№ ЛД -21 ИН

Federal state-funded educational institution of the higher education
"NORTH OSSETIAN STATE MEDICAL ACADEMY"
Ministries of Health of the Russian Federation

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DEPARTMENT OF CHILDREN'S DISEASES NO. 2

EEDUCATIONAL AND METHODICAL RECOMMENDATIONS OF DISCIPLINE PEDIATRICS

Part III

Vladikavkaz, 2020.

The methodical recommendations for out-of-class independent work for studentsof discipline pediatrics was approved at the meeting of the Department of Children's Diseases No.2 dated November 25, 2020, protocol No4.

The methodical recommendations for out-of-class independent work for studentsof discipline pediatrics was approved at the meeting of the central coordinating educational-methodical council dated December 04, 2020, protocol No2.

ORIGINATORS:

PhDs in Medicine. **S.K. Karyaeva,** MD, **Z.D. Kaloeva,** PhDs in Medicine. **K.M. Dzilikhova,** PhDs in Medicine. **Z.Yu. Sozaeva,** PhDs in Medicine. **I.G. Gussoeva,** PhDs in Medicine. **Z.K. Dzhagaeva.**

Reviewers:

Head Department of Pediatrics №1 FSBEI the KubGMU of the Ministry of Health of Russia m.d.s., professor V.A. Shashel

Head of the Department of Children's Diseases №1 FSBEI the SOGMA of the Ministry of Health of Russia, m.d.s., professor T.T. Boraeva

Class in a subject:

"BRONCHIAL ASTHMA AND CHRONIC NONSPECIFIC DISEASES OF LUNGS AT CHILDREN"

I. Scientific and methodical justification of a subject.

The problem of pulmonary pathology remains to the leader in pediatrics. Recently the tendency to increase in number of respiratory allergoses and HNZL in all advanced countries is noted. Beginning at children's age and progressing, bronchial asthma and HNZL lead to development of heavy pathology with permanent disability at mature age. Considering it, the medical student needs studying the reasons and conditions of forming of chronic pathology of a bronchopulmonary system, methods of clinical, laboratory and tool diagnostics, landmark treatment, questions of dispensary observation and continuity of medical care.

II. Purpose of activity of students.

The student has to know:

- anatomo-physiological features of respiratory organs at children;
- types of allergic reactions;
- etiology and forms of bronchial asthma;
- pathogenesis of an attack of suffocation;
- clinical picture of the criminal and mezhpristupny periods of bronchial asthma;
- features of pathogenesis and clinic of bronchial asthma at children of early age;
- diagnostics and the differential diagnosis of bronchial asthma with other diseases;
- landmark treatment;
- emergency treatment at an attack;
- medical tactics during the mezhpristupny period;
- actions of primary and secondary prevention of bronchial asthma;
- etiological factors and the causes of the chronic non-specific lung diseases (CNSLD) at children;
- questions of pathogenesis and pathomorphologic changes of a bronchopulmonary system at HNZL;

- classification of HNZL;
- principles of laboratory and clinical methods of inspection;
- features of treatment and medical examination of children at HNZL.

The student has to be able:

- to reveal ways of allergization;
- to examine the patient with a respiratory allergosis (features of the anamnesis, a clinical picture);
- to estimate laboratory, tool, radiological methods of inspection;
- to make the diagnosis according to the existing classification;
- to recommend the rational mode, a diet;
- to appoint treatment in the criminal period, to write prescriptions on the main medicines;
- to appoint treatment in the vnepristupny period;
- to carry out prevention of infectious and allergic diseases.
- to establish the factors and the reasons promoting emergence of HNZL;
- to examine the patient from HNZL (feature of the anamnesis, a clinical picture);
- to carry out the differential diagnosis of a mucoviscidosis;
- to make dispensary observation for children with a mucoviscidosis;

III. Content of training:

1. Etiology and forms of bronchial asthma.
2. Pathogenesis of an attack of suffocation.
3. Features of pathogenesis and clinic of bronchial asthma at children of early age.
4. Diagnostics and differential diagnosis of bronchial asthma with other diseases.
5. Features of a X-ray pattern of bronchial asthma.
6. Measures of emergency treatment at an attack of bronchial asthma.
7. Treatment during the vnepristupny period.
8. Primary and secondary prevention of bronchial asthma.
9. Pathogenesis and pathomorphologic changes in a bronchial system in chronic pneumonia.
10. Clinical picture of chronic pneumonia.

11. Laboratory, radiological and these bronkhoskopiya in chronic pneumonia.

12. Principles of antibacterial therapy (choice, method of administration of drugs, course duration).

13. Indications to sanatorium treatment at HNZL.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.

2. Educational medical documentation (case histories, laboratory researches, roentgenograms).

3. Technical means of training.

4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.

2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.

3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.

4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.

5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.

6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. Call the main anatomo-physiological features of a bronchopulmonary system at children.
2. Call types of allergic reactions.
3. Call forms of bronchial asthma.
4. What etiological factors matter in development of an attack of bronchial asthma?
5. Call the main pathogenetic mechanisms of an attack of suffocation.
6. Call features of the immune system at children.

VII. List of questions for check of final level of knowledge:

1. What allergic damages of upper airways meet at children?
2. Call features of pathogenesis and clinic of bronchial asthma at children of early age.
3. Describe a clinical picture of the criminal period of bronchial asthma.
4. Call clinical manifestations of bronchial asthma in the vnepristupny period.
5. Diagnostics and differential diagnosis of bronchial asthma with other diseases.
6. What features the X-ray pattern of bronchial asthma has.
7. Call etiological factors and clinical manifestations of an obstructive syndrome.
8. Call measures of emergency treatment at an attack of bronchial asthma.
9. What treatment of bronchial asthma is carried out in the vnepristupny period?
10. Call measures of primary prevention of bronchial asthma.
11. What is secondary prevention of bronchial asthma? How it is carried out?
12. What forecast in bronchial asthma?
13. Give classification of HNZL at children.
14. Call the purposes of bronchoscopic sanitation.
15. What methods of physiotreatment of HNZL are known to you?
16. List the basic principles of prevention and dispensary observation for children with HNZL.
17. Etiopatogeneticheskу aspects of a mucoviscidosis.
18. Call forms of a mucoviscidosis and the main clinical manifestations of a disease.
19. List the main differential and diagnostic criteria of a mucoviscidosis.
20. Call the principles of treatment and the forecast in a mucoviscidosis.

Information block

Bronchial asthma at children – the disease which is developing on the basis of chronic allergic inflammation of bronchial tubes, their giperreaktivost and characterized by periodically arising attacks of the complicated breath or suffocation as a result of the widespread bronkhoobstruktion caused by a bronkhokonstriktion, slime hypersecretion, hypostasis of a wall of bronchial tubes.

This definition excludes division on the allergic, infectious and allergic and mixed forms which was earlier widely used.

