old, has been suffering for 2 days. He is complaining on hyperaemia, pain in the lower third of the femur and in the knee joint. There is a trauma of the knee joint in his anamnesis. There is a local temperature rise, minor infiltration of soft tissues on examination. The local pain is boosted on percussion. The suggested diagnosis is acute hematogenous osteomyelitis of the lower third of the right femur. What is the pain caused by on the first days of osteomyelitis?
A. +Increased intraosseal pressure.
B. Trauma.
C. Increase intraarticular pressure.
D. Contracture of a joint.
E. Department of a periosteum.

18. A three-year-old child has got periodic massive bleedings from the lower evacuated ments intestine with no changings of blood. What is the most probable cause of such bleeding?
A. +Hemorrhoids.
B. Polyp of a rectum.
C. Anal fissure.
D. Ulcerative colitis.
E. Ulcer Meckel's diverticulum.

19. A cild, 3 years old, does not go potty by himself. It is determined failure to thrive, abdominal distension on examination. The diagnosis is Hirschprung's disease. What is the cause of Hirschprung's disease?
A. +Congenital stenosis of a colon.
B. Prelum embrional by cicatrix.
C. Chronic colitis.
D. Mucoviscidosis.
E. Congenital agangliosis of intestinal wall.

20. It is necessary to convey a newborn with an atresia of the oesophagus to a specialised hospital. What is necessary to do to prevent the aspiration during the conveyance?
A. +Hemivertical position of the child.
B. To make a tracheotomy.
C. To enter a constant catheter into the oesophagus.
D. To convey the child lying on the abdomen.
E. To enter a catheter into the oesophagus and to pump out slime everyone 15–20 minutes.

21. A baby of 3 weeks old has got anxiency, the body temperature rise up to 38.3°C. Objectively: there are increase and hyperaemia of the right mammary gland, painful on palpation. The diagnosis is mastitis. What local treatment should you administer to prevent any complication of activity of the mammary gland in future?
A. +Punctional method.
B. Notch by the "chess" scheme.
C. Arc-like cut (section, sectional view).
D. By radial cuts — notches.
E. Retromammarial introducing of antibiotics.

22. A child, 10 years old, is with acute hematogenous osteomyelitis of the lower third of the right femur in hospital. A considerable osteal tissue destruction, periostitis was determined on the X-ray. What measures are necessary to carry out preventing a pathological fracture?
A. +Fixing (fixation) of a limb with gypsum bandage from a foot up to lumbar area.

B. Skeletal traction.
C. Soft bandage.
D. Walking with skids.
E. Unloading vehicle.

23. Name the optimal volume of antishock complex on pelvic fractures?
A. +Intramuscular introducing of drugs.
B. Fixing on a board.
C. Fixing on a board in a position of "bellows", intrapelvical blockade on Shkolnikov–Selivanov's, infusion therapy.
D. Medical narcosis without immobilisation.
E. I/v introducing of narcotics, transfusion therapy

24. What diseases is the paradox inhalation sign pathognomonic for?
A. +Diaphragmal hernia.
B. Of congenital funnel-shaped deformation of thorax.
C. Aplasia of the diaphragm.
D. Cleavage of a breast bone.
E. Muscles aplasia of the forward abdominal wall.

25. What is the most often cause of asphixitic syndrome in children requiring a surgical intervention?
A. +Lobar emphysema.
B. Destructive pneumonia.
C. False diaphragmatic a hernia.
D. Congenital atelectasis of a lung.
E. Swelling of a mediastinum.

26. A child, 5 years old, wake up at night because of abdominal pain. There was vomiting with food. In the morning the child was week, refused eating, was complaining on colic pain in the right half of the abdomen. There was no stool more than 2 days. Your suggested diagnosis and managements is?
A. +Coprostasia. To make a cleansing enema.
B. An acute appendicitis. A cleansing enema. Common blood and urine test. Repeated examination of the surgeon.
C. Acute appendicitis. Operational treatment.
D. Coprostasia. To make to survey X-ray abdominal cavity.
E. Alimentary intoxication. A gastric lavage.

27. You are a doctor of the first aid. You were called to a child, 2 years old, which was complaining on abdominal pain. The hyperaemia of the face, nasal mucoid discharge, the body temperature rise up to 38° were revealed on examination of the child. The fauces was hyperaemic. The abdomen participated in breathing, painful on palpation. The child resisted to examination. Your managements is?
A. +Antibiotics. Antiinflammatory drops in a nose.
B. To hospitalise the child in a somatic hospital.
C. To hospitalise in a children's surgical hospital, consulting of the otorhinolaryngological doctor and pediatrist.
D. To hospitalise in otorhinolaryngological department.
E. To hospitalise in infectious department.

28. Parents of a girl, 2.5 years old, called you. Her disease began with temperature rise up to 38° C, repeated vomiting, and pain in the abdomen 2 days ago. The child slept badly at night, was moody, refused eating, the stool remained normal. The toxicosis phenomena were discernible in the girl. She did not go towards the contact, resisted to examination. Your suggested diagnosis is?
A. +Pleuropneumonia.
B. Ascariasis.
C. Primary peritonitis.
D. Intestinal obstruction.
E. An acute appendicitis.

29. A boy, 9 years old, is admitted to the medical admission unit of children's hospital with strong colic pains in the right half of the abdomen. He has been suffering for 12 hours, then pains, frequent emiction have appeared, and there is vomiting. The child's condition is moderate. There is anxiety, paleness of skin integument. The body temperature is 37.2°C, the pulse rate is 92/min, and the tongue is wet. The abdomen is uniformly inflated, sharply painful in the right half, Pasternatsky sign is positive on the right. Your suggested diagnosis is?
A. +Acute appendicitis.
B. Invagination.
C. A righted renal colic.
D. Mesadenitis.
E. Diverticulitis.

