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PART I

Pediatric surgery is rightfully called malformational surgery, because the majority of surgical diseases in childhood are congenital. Among these, abnormalities of the lungs, oesophagus, diaphragm, abdomen and urinary system, as well as the musculoskeletal system, are of greatest practical significance. The experience gained from our medical and educational work has dictated the necessity to write this chapter, which will combine most of pediatric surgical pathology — the surgery of deformities.

Global and Russian pediatric surgery (as a part of it), has achieved significant success in recent years, with improved diagnosis of disease, optimised preoperative preparation and anesthesia methods, and widespread use of high-tech techniques for treating most pediatric surgical conditions. However, a variety of clinical manifestations and emergence of new pathological entities lead to numerous diagnostic errors.

While working on this book, the authors aim to help foreign students at Russian medical universities to understand all the complexities of these issues.

Chapter 1
MALFORMATIONS OF THE NECK AND ORGANS OF THE THORACIC CAVITY

1.1. CONGENITAL CYSTS AND FISTULAE OF THE NECK

In pediatric practice, it is not uncommon to encounter conditions such as congenital cysts and neck fistulas. There are medial and lateral cervical cysts.

1.1.1. A median cervical cyst

It represents a remnant of the embryonic unobliterated thyroglossal duct. It is usually detected in preschool-aged children. The cyst is located on the anterior surface of the neck along the midline, visualised and palpated there as a rounded, even formation with a diameter of usually 1–2 cm. The median cyst is always connected by a fistula to the hyoid bone, so if a child is asked to tilt his head back and swallow saliva, the cyst will move up and down following the cartilages of the larynx. If the median cyst is diagnosed in the quiescent period (uncomplicated by suppuration), a planned operation is indicated: excision of the cyst with fistula and mandatory resection of the isthmus of the hyoid bone (otherwise, recurrence of the disease is possible). ***Staphylococcal aggression*** can lead to cyst suppuration. In this case (edema, hyperemia, soreness, hyperthermia), an opening and drainage of the abscess is indicated. Radical surgical treatment (cyst excision) should be postponed for 6 months.

1.1.2. Branchial cleft cyst

This represents a dysraphic anomaly due to non-closure of the branchial clefts or non-obliteration of the excretory duct of the thymus gland. The branchial cleft cyst is more often located on the side of the neck but it can also be located along the median line, and it is called "lateral" along the fistula. This fistula (usually thin, convoluted, long) connects the cyst to the oral cavity, opening either into the palatine tonsil or into the floor of the oral cavity at the root of the tongue. The branchial cleft cyst is palpated under the skin as an even rounded formation with a diameter of 1–2 cm. If a branchial cleft cyst of the neck is diagnosed in the cold period, a planned operation is indicated: excision of the cyst with fistula discharge. To avoid recurrence of the disease, it is important to stop the cyst communicating with the oral cavity, so if the fistula opens into the amygdala, the child is operated on together with an otolaryngologist — the surgeon excises the cyst and fistula from the neck, the otolaryngologist performs a tonsillec-

tomy. If a fistula opens at the root of the tongue, the surgeon should isolate it, explore it from the side of the oral cavity, turn the fistula into it, stitch, bandage, cross. With suppuration of the cyst, its opening and drainage are indicated. Radical surgical treatment, as in case of the median cyst of the neck, should be postponed for 6 months.

If median or lateral neck cyst communicates with environment, usually after suppurating and surgical treatment, then saliva discharges through the external opening, and the condition is termed a ***congenital cervical fistula***. Surgery for the latter is similar to that described above.

1.2. LUNG MALFORMATIONS

During its development, respiratory organs go through 5 stages.