Key provisions of definition of bronchial asthma

□vospalitelny process results in hyperreactivity of bronchial tubes, obstruction and emergence of respiratory symptoms**.**

the □obstruktion of airways happens four forms:

□ostry bronkhokonstriktion – owing to a spasm of unstriated muscles;

□podostry – because of hypostasis mucous airways;

** chronic – formation of a viscous secret, occlusive terminal department of bronchial tubes;*

** sclerous process of a wall of bronchial tubes.*

√ *the Atopy, genetically caused disturbances of synthesis of IgE.*

√ Asthma – a chronic persistent inflammatory disease

airways.

<u>Risk factors of developing of bronchial asthma</u>

Contribute to development of bronchial asthma:

● heredity;

● atopy;

● hyperreactivity of bronchial tubes.

The causative (sensibilizing) factors:

□bytovy allergens (house dust, pincers of house dust);

□allergena of animals, birds, allergens of cockroaches and other insects;

□gripkovy allergens;

□pyltsevy allergens;

□pishchevy allergens;

□lekarstvenny means;

□virusa and vaccines;

□khimicheky substances.

The factors promoting developing of bronchial asthma and aggravating action of causative factors:

+ viral respiratory infections;

+ pathological course of pregnancy at the child's mother;

+ prematurity;

+ irrational food;

+ atopic dermatitis;

+ various pollyutant;

+ tobacco smoke.

The factors causing exacerbation of bronchial asthma – triggers:

◊ allergens;

◊ viral respiratory infections;

◊ physical and psychoemotional activity;

◊ change of a meteosituation;

◊ ecological influence (xenobiotics, pungent smells);

◊ intolerable products, drugs, vaccines.

Mechanisms of development of bronchial asthma in children

Inflammation of airways in bronchial asthma

The hyperreactivity of large and small airways is shown by chronic inflammation. Inflammation is coordinated by CD4+ cells (T – helper) – Th2 lymphocytes. There are many applicants for a role of a starting link, including biologically active agents produced by mast cells, eosinophils.

Lymphocytes of Th2 cosecrete IL-4 and IL-5 cytokines playing a key role in development of allergic inflammation. Besides, IL-4 and IL-13 cytokines – too Th2 product of lymphocytes. They switch V-lymphocytes to synthesis of IgE-antibodies. IgE-antibodies contact receptors of mast cells. The mediators causing acute allergic manifestations and preparing development of late phase reaction when eosinophils which

are the second main effective cell of allergic inflammation are attracted are released. In the course of activation the cytokines supporting formation of Th2 cosecrete.

It is proved in recent years that lymphocytes can cause bronchial hyperactivity and without antigen challenge, do not use products of IgE.

Neurogenetic regulation of airways

The bronchial tone changes at disturbance of balance between systems:

√ *Exciting* – cholinergic, not cholinergic, α-adrenergic systems.

√ *Inhibiting* – β-adrenergic and neadrenenrgichesky systems.

Neuropeptids accompany and aggravate allergic inflammation which is initiated by reaginzavisimy reaction.

Vazointestinalny peptide (VIP) – the most powerful of bronchodilators known today which can resist to a bronkhopazm in asthma. Dysfunction in a VIP-system can occur in the course of inflammation of airways in asthma.

Endocrine regulation

Influence of an endocrine system in asthma is carried out through implementation of antistress effect and proper defense reaction of an organism against antigen. These effects are reached through systems:

+ *a hypothalamus – a hypophysis – adrenal glands;*

+ a hypothalamus – a hypophysis – a thymus gland;

+ a hypothalamus – a hypophysis – a thyroid gland.

Feature of bronchial asthma at children – existence of dissociative disturbances in a neuroimmunoendocrine complex.

Clinical picture, diagnosis and differential diagnosis

The first stage – diagnosis of bronchial asthma

Assessment of data of the anamnesis, clinical symptoms, allergological status.

The diagnosis "bronchial asthma" **is probable** *if:*

◊ the symptoms specified in an algorithm repeat;

◊ take place at children is more senior than 3 years;

◊ arise at night or early in the morning more often;

◊ are connected with allergen or physical activity;

◊ the seasonality of manifestation of symptoms is noted;

◊ cases of allergic diseases in family come to light.

The diagnosis "bronchial asthma" **is improbable** *if:*

◊ cough and/or goose breathing at early age;

◊ lag in physical development;

◊ persistent infection;

◊ permanent physical changes from lungs and cardiovascular systems;

◊ usual vomitings and poperkhivaniye.

Criteria of diagnosis of bronchial asthma

Clinical signs of obstruction of airways:

▪ incidental expiratory asthma; and/or paroxysmal cough, feeling of compression in a breast;

▪ auskultativno: the weakened breath with abundance of dry and damp rattles.

Features of the anamnesis:

□btyagoshchenny heredity on allergic diseases;

▪ presence of the accompanying allergic diseases (atopic dermatitis, Quincke's edema, small tortoiseshell, etc.);

▪ communication of symptoms of a bronkhoobstruktion with influence of allergens;

▪ improvement of a state after use of bronkhodilyatator.

Distinguish an asthmatic attack and the asthmatic status that represents the prolonged generalized obstruction resistant to use of bronkhospazmolitik and leading to acute respiratory insufficiency.

Clinic

The disease proceeds with the periods of aggravation and remission.

The Predpristupny period is characterized by symptoms of allergological rhinitis, vegetative dystonia, tussiculation. Suffocation attacks develop at night or in hours at dawn more often. The attacks arising in the afternoon are caused by contact with various

allergens and irritants. The attack of suffocation begins with painful cough with difficult departing phlegm, an expiratory asthma with participation of auxiliary muscles, remote rattles, cyanosis of lips, in some cases a Crocq's disease appear. In lungs various dry and mixed damp rattles, mainly on an exhalation on both sides are listened. There is tachycardia, arterial blood pressure increases.

Asthmatic attack – a suffocation attack with the dry rattles heard at distance. Breath with the complicated exhalation, the rattles and whistle audible at distance. The thorax is expanded, in the provision of a deep breath. The patient adopts the forced provision. During heavy attacks the face and skin of a tsianotichna, cervical veins are blown up. Percussionly over all surface of lungs – a box shade of a pulmonary sound. At first rattles on a breath and especially on an exhalation are listened multiple, high-pitch tone. Cough, a phlegm at the beginning and in the attack heat very poor, dense, viscous is possible, it is allocated hardly or at all does not separate.

Deaf cardiac sounds, tachycardia. The ABP raises.

Attack duration – of several minutes till several o'clock can also become the beginning of an asthmatic attack.

The asthmatic state can arise at any form of bronchial asthma and is characterized by the following main signs:

1. Fast increase of bronchial obstruction.
2. Lack of effect of introduction of sympathomimetics.
3. Increase of respiratory insufficiency

Distinguish three stages of an asthmatic state:

The I stage – an attack of bronchial asthma with full lack of effect of use of sympathomimetics.