30. A child, 12 years old, suddenly had collapse, profuse vomiting with dark blood not changed. On examination: there is pale skin integument, BP was 80/50 mm Hg. The abdomen is painful. There is omphalitis in the anamnesis. Your suggested diagnosis is?
A. +Chalasia of oesophagus.
B. A portal hypertension.
C. Peptic ulcer of abdomen.
D. Capillary toxicosis.
E. Bleeding from Meckel's diverticulum.

31. A boy, 10 years old, was operated 3 days ago because phlegmonous appendicitis got complicated by local peritonitis. The abdominal cavity is hardwired tightly. The body temperature is 37. 9°C; the wound is clean without inflammation. There is pain in the right ileal area, nausea. Muscles tension is marked in the right ileal area. Symptom of Shchotkin-Blumberg is doubtful. Your diagnosis is?
A. +Pyesis of postoperative wound.
B. Appendicular infiltrate.
C. Intestinal obstruction.
D. Abscess of the right ileac area.
E. Spread peritonitis.

32. A girl, 6 years old, got ill acutely, when pain appeared in the lower parts of the abdomen, which spread on all abdominal cavity. The body temperature was 39.4°C. Vomiting was 2 times. She became

week, refused eating, the tongue was coated, dry. The general condition is getting worse. Objectively: there is the tight abdomen, positive signs of peritoneum irritation, hyperaemia of fauces and mucous, maceration and discharges from vagina. Your suggested diagnosis is?

A. +ARVI.
B. Tonsillitis.
C. Acute appendicitis.
D. Pyelonephritis.
E. Primary peritonitis.

33. Point the causes of bleeding from the digestive tube in children from 1 year till 3 years.

A. +Doubling an intestine.
B. Meckel's diverticulum.
C. Hernia oesophageal opening of diaphragm.
D. Chalasia of oesophagus.
E. All numbered reasons.

34. You are a doctor of the first aid, who was called to a child of 8 months in 8 hours from the beginning of a disease. The mother told, that there were panic attacks, legs tapping, vomiting arisen among the full health. The stool was the day before. There were discharges from the rectum like crimson jelly on rectal exam. Your suggested diagnosis is?

A. +Acute appendicitis.
B. Capillary toxicosis.
C. Meckel's diverticulum.
D. Invagination.
E. Dysentery.

35. You are a doctor of the first aid, who was called to the boy of 10. He got ill acutely 6 hours ago, when sharp abdominal pains, vomiting have appeared. He is in the enforced position — genucubital. On examination the abdomen is moderately inflated, the peristalsis is increased, there is acute pain on palpation, positive signs of peritoneum irritation. Your suggested diagnosis is?

A. +Coprostasia.
B. Acute appendicitis.
C. Intestinal infection.
D. Acute intestinal obstruction.
E. Acute gastritis.

36. A child, 7 months old, is admitted to the department with complaints on paroxysmal anxiety, vomiting once in 8 hours after the beginning of a disease. There is a tumorous formation palpated in the right half of the abdomen. Some dark blood was on rectal exam. What disease is referred to?

A. +Doubling of intestine.
B. Invagination.
C. Tumour of abdominal cavity.
D. Ascaridas invasion.
E. Enterocystoma.

37. A child, who was operated 6 months ago because of phlegmonous appendicitis, has got colic pains in the abdomen, double vomiting after digestive disorder. The abdomen is soft, participates in breathing; the peristalsis amplification is determined. The suggested diagnosis is commissural obstruction. The X-ray of the abdominal cavity was made. What X-ray logical sign shows adhesive intestinal obstruction?

A. +Single different diameter's Kloiber's cup.
B. Multiple different diameter's Kloiber's cup.
C. Exuberant pneumatization of intestine.
D. Sign of "sickle".
E. Homogeneous black-out.

38. A child aged 6 months had paroxysmal pains in the abdomen, vomiting, and some dark blood in the stool after having eaten mashed vegetables. They suspected invagination at the medical admission unit. What signs are specific to this disease?

A. +"Vanka-vstanka" sign.
B. Sign of "scissors".
C. Dance's sign.
D. Filatov's sign.
E. Kocher's sign.

39. A child of 7 months is delivered to the clinic in 18 hours after the beginning of disease. There are complaints on paroxysmal abdominal pains, vomiting twice; there are fecal masses like blackberry jelly. The diagnosis is invagination. What management is necessary to be carried out?

A. +Laparoscopy.
B. Operating treatment.
C. Closed desinvagination.
D. Observation.
E. Siphon enema.

40. A boy, 10 years old, had strong colic pains in the abdomen, repeated vomiting after copious meal. There was no stool and gases. The child had laparotomy in occasion of a closed trauma of the abdomen 2 years ago. The abdomen is asymmetric, inflated in the left half; the peristalsis of the intestine is determined. On rectal exam there are incontinence sphincters, empty ampoule of the rectum. The suggested diagnosis is?

A. +Early commissural obstruction.
B. Late commissural obstruction.
C. Dynamic obstruction.
D. Early delayed obstruction.
E. Volvulus of a mean intestine.

