

CONTENTS

Participants of the publication	11
Abbreviations	13

PART I

Chapter 1. Malformations of the neck and organs of the thoracic cavity	17
1.1. Congenital cysts and fistulae of the neck	17
1.1.1. A median cervical cyst	17
1.1.2. Branchial cleft cyst	17
1.2. Lung malformations	18
1.2.1. Agenesis and aplasia of the lung and its lobes	19
1.2.2. Hypoplasia of the lung	20
1.2.3. Congenital solitary cyst	20
1.2.4. Congenital lobar pulmonary emphysema	22
1.2.5. Pulmonary sequestration	23
1.3. Malformations of the tracheobronchial tree	24
1.3.1. Williams-Campbell Syndrome	24
1.3.2. Siewert–Kartagener’s syndrome	25
1.3.3. Bronchiectasis	26
1.4. Oesophageal atresia	28
1.4.1. Chalazia cardia and GERD	31
1.5. Achalasia and congenital oesophageal narrowing	33
1.5.1. Achalasia of the oesophagus	33
1.5.2. Congenital oesophageal stenoses	34
1.6. Congenital diaphragmatic hernias	35
Chapter 2. Malformations of the anterior abdominal wall and abdominal organs	41
2.1. Malformations of the anterior abdominal wall	42
2.1.1. Gastroschisis	43
2.1.2. Umbilical hernia	44
2.1.3. Hernia of the linea alba	45
2.2. Congenital intestinal obstruction	45
2.2.1. Low intestinal obstruction	47
2.3. Congenital pyloric stenosis	49
2.4. Omphalomesenteric duct anomalies	51
2.4.1. Incomplete omphalomesenteric fistula	51
2.4.2. Complete omphalomesenteric fistula	51
2.4.3. Cyst of the omphalomesenteric duct	52
2.4.4. Meckel’s diverticulum	52
2.5. Malformations of the colon	53
2.5.1. Hirschsprung’s disease	55
2.5.2. Malrotations of the third period	60
2.6. Anorectal malformations	63
2.6.1. Epithelial coccygeal passage	67

2.7. Biliary tract malformations	67
2.7.1. Choledochal cyst	68
2.8. Anomalies of the pancreas and spleen	70
2.8.1. Pancreatic malformations	70
2.8.2. Defects of the spleen	72
Chapter 3. Malformations of the musculoskeletal system	74
3.1. Congenital pathology of hip joints	74
3.1.1. Classification of hip joint underdevelopment	74
3.1.2. Etiology and pathogenesis of the disease	75
3.1.3. Clinical presentation and diagnosis at different ages	75
3.1.3.1. Clinical presentation and diagnosis of pathology in newborns and children of the first three months of life	75
3.1.3.2. Clinical presentation and diagnosis in children aged 3–12 months	76
3.1.3.3. Clinical presentation and diagnosis of pathology in children older than one year.	77
3.1.4. Treatment of congenital pathology of hip joints.	79
3.1.4.1. Treatment of children under three months of age	80
3.1.4.2. Treatment of children older than three months.	80
3.2. Malformations of limbs	81
3.2.1. Reduction defects of the limbs	81
3.2.2. Syndactyly	82
3.2.3. Polydactyly.	83
3.2.4. Clubfoot	84
3.2.5. Clubhand	85
3.2.6. Flat feet (platypodia)	87
3.3. Deformities of the chest, neck and spine	88
3.3.1. Congenital deformities of the chest.	88
3.3.1.1. Pectus Excavatum	89
3.3.2. Congenital muscular torticollis.	92
3.3.3. Spinal curvatures	94
3.3.3.1. Kyphosis.	96
3.3.3.2. Lordosis	97
3.3.3.3. Scoliosis	98
3.4. Osteochondropathies.	101
3.4.1. Perthes' disease.	104
3.4.2. Osgood-Schlatter disease.	105
Chapter 4. Malformations of the urinary system	106
4.1. Urogenesis and the basis of its disorders.	106
4.2. Classification of urinary system disorders.	109
4.3. Kidney malformations.	110
4.3.1. Quantity anomalies	110
4.3.2. Position anomalies.	111
4.3.3. Anomalies of size value	113
4.3.4. Relationship anomalies.	114
4.3.5. Structural anomalies	115