1. ***In the embryonic period***, which begins in the third week of gestation, a ventral diverticulum forms from the lower part of the larynx-tracheal groove of the primitive intestine. From this, a primitive trachea forms, which divides into two primary lung buds in the fourth week.
2. From week 7–16, ***the false glandular stage*** occurs, during which 16–25 generations of airways form. At this stage, the lungs resemble glands and are composed of small tubes lined with epithelial cells and surrounded by connective tissue. Bronchioles form by the end of this phase.
3. From 16 weeks to 24, ***the canalicular stage*** lasts, leading to the formation of major functional lung structures.
4. From 24 week until birth, ***the terminal saccular stage*** occurs. This stage is characterised by continued morphogenesis and the transformation of sac lining cells into types I and II pneumocytes. However, the development of respiratory organs does not stop there.
5. ***The postnatal stage*** is marked by the formation of alveolar structures in the lungs. In general, this process is completed by age 8, and fully by age 25 (by which time the bronchial system has doubled and the alveoli have tripled).

Most congenital lung malformation are the result of disorders in their embryonic development during the 3rd to 6th week of gestation, and between the 6th and 16th weeks, that is, they are disorders in the first two stages of embryogenesis.

The first descriptions of these malformations were made in the 17th century and were mainly based on pathological anatomical studies. Vital diagnostics became possible with the advent of X-rays. According to the classification, these malformations can be divided into the following categories:

- malformations associated with preferential underdevelopment of the lung (agenesis, aplasia, hypoplasia, congenital lobar emphysema);
- malformations of the tracheobronchial tree (tracheobronchomegaly, Williams-Campbell syndrome, Kartagener's syndrome, bronchiectasis);
- malformations associated with the presence of excess dysembryogenetic masses (lung sequestration, lung cyst).

Among the causes of congenital lung deformations are:

- ***Exogenous***:
  1) physical (mechanical, thermal, radiation, electromagnetic);
  2) chemical (hypoxia, malnutrition, hormonal disorders, teratogenic toxins);
  3) biological (viral infection, exposure to bacterial toxins).

- ***Endogenous***:
  1) heredity;
  2) biological inferiority of gametes;
  3) age of parents.

1.2.1. Agenesis and aplasia of the lung and its lobes

Agenesis should be understood as the absence of both lungs at the same time, as well as the absence of their main bronchi. Aplasia refers to the absence of either one or both lungs in the presence of formed or rudimentary bronchi. Bilateral lung agenesis or aplasia are incompatible with survival. Lung agenesis and aplasia occur more commonly on the left side. The prognosis for survival in the absence of one lung is better than in case of the other. Mortality from this condition reaches 33 percent in the first year of life.

Clinical presentation may include respiratory failure in infants or chronic suppurative processes in older children. Infection in the rudimentary bronchus can lead to infection of the healthy lung, resulting in chronic bronchitis or bronchiectasis, worsening the patient's condition. Asymptomatic lung agenesis and aplasia may occur. External examination of patients may reveal deformity on one side of the chest due to decreased volume and narrowed intercostal space. There may be a marked lag in breathing on the affected side. Percussion may reveal displacement of mediastinal structures.

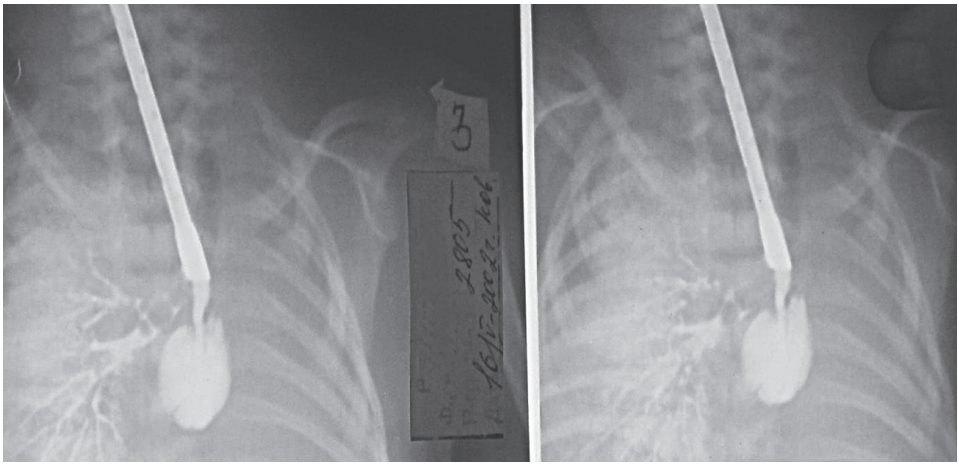

Fig. 1.1. Bronchography in aplasia of the left lung

Diagnosis is primarily radiologic, utilising chest X-rays, bronchoscopy, bronchography, angiography, and CT scans.