41. A child, 5 years old, was operated because of gangrenous appendicitis, spread peritonitis. Abdominal pains, distendion appeared on the 5th day after operation; there was vomiting two times. Dyspnea, tachycardia are marked. The abdomen is severe uniformly inflated; the single weak peristaltic sounds are heard on auscultation. There are multiple hydroaeric levels on the X-ray. Your diagnosis is?

A. +Peritonitis.
B. Invagination.
C. Adhesive paretic intestinal obstruction.
D. Dynamic obstruction.
E. Pneumonia.

42. A child, 7 years old, was operated for spread peritonitis. His general state got worsened on the 4th

day after the operation, when paroxysmal abdominal pains, frequent vomiting appeared, and there was no stool. The surgeon suspected early adhesive paretic intestinal obstruction. What measures are necessary to be initiated?

A. +Conservative treatment.
B. Operating treatment.
C. Observation.
D. Amplification of antibacterial therapy.
E. Cleansing enema.

43. A child, 7 years old, was operated on the 3rd day from the beginning of disease because of gangrenous perforated appendicitis, spread peritonitis. By the clinical signs he was referred to a group which can have development of commissural obstruction. When is it necessary to take measures for prevention of commissural process in the abdominal cavity?

A. +In operation.
B. Since 2 day after operation.
C. In 1 week after operation.
D. In 2 weeks after operation.
E. In one month after operation.

44. A child, 6 years old, had increased abdominal pains, vomiting on the 3rd day after operation of appendicular spread peritonitis. The abdomen is inflated. Weak peristalsis is tracked from time to time. Gentle peristaltic sounds are determined by auscultation. There is no independent stool. The diagnosis is early adhesive paretic intestinal obstruction. What X-ray signs are specific to this disease?

A. +Single hidroaeric level s.
B. Two hydroaeric level
C. Mute abdomen.
D. Multiple hydroaeric level s.
E. Sign of "ablation".

45. A child of 12 suddenly had strong colic pains in the abdomen, vomiting. There was no stool and no gases flatulated. The abdomen is asymmetric. The peristalsis is determined. Empty rectal ampoule was revealed on rectal exam. The child was operated because of spread appendicular peritonitis one year ago. The diagnosis is late adhesive intestinal obstruction. What X-ray signs are to confirm this disease?

A. +Single Kloiber's cups in the upper part of the abdominal cavity.
B. Two Kloiber's cups.
C. Mute abdomen.
D. Multiple cups Kloiber's in all abdominal cavity.
E. Sign of "ablation".

46. Survey X-ray examination of thorax was made to a child admitted. Sharp displacement of the heart contours was determined, and cellular cavities of irregular value were in the pulmonary area of the counter part. The diaphragm contours were not differentiated. What pathology is the X-ray given specific to?

A. +Polycystosis of lungs.
B. Congenital lobar emphysema.
C. Congenital false diaphragmatic hernia.
D. Congenital cyst of lung.
E. Parasitogenic cyst of lung.

47. The surgeon has suspected high congenital intestinal obstruction on examination of a child in maternity hospital. Which of the following signs are most typical for this pathology?

A. +Vomiting after birth during 1 day.
B. Even abdominal distension.
C. Spread closed loops intestine are seen through the anterior abdominal wall.
D. Visible peristalsis of the closed-loop obstruction.
E. Meconium evacuated in a small amount.

48. A newborn had intestinal contains and gases from umbilical ring after lose of umbilicus. What disease should be suspected?

A. +Gastroshisis.
B. Omphalocele umbilical funiculus.
C. A full fistula of a belly-button.
D. Incomplete fistula of a belly-button.
E. Disturbance of return development of urachus.

49. A newborn, 3 weeks old, is hospitalised in connection with vomiting, which began at the age of 2 weeks. There is projectile vomiting, stagnant acidic contains, plentiful constipation, infrequent emaciation, exhausting. Your diagnosis is:

A. +Adrenogenital syndrome.
B. Pylorostenosis.
C. Pylorospasm.
D. Obstetric craniocerebral trauma.
E. Invagination of intestine.

50. A newborn has got a tumorous formation in the area of the umbillicus (size about an apple), boarded by smooth, brilliant, translucent white membrane, through which one the organs of the abdominal cavity is visible. What disease should you think about?

A. +Umbilical a hernia of large size.
B. Aplasia of anterior abdominal wall.
C. Omphalocele umbilical funiculus.
D. Anomaly of development omphaloentericus duct.
E. Anomaly of development of urachus.

51. Children's surgeon has been called in maternity hospital to exam a newborn of 2 days old suspected intestinal obstruction. What examination should be done at first term?

A. +FGS.
B. FGS.
C. The ultrasound of the abdominal cavity.
D. The survey X-ray of the abdominal cavity.
E. Irrigography of the small intestine.

52. An infant at the age of 3 weeks entered the medical admission unit with suspected Congenital pylorostenosis diagnosis. What disease is it necessary to work-up differential diagnosis with?

A. +Invagination of intestine.
B. A pseudopylorostenosis.
C. Meconial obstruction.
D. Intestinal obstruction.
E. Gastroenterocolitis.

53. A newborn at the age of 3 day has got vomiting with addition of bile excretion. What kind of diseases are observed with this pathology?
A. +Atresia of a small intestine.
B. Atresia ileal intestine.
C. Volvulus of a mean intestine.
D. Syndrome Ladd's.
E. All named diseases.