4.4. Malformations of the ureteral embryo	117
4.4.1. Hydronephrosis	117
4.4.2. Vesicoureteral reflux (VUR)	119
4.4.3. Obstructive megaureter	121
4.5. Malformations of the bladder, urethra and foreskin	122
4.5.1. Bladder exstrophy	122
4.5.2. Epispadia	123
4.5.3. Hypospasia	124
4.5.4. Congenital infravesical obstruction syndrome	124
4.5.5. Phimosis	125
4.6. Disorders of testicular position	125
4.7. Malformations of the urinary duct	127
4.8. Varicocele	128
4.9. Urolithiasis in children	131
Chapter 5. Features of childhood oncology fetal tumors	138
5.1. Features of childhood oncology	138
5.2. Fetal tumors	140
5.2.1. Wilms' tumor (WT)	141
5.2.2. Neuroblastoma	143
5.2.3. Germ cell tumors	147

PART II

Chapter 6. Specificity of surgical infection in children	153
Chapter 7. Purulent-inflammatory diseases of skin and soft tissues.	155
7.1. The furuncle (boil). The carbuncle	155
7.2. Lymphadenitis	156
7.3. Panaritium	157
7.4. Mastitis	157
7.5. Erysipelas	158
7.6. Paraproctitis	159
7.7. Balanoposthitis	159
7.8. Omphalitis	159
7.9. Hydradenitis	161
7.10. Bartholinitis	161
7.11. Purulent wound and wound process	162
7.12. Necrotic phlegmon of newborns	164
Chapter 8. Hematogenous osteomyelitis.	165
8.1. Acute hematogenous osteomyelitis	165
8.1.1. Etiology and pathogenesis	165
8.1.2. Clinic and diagnostics	166
8.1.3. Additional diagnostic methods	168
8.1.4. Differential diagnosis	169
8.1.5. Principles of treatment	170
8.2. Epiphyseal osteomyelitis	173

8.3. Chronic hematogenous osteomyelitis	175
8.3.1. Secondary chronic osteomyelitis.	175
8.3.2. Primary chronic forms of osteomyelitis	177
Chapter 9. Purulent-inflammatory diseases of the abdomen	179
9.1. Acute appendicitis	179
9.2. Inflammatory complications of acute appendicitis	184
9.2.1. Appendicular infiltration	184
9.2.2. Abdominal abscesses	184
9.2.3. Appendicular peritonitis	185
Chapter 10. Purulent-inflammatory diseases of the thoracic cavity	186
10.1. Destructive pneumonia	186
10.1.1. BLD classification	186
10.1.2. Etiology and pathogenesis of BLD	187
10.1.3. Clinic, diagnosis, principles of treatment of various pneumonia complications forms	188
10.1.3.1. Exudative pleurisy and pyothorax.	188
10.1.3.2. Pyopneumothorax.	190
10.1.3.3. Lung abscesses	191
10.1.3.4. Pyopneumomediastinum	192
Chapter 11. Purulent-inflammatory diseases of the urinary system.	194
11.1. Pyelonephritis	194
11.1.1. “Purulent kidney”	196
11.2. Cystitis	197
Chapter 12. Surgical sepsis in children	199
12.1. Diagnosis of sepsis in children.	199
12.2. Diagnosis of septic shock in children	202
12.2.1. Multiple organ failure	203
12.3. Laboratory diagnosis of sepsis in children	204
12.4. Treatment of sepsis in children	204
Chapter 13. Rational antibiotic therapy	207
13.1. Specific features of surgical infection pathogens	207
13.2. Bactericidal antibiotics	209
13.3. Bacteriostatic antibiotics	210
13.4. Tactics of antibacterial therapy	211

PART III

Chapter 14. Acquired intestinal obstruction	215
14.1. Adhesive obstruction	215
14.1.1. Adhesive disease.	218
14.2. Intestinal invagination.	221
14.3. Obstructive obstruction	223
14.4. Functional obstruction	225