The II stage – the increasing respiratory insufficiency, emergence of zones of "a mute lung" - sites over lungs where completely there is no vesicular breath sound; reduction of number of dry rattles, emergence of sites where rattles are not listened that is connected with bronchial obstruction.

The III stage – a giperkapnichesky coma or a hypoxemic coma – tension $WITH_2$ increases up to 80 - 90 mm Hg., and oxygen tension sharply falls up to 40 – 50 mm Hg. The patient faints, breath deep, with the extended exhalation.

About weight of a course of bronchial asthma it is possible to judge by the frequency, duration, the nature of attacks of suffocation. A state in the mezhpristupny period, development of asthmatic states. At an easy course the attacks arise 2 – 3 times a year, proceed not hard, are short-term, are easily stopped by antispasmodics in powders and tablets.

At a medium-weight course the frequency of attacks of suffocation is 5 and more in a year, they are more long, for their stopping aerosols and injections of bronkhospazmolitichesky means are required.

The heavy course of bronchial asthma is characterized monthly, sometimes in addition more frequent and quite often long attacks demanding intravenous administration of broncholitic means and is frequent – glucocorticoid hormones. At a heavy course of bronchial asthma also asthmatic states, quite often life-threatening develop.

The clinical parameters characterizing weight attack of bronchial asthma

√ Respiration rate.

√ Participation of auxiliary muscles in the act of breath.

√ Intensity of goose breathing.

√ thorax Swelling.

√ Character and carrying out breath in lungs (at auscultation).

√ Heart rate.

√ the Forced situation.

√ Change of behavior.

√ Extent of restriction of physical activity.

√ therapy Volume (drugs and methods of their maintaining), used for stoppings of an attack.

Gradation:

* easy attack;
* moderately severe attack;
* heavy attack;

* extremely heavy attack (asthmatic status).

On duration of a course allocate 3 options of a course of bronchial asthma.

Intermittent with rare attacks

Attacks of suffocation are more rare than 1 time of 4 - 6 weeks; the whistling rattles after big physical activity. Normal function of lungs and lack of symptoms during the mezhpristupny period. Does not need therapy.

Intermittent with frequent attacks

Attacks Bol frequent, but more rare than once a week. The whistling rattles after moderate physical activity. In the mezhpristupny period normal or almost normal functions of lungs. Preventive therapy, as a rule, is necessary.

Persistent asthma

About 5% of children suffer from this option of asthma. Frequent attacks, the whistling rattles after the slightest loading. Function of lungs is reduced also during the mezhpristupny period. β_2 – agonists are applied more often than 3 times a week. Preventive therapy is obligatory.

Basic principles of therapy

Basic therapy of bronchial asthma – **anti-inflammatory therapy.** At exacerbation of bronchial asthma, bronkhospazmolitichesky means are connected.

• Anti-inflammatory drugs

Inhibit an early phase of the allergic answer, a late phase of allergic reactions at chronic inflammation and reduce bronchial hyperreactivity.

Non-steroidal anti-inflammatory drugs

The inhalation method of introduction is used.

□kromoglikat *sodium (intal, kromolin-sodium).* For prevention of an early phase of allergicheky reaction the only preventive dose can be sufficient. The course not less than 1.5 … 2 months on 1 … 2 inhalations 3 … 4 times a day is necessary for impact on bronchial hyperreactivity.

* *Nedokromil of sodium (tayled).* More active, than intal.

Drugs of this group are effective at children with slight and medium-weight bronchial asthma, especially at the initial stages of a disease.

Inhalation corticosteroids

Modern inhalation steroids – beclomethasone, budezonid, flunisolid, flutikazon. At a heavy course the prolonged use – not less than 6 … is shown 8 months. Are appointed after elimination of the main symptoms of acute respiratory insufficiency, recovery of bronchial passability.

At a medium-weight and heavy course of bronchial asthma for increase in activity of anti-inflammatory therapy the prolonged bronchial spasmolytics – theophyllines of long action or prolonged β2-агонисты are added.

* *Theophyllines of long action*

are used in a complex with anti-inflammatory therapy and for prevention of emergence of attacks, especially night asthma. A daily dose – 12 - 15 mg/kg of body weight; at a heavy course – 11 - 12 mg/kg of body weight.

* *inhalation β2-агонисты long action*

Provide bronkhodilatiruyushchy effect till 12 o'clock. Are appointed for reduction of number of the arising attacks of bronchial asthma.

Step approach to basic (long-term treatment)

bronchial asthma

At prescription of medicines "*the step approach*" - increase in quantity and frequency of drug intake in process of increase of weight of a course of asthma is applied.

Course	Step 1	Step 2	Step 3
	Lung	**Medium-weight**	**Heavy**
	Basic therapy (directed to prevention attack		
Anti-inflammatory	Kromoglikat of sodium 4 times a day or nedokromit sodium 2 times a day	Kromoglikat of sodium 4 times a day or nedokromit sodium 2 - 4 times a day At insufficient efficiency within 6-8 weeks to replace with inhalation are corticosteroid in sredneterapevtichesk y doses	Inhalation corticosteroids in high doses At insufficient efficiency: + oral corticosteroids short course
Bronkhodilyata tora for prolonged use	Are not shown	Theophylline of the prolonged action or β-agonists $_2$ of the prolonged action	Theophylline is prolonged - leg of action or β-agonists $_2$ of the prolonged action

Independent work of students.

Scheme of inspection of the patient.

When collecting the anamnesis to pay attention on:

- allergic status of close relatives;

- features of a course of pregnancy (diseases, treatment, pregnant woman's food, allergic manifestations);
- neonatality period (resistant intertrigo, nature of an enanthesis);
- feeding (presence of food allergy);
- existence of complications after vaccination;
- frequency of diseases of ORZ, course duration;
- age at which the disease, a course, aggravation frequency, results of therapy is for the first time revealed;
- observation and treatment out of aggravation;
- what aggravation of symptoms, treatment before hospitalization is connected with.

At the general survey to pay attention on:

- general condition of the child
- physical and psychological development;
- presence of intoxication, degree of respiratory insufficiency, cardiovascular insufficiency;
- condition of integuments and mucous membranes (presence of allergic rashes, centers of persistent infection);
- changes from skeletal system (a shape of a facial skull, a thorax, fingers);
- asthma and its character;
- these palpations, percussions, auscultations of a bronchopulmonary system;
- condition of a cardiovascular system (circulatory unefficiency, heart borders, tones, noise);
- condition of abdominal organs.

At assessment of paraclinic methods of a research to pay attention on:

- blood test (maintenance of erythrocytes, Hb, leukocytosis, leukocytic formula, increase in SOE);
- biochemical analysis of blood (disproteinemia, hypergammaglobulinemia, hyper alpha-2-globulinemiyu);
- research of immunoglobulins (increase in maintenance of IgE);
- changes on roentgenograms of a thorax to give their assessment;

- the nature of changes on bronkhogramma;
- assessment of a bronchoscopic picture at HNZL at children;
- phlegm research (bacteriological, cellular structure);
- assessment of function of external respiration, pikfloumetriya.