54. A girl aged 1,5 month is transferred from district hospital with the pylorostenosis diagnosis. She has been ill for 2 weeks, when the nonconstant projectile vomiting with bile excretion, diarrhoea, adynamia, food refusal, weight loss have appeared. The girl's condition is moderate. She is weak, acyanotic; skin integument is dry, and the abdomen is inflated in epigastrium. There is swelling of the mammary gland, as well as pubescence is marked. Your suggested diagnosis is?
A. +Pylorostenosis.
B. Pylorospasm.
C. A pseudopylorostenosis.
D. Prepyloric diaphragm.
E. Ladd's Syndrome.

55. A child at the age of 2 months entered the medical admission unit with the complaints on stool and gas retention, abdominal distension, vomiting twice. Handicapping of passage of flatus and stool is marked from birth. Now they do not make cleansing enemas and not insert a colonic tube for facilitation. Your suggested diagnosis is?
A. +Ladd's syndrome.
B. Volvulus of the mean intestine.
C. Enterocystoma.
D. Hirschprung's disease.
E. Dolichosigma.

56. In maternity hospital there was a newborn (weight — 3200 g, growth — 51cm) who was revealed with anal orifice escalation and rectovestibular fistula, through which Meconium freely separated. What examination is it necessary to conduct verifying the diagnosis?
A. +The ultrasound.
B. Fistulography.
C. Survey X-ray of the abdominal cavity.
D. Research on Wangensteen.
E. Colonoscopy.

57. A girl, 9 years old, was hospitalised to Surgery with complaints on the body temperature rise up to 38°C, pain in the low third of the right femur, limitation of motions in the right knee joint on the 3rd day. Objectively: the low third of the femur and the knee joint are edemed; there is hyperaemia on the flank surface, and painful metaphysis when percussing. What disease has got such signs?
A. +Inflammation of a knee joint.
B. Phlegmon of soft tissues.
C. Osteomyelitis of a femur.
D. Distorsio of a knee joint.
E. Lymphadenitis.

58. A child, 8 years old, was hospitalised to surgery suspected AHO of the right femur. The child has been suffering for 2 days. You should select the most authentic method of diagnostic:
A. +Diagnostic punction of soft tissues.
B. Phlebography.
C. Electroroentgenography.
D. Determination of intraosseal pressure.
E. Osteoperforation.

59. A child, 4 years old, has got the clinic of AHO of the low third of the femur, a septicopyemic form. It's the 2nd day of the disease. Pus hasn't been obtained in punction of soft tissues. Your managements is:
A. +Section of soft tissues.
B. Osteoperforation + antibiotics in centre of inflammation.
C. Exposure of soft tissues. Osteoperforation by a milling cutter.
D. Antibiotics intravenously.
E. Exposure, osteoperforation, rubber venting.

60. In the neonatal pathology unit there is a newborn, 26 days old, with the umbilical sepsis diagnosis. Oedema of the left shoulder, absence of fissile motions in the shoulder joint, with the body temperature of 38° appeared 2 days ago. The diagnosis is?
A. +Metaephyseal osteomyelitis.
B. Fracture.
C. Phlegmon.
D. Duchenne–Erb paralysis.
E. Traumatic epiphysiolyisis.

61. A child aged 1 year and 8 month, was treated for destructive pneumonia. She began walking only at 12 months, and thus limped the right leg. Objectively: the right femur is shortened, the driving back is limited. On the X-ray examination: the left hip joint is without features. The femur dislocation is on the right, its neck is missing. The diagnosis is?
A. +A pathological dislocation of femur.
B. Congenital dislocation of femur.
C. Traumatic epiphysiolyisis.
D. Pathological epiphysiolyisis.
E. Children's cerebral paralysis.

62. A newborn had the body temperature up to 38.7°C, anxiety to the end of the second week. Objectively: there is hyperaemia, oedema in the area of the umbillicus. It is painful when palpated. The diagnosis is?
A. +Omphalitis.
B. Necrotic phlegmon.
C. Water-omphalitis.
D. Adiposonecrosis.
E. Scleroma of newborns.

63. A child, 8 years old, is hospitalised with pains in the left calf. The body temperature is 37.3°C. He had AHO of the left tibia bone 14 months ago. A bruise of the left calf appeared 3 days before the disease. Locally: there is oedema, hyperaemia, and severe painfulness of soft tissues on the left calf ante-

rior surface. On the X-ray examination: two steal cavities are determined in the middle third of the left tibia bone because of sclerosis. Your diagnosis is?

A. +Phlegmon of soft tissues.
B. Osteal sarcoma.
C. Tuberculosis.
D. Pyesis of hematoma.
E. Chronic osteomyelitis.

64. A patient, 10 years old, is complaining on periodic pains (more at night) in the upper third of the right calf. He has been suffering for 3 months. He connects the disease to a trauma. Objectively: the limb doesn't differ from the healthy one. The motions are painless in the knee joint. On the X-ray examination: there is a spherical cavity of 2 sm sized, with legible edges of the tibia bone proximal metaphysis. The diagnosis is?

A. +Brodi abscess.
B. Tuberculosis.
C. Fibroseal dysplasia.
D. Cyst.
E. Ostoclastoma.

65. The right hand of a newborn is hanging down fixed along a trunk: there is a tumescence, deformation, and painfulness on palpation in the middle third of the right shoulder. What pathology should be suspected?

A. +Obstetrics paralysis by Duchenne–Erb.
B. Fracture of a shoulder in a middle third.
C. Pathological fracture.
D. Acute haematogenous osteomyelitis.
E. Children's cerebral paralysis.

66. A child, 3 years old, was transported to traumatology with the suggested diagnosis "Sublaxation of the head of radius". Which of the following signs is most typical for this trauma?