Chapter 15. Gastrointestinal bleeding. Portal hypertension	227
15.1. Features of gastrointestinal bleeding in children	227
15.2. Portal hypertension	231
15.3. Peptic ulcer of the stomach and duodenum	234
15.4. Polyps and intestinal polyposis	235
15.4.1. Single polyps of the colon	236
15.4.2. Multiple polyps of the colon	236
15.4.3. Diffuse colon polyposis	237
15.5. Anal fissures and haemorrhoids.	239
Chapter 16. Necrotizing enterocolitis	241
16.1. Issues of etiology and pathogenesis.	241
16.2. Clinical presentation and diagnostics	242
16.3. Modern principles of treatment	246
Chapter 17. Cholecystitis. Nonspecific ulcerative colitis. Crohn's disease	249
17.1. Acute cholecystitis.	249
17.2. Chronic calculous cholecystitis.	251
17.3. Nonspecific ulcerative colitis	254
17.4. Crohn's disease.	255
Chapter 18. Emergency urology-andrology	258
18.1. Renal colic	258
18.2. Acute urinary retention	259
18.3. Acute scrotum syndrome.	260
18.4. Complicated inguinal hernias	263
18.5. Acute renal failure	264
Chapter 19. Chylothorax. Chyloperitoneum	267
19.1. The problem and possible causes of conditions	267
19.2. Clinical presentation, diagnosis, principles of treatment.	267

PART IV

Chapter 20. Skeletal injury in children	271
20.1. Features of injuries to the musculoskeletal system in children	271
20.2. Injuries to the upper limb girdle	276
20.2.1. Broken collarbone	276
20.2.2. Fracture of the humerus	277
20.2.3. Fractures of forearm bones	282
20.2.4. Fractures of hand bones.	285
20.2.5. Dislocations of the bones of the upper limb	286
20.3. Lower limb injuries	288
20.3.1. Fractures of the femur	288
20.3.2. Knee joint injuries	291
20.3.3. Tibial fractures	293
20.3.4. Fractures of the foot bones	294
20.4. Fractures of the pelvis and spine	295
20.4.1. Fractures of the pelvic bones.	295
20.4.2. Spinal fractures	298

Chapter 21. Traumatic brain injury in children	300
21.1. Types and periodisation of TBI.	300
21.2. Classification of TBI in children (A.A. Artaryan, L.B. Likhterman in modification)	301
21.3. Clinical forms of TBI.	301
21.4. Fractures of the arch and base of the skull	304
21.5. Diagnosis of traumatic brain injury.	306
21.6. Treatment of traumatic brain injury	307
21.7. “Shaken baby” syndrome (SBS)	316
21.8. Cephalohematoma.	319
Chapter 22. Abdominal injury in children	320
22.1. Abdominal injury	320
22.1.1. Injury to the anterior abdominal wall	320
22.1.2. Injury to internal organs	320
22.1.3. Injury to the spleen	322
22.1.4. Liver damage	324
22.1.5. Damage to the pancreas	325
22.1.6. Stomach injury.	326
22.1.7. Intestinal injury	326
22.1.8. Injury to the colon	327
22.1.9. Rectal injury.	328
22.2. Chest injury	329
22.2.1. Damage to the chest wall.	330
22.2.2. Lung contusion	330
22.2.3. Intrapleural injuries	330
22.2.4. Traumatic asphyxia	331
22.2.5. Damage to the diaphragm	332
22.3. Injury to retroperitoneal organs.	332
22.3.1. Blunt trauma to the kidney	333
22.3.2. Injury to the ureter.	336
22.3.3. Damage to the bladder.	336
22.3.4. Urethra injury.	337
Control and training block.	339
Test tasks to part I	339
The standards of answers to the test tasks of part I.	379
Test tasks to part II.	381
Standards of correct answers to part II	393
Test tasks to part III.	393
Standards of correct answers to part III	404
Test tasks to part IV	404
Standards of correct answers to part IV	418
Conclusion	419
Recommended reading	420
Index.	422

PARTICIPANTS OF THE PUBLICATION

Authoring Team

Mikhail Alexandrovich Akselrov — Head of the Department of Pediatric Surgery at Tyumen State Medical University, Doctor of Medical Sciences, pediatric surgeon of the highest qualification category, winner of the All-Russian competition “Best Doctor of the Year” in the nomination “Best surgeon”, Honored Doctor of the Russian Federation, Associate Professor

Igor Nikolaevich Gerasimenko — Associate Professor of the Department of Pediatric Surgery with a course in Advanced Medical Education at Stavropol State Medical University, Doctor of Medical Sciences, pediatric surgeon of the highest qualification category, Associate Professor

Leonid Vadimovich Kolotilov — Professor of the Department of Anesthesiology and Intensive Care Medicine at St. Petersburg State Pediatric Medical University, Doctor of Medical Sciences, anesthesiologist-intensive care specialist of the highest qualification category, Professor