Treatment is predominantly conservative, beginning in the newborn period. Measures include removing sputum from the bronchial tree, breathing exercises, chest massage, and therapeutic physical therapy to strengthen chest muscles. Sputum is suctioned from the oral cavity. For children over three years old, mucolytic and expectorant agents are prescribed. Antibacterial therapy prevents infections. Surgical treatment is considered if bronchiectasis is present.

1.2.2. Hypoplasia of the lung

Hypoplasia of the lung refers to underdevelopment of all its structural elements. Congenital underdevelopment of vessels means blood circulation in the hypoplastic lung occurs through systemic circulation. Hypoplasia is more frequent on the left side and is somewhat more common in girls. A lung with diaphragmatic hernia is a common example.

Hypoplasia can be categorised into:

1. ***Simple hypoplasia at the stage of lobular bronchi formation.***
2. ***Simple hypoplasia at the segmental bronchi stage,*** known as "rudimentary lung."
3. ***Cystic hypoplasia,*** involving impaired development at the subsegmental bronchi level:
   - predominant underdevelopment of parenchyma with cystic changes in the bronchial tree;
   - predominant underdevelopment of the bronchial tree with cystic changes in lung tissue.

Cystic hypoplasia is also called ***congenital polycystic lung disease.***

The clinical presentation of simple hypoplasia varies depending on the severity of the lesion, as well as the presence or absence of inflammatory changes in malformed or neighboring organs. In some cases, there may be symptoms of respiratory failure, asymmetry of the chest and respiratory system, and displacement of mediastinum towards the affected lung. Recurrent inflammatory lung disease is often a sign that the child may have this condition. Weakened breathing, crackles, and changes in lung tissue transparency on X-rays can also be detected.

The clinical picture of cystic hypoplasia is not significantly different from that of simple hypoplasia. However, on X-ray, multiple thin walled air spaces (usually without liquid) can be seen in the region corresponding to the lesion. Accumulation and stagnation of bronchial secretions in these spaces, as well as infection, can lead to a purulent inflammatory process. Symptoms of intoxication and moist cough with sputum may appear in such children.

Treatment for hypoplasia involves surgery, as 1) hypoplastic lungs are practically unable to participate in gas exchange; 2) the risk of infection in underdeveloped lungs is high; and 3) healthy lungs may become infected with purulent infections. Surgical treatment involves removing the affected portion of the lung

1.2.3. Congenital solitary cyst

This defect is characterised by the presence of cystic formations located centrally in the root zone, or closer to the peripheral region of the lung. Other names for this condition include "bronchogenic cysts" and "bronchial cysts". Microscopic examination reveals elements of the bronchial wall in the cavity. The embryogenesis of these cysts is linked to the formation of additional hypoplastic lobes (segments, subsegments), which are separated from the trachea-bronchial tree completely or maintain communication with it.

Clinical manifestations vary. Uncomplicated cysts are asymptomatic in 15% of cases and are detected during chest X-rays. Inflammation of cysts develops in 60%, and the

clinical picture resembles that of a lung abscess, with cough, purulent sputum production, fever, and chest pain. Suppurative cysts have a more favorable course than abscesses, with less severe symptoms and slower progression. Radiographs show no perifocal infiltrates around the cysts, and a round or oval air mass with thin walls is visible.