A. +The child holds an ingured arm by a healthy arm.
B. The arm is pronationated, is prolated along a trunk, is slightly bent in elbow.
C. There are motions in elbow.
D. Deformation of elbow.
E. There is tumescence in the field of elbow.

67. While swimming, a boy of 13 struck his head against the bottom, then he felt a severe pain in the area of the neck. On examination: the head is holding unnaturally directed; the chin is supported by hands; the head rotation is hindered. What damage could the patient have?

A. +Dislocation of a mandible.
B. Dislocation-fracture of cervical part of a backbone.
C. Bruise of a spinal cord.
D. Stretching ligament of cervical part of a backbone.
E. Intermuscular hematoma.

68. A child, 8 years old, was delivered to traumatology with the diagnosis of closed oblique fracture of bones in the middle part of the right calf without displacement. What treatment management for the fracture should you prescribe?

A. +Opened reposition.
B. Plaster bandage.
C. Ilizarov's apparatus.
D. Plaster extension.
E. Skeletal traction.

69. A boy, 10 years old, after a dip felt a severe pain in the hip, where it was detected a tumescence, deformation. He can't stand on the leg. Which of the following signs refers to the fracture authentic signs?

A. +Tumescence.
B. Deformation.
C. Crepitation parts of fracture.
D. Pain on palpation.
E. Disorders of function.

70. A boy of 9 years was admitted to the first-aid station with complaints on pain, absence of movements in the left shoulder joint. When he was riding, he felt on his left hand. Objectively: there is swelling of the upper third of the left shoulder, painfulness, and deformation. On the X-ray there is epiphysiolyisis of the head of the left shoulder bone with displacement. What kind of treatment is to be assigned at this pathology?

A. +Closed reposition, Dezo bandage.
B. Opened reposition.
C. Closed reposition, Gromov bandage.
D. Skeletal traction.
E. Plaster extension.

71. In children's traumatology there are a lot of fractures caused by anatomic-physiological features of a skeleton. You should point, after what type of fractures there can appear brittleness of a limb as complication?

A. +Subperiostal fracture.
B. Fracture by the type of "green tree".
C. Epiphysiolyisis, oysteoepiphysiolyisis.
D. Fracture with many parts.
E. Apophysiolyisis.

72. A child of 10 years old had pain, swelling, deformation in the low third of the left forearm after landing on the arm. There was revealed a chisel fracture on the X-ray examination. What type of dislocation on the fracture is it possible to consider exceptable in this case?

A. +Dislocation lengthways up to 1 sm.
B. Dislocation bevel way up to 10°.
C. Dislocation bevel way more than 10°.
D. Dislocation by width (on 1/3 diameters).
E. Dislocation by width in all diameter.

73. A child was in a motor vehicle accident. He was complaining about pains in the down part of the abdomen, vast hematoma in the region of the symphysis. Horizontal branches fracture of the pubic bone was revealed in the X-ray examination. What clinical sign is specific to this fracture?

A. +Limitation of flexion in p/f joints.

B. Verneil's Sign.
C. Outside rotation of limb.
D. Sign of "adhered heel".
E. Limitation of driving back in hip joint.

74. A child is under treatment in traumatology with the diagnosis of a symphysis. You should determine medical managements at a symphysis gap with the divergence:
A. +Imposing of a skeletal traction.
B. Fixing on a board in a position of bellows.
C. Juncture of a pubic articulation.
D. Fixing by a plaster bandage.
E. Fixation in bed.

75. A child received a craniocerebral trauma accompanying with an increasing headache, repeated vomiting, psychomotor restnessless, and confusion of consciousness. The subarachnoidal haemorrhage is suspected. What examination is most necessary for the diagnostic verification?
A. +Back-cerebral Punction.
B. X-ray of bones of a skull.
C. Trepanation of bones of a skull.
D. Analysis of a blood.
E. Determination of intracranial pressure.

76. A child, 10 years old, was admitted to the injury care center with the diagnosis of an open fracture of the right calf in middle third with dislocation, syndrome of a prelum of soft tissues. The trauma was received on building, where concrete design had pressed down the leg. Objectively: there is an oedema of soft tissues in the middle and low third of the right calf, a lacerated wound on the anterior surface, paleness of skin integument of the calf; the toes are warm, coloured normally. What is the plan of local treatment to this patient?
A. +Closed reposition, plaster bandage, FSI.
B. Skeletal traction, primary surgical treatment of a wound.
C. Opened reposition of parts metalostoesynthesis.
D. Closed reposition, FSI, Plaster extension.
E. FSI, processing of a skin on Krasovitov's, imposing of the Ilizarov's apparatus.

77. Parents of a 12-year-old child, who had a fracture of the femur 2 months ago, have applied for consulting aid. The boy was treated with a skeletal traction at regional hospital. Objective: there is deformation in the middle third of the femur; the limb is shortened. There are osteal callosities, angular dislocation of about 30°; deformation by width on half of diameter seen on the X-ray examination. What types of dislocated parts of the fracture are being corrected during a child's growth?
A. +Dislocationby width is corrected completely (up to cortical layer).
B. Dislocationlengthways up to 1cm.
C. Dislocationbevel way 15°.
D. All types of dislocationare corrected during growth.
E. Residual dislocationisn't corrected.

78. A child, 10 years old, is complaining about pain in the region of the left clavicle after falling from a tree. On examination there is swelling, crepitating on palpation. There is a fracture of the clavicle in the middle third with dislocation lengthways and by width on all the diameters seen on the X-ray examination. The reposition and imposing of immobilising bandage is ordered. What is the type of a bandage used in this dislocation?
A. +Bandage by Deso.
B. Imposing of rings.
C. Trunk Alberts.
D. Thoracic-brachial bandage.
E. Bandage such as Kuzminskii's trunk.

