MINISTRY OF HEALTH OF UKRAINE

ODESA NATIONAL MEDICAL UNIVERSITY

Departments of Pediatrics №2

CONFIRMED by

Vice-rector for research and educational work

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METHODOLOGICAL RECOMMENDATIONS ON PRACTICAL CLASSES FOR STUDENTS

International Medical Faculty, course 6

Educational discipline "**PEDIATRICS**"

Approved
at the meeting of the department of Pediatrics №2
Protocol No. 11 dated 28/08/2022

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1. **Topic No. 13**

   Differential diagnosis of hereditary and congenital diseases of the bronchopulmonary system in children. Leading clinical symptoms and syndromes in hereditary and congenital diseases of the bronchopulmonary system (cystic fibrosis, idiopathic pulmonary hemosiderosis, primary ciliary dyskinesia, Wilms-Campbell syndrome, bronchomalacia, aplasia and hypoplasia and instrumental studies in hereditary and congenital diseases of the bronchopulmonary system and their complications. Patient management tactics in different clinical variants of the course of broncho-obstructive syndrome and its complications in children. Emergency care for asthmatic conditions. Helping children with bronchial obstruction syndrome according to the Integrated Management of Childhood Illness system. Prevention of bronchial asthma and bronchial obstruction syndrome in children of different ages. Medical supervision.

2. Relevance of the topic. Since childhood, chronic respiratory disease is the most common cause of deterioration in the quality of life of children and premature disability. Chronic nonspecific lung diseases occupy an important place in the pathology of children (they are diagnosed in 1% of children), while hereditary and congenital diseases occur in 10% of patients.

A chronic disease of the bronchopulmonary system should be understood as a hereditary or acquired disease of the lungs, accompanied by the development of pneumosclerosis in one or more segments of the lungs and varying degrees of deformation and chronic inflammation of the bronchi. The introduction of biochemical, immunological studies revealed genetically determined metabolic disorders (cystic fibrosis), various forms of immunodeficiency states, deficiency of -1-antitrypsin, etc. in a number of patients with chronic inflammatory process in the lungs. This made it possible to distinguish from the group of chronic lung diseases a number of nosologies in which the chronic inflammatory process in the lungs is secondary. Despite the fact that this group of diseases is infrequent, a heavy progression of the disease, the duration of the process, the possibility of disabling the patient, attach great importance to this topic. This determines the relevance of the topic and its importance for the therapist and paediatrician. Over the last two decades, the spectrum of chronic broncholegenic pathology has undergone significant changes, which have decisively affected the current structure. The high prevalence of bronchopulmonary diseases among the child population, the tendency towards a protracted, recurrent and chronic course also determine the relevance of the problem of prevention, rehabilitation and medical examination of children with respiratory pathology. These issues are of particular importance in connection with the deterioration of environmental conditions, contributing to the growth of chronic lung diseases. Therefore, the problem of differential diagnosis of hereditary, congenital and chronic diseases of the bronchopulmonary system in children is a highly urgent medical and social problem.

3. Lesson objectives:

3.1. *General goals:* to identify and evaluate syndromes in chronic diseases of the bronchopulmonary system on the basis of complaints, anamnesis and objective examination data, draw up a differential diagnostic algorithm, analyze the results of additional research methods in chronic diseases of the bronchopulmonary system, establish a preliminary clinical diagnosis of chronic diseases of the bronchopulmonary system in children, determine the tactics of patient treatment.

3.2. *Educational purposes:* to get acquainted with the recommendations of the World Health Organization, statistics of morbidity and mortality from diseases included in COPD; reduce

mortality and the frequency and severity of illness and disability, and contribute to improving the physical development of the child.

3.3. *Specific objectives:*

- *to know:*

1. The modern concept of COPD
2. Etiology and pathogenesis of COPD in children
3. Modern classification of chronic nonspecific lung diseases (COPD) in children

3.4. Based on theoretical knowledge of the topic:

- *master the techniques / be able to /:*

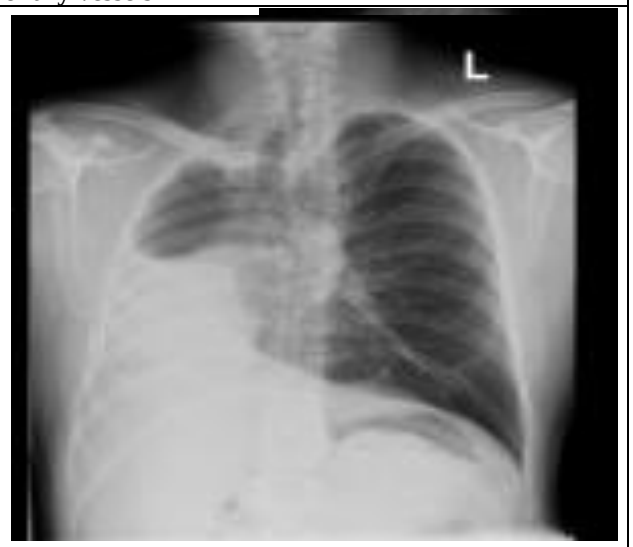
1. Recognize clinical manifestations, syndromes accompanying COPD in children
2. Diagnose and treat conditions and diseases included in COPD
3. Have the skills of communicating with the parents of a child with COPD
4. Differentiate the conditions and diseases included in COPD, even in the most difficult cases
5. Draw up a plan for the treatment and prevention of diseases related to COPD

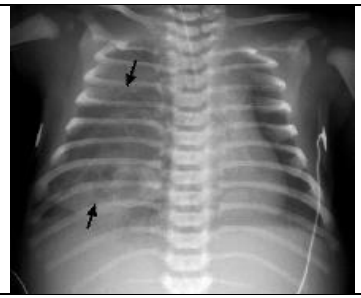
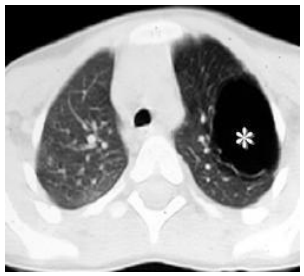
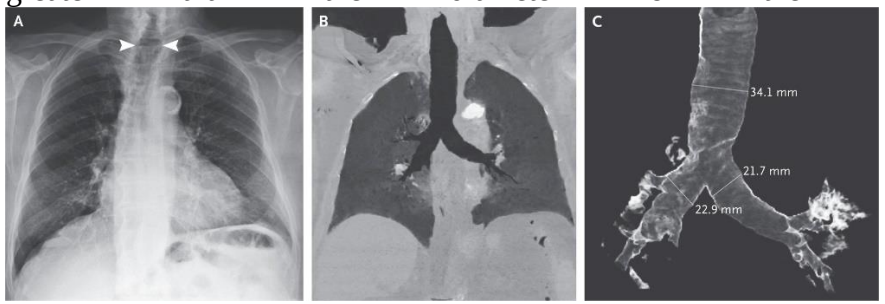
4. Materials for classroom self-preparation (interdisciplinary integration).