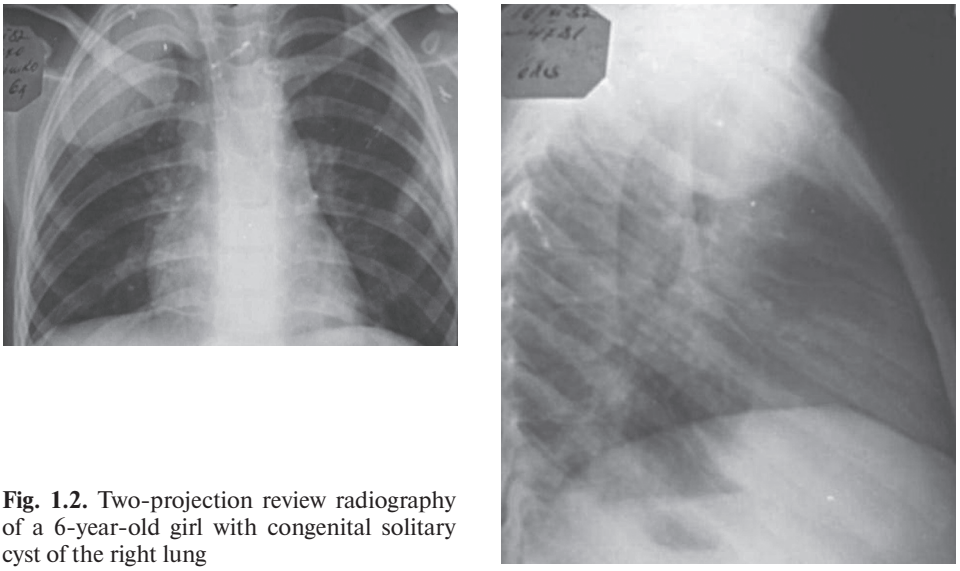

Fig. 1.2. Two-projection review radiography of a 6-year-old girl with congenital solitary cyst of the right lung

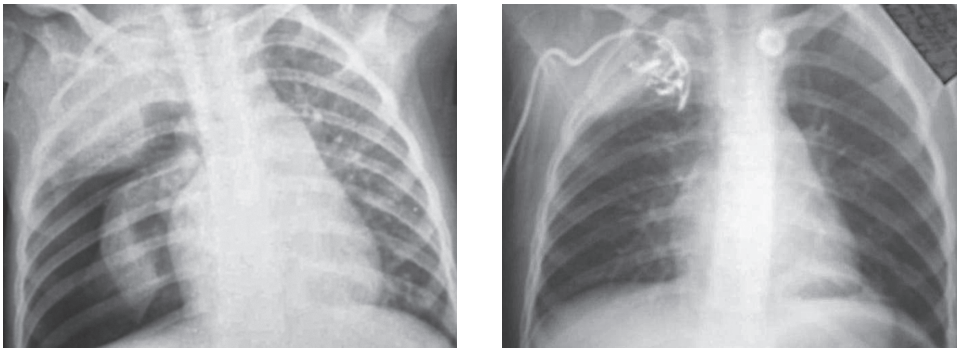

Fig. 1.3. Diagnostic pneumothorax and cyst contrast after Monaldi drainage in the same child

Tension within the cyst develops in approximately 20% of patients and cyst rupture into the pleural cavity, with the development of pneumothorax, occurs in 5%. In this case the clinic of intra-thoracic tension and respiratory failure develop.

Characteristic epidemiological history, positive serological reactions are grounds for ultrasound examination of the liver in patients suspected of pulmonary echinococcosis due to the frequent combined lesions in these organs. The difficulty of differential diagnosis between cysts and tumors, the lack of ability to predict the progression of the disease (enlargement of the cyst, suppuration, and rupture) determines the need for surgical treatment of solitary cystic lesions in a planned manner.

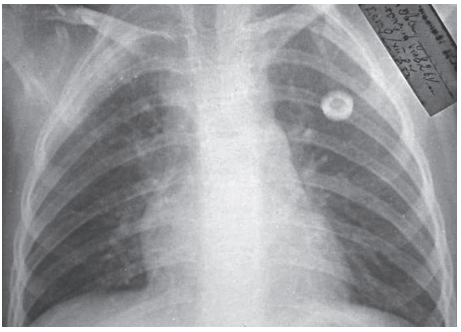

Fig. 1.4. Radiography after surgical treatment (lobectomy of the upper lobe of the right lung with a cyst) of the same child

Treatment. More frequently, uncomplicated cystic lesions are removed along with a portion of the lung tissue (lobectomy, segmental resection, atypical excision). Therapeutic interventions for complicated cystic lesions are classified as emergency. In cases of tense cystic lesions, drainage according to the Monaldi method is indicated. In the event of a developed pneumothorax, drainage of the pleural space according to the Bülow method is recommended.