79. A boy, 13 years old, fell on the arm from some height, bent in the elbow. On examination: there is a signed swelling in the field of elbow; Marks' sign is disturbed. The passive motions are possible. The suggested diagnosis is:
A. +Bruise of the elbow.
B. Dislocation of the forearm bones.
C. Transepicondylar fracture.
D. Fracture of neck of radius.
E. Intermuscular hematoma.

80. A child, 6 months old, is conservatively being treated for congenital talipes of the 3rd degree, however, deformation of foot is not cured. What treatment should be prescribed to the child?
A. +To prolong plaster bandages.
B. Operating treatment.
C. Correction orthopaedic bandage.
D. Medical gymnastics and massage.
E. Nothing to undertake till 1 year.

81. Congenital muscle torticolis is suspected in a child, 3 months old. What of the following signs can be used for differential diagnostic?
A. +Asymmetry of facing skull.
B. High localisation of forearm.
C. Tilt of a head.
D. Limitation of motions in cervical part of column vertebras.
E. Linear shortness and thickness of sternoclaidimastoideus muscle.

82. A child, 4 years old, is complaining about frequent fractures of long tubular bones. Imperfect osteogenesis is suspected. What of the following signs is specific to this disease?
A. +Low growth.
B. Blue colour of sclera.
C. Disproportion of a body build.
D. Mongoloid section of eye.
E. High palate.

83. A child, 5 months old, was born with asphyxia. Examining lying down, driving back in the hip joints is limited, as well as the back creasing of feet is limited. Decussation of the lower extremity, equine position of feet were observed upright. The suggested diagnosis is:
A. +Congenital dislocation of hip.

B. Congenital talipes.
C. Children's cerebral paralysis.
D. Congenital two-sided varus deformation of a femur.
E. Physiological hypertonus of muscles.

84. A child, 3 years old, is complaining about asymmetry of the face, a head tilt to the right, which have not been treated before. On examination there is asymmetry of facing and cerebral skull; the head is tilted to the right; the face is turned around to the left, swollen and linear shortening of the right sternoclaidomastoideus muscle is determined. The diagnosis is congenital right muscle torticolis. What treatment plan would you assign?
A. +Treatment physical training, massage.
B. Wearing bandage for correction.
C. Operative measures exigent.
D. Operative measure after 5 years.
E. Extension by a closed Glisson loop.

85. A newborn has got syndactylia III–IV of the right hand fingers without deformation. Motions in the interphalageal joints are free. At what age is it expedient to operate syndactylia?
A. +In period of newborn.
B. Till 1 year.
C. At the age of from 3 till 6 years.
D. At school age.
E. After 6 months.

86. Parents of a child, 1.5 years old, with the diagnosis of talipomanus applied for consultation to polyclinic in connection with underdevelopment of the left radial bone. What would you recommend for the child's treatment?
A. +The operative measure is urgent.
B. To conduct massage, physical training treatment.
C. Conservatively correction with bandage, operating treatment after 5 years.
D. To not undertake anything.
E. Ablation with the subsequent prosthetic repair.

87. The diagnosis of dysplastic dislocation of the femur was set in maternity hospital when examining newborns. What of the following signs is authentic for making the diagnosis?
A. +Relative linear shortening of a limb.
B. Asymmetry of dermal folds.
C. Increase of outside rotation of a limb.
D. Sign of "click".
E. Limitation of driving back of hip.

88. A child, 3 years old, was applied to polyclinic with complaints on lameness. There are linear shortening of the left lower limb, limited driving back in the left hip joint, positive Trendellenburg's sign on the left, detected on examination. What disease can cause these signs?
A. +Acute hematogenous osteomyelitis.
B. Polyarthritis.
C. Dislocation of hip.
D. Swelling of bone.
E. Perthes disease.

89. The diagnosis of dysplastic dislocation of the left-hand femur is revealed in a 3-month-old child for the first time. What treatment plan would you advise?
A. +Extension in a vertical plane.
B. Massage, physical training treatment.
C. Operative measure.
D. Removal of dislocation on the assigning trunk (stapeses).
E. Removal of dislocation, plaster bandage.

90. Asymmetry of dermal folds, limitation of driving back in the right hip joint were detected during preventive examination of a 3-month-old child. The radiography of coxofemoral joints was made. What is the norm of Acetabular index for a child aged 3 months?
A. +10°.
B. 45°.
C. 60°.
D. 90°.
E. 25–30°.

91. A surgeon revealed asymmetry of dermal folds on preventive examination of a child, 3 months old. The X-ray is made. Acetabular angle is 26. There are no ossification cores of heads of femoral bones. When do ossification cores of heads of femoral bones appear in the norm?
A. +On the first month of life.
B. After one year.
C. From 3 about 5 months.
D. After 2 years.
E. During a puberty.

92. Dysplastic dislocation on the right was diagnosed in a 5-year-old child. Point out the authentic clinical sign at this defect of development.
A. +Pain.
B. Linear shortening of limb.
C. Sign of "nonvanishing" pulse and retraction in Scarp's triangle.
D. Lameness.
E. Limitation of driving back.