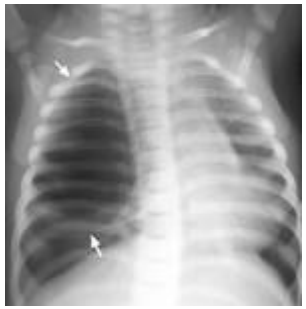
№	Disciplines	To know	To be able to do
1	Previous disciplines (normal physiology, propaedeutics of childhood diseases, faculty pediatrics, hospital pediatrics)	Respiratory mechanisms, anatomical and physiological features of the bronchopulmonary system in children, pathogenetic mechanisms of respiratory disorders in young children, clinical picture, modern methods of diagnosis and treatment of diseases of the pulmonary system in children.	Conduct differential diagnosis of diseases, and proceed with the syndrome of respiratory failure and prescribe differential treatment.
2	Internal subject integration (topics: "Differential diagnosis of pneumonia in children", "Differential diagnosis of broncho-obstructive syndrome in children")	Criteria for the diagnosis of inflammatory lung diseases in children of different ages.	Conduct a clinical examination of a child with an inflammatory disease of the bronchopulmonary system. To be able to draw up an algorithm for the differential diagnosis of a disease. Assess the

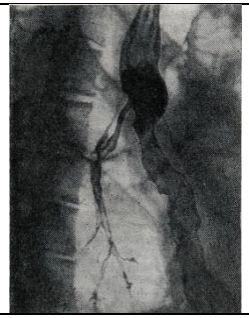
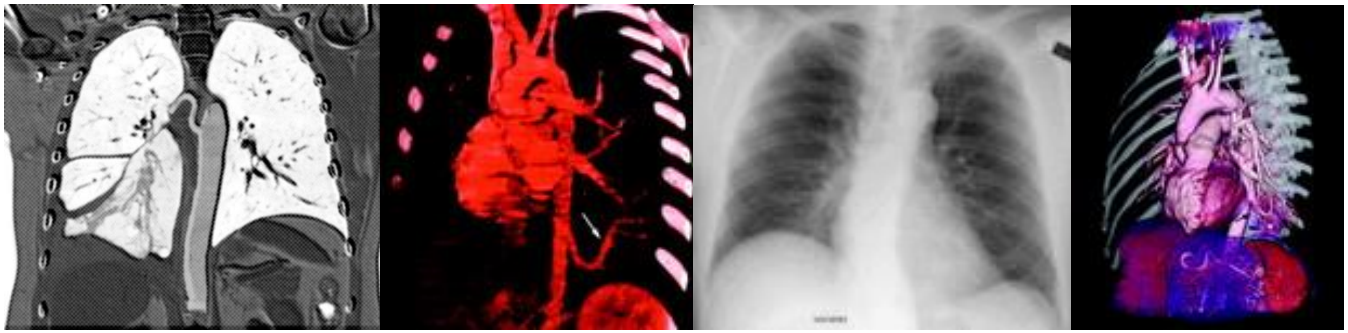
			severity of the sick child's condition.

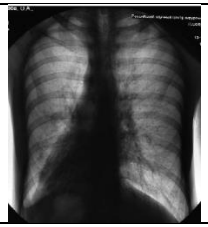
5. The content of the topic (text or theses) of the lesson.

<table>
<tr><th colspan="5">Classification and clinical characteristics of malformations of the bronchi, lungs and pulmonary vessels</th></tr>
<tr><td rowspan="3">Underdevelopment of bronchopulmonary structures</td><td colspan="2">Lung agenesis</td><td>Absence of a lung along with the main bronchus</td><td rowspan="2">Lagging in physical development. The chest on the side of the lesion is flattened, scoliosis with a bulge in the healthy side, deformation of the terminal phalanges of the fingers is formed.
Dullness of percussion sound, weakening of breathing, pronounced shift of the mediastinum to the sore side, shortness of breath.
On X-ray examination: narrowing of the pulmonary field, displacement of the shadow of the heart and high standing of the diaphragm, the contours of which can merge with intense darkening. Mediastinal pulmonary hernia is characteristic.
RF: decreased lung volumes, hypoxemia.
Bronchography: displacement of the trachea, which directly passes into the main bronchus of a healthy lung with agenesis or into the stump of the main bronchus on the affected side with aplasia of the lung.
  </td></tr>
<tr><td colspan="2">Lung aplasia</td><td>Lack of lung tissue in the presence of a rudimentary main bronchus</td></tr>
<tr><td>Lung hypoplasia</td><td>Simple</td><td>Simultaneous underdevelopment of the bronchi and pulmonary parenchyma</td><td>Uniform underdevelopment of the entire lung, lobe or segments with reduction of the bronchial tree (up to 5-10 generations instead of 18-24 in the norm).
The first signs of the disease in the form of acute pneumonia or bronchitis at 1-2 years of age with pronounced signs of DN and a protracted course. Repeated episodes of bronchitis or pneumonia 3-6 times a year. The presence of a moist persistent cough with purulent sputum, moist wheezing against a background of weakened breathing. Deformation of the chest, flattening over the affected area, scoliosis. Obstructive syndrome.
Radiological changes: deformation of the pulmonary pattern, decreased perfusion in the affected area, displacement of the mediastinum in this direction, compensatory emphysema of the segments of the affected lung.
Surgical treatment.</td></tr>
</table>

		Cystic		Underdevelopment of the lung with cystic degeneration of the respiratory department. The clinical manifestations of CG are similar to those in PH. Radiographs reveal multiple thin-walled cavities in the area of the lung, a decrease in volume. Surgical treatment.	 
Malformations of the wall of the trachea and bronchi.	**Распространенные пороки**				
	Tracheobronchomegaly (Munier-Kuhn syndrome)		Congenital excessive expansion of the trachea and large bronchi.	Violent cough of a vibrating character with purulent sputum, noisy "purring" breathing. Asthma attacks are regarded as bronchial asthma. Sufficient for suspicion of TBM in adults is the diameter of the trachea more than 25 mm, and the diameter of the right and left main bronchi is 23 and 20 mm, respectively. In children, the diagnosis of TBM is likely if the diameter of the trachea on a direct radiograph is equal to or greater than the diameter of the thoracic vertebrae.  M.A. Fadeyeva, L.V. Simonova.
	Tracheobronchomalacia		Increased mobility of the walls of the trachea and bronchi is narrowing of the lumen on exhalation and expansion during inhalation	Noisy breathing, obstructive changes, shortness of breath. Symptoms increase with the addition of acute respiratory infections, persist after recovery. The cartilaginous framework of the trachea and bronchi strengthens with age in young children, signs of tracheobronchomalacia disappear most often with the age of 1-2 years without treatment. Treatment: with pronounced signs of tracheal stenosis, intubation and mechanical ventilation, surgical treatment, endoprosthetics using stents are performed.
	Williams-Campbell syndrome		Underdevelopment of the cartilaginous rings of the bronchi of the 3-8 orders	More common is in boys in the first 3 years of life: pneumonia, bronchitis, prolonged, with obstructive syndrome. Moist persistent cough with purulent sputum, deformation of the chest, the terminal phalanges of the fingers are transformed. Delayed physical development. On auscultation: scattered moist, mid-mucus rales in both lungs and dry wheezing rales predominantly on expiration,