1.2.4. Congenital lobar pulmonary emphysema

This malformation is characterised by the stretching (emphysema) of part of the lungs (more often, one lobe). This is the most common type of lung malformation among children. The exact causes of this malformation are not fully understood, but it has been found that its occurrence is linked to aplasia of cartilage elements in the bronchi and hypoplasia in elastic fibers and muscle elements in terminal and respiratory bronchi. These factors lead to the development of a valve mechanism that causes excessive ballooning in the affected part of the lung, resulting in less air being removed during exhalation compared to what is supplied during inspiration. The malformation tends to occur in ***the upper left lobe***, and the affected lobe becomes severely inflated and can reach enormous sizes. Other parts of the lungs become atelectatic and disconnected from respiration, while the mediastinal structures are displaced towards the unaffected side. In severe cases, the enlarged lobe can expand into the anterior mediastinum and move to the opposite side of the chest.

The main signs and symptoms of congenital lobular emphysema include progressive shortness of breath, difficulty breathing and widening of the intercostal space on the affected side. Coughing and cyanotic episodes may also occur. The severity of these symptoms depends on the degree of respiratory and circulatory insufficiency. There are three types of this condition.

Decompensated forms are the most serious threats to the lives of children. Newborns are admitted to hospitals in serious condition. Auscultation reveals that breathing is not heard on the affected side. Intercostal space widens, heart tones shift sharply to the opposite side, and there may be loss of consciousness or hypoxic seizures. In undetected cases, an asphyxic attack can lead to cardiac arrest or death.

Subcompensation occurs when symptoms appear in the early weeks of life, or near the end of newborn period. The main symptom is shortness of breath, and asphyxial attacks are not always noticed. Coughing may occur, and in some cases, the disease progresses to decompensation. Children are susceptible to frequent lung infections.

The compensated form is typical of infancy, with poor manifestations of the disease. The main symptoms are cough and short attacks of difficulty breathing. Symptoms of-

ten occur by the end of the first year. There is no progression of the disease as children grow older. Children often fall behind in physical development and sometimes develop secondary chest deformities.

The basis for diagnosing lobar pulmonary emphysema is a radiological examination. In CLE, lung field lucencies are determined, as well as impoverished pulmonary patterns. A "bloated" lung is shifted towards the opposite side and the dome of the diaphragm is flattened on the affected side, while compression of healthy lung parts (triangular shadow of atelectasis) is also detected.

Diagnostic errors can occur — it is possible that lobar emphysema may be mistaken for pneumothorax, leading to drainage of the pleural space. In such cases, air is initially released through the drain, but then the air release stops and no improvement is seen on the control X-ray. CT and radioisotopic studies of the lungs can be successfully used in the diagnosis of CLE.

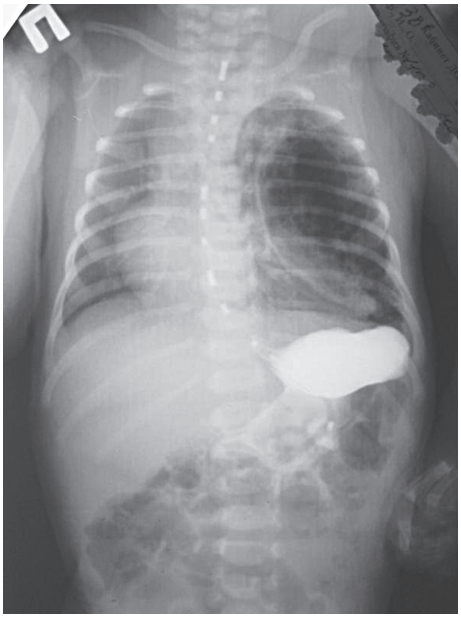

Fig. 1.5. X-ray of a newborn with congenital lobar emphysema of the upper lobe of the left lung

Surgical procedure is the only appropriate treatment for congenital lobar emphysema. The surgery involves the removal of a malformed lung lobe. Modern treatment technologies allow for lobectomy to be performed thoracoscopically, which is a minimally invasive procedure.