Task No. 1

Girl of 6 years. The district doctor visited the child on the asset received from the emergency doctor at home. Complaints to paroxysmal cough, goose breathing.

The girl from the first normally proceeding pregnancy, births in time. Weight at the birth 3400 gr., length of 52 cm. The period of a neonatality proceeded without features. On artificial feeding since 2 months. Till 1 year of life had children's eczema. Does not transfer chocolate, strawberry, eggs (on skin rashes develop). Family anamnesis: mother of the child has a recurrent small tortoiseshell, the father has a peptic ulcer of a stomach.

At the age of 3 and 4 years, in May, in the country the girl had suffocation attacks which were independently stopped when moving to the city. The real attack arose after consumption of chocolate. The emergency doctor held the emergency events. The attack is stopped. The asset is transferred to the district doctor.

At survey: moderately severe state. Pale integuments, blue under eyes. On cheeks, behind ears, in natural folds of hands and legs dryness, peeling, raschesa. "Geographical" language, perleches in mouth corners. Breath whistling, audible at distance. The exhalation is extended. ChD – 28 in 1 min. Over lungs a percussion sound with a box shade, auskultativno: a lot of dry rattles on all surface of lungs. Heart borders: right – on 1 cmof a knutra from the right edge of a breast, left – on 1 cmof a knutra from the left average and clavicular line. Tones are muffled. ChSS – 72 beats/min. Soft, painless stomach. A liver + 2 cmfrom under edge of a costal arch. The spleen is not palpated. The daily chair issued.

General blood test: Ayr - 4.3xl012/l, Hb - 118 g/l, Leyk - 5.8x 109/l, p.b. - 1%, with - 48%, e/f - 14%, l/c - 29%, m/c - 8%, SOE - 3 mm/hour.

General analysis of urine: quantity – 100.0 ml, relative density – 1016, there is no slime – leukocytes – 3-2-3 in p/z, erythrocytes do not.

Roentgenogram of a thorax: pulmonary fields of the increased transparency, strengthening of the bronchopulmonary drawing in radical zones, there are no focal

shadows.

Task:

1. Your diagnosis? Justification of the diagnosis.
2. Etiology of this form of a disease?
3. Urgent actions necessary in this case?
4. Appoint the treatment necessary in the mezhpristupny period.

Task No. 2

The boy of 8 years, came to hospital with complaints to the complicated breath.

From the third pregnancy (children from the first and second pregnancy died in the neonatal period of intestinal impassability).

It is sick since the birth: constant cough was noted, on the first year of life transferred pneumonia three times. In the next years it was repeatedly hospitalized with complaints to high temperature, an asthma, cough with difficult separated phlegm,

At receipt condition of the boy very heavy. Body weight of 29 kg., height of 140 cm. Pale integuments, cyanosis of a nasolabial triangle, Are expressed symptoms of "hour glasses" and "drum sticks". ChD - 40 1 minute, ChSS - 120 beats/min. ABP of 90/60 mm Hg. Thorax of a barrel-shaped form. A percussion sound over lungs with a timpanichssky shade. Auskultativno: on the right breath is weakened, at the left – rigid. Mixed damp and dry rattles are listened, it is more at the left. Cardiac sounds are muffled, systolic noise on a top of weak intensity. The liver on 5-6 cmacts from under edge of a costal arch. The spleen is not palpated. A plentiful chair, with a greasy luster, zamazkoobrazny.

Complete blood count test: Ayr - 3.5Õ1012/l, Nv - 100 g/l, C. the item - 0.85, Leyk - 7.7x10%, p.b. - 8%, with - 54%, e - 3%, l - 25%, m - 10%, SOE of-45 mm/hour,

Biochemical analysis of blood: crude protein - 60 g/l, albumine - 46%, an alpha 1 globulins - 9%, an alpha 2 globulins - 15%, beta глобулины10.5%, gamma-globulins - 19.5%, thymol turbidity test - 9.0, SRB ++, SF - 850 Ud/l (norm - 220-820), ALT - 36 Pieces/l, ACT - 30 Pieces/l.

Pilokarpinovy test: sodium - 132 mmol/l, chlorine - 120 mmol/l.

Koprogramma: large amount of neutral fat.

Roentgenogram of a difficult cell: strengthening and sharp bilateral deformation of the

bronkhososudisty drawing, mainly in radical zones, dense fibrous tyazh. In the field of an average share on the right considerable lowering of transparency. Expansion of a cone of a pulmonary artery, "pendulous heart" is noted.

Ultrasonography of abdominal organs: the liver is increased at the expense of the left share, is condensed, non-uniform, the vascular drawing on the periphery is grown poor, moderate growth of connective tissue; a pancreas – 15x8x25 mm. it is increased, it is diffuzno condensed, has indistinct contours (gases); a gall bladder of a S-shaped form, with dense walls; the spleen is increased, condensed, walls of vessels dense, a splenic vein of the izvit.

Task

1. Give an assessment of the given laboratory and tool methods of a research.
2. Make the plan of further inspection of the child.
3. Formulate the diagnosis to this patient.
4. What etiology and pathogenesis of a basic disease?
5. Appoint to the patient treatment.

Test control

1. What type of allergic reaction is characteristic of an atopic form of bronchial asthma:

a) The I type (immediate)

b) The II type (cytotoxic)

c) The IV type which is (slowed down)

2. What type of allergic reaction is characteristic of an infectious and allergic form of bronchial asthma:

a) The I type (immediate)

b) The II type (cytotoxic)

c) The IV type which is (slowed down)

3. Bronchial asthma is:

a) chronic inflammation of airways

b) chronic inflammation of airways against the background of hyperreactivity of bronchial tubes

c) the chronic inflammation of airways against the background of hyperreactivity of bronchial tubes which is shown suffocation attacks, the asthmatic status or respiratory discomfort

4. At irritation of β-adrenoceptors $_2$of bronchial tubes:

a) extend

b) are narrowed

c) do not change

5. The hyperreactivity of bronchial tubes is observed at:

a) to blockade of β-adrenoceptors$_2$

b) to blockade of α-adrenoceptors

c) hypoxias

d) passive huts

6. Treat immune forms of bronchial asthma

a) infectious and allergic

b) atopic

c) "aspirinovy"

d) neurogenetic

e) asthma of physical tension

e) mixed

7. The clinic of typical bronchial asthma is:

a) expressed attacks of suffocation

b) persistent spastic cough

c) acute emphysema of lungs

d) asthmatic bronchitis

e) allergic bronchitis

8. The Predpristupny period of bronchial asthma is characterized:

a) irritability

b) the whistling rattles

c) itching of a nose

d) the forced position of a body

e) expiratory asthma

e) dacryagogue

g) sensation of fear

h) painful dry cough

i) expectoration of a phlegm

9. The criminal period of bronchial asthma is characterized:

a) irritability

b) the whistling rattles

c) itching of a nose

d) the forced position of a body

e) expiratory asthma

e) dacryagogue

g) sensation of fear

h) painful dry cough

i) expectoration of a phlegm

10. The Poslepristupny period of bronchial asthma is characterized:

a) irritability

b) the whistling rattles

c) itching of a nose

d) the forced position of a body

e) expiratory asthma

e) dacryagogue

g) sensation of fear

h) painful dry cough

i) expectoration of a phlegm

11. Possible complications during an attack of bronchial asthma are:

a) deformation of a thorax

b) atelectasis of lungs

c) pneumosclerosis

d) hypodermic emphysema

e) chronic pulmonary heart

e) acute heart failure

g) asphyxial syndrome

12. Treatment of the child in an attack of bronchial asthma includes (define the sequence of actions):

- to air the room
- intravenously by drop infusion Euphyllinum
- to calm the child
- intravenously Prednisolonum
- salbutamol aerosol