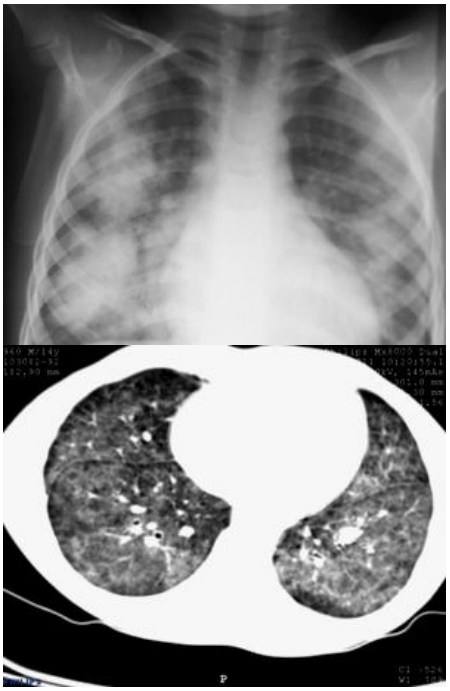
				lengthening of expiration. Violation of ventilation: obstructive type - a symptom of an air trap, with samples with forced breathing. Plot of plateau on the FEV curves. Quite often the spirographic manifestation of the "wavy exhalation" phenomenon. Hyperimmunoglobulinemia G, High level of secretory immunoglobulin A.
	Bronchomolation	Expansion of the bronchi Proximal and mixed type		Early onset, moist persistent cough with mucous or mucopurulent sputum. Frequent pneumonic and bronchitis episodes from 2-3 years of age quickly, the formation of a chronic process. Lag in physical development, chest deformity. Obstructive syndrome. Dysfunction of external respiration, obstructive or combined. Signs of an overload of the right heart
	Limited malformations of the walls of the trachea and bronchi			
	Congenital tracheal stenosis	organic	Primary changes in the tracheal wall	Expiratory stridor, which manifests itself immediately after the birth of the child. Noisy breathing, "wheezing", "crackling", "sawing", sometimes - persistent, treatment-resistant "spastic bronchitis" with a corresponding physical picture in the lungs. Surgical treatment.
			Secondary (compression)	
		functional		
	Congenital lobar emphysema	The valve mechanism of narrowing of the bronchus leads to hyperinflation of the corresponding section of the lung. Stenosis of the bronchus is more often associated with a defect in the cartilage of its wall		Acute attacks of respiratory failure, especially with ARVI, feeding and physical exertion. In older children: recurrent pneumonia and bronchitis. Swelling and lagging of the chest during breathing on the affected side, a section of the box sound during percussion and weakening of breathing. A characteristic shift of the mediastinum to the healthy side. X-ray signs: increased transparency of the affected area with depletion of the pulmonary pattern, widening of the intercostal spaces, displacement of the mediastinum to the healthy side. Surgical treatment 
	Diverticula of the trachea and bronchi	Single or multiple protrusions of the wall of the trachea and / or bronchi of any etiology, clinically they are not manifested		

	Tracheobronchoesophageal fistula		There are severe attacks of suffocation, coughing and cyanosis with short and wide fistulas, at the very first feeding of the child. Severe aspiration pneumonia, usually fatal. With narrow joints, recurrent bronchitis or pneumonia is observed. Increased discharge of mucus when coughing. 
Lung cysts	Air or liquid-filled cavities	Acquired Congenital	Changes in uncomplicated cysts are absent, there is a shortening of percussion sound, weakened breathing and a relatively small amount of wet wheezing with pneumonic exacerbations. In the period of remission, the condition of the patients is satisfactory, there are no symptoms of intoxication and a decrease in indicators of physical development.
Lung sequestration	Lack of connection of the lung area with the bronchial system + blood supply from the abnormal artery extending directly from the thoracic or abdominal aorta or its main branches.	Extralobar Intralobar	Clinical manifestations of deficiencies may be absent for a number of years to the development of infection in the sequestering site, proceeds with a clinical picture of pneumonia, often recurrent. Diagnosis of lung sequestration is difficult, since the clinical and radiological symptoms of other diseases and malformations (polycystic and bronchiectasis, solitary cyst and lung abscess, etc.) are very similar. Only the identification of an abnormal vessel, the shadow of which can sometimes be detected by computed tomography and in most cases by aortography, allows a diagnosis to be made before surgery. In the picture, sequestration looks like a more or less homogeneous darkening, in some cases with cystic changes. 
Kartagener's syndrome (ciliary dyskinesia)	Defect of the ciliary epithelium is the absence of dynein		Stagnation of secretions in the respiratory tract, infection and the formation of a chronic inflammatory process. The opposite arrangement of organs is bronchiectasis and chronic sinusitis. Dysfunction of the ciliary epithelium is combined with immovable sperm in men and dysfunction of the epithelium of the fallopian tubes in women.синусит.

	arms in the eyelashes of the ciliary epithelium	
Pulmonary arteriovenous malformation	Agenesis and hypoplasia of the pulmonary artery and its branches	
	Agenesis and hypoplasia of the pulmonary artery and its branches	
	Abnormal (transposition) confluence of the pulmonary veins	

Hereditary lung diseases:

- Idiopathic pulmonary hemosiderosis;
- Primary pulmonary hypertension;
- Alpha-one-antitrypsin deficiency (AAT)
- Cystic fibrosis.



Idiopathic hemosiderosis of the lungs is a rare disease, characterized by recurrent intraalveolar hemorrhages, manifested by hemoptysis of varying intensity, secondary iron deficiency anemia, has a wavy recurrent course, leads to widespread pneumofibrosis

The disease was first described in 1960 under the name "brown induration of the lungs."

With an exacerbation, a cough appears, the cough is accompanied by hemoptysis, the appearance of blood in sputum in older children. Small children do not cough up phlegm, they swallow it. With profuse pulmonary bleeding, they may vomit with an admixture of vomit with swallowed blood. The severity of the clinical picture from the lungs depends on the amount of blood poured into the alveoli. Together with cough and fever, shortness of breath and cyanosis appear. In the lungs, wheezing is heard. A feature of radiologically detected changes is the rapid reverse development of foci of darkening. In some cases, scattered small shadows in both lungs are noted on chest x-rays, which is the reason for the erroneous diagnosis of miliary pulmonary tuberculosis. Changes in the lungs detected by radiography can vary widely: from small infiltrates to massive shadows, accompanied by atelectasis, emphysema and a reaction from the lymph nodes of the roots of the lungs.