1.2.5. Pulmonary sequestration

Lung sequestration is a malformation that is characterised by the presence of a section of lung tissue that is located inside or outside of the normal lung. This section has its own bronchial system, which is supplied by an abnormal blood vessel that branches off from the aorta. The abnormal section of lung is isolated from the rest of the bronchial tree.

There are three types of sequestrated lung: intra-lobar, extrathoracic, and extrathoracically located. Intra-lobar sequestration occurs when the abnormal area is located within the lobe of the lung, while extrathoracic sequestration refers to an area that is surrounded by pleural tissue. Extrathoracally located sequestration can occur in the abdomen.

In children, ***sequestrated lungs can present*** with symptoms such as infection and inflammation of the affected area. If the sequestrum is not connected to the bronchial system, it may remain asymptomatic for a prolonged period of time and can only be detected through imaging studies. However, if there is communication between the sequested area and the bronchus, symptoms may include fever, cough, and chest pain.

Diagnosis of lung sequestration can be difficult. During the examination, a CT scan is done, which reveals cystic formations and may show shadows of abnormal blood vessels leaving the aorta. Bronchograms show the bronchial tubes wrapping around the cyst, and if they are connected to the bronchus, contrast can be seen flowing into cystic cavities.

Surgical treatment involves resection of the affected lung lobe.

1.3. MALFORMATIONS OF THE TRACHEOBRONCHIAL TREE

Mounier-Kuhn Syndrome ***is tracheobronchomegaly*** caused by the underdevelopment or degeneration of the cartilaginous, elastic and muscle structures in the TBT. This occurs infewer than one in 100,000 people, and its etiology remains unclear. It is characterised by ***a biphasic, irritating cough that resembles a goat's bleat***; recurrent tracheobronchial inflammation; purulent sputum; wheezes of various calibres due to accumulated chronic infection; and possible bronchiectasis formation. Additionally, respiratory insufficiency, low-grade fever, **growth retardation**, and **haemoptysis** may occur. Bronchoscopic examination reveals dilated tracheal lumens, possible diverticulae, posterior tracheal protrusion, and varying degrees of endobronchial involvement. Tracheobronchography reveals ***a "saw tooth" appearance*** (contour of the tracheal walls). Treatment is typically conservative, with surgical intervention rarely used due to its limited effectiveness.

1.3.1. Williams-Campbell Syndrome

Williams-Campbell Syndrome is a congenital malformation characterised by generalised underdevelopment of cartilage tissue in segmental and sub-segmental bronchi, from ***the third to the eighth order***, followed by secondary formation of bronchial ectasia. The defect is predominantly symmetrical, with a greater predilection for the lower lobes, and is caused by the morphological absence of or underdevelopment of cartilagenous rings in bronchial walls, leading to excessive airway dilatation during inspiration and collapse during expiration, causing impaired bronchial drainage, secondary inflammation, and eventually bronchiectasis and pneumosclerosis. The clinical course may remain latent for a period owing to compensated drainage function.

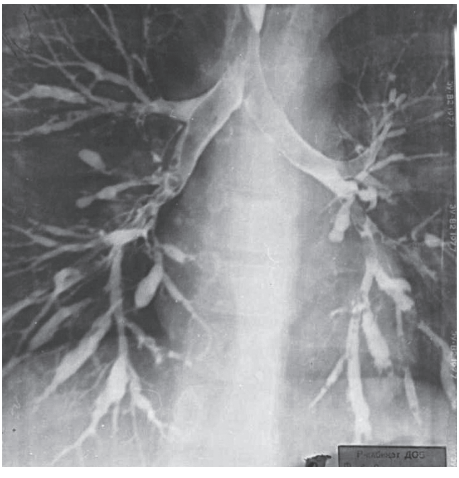

Fig. 1.6. Bronchography of a child with Williams-Campbell syndrome

The onset of disease often mimics severe pneumonia, but persistent "bronchial obstruction" and chronic intoxication signs (such as clubbing and watch-glass nails) suggest a congenital origin.