13. Euphyllinum intravenously enter in a look ____ solution %.

14. Apply to prevention of an attack of bronchial asthma:

a) Suprastinum

b) intal

c) salbutamol

d) astafen

e) Theophedrinum

e) Ketotifenum

15. In the poslepristupny period of bronchial asthma in complex treatment appoint:

a) hypoallergenic diet

b) hardening

c) Euphyllinum

d) massage of a thorax

e) vitamin B_6

e) vitamin E

g) specific desensitization

h) LFK

16. Allergic rhinitis is characterized:

a) sudden emergence

b) high temperature

c) sneezing

d) general malaise

e) vomiting

e) loss of appetite

g) in pale cyanotic color mucous at a rinoskopiya

h) neutrocytosis in peripheral blood

17. Allergicheky laryngitis is shown:

a) the "barking" cough

b) aphonia

c) high fever

d) neutrocytosis in blood

e) the accelerated SOE

e) inspiratory asthma

18. Radiological in an allergic bronchitis it is noted:

a) increase in transparency of pulmonary fabric

b) high standing of a diaphragm

c) expansion of borders of heart

d) strengthening of the vascular drawing

e) perivascular infiltration

19. Carrying out preventive inoculations to children with respiratory allergoses:

a) without restrictions

b) with preliminary preparation

c) it is forbidden

20. Has to carry out observation of children with respiratory allergoses:

a) local pediatrician

b) allergist

c) pulmonologist

d) cardiologist

e) ENT specialist

Class in a subject:

"RHEUMATISM AT CHILDREN"

I. Scientific and methodical justification of a subject.

According to WHO data, about 30% of all diseases of adults are the share of group of rheumatic diseases, every 10th disabled person suffers from one of these diseases. Diseases of this group quite often begin at children's and youthful age, are difficult for early diagnostics, proceed heavier, than at adults. Early diagnosis of these diseases and the corresponding therapy have basic value for the forecast of a disease and the patient's life.

II. Purpose of activity of students.

The student has to know:

- the main questions of an etiology and pathogenesis of rheumatism, the contributing factors leading to disturbance of an immunogenesis;
- morphological changes of connective tissue in rheumatism;
- features of a course of rheumatism at children;
- clinical, laboratory and tool and graphic diagnostic criteria of rheumatism;
- classification of rheumatism;
- the basic principles of treatment and prevention of rheumatism at children.

The student has to be able:

- to purposefully collect the anamnesis and to perform objective examination of the child;
- to reveal diagnostic and differential and diagnostic criteria and also possible etiological and pathogenetic mechanisms of a disease;
- to carry out the differential diagnosis of rheumatism with infectious and allergic myocarditis, a septic endocarditis, a pseudorheumatism;
- to make the diagnosis according to the existing classification, to establish a degree of activity and degree of a circulatory unefficiency;
- to make the plan of medical and preventive actions (primary and secondary).

III. Content of training:

1. Rheumatism etiopathogenesis. Features of rheumatism at children.
2. The main pathomorphologic changes in rheumatism.
3. Clinical manifestations and diagnostic criteria of rheumatism (the main, additional).
4. Classification of rheumatism.

5. Clinical signs of rheumatic heart diseases (insufficiency of the mitral valve, stenosis of an atrioventricular opening, insufficiency of the aortal valve, etc.).
6. Characteristic and outcome of a long and sharp course of rheumatism.
7. Diagnostic criteria of degrees of a circulatory unefficiency.
8. Clinical and laboratory, morphological indicators of activity of rheumatic process.
9. Treatment in rheumatism of various degree of activity.
10. Treatment of a circulatory unefficiency. Performing treatment in a circulatory unefficiency at children I, II, III St.
11. Differential diagnosis of a rheumatic carditis and nonspecific carditises, septic endocarditis, congenital heart diseases, functional cardiac disturbances.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.
3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.
4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.
5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.
6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. Call AFO of bodies of blood circulation at children.
2. Define heart borders at children of different age groups.
3. What features of immunity and nonspecific reactivity at children.
4. Provide the scheme of an etiopathogenesis of rheumatism.
5. List the main pathomorphologic changes in rheumatism.

VII. List of questions for check of final level of knowledge:

1. Features of rheumatism at children.
2. Call diagnostic criteria of rheumatism (the main, additional).
3. Classification of rheumatism.
4. Call clinical signs of rheumatic malformations of heart (insufficiency of the mitral valve, a stenosis of an atrioventricular opening, insufficiency of the aortal valve, etc.).
5. Characteristic and outcome of a long and sharp course of rheumatism.
6. Call exudate symptoms in a pericardium cavity.
7. Diagnostic criteria of degrees of a circulatory unefficiency.
8. Clinical and laboratory, morphological indicators of activity of rheumatic process.
9. To call treatment in rheumatism of various degree of activity. Cardiac glycosides, dosages at children's age and symptoms of initial toxic action.
10. Treatment of a circulatory unefficiency. Performing treatment in a circulatory unefficiency at children I, II, III St.
11. Differential diagnosis of a rheumatic carditis and nonspecific carditises, septic endocarditis, congenital heart diseases, functional cardiac disturbances.
12. Symptoms of rheumatic polyarthritis at children (clinical, laboratory).
13. Call symptoms and the applied tests of early diagnostics of a chorea.

14. List clinical and tool and graphic signs of a carditis.

15. Call forms of damage of skin in rheumatism.

16. Plan of treatment: at a sharp, long, continuous recurrent course of rheumatism at children.

Information block.

ACUTE RHEUMATIC FEVER

ORL – one of current problems of clinical medicine in general and pediatrics in particular in which the interests of cardiologists, immunologists, surgeons, etc. intertwine. Having begun in the childhood, rheumatism increases number disabled among adult population.

Etiology and pathogenesis.