Treatment: the appointment of corticosteroid drugs and symptomatic treatment.

Some authors recommend excluding cow's milk from the diet before determining precipitins in the blood serum of patients.

Prednisolone is prescribed at the rate of 1-1.5 mg / kg until clinical and laboratory remission is achieved. There are reports that persistent long-term remission occurs after splenectomy.

Primary pulmonary hypertension is a rare disease of unknown etiology, characterized by a pronounced increase in total pulmonary vascular resistance (PVR) and pulmonary artery pressure, often a progressive course with a rapid development of right ventricular decompensation, and a fatal prognosis.

Synonyms: primary pulmonary hypertension, Aerz-Arilago syndrome, Aerz's disease, Escudero's disease.

Diagnostic criteria:

1. Mean pulmonary artery pressure (MPAP) > 25 mm Hg. at rest and > 30 mm Hg during physical exertion
2. Normal pressure of wedging in the pulmonary artery (PAWP) is up to 10-12 mm Hg.
3. Absence of possible causes of pulmonary hypertension (PH) - diseases of the heart, lungs, chronic pulmonary embolism, etc.

The clinical symptoms of IPH are nonspecific, which significantly complicates the early diagnosis of the disease (shortness of breath, chest pain, dizziness, cough, hemoptysis, palpitations and heart failure).

The PH diagnosis algorithm includes the following steps:
1. Suspicion of the presence of PH in the patient (preliminary diagnosis).
2. Verification of the diagnosis of PH (catheterization of the right heart and PA, ECG, VKG, PCG, Echo-KG)
3. Establishment of the clinical class of PH (determined by FVD, CT, MRI, APG)
4. Assessment of LH (type, functional class) - (blood tests (general, biochemical, immunological), HIV test: ultrasound of internal organs; 6-minute walk test / cardiopulmonary test; FC assessment; lung biopsy).
Treatment
1. General recommendations: exclusion of excessive stress on CVS and DS, timely vaccination, regular physical activity (aerobic exercise of low intensity, for example, walking at a moderate pace, if the patient tolerates them normally), a diet with salt restriction and control of fluid intake.
2. Drug therapy:
• Calcium channel blockers - nifedipine, diltiazem, amlodipine
• Prostanoids - epoprostenol, treprostinil, iloprost
• Endothelin receptor antagonists - bosentan
• Phosphodiesterase-5 inhibitors - sildenafil, tadalafil
• Diuretics - loop diuretics: furosemide 20–120 mg / day, ethacrynic acid 50–100 mg / day, torasemide 5–20 mg / day. It is advisable to add adosterone antagonists: veroshpiron 25–150 mg, eplerenone 20 mg. In all cases of prescribing diuretics, it is necessary to carefully monitor the levels of blood electrolytes, as well as the state of renal function.
• Oxygen therapy

2. Patients with PH are indicated for surgical treatment when, against the background of active pharmacotherapy (including drugs specific for the treatment of PH and inotropic drugs), the disease progresses and there is a poor prognosis. In such cases, bilateral lung transplantation is performed (if necessary, with simultaneous elimination of the heart defect) or the heart-lung complex.

Alpha-one-antitrypsin (AAT) deficiency is inherited in an autonomously recessive manner. Due to the low activity of AAT, protease circulating and settled in the lungs of granulocytes and monocytes (chemotrypsin, trypsin, elastase, neutral protease), they destroy lung tissue, which leads to thinning and rupture of the alveolar septa and further, to emphysema. More often, the defect manifests itself in adolescents and adults, but often in the first years of life in the form of pulmonary or combined pulmonary-hepatic pathology. Hepatic pathology is cholestatic jaundice, liver cirrhosis. Pulmonary pathology forms COPD. Patients are described in whom AAT deficiency is manifested by shortness of breath, emphysema without prior recurrent bronchitis. The diagnosis is established after preliminary determination of the level of alpha-one-antitrypsin by immunodiffusion (3.2-3.7 g / l in healthy people in serum) or antitrptic activity of serum. It is advisable to investigate the level of alpha-one-globulins, since alpha-1-antitrypsin makes up 70-90% of this fraction. In the treatment, purified and recombinant AAT preparations are used in aerosol administration or IV drip 4 g once a week. There is an improvement after the use of proteolytic enzymes (trasylol, contrikal, gordox), the anabolic steroid danazole, which increases the synthesis of AAT.

Cystic fibrosis. The most common hereditary disease in children. This is a systemic exocrinopathy. Mimicry is autosomal recessive. The prevalence of this pathology is 1: 2000

newborns. A defective gene (CF gene), which is located on the long arm of chromosome VII, leads to the defeat of the exocrine glands. The gene mutation causes structural and functional disorders of the regulatory protein CFTR.

More than 500 types of mutations are known now. The most common mutation is F508.
The pathogenesis is based on the violation of ion transport across the apical membrane of epithelial cells, which leads to dehydration and thickening of the secretions of exocrine glands, including bronchial ones. Blockage of the excretory ducts of the pancreas occurs in utero, causes its cystic degeneration, and as a result there are significant disorders of digestion and absorption.
The thick bronchial secretion of patients with cystic fibrosis is a very concentrated supersaturated solution, inhibits the movement of the cilia of the bronchial epithelium. This leads to a violation of mucociliary clearance, as well as the reproduction of pathogenic flora. DNA and lipids contained in bacteria, as well as immunoglobulin M, further thicken the sputum. Inflammation leads, in turn, to swelling of the mucous membrane, bronchospasm, increased obstruction. An increase in the production of viscous bronchial secretions in such conditions causes clogging of the bronchi with mucopurulent plugs. Thus, a vicious circle of bronchial obstruction occurs. Secondary chronic bronchitis gradually develops.
Clinic. The leading diagnostic criteria for mucovicidosis are: meconial intestinal obstruction, steatorrhea, malabsorption, whooping cough, recurrent bronchopulmonary diseases, retention jaundice, salty sweat, cirrhosis of the liver, nasal polyps, rectal prolapse, hot pansinusitis, lethargy during the year.
The main forms of cystic fibrosis are with pancreatic insufficiency, without pancreatic insufficiency.
Lung damage occurs in 95% of patients with cystic fibrosis. The main respiratory manifestations are debilitating whooping cough, bronchorrhea, and shortness of breath. A boxed sound and areas of shortening of the percussion sound are found in most patients with percussion of the lungs. Most often, scattered, varied and dry rales are heard over the lungs against the background of hard breathing. Leading in the X-ray picture are changes in the interstitial tissue together with the cells of the eclipse, which lead to the perforation of the picture.
Diagnosis of cystic fibrosis is based on typical clinical symptoms, genetic history, high concentration of electrolytes in sweat, decreased activity of pancreatic enzymes in duodenal secretions, molecular genetic research.
The sweat test is used to determine the concentration of chlorine ions in sweat. Sweat is collected with filter paper after pilocarpine electrophoresis. The concentration of chloride in sweat exceeds 60 mmol / l in children with cystic fibrosis. The amount of chlorides in sweat does not exceed 40 mmol / l in healthy children. The concentration of chlorides in the range of 40-60 mmol / l does not allow either to confirm or exclude the diagnosis of cystic fibrosis and requires molecular genetic research.
Treatment is largely symptomatic. Adequate therapy, especially early started, gives fairly reliable results, significantly increases the life expectancy of patients.