93. A child, 12 years old, is admitted to hospital with complaints on pain in the left knee joint, lameness, limitation of driving back in the left hip joint. What disease should you suspect?
A. +Congenital dislocation of a femur.
B. Perthes disease.
C. Osteomyelitis.
D. Synovitis of a knee joint.
E. Tuberculosis of a hip joint

94. Parents applied to polyclinic with complaints of lameness of a child, 4 years old. It's known form the anamnesis that the child started walking late, at the age of 1.5 years. On examination: he is developed according to the age; goose gait, Trendelenburg's sign is positive; driving back in the hip joints is limited on both sides; large condylus is 2 cm higher than Roser–Nelaton's lines place. In the X-ray of hip joints: there is reduction of neck-diaphyseal angles up to 85°. Your diagnosis is:

A. +Congenital dislocation of hip.
B. Children's cerebral paralysis.
C. Two-sided congenital Perthes disease.
D. Two-sided congenital coxa vara.
E. Spondyloepyphiseal dysplasia.

95. Two-sided dysplastic dislocation of a hip was diagnosed in a 5-year-old child. Disturbance of posture is detected on examination. What type of posture is specific to this disease?
A. +Round spine.
B. Flat spine.
C. Scoliotic spine.
D. Plano-concave spine.
E. Lordotic posture.

96. A child, 10 years old, came to polyclinic with the diagnosis of scoliosis of the 2nd degree. What sing is most characteristic for this diagnosis?
A. +Asymmetry of shoulders.
B. Asymmetry of blades.
C. Presence of a rib back hollow.
D. Kyphosis of thoracic part of spine.
E. Asymmetry of triangles of waist.

97. A boy, 6 years old, has got scoliosis of the spine. Deformation was noted at the age of 2 years. Progressing of the curvature has been marked for the last 2 years; a rib back hollow has appeared on the right. On the X-ray of a vertebral column: there is padding wedge-shaped hemivertebra between the VI–VII thoracic vertebras. What type of scoliosis does this deformation concern to?
A. +Myogenic scoliosis.
B. Neurological scoliosis.
C. Congenital scoliosis.
D. Idiopathic scoliosis.
E. Dysplastic scoliosis.

98. A child, 7 years old, is complaining about deformation of the thorax. It is known from the anamnesis that the thorax deformation has been marked since birth; there was a respiratory paradox at the age of about one year, and frequent catarrhal disease; and he avoids physical exertion. On examination: there is recess of the breastbone and ribs; the deformation is symmetrical, up to 2,5 cm deep. Your managements is:
A. +Operating treatment.
B. Conservative treatment.
C. Wearing bandage.
D. Massage, physical training treatment.
E. Observation.

99. A child, 6 years old, is on "D" registration connecting with funnel-shaped deformation of the thorax of the 1st degree. You should determine a treatment plan.
A. +Bandage-therapy.
B. physical training treatment, massage.
C. Operating treatment.
D. Further observation.
E. Kinesotherapy, sailing.

100. Symmetrical funnel-shaped deformation of the thorax of the II degree was detected on examination of a child, 7 years old. Point out the depth of deformation of the II degree.
A. +0–1 cm.
B. 1–2 cm.
C. 2–4 cm.
D. 4–5 cm.
E. It is more than 5 cm.

101. A child, 12 years old, is followed by in polyclinic because of "S"dysplastic — figurative thoraco-lumbar scoliosis of a vertebral column of the II degree. Point out the sings of scoliosis possible progressing.
A. +High scale of dysplastic.
B. Age.
C. Disturbance of exchange of a collagen.
D. Difference between angles of rachiocampsis lying and standing is more 10°.
E. All named tags.

102. The parents have noted, that their child, 8 years old, abnormally sits at the desk, making a hometask. An orthopaedist has determined the diagnosis of scoliotic posture. Point out the curvature of posture with scoliosis.
A. +Angle of a curvature 5–10 degree.
B. Presence of a muscle cylinder.
C. Does not fade in lying down.
D. Fades in position of lying.
E. Rotation of bodies of vertebrae.

103. A child, 6 years old, has been complaining on pain in the left calf for one month. Some moderate increase of all calf is determined on examination. The skin is of a normal colour. On the X-ray examination: there is cellular destruction of all the tibia bone, linear periostitis. What additional examinations are necessary to verify the diagnosis?
A. +Common blood test.
B. An aspiration biopsy.
C. Punction biopsy.
D. X-ray of lungs.
E. Punction of a breast bone.

104. A pathological fracture of the shoulder bone is diagnosed in its mean third in a child of 5 years old. On the X-ray examination: there is sharp thinning of the cortical layer with resorption of all parts of osteal pattern. What disease does the X-ray picture indicate?
A. +Cyst of a bone.
B. Ostoclastoma.
C. Osteosarcoma.
D. Acute hematogenous osteomyelitis.
E. Tuberculosis of a bone.

105. A child, 12 years old, is complaining about megalgias in the upper third of the right femur, lingered under influence of analgetics, for one month. On the X-ray examination: there is spindle-shaped hyperostosis with obliteration of the medullar chan-

nel. Changes are not detected in the blood test. What disease is it possible to consider?

A. +Osteosarcoma.
B. Osteoid osteoma.
C. Sarcoma Ewing's.
D. Osteoma of a bone.
E. Chronic osteomyelitis.

106. A child, 12 years old, has been complaining about pain in the lower third of the right femur for one month (mainly at night time). There are increase of soft tissues volume, the local temperature rise. The osteosarcoma is suspected. Which of the following X-ray signs will be decisive in diagnostic?

A. +Destruction of osteal tissue.
B. Sclerosis of a bone.
C. Needle-like periostitis.
D. Osteoporosis.
E. Eccentric subperiosteal defect.