Most of authors attach to a β-hemolytic streptococcus of group A crucial importance in a rheumatism etiology. In favor of the streptococcal theory developing of rheumatism in 2-8 weeks after a streptococcal infection, high antiserum capacities to a streptococcus in blood of patients in an active phase, discharge from blood at 60-75% of patients of streptococcal antigen, sharp decrease in incidence and a recurrence testify at a bitsillinoprofilaktika. A recurrence of rheumatism arises 2.5 times more often at persons with an adenoid disease, than without it. Latent and chronic forms of rheumatism are explained with transformation of a streptococcus into casing-free L-forms now; the last are found after a long-term bitsillinoprofilaktika, persistirut in bodies and fabrics as do not give in to phagocytosis and are resistant to antibiotics.

Now recognize value of social and hygienic conditions, home contacts, i.e. a streptococcal environment. Patients with rheumatism are a streptococcus nursery, relatives of the children sick with rheumatism more often have an adenoid disease, than in population. Rheumatism arises at persons with genetic insufficiency, i.e. it is a disease of hereditary predisposition. A polygenic mode of inheritance at which along with genetic a part is played by environmental factors.

Not less disturbance of an immunological homeostasis, i.e. rheumatism – a kliniko-immune disease is important. The streptococcus influences an organism the toxins (streptolysin - Oh, DNA-ase, proteinase, hyaluronidase, streptokinase) which have

properties of antigen, and to them are produced antibodies (antistreptolysin Oh, of an antistreptogialuronidaz, etc.). At sensibilized children with an adenoid disease the new exacerbation of a streptococcal infection leads to accumulation in the increased quantity of cell-bound immune complexes (streptococcal antigen + an antibody + a complement). Circulating in the vascular system, they are fixed in a wall of vessels of a microcirculation and damage them. The last facilitates intake of antigens and proteins in connective tissue, promoting its destruction (allergic reactions of immediate type). Because of community of the antigenic structure of a streptococcus and connective tissue of heart (phenomenon of a molecular mimicry) the immune responses in covers of heart damage them with formation of autoantigens and autoantibodies. Autoantigens have high specificity and big destructive action on an endomyocardium, than one streptococcal antigen. Autoantibodies in rheumatism are called anti-cardial antibodies (AKA). Immunnokompleksny reaction leads to chronic inflammation of heart.

In rheumatism also cell-mediated immunity suffers, at the same time the clone of the sensibilized lymphocytes killers bearing on themselves the fixed antibodies to a cardiac muscle and endocardium and damaging them (allergic reactions of the slowed-down type) is formed. Reliable increase in number of V-lymphocytes, T lymphocytes and T-helper inductor cells (CD4), reduction of percent of T-suppressor cells (CD8) is revealed that demonstrates immunoregulatory deficit.

Clinical patomorfologiya.

In rheumatism the damage of a microcirculation (vasculitis), its connective tissue structures is of particular importance (disorganization) after what, pathological reactions in connective tissue of heart, lungs, a liver, joints, central nervous system and other bodies with the subsequent involvement of parenchymatous elements are developed.

A.I. Strukov allocated 4 stages of disorganization of connective tissue:

1) mucoid swelling,

2) fibrinoid swelling,

3) granulematozny,

4) sclerous.

Discharge of a stage of mucoid swelling as at early the begun treatment involution of pathological process is possible is important for clinical physicians.

At fibrinoid swelling there is deeper destruction of connective tissue, and, therefore, the probability of involution is less. Both stages always demonstrate active rheumatic process and morphologically are expressed by a nonspecific exudative component that is most characteristic of a children's organism. Weight of clinical manifestations of rheumatism (myocarditis, polyserosites, a chorea) is connected with extent of development of a nonspecific exudative component. Ashof-Talalayevsky granulomas are found in pristenochny and valve endocardium, a pericardium, walls of vessels and connective tissue formation of other bodies. A granuloma development cycle – 3-4 months. The granulematozny reaction localized mainly in heart corresponds in clinic to a latent course of rheumatism.

Classification.

The most convenient doctor in daily practice the classification of rheumatism accepted on a symposium of All-Union scientific organization of rheumatologists in 1964, allowing to estimate all possible manifestations of rheumatism is. The active phase is defined by activity (maximum, moderate, minimum). It is possible to speak about an inactive phase not earlier than in 6 months after disappearance of clinical and laboratory signs of activity of process as morphological changes remain much longer, than clinical laboratory.

The course is defined by reactivity of the patient, features of the beginning and a course of the disease, its duration are considered.

– *sharp:* bright, rough clinical manifestations, a polisindromnost, laboratory indicators reflect high activity of process, positive dynamics of a disease within 2-3 months, heart disease is formed less often, such course meets more often in primary rheumatism;

– *subacute:* development of clinical symptoms more slowly, is less bent to a polisindromnost, duration of a disease of 2-6 months, the effect of antirheumatic therapy is less significant, heart disease is more often formed;

– *long and sluggish*: the torpid course, a disease lasts more than 4-6 months, without the significant aggravations, but also without full remissions. As a rule, it is a rheumatic carditis with moderate or minimum activity, heart disease, despite the carried-out treatment is often formed.

– continuous and recurrent: the heaviest course, meets more often at children of advanced age and is characterized by bright aggravations, a polisindromnost; under the influence of treatment there occurs incomplete remission with the subsequent deterioration;

– latent – is not present, and was not in the past of an active phase, there is no rheumatic anamnesis, heart disease is defined at once, insufficiency of the mitral valve is more often.

WORKING CLASSIFICATION OF RHEUMATISM

Phase	**Kliniko-anatomichesky characteristic of defeat**		***Course***	***TAX CODE***
	Hearts	***Other systems and bodies***		
Active ***Activity*** of I, II, III degrees	a) primary rheumatic carditis; b) rheumatic carditis returnable (without defect of valves, with defects of valves); c) rheumatism without obvious changes	Polyarthritis, serosites (pleurisy, peritonitis, abdominal syndrome), chorea, encephalitis, encephalomeningitis, cerebral vasculites, nephrite, hepatitis, pneumonia, damages of skin, iritis, iridocyclitis, thyroiditis	Sharp, subacute, long and sluggish, continuous and recurrent, latent	$_{about}$There is no N - N $_{1}$- the I St. N-II $_{II}$of St. H-III $_{III}$of St.
Inactive	a) (what) heart disease; b) rheumatic myocardiosclerosis	Consequences and the residual phenomena of the postponed extracardiac defeats		

DIAGNOSTIC CHARACTERS OF RHEUMATISM

KISELYA-DZHONSA-NESTEROVA

The main	***Additional***
• Carditis • Polyarthritis • Chorea • Rheumatic small knots • Anulyarny rash • Communication with the postponed streptococcal diseases • Efficiency of antirheumatic therapy	• Fever • Arthralgias • Leukocytosis, increase in SOE • Lengthening of an interval SOLUTION on the ECG • Serological and biochemical indicators • Hyperpermeability of capillaries • Fatigue, abdominal pain, nasal bleedings

A number of authors consider that the diagnosis of rheumatism is probable at two main or one basic and two additional criteria. Laboratory indicators remain not specific, reflecting weight of any infectious process.