Scheme of complex therapy for cystic fibrosis

Pathogen	Antibiotic	Daily dose (mg/kg)
Staphylococcus	Cloxacillin	25
	Dicloxacillin	25
	Fluklosacillin	25
	Cephalosporins of the 1st generation	100
	Carbenicillin	450-800
	Azlocillin	450-600

Pseudomonas aeruginosa	Ceftazidim	100-240
	Gentamicin	8-12
	Tobramycin	10-20
	Amikacin	15
	Imipenem	30-90
	Ciprofloxacin	15-50

Correction of pancreatic insufficiency for all patients with cystic fibrosis should be carried out with enzyme preparations (creon, pancitrate, prolipase with a daily dose of 10-20 thousand units of lipase activity per 1 kg of weight). This dose of enzymes allows you to expand your diet. Sick children should receive regular food, without special restrictions, but with increased calorie content (by 20-50% of the norm), more protein, salt.
Measures to improve lung function. Measures aimed at the evacuation of bronchial secretions:
• Physical: Massage, exercise therapy, postural drainage, aerosol therapy
• Chemical: DNase, Mukomist, Ambroxol
• Instrumental: bronchoscopy with sanitation of the bronchial tree
Activities to improve liver function: ursodex lipoic acid
Hepatoprotectors: Hepabene, lipostabil, lipoic acid

The criteria for the effectiveness of therapy are the absence of shortness of breath, easy sputum discharge, stool once a day, normal coprogram values, and normal abdominal contour.
The prognosis is unfavorable. Only 50% of patients live up to 30 years, in rare cases - they live for a long time.
Clinical examination. Patients are under the supervision of a local pediatrician, regional pulmonologist, in the regional center of cystic fibrosis. They are examined every 1-2 months.

It is necessary to consider the **problem of providing palliative care to children**, considering that the prognosis for many patients with congenital malformations of the lungs, COPD and congenital pulmonary diseases is poor.
According to the estimates of international experts, about 500,000 patients need palliative care in Ukraine every year, of which more than 15,000 are children. All over the world, palliative care for children is an integral part of quality medical care, which includes comprehensive medical, social, psychological and spiritual support for critically ill patients and their families.
In Ukraine, the system of children's palliative care is just being created. An acute shortage of qualified medical personnel, the lack of effective methods of providing care, the lack of opportunities to study best practices in this area, as well as the lack of state support at the regional and national levels are the main factors hindering the organization of a system for providing palliative care to children in Ukraine. According to the WHO, palliative care aims to improve the quality of life of children and their families, who experience all the clinical, psychological, ethical and spiritual problems associated with unnecessarily exultant illnesses. Despite the rapid development of research in the field of palliative care for children in recent decades, there are “blind spots” in the professional training of pediatricians and in the education of specialists, the specifics of organizing research and providing patient and family support, which are too often underestimated and ignored.
Definition of palliative for children: need is benefit. Palliative care is defined by WHO as an approach that improves the quality of life of patients (children and adults) and their families facing life-threatening illness by preventing and alleviating suffering through early detection, careful

assessment and treatment, pain and other physical symptoms; and the provision of psychosocial and spiritual support. The provision of palliative care is based on the principle of respect for the decisions of patients and is aimed at providing practical support to their family members during illness and in the event of a patient's death in order to overcome grief in connection with the loss of a loved one.

Philosophy, levels and organization of palliative care for children.

The provision of palliative care contributes to the improvement of the quality of life of patients (for example, by reducing the severity of symptoms of the disease) and their family members, and is useful for health care systems itself, since it can significantly reduce the number of patients in hospitals and reduce the workload on staff. Most of the evidence for the effectiveness of palliative care comes from studies in high-income countries, but there is growing scientific evidence that the provision of palliative care in a culturally sensitive and participatory manner can lead to positive outcomes in low- and middle income countries.

Palliative care was first introduced in 1990 by WHO and is defined as “an approach to care that improves the quality of life of patients and their families with life-threatening illnesses through prevention, assessment and treatment of pain, prevention of physical, psychological and spiritual problems "

DPP philosophy. Despite the existing differences in approaches to palliative care in different countries, palliative care specialists have identified some common values: respect for patient autonomy and dignity, the need for individual planning and decision-making, a holistic approach. Patient autonomy, first of all, implies recognition of the value of each person and respect for him as a unique person. Dignity preservation presupposes the attitude of the staff towards the patient with respect, openness, responsiveness, taking into account his personal, cultural and religious values, hopes and attitudes. Therefore, one of the philosophical approaches to palliative care is the relationship between the patient and the medical staff. Staff should maintain a cooperative relationship with patients and their families, who are important partners. In the process of providing assistance, it is important to observe the aspect in which all participants in the provision of assistance are aimed at developing the child's so-called resilience, namely the ability to live with incurable diseases, psychologically accept the inability to change plans for the future. Yet, the main goal of DPP is to achieve, support, preserve and improve, as far as possible, the quality of life. The philosophical issue of palliative care is the relationship to life and death; so many definitions of palliative care contain a position in relation to life and death. Death is regarded as a phenomenon of the same importance as life; it is inseparable from life and is a part of it. Acts of euthanasia or suicide with the assistance of a physician should not be part of the scope of palliative care. “The value of life, the naturalness of the death process and the realization that life and death provide a person with opportunities for personal growth and self-realization” must be recognized. To provide palliative care of adequate quality, it is necessary that staff have effective communication skills - the interaction between the child and the health-care workers - us, between the child and their relatives, between the various categories of health-care workers and the staff of the services involved in the provision of care. Often, grievances and complaints are due to ineffective communication. Effective communication has been shown to improve the quality of care provided. In the process of communication, one has to discuss difficult and sometimes sensitive issues that require a certain amount of time, participation and growth. Palliative care staff face a difficult task: on the one hand, to provide the patient with honest and complete information, and on the other hand, to maintain respect for his hopes for a favorable prognosis and / or survival, despite the approach of death. Raising public awareness involves ensuring the development of the palliative care system, which will allow members of future generations to be less afraid of death and worry about bereavement, which is inevitable for all of us. The philosophical approach to the

provision of DPP is a multiprofessional and interdisciplinary approach, as teamwork is considered the foundation of palliative care. Multiprofessional teams should include a variety of clinical disciplines. There is compelling evidence that palliative care teamwork brings more benefits to children, reduces the overall cost of care by reducing the time the patient spends in emergency care settings that are effective in treating pain and other distressing symptoms.