Victoria Anatolyevna Makhneva — Associate Professor of the Department of Pediatric Surgery at Kirov State Medical University, Ph.D. in Medicine, pediatric surgeon of the highest qualification category, Associate Professor

Sergey Viktorovich Minaev — Head of the Department of Pediatric Surgery with a course in Advanced Medical Education at Stavropol State Medical University, Doctor of Medical Sciences, pediatric surgeon of the highest qualification category, excellent healthcare worker of the Russian Federation, Full member of the Russian Academy of Medical Sciences and the European Association of Pediatric Surgeons (EUPSA), Member of the Board of UEMS, Professor

Maxim Petrovich Razin — Head of the Department of Pediatric Surgery at Kirov State Medical University, Doctor of Medical Sciences, pediatric surgeon of the highest qualification category, twice winner of the Docendo discimus Award for the best educational publication for Russian students, winner of the Kirov Region Prize in healthcare, Professor

Alexander Yurievich Razumovsky — Head of the Department of Pediatric Surgery at the Pirogov Russian National Research Medical University, Doctor of Medical Sciences, pediatric surgeon of the highest qualification category, chief freelance specialist-expert pediatric surgeon of the Moscow Department of Health, multiple winner of the Moscow Government Award in the field of healthcare, President of the Russian Association of Pediatric Surgeons, full member of EUPSA and UEMS, Professor, Corresponding member. RAS

Valentin Alexandrovich Skobelev — Associate Professor of the Department of Pediatric Surgery at Kirov State Medical University, Ph.D. in Medicine, pediatric surgeon of the highest qualification category, Associate Professor, winner of the Kirov Region Prize in Healthcare, excellent healthcare worker of the Russian Federation

Nikolai Konstantinovich Sukhikh — Associate Professor of the Department of Pediatric Surgery at Kirov State Medical University, Ph.D. in Medicine, pediatric sur-

geon of the highest qualification category, excellent healthcare worker of the Russian Federation, Associate Professor

Sergey Ivanovich Timofeev — Assistant of the Department of Pediatric Surgery at Khabarovsk State Medical University, Ph.D. in Medicine, pediatric surgeon of the highest qualification category

Ivan Alexandrovich Turabov — Head of the Department of Pediatric Surgery at Northern State Medical University, Doctor of Medical Sciences, pediatric oncologist of the highest qualification category, Honored Doctor of the Russian Federation, Professor

Reviewers

Yuri Yurievich Sokolov, Doctor of Medical Sciences, Professor, Head of the Department of Pediatric Surgery at the Federal State Budgetary Educational Institution of Additional Professional Education “Russian Medical Academy of Continuing Professional Education” of the Ministry of Health of the Russian Federation

Andrey Valerievich Pisklakov, Doctor of Medical Sciences, Professor, Head of the Department of Pediatric Surgery, Reproductive Medicine of Children of the Federal State Budgetary Educational Institution of Higher Education “Omsk State Medical University” of the Ministry of Health of the Russian Federation

Nikolay Sergeevich Grachev — General Director of the Federal State Budgetary Institution Dmitry Rogachev National Medical Research Center of Pediatric Haematology, Oncology and Immunology of the Ministry of Health of the Russian Federation, Doctor of Medical Sciences, Professor

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 musculo-skeletal 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.

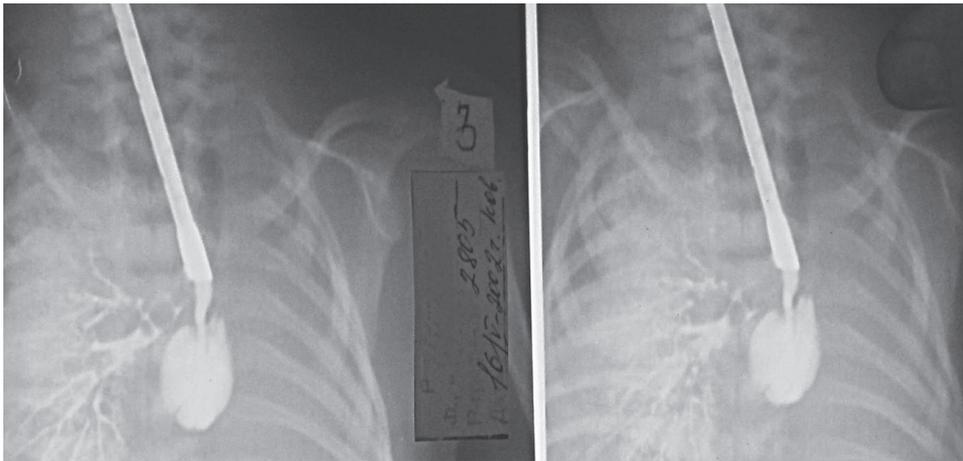


Fig. 1.1. Bronchography in aplasia of the left lung

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.