PRIMARY RHEUMATISM

The sharp or subacute course is peculiar to primary rheumatism. In 2-3 weeks after a nasopharyngeal infection (SARS, a tonsillitis, pharyngitis), less often without it the first symptoms of rheumatism appear. It is possible to call this stage the period of harbingers as after a tonsillitis remain slackness, an indisposition, complaints to a headache, increased fatigue, malassimilation of a training material, perspiration, nasal bleedings, subfebrile condition remains. At children it is necessary to refer sudden temperature rise, symptoms of intoxication, arthralgia or polyarthritis to initial manifestations of rheumatism. Changes of joints meet at 1/2 - 1/3 patients with primary rheumatism. The expressiveness of this sign gave the grounds to doctors of last years to call a disease of "acute articulate rheumatism". In rheumatic polyarthritis generally large and average joints (knee, talocrural, elbow), usually symmetric are surprised. The flying defeat migrating – in 1-7 days different joints are covered, one joint is less often changed. Joints the swelled-up, their contours are maleficiated, passive and active movements are sharply limited, skin

over them is hyperemic, to the touch they are hot. However, despite sharpness of the defeat caused by serous inflammation under the influence of treatment the joint syndrome quickly disappears, all changes take place completely. The increased temperature sticks to 3-5 days, it is normalized together with fading of an articulate syndrome.

Along with damage of joints come to light and become conducting symptoms of a rheumatic carditis. In a rheumatic carditis three areas are involved in process, and depending on prevalence of any given clinical symptoms it is possible to speak about primary involvement of one of them, but not about the isolated endocarditis, myocarditis and a pericarditis. Weight of clinical manifestations is defined by damage of a myocardium. Myocarditis can be more or less diffusion that depends on a nonspecific exudative component.

In pronounced myocarditis the serious general condition of children, pallor of integuments, an asthma, pains in heart, heartbeat are noted. The apical beat is weakened, limits of warm dullness are expanded, it is more to the left, tones are muffled, often there is bradycardia till 50-60 1 min. or tachycardia, arrhythmia in the form of premature ventricular contraction is seldom listened.

Systolic noise in rheumatic myocarditis can be various. It is soft, not rough noise with a maximum in the 5th point and on a top, without carrying out zone (noise of dysfunction, hypotonia of papillary muscles) or the "emission noise" on the basis of heart connected with change of speed of a blood-groove, pulmonary pressure, change of viscosity of blood, increase in its volume, compensatory increase of force of reductions of a myocardium of ventricles. The intensity of such noise decreases upon transition to vertical position or at physical activity. On FKG these noise differ from organic in the fact that they are not connected with 1 tone, change the form and amplitude in different cardiocycles. Also noise of relative insufficiency of MK is possible. In diagnosis of myocarditis the dynamism of auskultativny symptoms against the background of treatment is of great importance.

To diffusion damage of a myocardium in a clinical picture heart failure in the form of an asthma can accompany, to tachycardia, increases in a liver.

On the ECG the following changes are possible:

a) disturbance of rhythmic activity in a look takhi- or bradycardia, migration of a pacemaker, atrioventricular dissociation, is more rare – premature ventricular contraction;

b) disturbance of atrioventricular conductivity in the form of lengthening of an interval SOLUTION which arises at 10-30% of patients with rheumatism;

The increased lability of the autonomic nervous system with a vagotonia (quickly passing lengthening of R-Q) or defeat by inflammatory process of atrioventricular connection can be the cause of this phenomenon (keeps is longer).

c) changes of a tooth of T are insignificant in moderate myocarditis, at the same time it can be a little smoothed or high, pointed (vagotonia) in the left chest assignments. Against the background of treatment positive dynamics is noted.

d) Decrease in a voltage of the QRS complexes meets infrequently and testifies to weight of process.

On FKG it is possible to find:

a) reduction of amplitude of 1 tone on a top that SOLUTION can be connected both with damage of a myocardium, and with lengthening of an interval;

b) increase in amplitude of 3 and 4 tones;

c) functional or organic systolic noise (connected with 1 tone, mid-frequency).

Radiological inspection informatively in brightly or moderate carditis (increase in a cavity of a left ventricle, decrease in amplitude of a pulsation of his myocardium) has also no diagnostic value at small activity.

At insufficiency of the mitral valve the weakened 1 tone, the rough, blowing systolic noise with a maximum on a heart top are listened, begins along with 1 tone, is well carried out to the left underarm, and is frequent also on a back; the noise melody slightly changes upon transition to a standing position. On FKG it is high-frequency apex murmur, occupies 2/3 systoles or all systole, has various amplitude, decreasing, taenioid is more rare, it is connected with 1 tone. At considerable mitral regurgitation on a top of heart the mesodiastolic noise connected with a relative stenosis of MK is listened; during a diastole the large volume of blood equal to arrived normal on pulmonary veins, and additional volume because of insufficiency of MK returns to a left ventricle from the left auricle; it causes discrepancy of the sizes of an antrioventrikulyarny ring with amount of

the proceeding blood. On the ECG there can be signs of the increased bioelectric activity of a myocardium of a left ventricle.

At many patients at X-ray inspection the heart of a mitral configuration decides on increase in both left heart cameras 1-2 degrees, the dynamism of these changes against the background of treatment is characteristic.

At defeat of the aortal valve the flowing protodiastolic noise along the left edge of a breast, as much as possible in the third or fourth mezhreberye is listened at the left. On FKG the noise is connected with the 2nd tone, has the fading form, occupies all diastole or 2/3 it. On the roentgenogram the aortal configuration of a warm shadow is possible.

Clinically in a rheumatic carditis most less often the pericardium is involved in inflammatory process. The pericarditis testifies to a pancarditis. As a rule, the pericarditis is combined with involvement of a pleura. Clinically there are complaints to sharp pains in heart, dry and persuasive cough, children hold in a bed the forced sitting position with an inclination forward. In a dry shaggy pericardium over all area of heart or over any one site the pericardial rub reminding a snow crunch is listened (noise is listened in a systole and a diastole, it is not connected with cardiac sounds). It can be non-constant, be listened only several hours, disappear and again appear.

At accumulation of an exudate in a cavity of a pericardium it is observed:

a) disappearance or reduction of earlier noted pulsation in heart and pericardial rub;

b) fast increase in heart;

c) rapprochement of limits of relative and absolute dullness;

d) change of a configuration of a warm shadow in the form of a trapeze on the roentgenogram;

e) decrease in a voltage of the QRS complexes, rise is higher than the isoline of a segment of QT with deformation of a tooth of T on the ECG.

At the same time weight of a state increases, appear swelling of cervical veins, an asthma, the patient holds the forced position. There can be abdominal pain, hypostases, the liver becomes big, dense, painful. The serous exudate accompanying the most acute forms of rheumatism usually is not plentiful and quickly resolves. Fibrinous exudate resolves slowly, being exposed to the organization (cavity obliteration). Detection on roentgenograms of illegibility, roughness, deformation of contours of a shadow of heart

and diaphragm at 40% of patients with primary rheumatic carditis demonstrates early involvement in process of serous covers which clinically in due time is not distinguished.