DPP levels. Palliative care for children is a special, albeit closely related to adult, section of palliative care. There are two levels of palliative care: the palliative approach and specialized palliative care. The palliative approach to the provision of medical care is implemented in institutions and services that only periodically deal with the provision of care for "palliative" patients. In contrast, a trained team of doctors, nurses, social workers, church leaders, psychologists, volunteers, and others with experience in helping children improve their quality of life deliver specialized palliative care.

There are two main definitions of conditions in DPP: conditions that limit life expectancy and conditions that are life threatening. Life-limiting conditions in children are those conditions in which premature death usually occurs; for example, Duchenne muscular dystrophy. Life-threatening diseases are diseases in which there is a high probability of premature death due to the severity of the disease, but in which there is also a chance of survival and the child can live to adulthood. For example, children who are about to rhyme with cancer treatment or are admitted to the intensive care unit after an acute trauma. Palliative care for children is characterized by a diverse spectrum of conditions, among which non-oncological diseases account for a larger share than in the case of palliative care for adults.

In general terms, palliative care for children and adolescents can be divided into **4 categories, based on the Guidelines for the Development of Palliative Care Services for Children** prepared by the Children's Palliative Care Association and the Royal College of Pediatricians and Children's Health in the UK:

Group 1: Life-threatening diseases in which radical treatment can be carried out, but often does not work. The use of a palliative care service may be necessary in parallel with attempts at radical treatment and / or in case of failure.

Group 2: Conditions in which premature death is inevitable, but long-term intensive treatment can increase the child's life expectancy and maintain quality and physical activity (for example, cystic fibrosis).

Group 3: progressive incrabable conditions, usually lasting many years, when only palliative therapy is possible, for example, Batten's disease (neural waxy lipofuscinosis / Tay-Sachs disease) or muscular dystrophy.

Group 4: Irreversible / incurable, but not progressive conditions that lead to severe disability of the child, a tendency to frequent complications and the possibility of premature death. Examples include severe cerebral palsy; multiple severe consequences of hemorrhages in the brain or spinal cord.

The DPP models are defined as follows:

1. *Outpatient pediatric palliative care.* Very often, palliative care for children is provided as part of a general palliative care program (for example, by specialists working with both adults and children on an outpatient basis or as part of a mobile team). Advantage: the best way to bring the provision of palliative care as close as possible to the patient's life, which is extremely important in the presence of a small number of children's programs located at a great distance from the child's place of residence. Disadvantage: The number of staff trained in palliative care for children may be insufficient, or the number of children requiring this assistance may not be sufficient.

2. Inpatient palliative care units. Specialized palliative care units can function as independent organizations, hospices, or as part of a hospital or other healthcare facility. They offer family

members social rest as well as symptomatic control and an end-of-life (terminal illness) assistance program. Advantage: Children who are in serious condition can be hospitalized to correct symptomatic therapy. Disadvantage: inpatient care is a rather expensive service and may be located far from the child's place of residence.

3. Day care programs (day centers, day hospitals) can only provide assistance to those children who live close to this service. Advantage: in day centers it is possible to provide children with some additional types of assistance besides receiving them at home. During this time, the parents / guardians of the sick child have the opportunity to rest. Disadvantage: such centers can be located far from the place of residence of children, thus complicating the provision of palliative care.

Thus, the services and institutions for the DPP are allocated. Non-specialized palliative care institutions are: - district / district nursing services; - general practitioners; - outpatient nursing services; - departments of general hospitals; - orphanages and boarding schools.

Specialized care services are: - inpatient palliative care units; - inpatient hospices; - teams (support) for palliative care in hospitals; - teams that provide palliative care at home; - hospice teams that provide assistance at the patient's place of residence: - day hospices; - "hospitals at home"; - outpatient clinics.

Criteria for the appointment of palliative care (one or more of the following)

Conflicts over the use of medical nutrition / hydration in cognitively impaired, critically ill or dying patients	Automatically
Or	
A new diagnosis of a life-limiting or life-threatening illness Three or more hospitalizations within 6 months Difficulty getting over pain and symptoms Uncertainty about the patient, family, or doctor as predicted Family with limited social support The order "Let me die a natural death" / "Do not reanimate" and other ethical conflicts Complex coordination of care and / or household needs Long-term hospitalization for more than 3 weeks The need to use hospice resources	Recommended
Criteria for malignant diseases	
Having a malignant disease in combination with one of the following: Progressive metastatic cancer Bone marrow / stem cell transplant Diffuse congenital glioma of the pons varoli Stage IV neuroblastoma Recurrent malignant disease after stem cell / bone marrow transplant	Automatically
Any malignant disease first diagnosed with survival	Recommended
The presence of pulmonary abnormalities in combination with one of these	
Patients with cystic fibrosis (CF) for whom lung transplantation is being considered / or are in the process of transplantation Patients with CF with FEV1 (forced expiratory volume in 1 second) <30% Patients with CF who are dependent on a ventilator or are not suitable for lung transplantation obliterating bronchiolitis	Automatically
CF patients with multiple hospitalizations Patients with CF suffering from pain, shortness of breath or others, and how symptomatic therapy would be in favor Central hypoventilation syndrome Patients chronically dependent on a ventilator	Recommended

6. Materials for methodological support of the lesson.

6.1. Tasks for self-examination of the initial level of knowledge and skills / with the submission of standards of answers at the end of the block of tasks - tasks of the II level; tests of various types also with response standards /:

1. The modern definition of chronic nonspecific bronchopulmonary diseases (COPD).

2. Etiopathogenesis, clinical course and additional methods for diagnosing congenital malformations and COPD in children.

3. Risk factors for the development of COPD in children.

4. Consider the principles of rational therapy, issues of organizing rehabilitation measures for COPD in children

6.2. The information necessary for the formation of knowledge and skills can be found in the textbooks: / services, main literary sources, indicating pages /:

- *basic:*

1. Volosovets O.P, Snisar V.I. Recommendations for heart-healthy reanimation for children. A methodical colleague. Dnipropetrovsk: ART-PRES, 2015.48 p.
2. D 362 State form of medicines, tenth edition. Kyiv, 2018 https://moz.gov.ua/uploads/1/5052-dn_20180510_868_dod_2.pdf