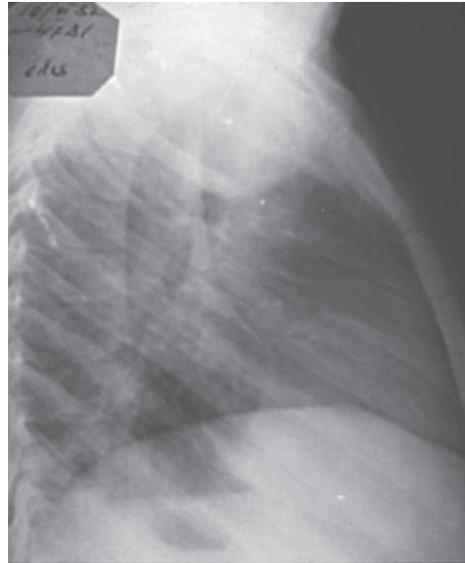
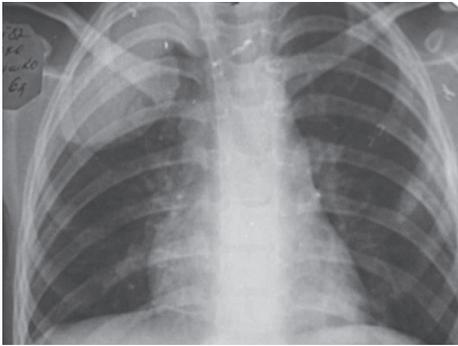


Fig. 1.2. Two-projection review radiography of a 6-year-old girl with congenital solitary cyst of the right lung

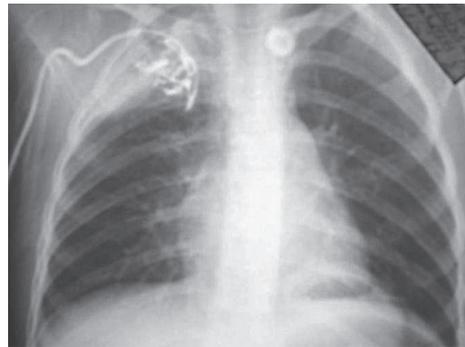
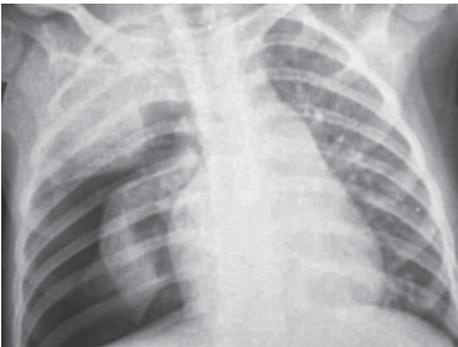


Fig. 1.3. Diagnostic pneumothorax and cyst contrast after Monaldi drainage in the same child

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.

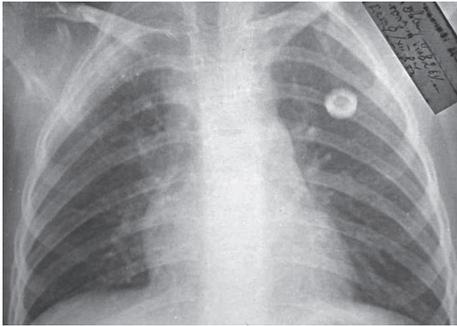


Fig. 1.4. Radiography after surgical treatment (lobectomy of the upper lobe of the right lung with a cyst) of the same child

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.

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.