107. A child, 7 years old, is followed by in polyclinic with osteoarticular exostoses of the lower third of the right femur and the upper third of the right calf. What type of medical managements should be recommended to the patient?

A. +Operating treatment.
B. Chemotherapy.
C. X-ray-therapy.
D. Observation.
E. Hormonal therapy.

108. A child, 13 years old, has been complaining about pain in the upper third of the calf for 10 months mainly at nighttime. The calf is increased in a volume, painless on palpation. On the X-ray examination: there is spindle-shaped sclerosis, on the background of which an oval locus of enlightenment of 1×0,5 cm is seen. What disease is it possible to consider?

A. +Osteosarcoma.
B. Tuberculosis of a bone.
C. Chronic osteomyelitis.
D. Osteoid osteoma.
E. Sarcoma Ewing's.

109. A boy, 10 years old, has twisted a leg, and is complaining about pain in the region of the talocrural joint. The X-ray examination shows there is an oval focus with sharp thinning of the cortical wall in the lower third of the tibia bone. The diagnosis is osteal cyst. What complications can this pathology be accompanied by?

A. +Bleeding.
B. Pathological fracture.
C. Deformation of a bone.
D. Gap of a cyst.
E. Germination of cyst into epiphysis.

110. A child, 11 years old, has been complaining about pain in the field of the lower third of the femur within 3 weeks after trauma (falling from a height). There are limitation of functions in the knee joint, swelling of soft tissues of the lower third of the femur, the local temperature rise. The suggested diagnosis is a tumour of the lower third of the right femur, primary — chronic osteomyelitis. What method will be most informative for making a differential diagnostic?

A. +Tomography.
B. Aspiration biopsy.
C. Radiography.
D. Biochemical Methods.
E. Angiography.

111. Parents have applied with a complaint of absence of testicles in a child's scrotum. The child is 5 years old. The testicles were found in both sides in the region of the outside inguinal rings on palpation. That possibly led them in the scrotum easily. Your suggested diagnosis is:

A. +Bilateral inguinal lymphadenitis.
B. Bilateral cryptorchidism.
C. Bilateral false cryptorchidism.
D. Bilateral inguinal hernia.
E. Bilateral cryptorchidism. The hormonal therapy is prescribed.

112. A child, 6 years old, is delivered to a medical admission unit with complaints of severe pains in the right inguinal area, where a severe painful formation of the spherical form is palpated. Your suggested diagnosis is:

A. +Right-handed lymphadenitis in stage of abscess.
B. Acute oedema of the right testicle.
C. Acute funicular hydrocele.
D. Strangulated inguinal hernia.
E. Volvulus of the right testicle.

113. Absence of testicles in a scrotum is detected in a child, 4 years old, from the birth. They are not palpated in the inguinal channel. The genitals are developed for the man's type, though the scrotum is underdeveloped. Your management is?

A. +Overseeing by the child till 14 years.
B. Operational treatment after 2 years.
C. Conservative treatment (hormonal).
D. Operational Treatment at the age of 10–14 years.
E. Operational treatment at the age of 5–10 years.

114. Parents of a boy, 13 years old, have applied to you with complaints of increase of the right half of a scrotum, that appeared 2 months ago. The child is complaining of weakness, heightened fatigability. The testicles are palpated in the scrotum, but expanded veins of the funiculus spermaticus are palpated in the left half of the scrotum. When lying down the bloodstream decreases insignificantly. Your suggested diagnosis is:

A. +Oedema of right testicle membranes.
B. Adhesive oedema of left testicle membranes.
C. Varycocele at the left of Ist degree.
D. Varycocele at the left of IInd degree.
E. Inflammation of the left testicle (orchitis).

115. A girl, 3 months old, got operated with an inguinal hernia 2 hours ago. There wasn't nausea or

vomiting. The child is healthy somatically. Your management is?

A. +Home observation by the local pediatrist.

B. Hospitalisation in somatic department of hospital.

C. Hospitalisation to surgery hospital. Observation.

D. Hospitalisation to surgery hospital. Operating treatment.

E. Hospitalisation to surgery hospital, and after not successful therapy — operating treatment.

116. A boy, 2 years old, is suffering from measles in the severe form; a tight oedema of testicle membranes is observed. The pressure is increasing. Your management is?

A. +Antibiotic therapy.

B. Punction.

C. Scheduled operating treatment.

D. Urgent operating treatment.

E. Observation of the surgeon.

117. A 12-year-old child's penis is curved as a becket at the time of erection. The outside opening of the urethra opens on the prostate gland. Your diagnosis is:

A. +Extrohpyas of a urinary bubble.

B. Epispadias.

C. Hypospadias (head's form).

D. Hypospadias (corporal form).

E. Hypospadias (scrotal form).

118. Bathing her 4-year-old child, a mother found a very hard, painless tumorous formation of 12×8×5 cm by sizes in the abdominal cavity. There was enlargement of the left kidney. Its very thin parenchyma, its expanded collector system were determined in the ultrasound. Your suggested diagnosis is:

A. +Nephroptosis.

B. William's tumour.

C. Pyelonephritis.

D. Hydronephrosis.

E. Vesicoureteral reflux.

119. The most typical sign of intestinal polyps' presence is:

A. +Petition on abdominal pain.

B. Signs anaemia.

C. Intestinal bleeding.

D. Unstable stool.

E. Pain during defecation.

120. The most radical method of treatment of hemangiomas is:

A. +Cryotherapy.

B. Surgical.

C. Sclerosing.

D. Electrocoagulation.

E. Hormonetherapy.

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