3. Differential diagnosis of the most common diseases of childhood. Textbook / ed. V.M. Dudnyk, 1st edition. Vinnytsia: Nilan Ltd., 2017. 560 p.
4. Karen J. Markdante, Robert M. Kligman. Fundamentals of Pediatrics according to Nelson: translation of the 8th English. edition: in 2 volumes. Volume Kyiv: VSV "Medicine", 2019. XIV, 378 p.
5. Karen J. Markdante, Robert M. Kligman. Fundamentals of Pediatrics according to Nelson: translation of the 8th English. edition: in 2 volumes. Volume Kyiv: VSV "Medicine", 2019. XIV, 426 p.
6. Kryuchko T.A, Abaturov A.E, Kushnereva T.V Pediatrics: textbook (University IV level. A); under ed. AND. T.A. Kryuchko, A.E. Abaturov. Kiev: VSI "Medicine", 2020. 224 p.
7. Emergencies in pediatric practice: Textbook. way. for students. med. ZVO, interns. - 2nd type. Recommended by the Ministry of Education and Science, Recommended by the Academic Council of NMU. O.O. Bogomolets / Marushko Y.V, Chef G.G etc. Kyiv: VSV "Medicine", 2020. 440 p.
8. Pediatrics: a national textbook: in 2 volumes / Ed. prof. Berezhnogo V.V Kyiv, 2013. Vol.1. Kyiv, 2013. 1040 p.
9. Pediatrics: a national textbook: in 2 volumes / Ed. prof. Berezhnogo V.V Kyiv, 2013. Vol.2. Kyiv, 2013. 1024 p.
10. Pediatrics: a textbook for students. higher education institutions IV level of accreditation. / for ed. prof. O.V Severe. View. 5th, ed. and add. Vinnytsia: Nova Kniga, 2018. 1152 p.: ill.
11. Order of 13.01.2005 №18. On approval of protocols for providing medical care to children in the specialty "Children's Pulmonology"

- *additional:*

1. Okhotnikova E.N. Urgent issues of rational antibiotic therapy of inflammatory diseases of the lower respiratory tract in children's practice / E.N. Okhotnikova, E.V. Ponochevnaya, E.V. Sharikadze [and others] // - "Child's Health" magazine. - No. 2 (t 61). - 2015. - P.16-20.

2. Gonchar M. O., Riga O. O., Penkov A. Yu. Principles of providing palliative assistance to children / M. O. Gonchar, O. O. Riga, A. Yu. Penkov. - Kharkiv: KhNMU, 2016 .-- 112 p.
3. Belenkaya O.I, Afanasyeva N.I, Yudin A.L Swire-James syndrome (McLeod's syndrome) // Radiology-practice. - 2009. - No. 3. - S. 21-24.

6.3. Orientation map for independent work with literature on the topic of the lesson.

№	Basic tasks	Guidance	Answers
1	2	3	4
1.	Learn concepts: COPD.	To give the definition of COPD.	To indicate what lies at the heart of COPD.
2.	Etiology.	To indicate the reason for the development of the disease	To note that congenital malformations of the lungs, hereditary diseases of the lungs are essential
3.	Pathogenesis.	To highlight the main links of pathogenesis.	
4.	Clinic.	To characterize the clinical manifestations in COPD.	To determine the clinical manifestations of various conditions that are classified as COPD.
5.	Differential Diagnosis.	To characterize the most common disease with which it is necessary to carry out differential diagnosis.	
6.	Treatment.		To know the principles of treatment of various diseases related to COPD
7.	Prevention.	To point out the basic principles of prevention.	To know the principles of prevention of various diseases related to COPD

7. Materials for self-control of the quality of training.

A. Questions for self-control:

1. Definition, classification, predisposing factors, signs of chronic lung disease in children.
2. Chronic bronchitis: risk factors, classification, pathogenesis, diagnostic criteria (clinical, laboratory, instrumental).
3. Bronchiectasis: definition, etiology and pathogenesis, clinical picture.
4. Bronchiectasis: diagnostics (laboratory, instrumental, bronchoscopic examination)
5. Cystic fibrosis: definition, etiology and pathogenesis, clinical picture, diagnostic criteria.
6. Congenital malformations of the respiratory system: classification, malformations associated with underdevelopment of bronchopulmonary structures (agenesis, aplasia, hypoplasia of the

lungs), malformations of the lungs (congenital lung cyst, lung separation, congenital bronchiectasis).
7. Often malformations of the wall of the trachea and bronchi:
- tracheobronchomalacia (Munier-Kuhn syndrome) tracheobronchomalacia;
Williams-Campbell syndrome (ballooning bronchiectasis syndrome) limited developmental defects of the wall of the trachea and bronchi: congenital tracheal stenosis; congenital lobar emphysema; diverticula of the trachea and bronchi tracheo- and bronchoesophageal fistula.
8. Kartagener's syndrome.
9. Differential diagnosis of COPD and BA, the principles of basic therapy in COPD.

B. Tests for self-control with reference standards:
1. A 12-year-old child has been diagnosed with chronic bronchitis after suffering whooping cough. At this time, there is a period of exacerbation. A hemophilic bacillus is inoculated from the sputum at a concentration of 105 in 1 ml. Do you need an antibiotic?
A. Penicillin
B. Oxacillin
C. Cloxacillin
D ampicillin
E. Lincomycin
2. Valya K. suffers from cystic fibrosis. Infiltration of the lower lobe of the right lung with a clear border from above, with displacement of the mediastinal organs to the right was detected with the next exacerbation on the roentgenogram. What complication should be diagnosed?
A. Atelectasis of the lower lobe of the right lung
B. Atelectasis of the lower lobe of the left lung
C. Right-sided pneumothorax
D. right-sided pneumonia
E. Right-sided pleurisy
3. Katya S. is 7 years old. From the age of 3, after starting attending kindergarten, she suffers from frequent repeated bronchitis and pneumonia, from the age of 5 she was diagnosed with chronic sinusitis. A deformation of the bronchovascular pattern in the lower sections is noticeable on the roentgenogram, the heart is dextracardia. What is your likely diagnosis?
A. Cystic fibrosis
B. Bronchial asthma
C. tracheobronchomegaly
D. Kartagener's syndrome
E. Tetralogy of Fallot
4. The child is 11 months old. From the first days of life he is worried about a constant cough, sputum is hard to come out. Lagging behind in physical development, cyanosis of the nasolabial triangle, there is a large number of wet and dry wheezing over the lungs. Sweat electrolytes is 130 mmol / l. What is your likely diagnosis?
A. Chronic bronchitis
B. Chronic pneumonia
C. Cystic fibrosis
D. Acute obstructive bronchitis
E. Bronchiolitis
5. Nikolai K. is 13 years old. From 3 years of age, right-sided lower parts of pneumonia were observed 2-4 times a year. Until the age of 3 he was not sick. After 7 years, he constantly complains of cough with discharge of a large amount of purulent sputum in the morning. Above the lungs - the sound in the projection of the lower lobe is constantly heard, small and medium bubbles, moist rales. What is your likely diagnosis?
A. Chronic bronchitis
B. Chronic pneumonia