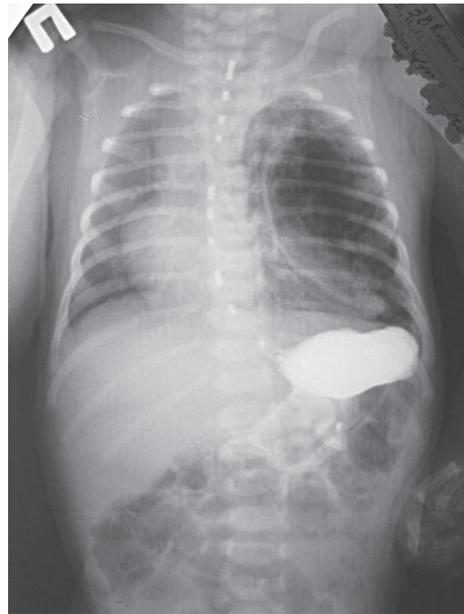


Fig. 1.5. X-ray of a newborn with congenital lobar emphysema of the upper lobe of the left lung

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 sequestered lung: intra-lobar, extrathoracic, and extra-thoracically 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. Extrathoracically located sequestration can occur in the abdomen.

In children, *sequestered 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 sequestered 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

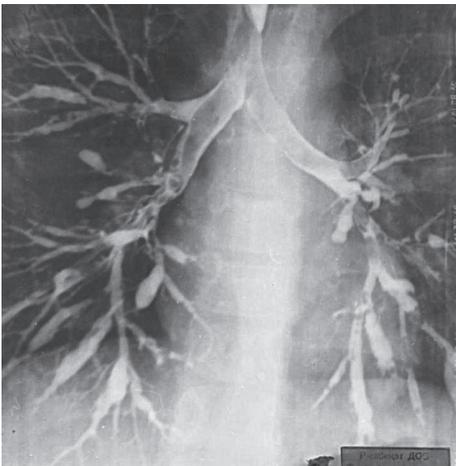


Fig. 1.6. Bronchography of a child with Williams-Campbell syndrome

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.

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.

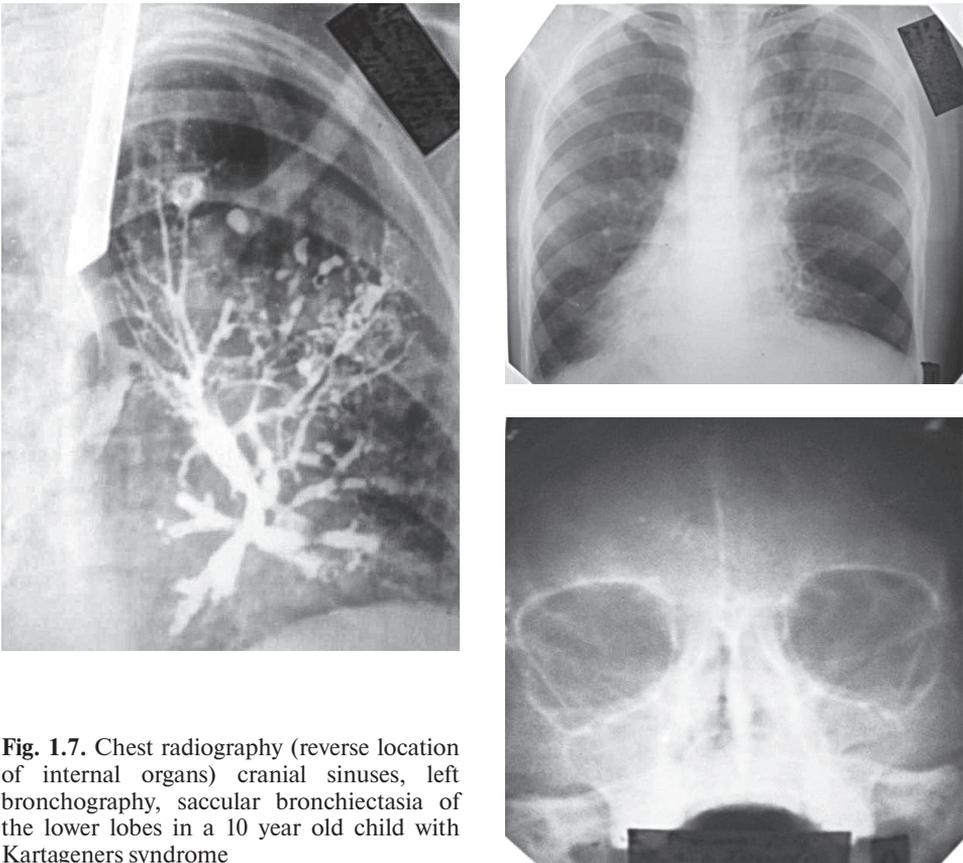


Fig. 1.7. Chest radiography (reverse location of internal organs) cranial sinuses, left bronchography, saccular bronchiectasia of the lower lobes in a 10 year old child with Kartageners syndrome