Diagnosis is made on the basis of bronchoscopy and bronchography, which show a characteristic pattern: **a normal bronchus, saccular bronchiectasis, and a normal bronchial tree.** Patients with generalised forms are treated conservatively, often in hospital settings, following the doctor's instructions for sanitation and anti-inflammatory therapy. Localised forms may require surgical intervention, involving resection of the affected lung tissue (lobectomy, bilobectomy).

1.3.2. Siewert–Kartagener s syndrome

The malformation is a combination of: 1) visceral situs inversa, 2) bronchial ectasia, 3), rhinosinusitis. It occurs in approximately one in 50,000 individuals. The syndrome is a clinical form of primary ciliary dyskinesia ***("immobile cilia" syndrome***). This is a defect at the cellular level. Immobile cilia are unable to generate the oscillatory movement necessary to propel the mucous component of bronchial secretions, so the mucus cannot "swim" along with dust particles and allergens and microorganisms that have entered it to the central parts of TBT. The mucus stagnates and the defect causes rhinosinusitis, otitis, bronchitis and bronchiectasis, tracheitis, etc.

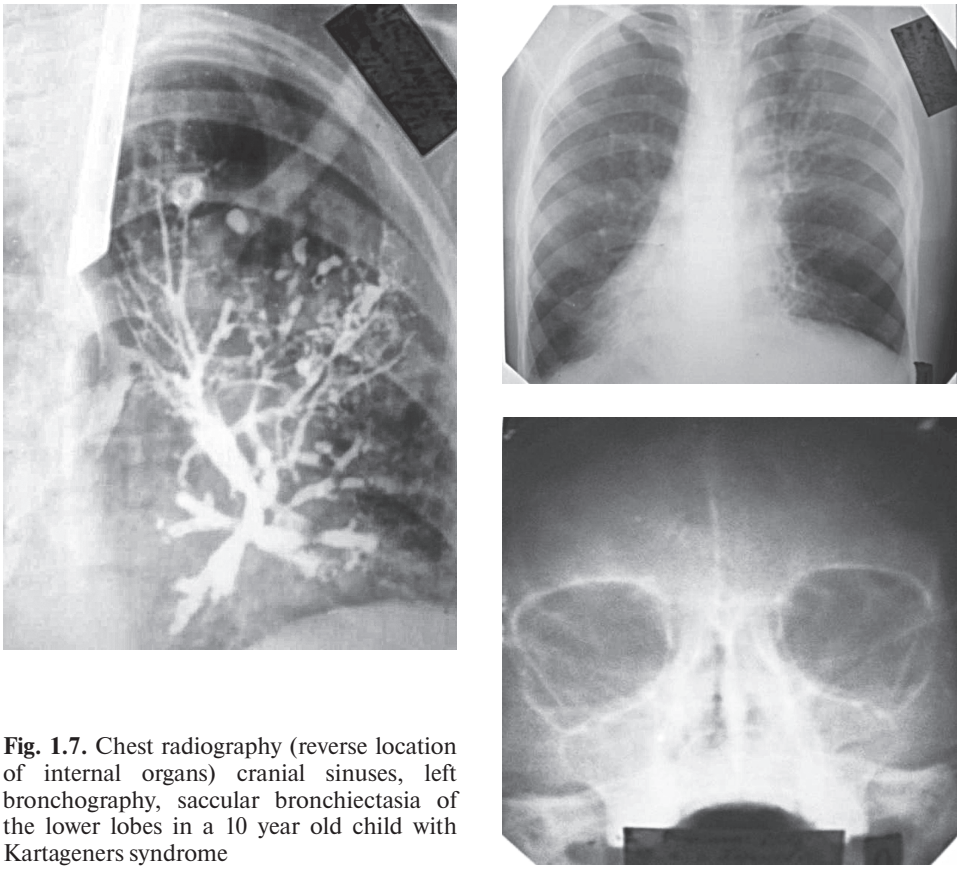

Fig. 1.7. Chest radiography (reverse location of internal organs) cranial sinuses, left bronchography, saccular bronchiectasia of the lower lobes in a 10 year old child with Kartageners syndrome