C. Cystic fibrosis
D. Whooping cough
E. Bronchiectasis
Correct answers (1-D, 2-B, 3-D, 4-C, 5-E)

B. Self-control tasks with answers:

<table>
<tr><td colspan="2">Task 1. An 8-year-old patient complains of subfebrile condition for 1 month, dry cough, shortness of breath during exertion. X-ray revealed the presence of pleural effusion. It was found in the study of pleural effusion: specific gravity - 1.026, protein level - 38 g / l. during cytological examination, lymphocytes, single mesothelial cells predominate.</td></tr>
<tr><td>Exercise
1. Make a diagnosis
2. Specify the radiological signs characteristic of pleurisy.
3. Characterize the effusion.
4. Indicate the etiology of the disease for which these parameters are characteristic.
5. What is the further tactics of treatment.</td><td>Answer
1. Exudative pleurisy.
2. Intense eclipse, indistinctness of the sinuses, displacement of the mediastinum to the healthy side.
3. The nature of the effusion is serous exudate (increased density, high protein level) and cytological studies indicate the process of tuberculous etiology.
4. It is possible to envisage the process of tuberculous etiology.
5. Appointment of specific therapy.</td></tr>
<tr><td colspan="2">Task 2. A child, 5 years old, has left-sided lower-frequency pneumonia 2-3 times a year. There are no indications for foreign body aspiration, no allergic reactions. Objectively: asymmetry of the chest (on the left, sunken in the anterolateral region), on the left - in the projection of the lower lobe, breathing is weakened, fine bubbling rales. On the Rg-gram - there is a decrease in hemithorax, a decrease in pneumatization, a higher level of the diaphragm on the left. IgE - 29 IU / ml, sweat chlorides - 38 mmol / l.</td></tr>
<tr><td>Exercise
1. Determine the most likely diagnosis.
2. What additional research methods will confirm the diagnosis?
3. Indicate the principles of treatment.
4. What are the tactics of a family doctor?
5. Determine the methods of medical examination and rehabilitation</td><td>Answer
1. Hypoplasia of the lower lobe of the left lung
2. Bronchography
3. Operational
4. Prevention of COPD
5. Respiratory gymnastics, exercise therapy, aerofitotherapy, immunomodulators</td></tr>
<tr><td colspan="2">Problem 3. Vasya M., 2 years old. From the first pregnancy, which took place with toxicosis of the second half, gentle timely, with weakness in labor, was born in asphyxia (2 points according to the Apgar school), was on artificial lung ventilation for 3 days. There are frequent recurrent obstructive bronchitis, pneumonia. In the last six months, during the period of remission, tachypnea, cyanosis of the nasolabial triangle has been noted. There is a clear deformation of the bronchovascular pattern on the roentgenogram. Sweat chlorides is 30 mEq</td></tr>
<tr><td>Exercise
1. What is your diagnosis?
2. Carry out differential diagnostics.</td><td>Answer
1. Bronchopulmonary dysplasia.
2. Cystic fibrosis, Hammen-Rich syndrome, Williams-Kembel, Kartegener syndrome.</td></tr>
</table>

3. What additional examination methods are required to confirm the diagnosis? 4. What is your therapeutic tactics? 5. Methods of medical examination and rehabilitation.	3. Bronchoscopy, bronchography, tomography of the lungs. 4. Symptomatic therapy, treatment of intercurrent infections. 5. "D" accounting, drugs with a mild vaccination effect (ribomunyl, bronchomunal), aerophytotherapy, aromatherapy, reflexotherapy.

8. Materials for classroom self-study:

8.1. *The list of educational practical tasks that must be completed during the practical (laboratory) lesson:*

1. To collect anamnesis, highlight data that indicate the disease.
2. To identify the most informative signs of the disease during an objective and laboratory and instrumental examination of the patient.
3. To establish a clinical diagnosis according to the modern classification.

9. Guidance materials for mastering professional skills:

9.1. *Methodology for performing the work, stages of implementation:*

1. Evaluate the data obtained from the anamnesis of life and illness, highlight risk factors
2. Conduct a clinical examination of the patient.
3. Draw up a plan for additional examination.
4. Evaluate the results of laboratory and instrumental examination.
5. Formulate a clinical diagnosis according to the classification.
6. Prescribe treatment that is appropriate for the specific situation

10. Materials for self-control of mastering knowledge, abilities, skills provided for by this work.

10.1. Tests of different levels (or tests that are part of the bank for the rector's control):

1. Child is 5 years old. It is the 5^{th} day of the disease with right-sided lower lobe pneumonia. The child's condition deteriorated: shortness of breath and abdominal pain appeared when breathing, a sharp rise in temperature up to 39.5°C. Objectively: a serious condition due to intoxication and respiratory failure, dyspnea of a mixed nature up to 35 per minute. The child lies on the right side, heart rate is 110 per 1 min. Percussion dulling of sound deals with the upper border, which goes from the spine outward to the inner corner of the scapula, where breathing is not heard. Leukocytosis, neutrophilic shift to the left, accelerated ESR was found in the blood. What is your diagnosis?

A Pneumothorax
B Lung abscess
C Pleurisy *
D Tuberculosis
E Bronchiectasis

2. A child is 8 years old. The condition is severe, lethargic, pale, perioral cyanosis. Expiratory dyspnea. Accessory muscles take part in the act of breathing. Percussion over the lungs is a boxed sound. Breathing is sharply weakened. Dry wheezing rales. BH is 40 per minute The boundaries of cardiac dullness are not expanded. Heart sounds are muffled. HRC is 120. Blood pressure is 105/65 mm. Hg Liver is + 1 cm. Diuresis is according to age. What causes the severity of the condition?

A vascular insufficiency
B Heart failure
C respiratory failure *
D renal failure

E liver failure
3. A 10-year-old girl has bilateral pneumonia. Dyspnea is increased, body temperature is risen to 39.7°C. Objectively: the right half of the chest lags behind in the act of breathing, the intercostal spaces are smoothed. Percussion tympanitis, auscultatory breathing over this area is absent. The borders of the heart are shifted to the left. In the blood, the total number of leukocytes is 27.5 g / l. What is the most likely complication?
A Atelectasis
B Pyopneumothorax *
C Hydrothorax
D Pneumothorax
E bronchiectasis
4. Child is 5 years old. 6-7 times a year, he suffers from acute respiratory infections, bronchitis, from the age of 4 he has sinusitis. Dextracardia, deformation of the bronchopulmonary pattern are on the R-gram of the chest organs. Determine the most likely diagnosis:
A Kartagener's syndrome *
B Allergic bronchitis
C Tetrad of Fallot
D A1-antitrypsin deficiency
E Fibrosing alveolitis
5. In an 11-month-old child who was admitted to the hospital for pneumonia, there was swelling of the cervical veins against the background of cystic fibrosis in a serious condition against the background of the progression of deterioration. On examination, pronounced cyanosis, pasty legs, tachycardia, rhythm disturbances, an emphasis of the II tone on the pulmonary arteries were revealed. The borders of the heart are sharply increased (more to the right), the liver is +4 cm. On the ECG, there are signs of right deviation. What complication can you think of?
A Acute heart failure *
B Acute coronary insufficiency
C Myocardial dystrophy
D Myocarditis
E Neurotoxicosis