Besides damage of heart and joints, in rheumatism **the central nervous system** (chorea, an encephalomeningitis, encephalopathy, a neurorhematism) is involved in process. Sydenham's chorea most often meets at children (in 11-13% of cases of primary rheumatism). Usually children of 5-10 years are ill.

At an objective research it is possible to establish **a typical triad of symptoms**:

1) involuntary distal wide hyperkinesias;

2) hypomyotonia;

3) ataxia.

At the same time there are manifestations of vegetative dysfunction and psychopathological lines. In the neurologic status are noted increase in tendon jerks, especially knee (a positive reflex of Gordon), the Common people symptom (retraction of a front wall of a stomach during an exhalation), instability in Romberg's pose (simple and complicated), negative paltsenosovy, calcaneal and knee tests, positive symptoms of "flabby shoulders", "an eye and Filatov's language" (the patient cannot long hold eyes densely screwed up, twitching of the put-out tongue is noted), etc.

Involution of symptoms occurs within 1.5-3 months. The chorea recurs, and on its background the heart disease is often formed. At the same time the choreic phenomena pass completely, but can is long to remain an asthenic syndrome.

Annulyarny rash and rheumatic small knots belong to more rare symptoms of rheumatism. Annulyarny rash in rheumatism represents roundish pinkish-red spots, in the center pale also are located more often on the side surfaces of a thorax, stomach, the internal surface of shoulders, hips, a neck, a back. Rash develops quite often at the very beginning of a disease, keeps not for long (therefore it is often passed), disappears completely (without pigmentation and peeling). It meets also at other states (adenoid disease, a SARS) therefore the diagnostic value of it signs is small, it matters only in combination with other symptoms of rheumatism.

Even less often now it is possible to see rheumatic small knots which were found at a heavy, continuous and recurrent course of rheumatism earlier. Small knots represent the

roundish dense, single or multiple, painless formations of 2-8 mm. in size which are located in sinews, aponeuroses, remaining from several days to several months.

Sometimes rheumatism begins with an abdominal syndrome. The abdominal syndrome is result of sharply arisen serous inflammation of a peritoneum (vasculitis) with the subsequent aseptic exudative peritonitis, it is more rare it is caused by irradiation of pains from bodies of a chest cavity (pleurisy, a pericarditis) or connected with liver capsule stretching (rheumatic hepatitis, a stagnant liver). The abdominal syndrome completely passes within several hours or days against the background of antirheumatic therapy.

Other rare form of a rheumatic polyserositis is pleurisy (dry and exudative). Pleurisy is often combined with a pericarditis, usually not heavy and quickly passes at treatment. The same positive dynamics is undergone also by the rheumatic pneumonia interstitial on character.

Damage of kidneys in rheumatism is expressed by an uric syndrome: the moderate proteinuria, a leukocyturia, is more rare an erythrocyturia. These changes benign are also caused by involvement in process of tubulointerstitsialny fabric. Extremely seldom there is rheumatic nephrite.

Besides clinical signs, also characteristic changes of laboratory indicators are noted. At active rheumatic process of SOE makes more than 30 mm/h, but pretty fast decreases against the background of hormonal therapy. In a blood count the shift is noted to the left (increase in quantity of stab neutrophils). The quantity of leukocytes can be increased. The hemoglobin content and a color indicator are reduced. In analyses of urine the quantity of erythrocytes can be 10-12 and in rare instances 50-80 under review (nephrite); the amount of protein lost with urine, as a rule, insignificant also corresponds to weight of a rheumatic infection, perhaps moderate leukocyturia. Immunobiochemical analyses of blood: increase in a caption of ASL-O, of ACK, ACH (norm 1:250-500), indicators of difenilaminovy reaction (norm up to 0.210 units of optical density), seromucoid (norm up to 0.180 units), detection of S-reactive protein (normal is absent), raised appears fibrinogen level (normal up to 9.2 μmol/l) at decrease in fibrinolytic activity of blood. Changes of protein fractions: decrease in albumine, increase α_2- and β- globulins are inherent to active rheumatic process.

RETURNABLE RHEUMATISM

Exacerbation of rheumatism is demonstrated by again appeared symptoms of intoxication, temperature rise or subfebrile condition, a joint syndrome (there can be both arthralgias, and polyarthritis), chorea symptoms, annulyarny rash can recur. With each new attack of rheumatism the noncardiac manifestations will become less bright, but all of them demonstrate process activization. Repeated it is possible to consider the attack which arose not earlier than in 10-12 months. At aggravation in earlier terms against the background of incomplete remission the rheumatism needs to be considered continuous and recurrent.

The crucial importance in returnable rheumatism is gained by changes of heart: expansion of limits of relative warm dullness, increase of dullness of cardiac sounds, emergence of new organic noise or strengthening of already available.

As a result of the repeated attacks of rheumatism at children heart diseases are formed. If after the first attack of floggings it is found only in 14-18% of patients, then after the second, third – in 100%. The recurrent carditis without forming of heart disease has to call the diagnosis of rheumatism into question.

Heart diseases should be divided into simple (the isolated defeats of one valve), combined (a stenosis and insufficiency of one valve) and combined (2 and 3 valves). On involvement frequency the mitral valve, on the second – aortal, on the third – their combination is on the first place, then – defects of the three-leaved valve (is more often with defeat of mitral or aortal valves, arises separately less often), seldom are surprised valves of a pulmonary artery. In a returnable rheumatic carditis the disease severity will be defined not only by activity of process, but also expressiveness of valve deformation. The last can be 1 degrees (insignificant), 2 degrees (moderate) and 3 degrees (sharp).

Treatment.

The modern rheumatology achieved indisputable success in treatment of RL. A big merit of domestic researchers was development of methods of the complex landmark treatment based on knowledge of the major etiopatogeneticheskiy features of this disease and providing:

– treatment of an acute period of a disease in a hospital;

– aftercare in local rheumatologic sanatorium;

– dispensary observation in a cardiorheumatologic office of policlinic.

This system passed test time and remains still.

The modern doctor has a powerful arsenal of antirheumatic means which appoint from the first day of establishment of the diagnosis of RL.

In view of distinct anti-inflammatory and giposensibiliziruyushy effect of steroid hormones, pediatricians unanimously come to conclusion that corticosteroids and in modern conditions are shown to children at RL with a distinct component of inflammation, i.e. in brightly and moderate carditis, at the maximum or moderate degree of activity of process at sharp less often – a subacute course of the disease.

At the minimum degree of activity or a latent course of RL hormonal drugs have no significant therapeutic activity. At these forms the use of non-steroidal anti-inflammatory drugs (NPVP) is more proved.

Considering a streptococcal etiology of RL, the principle of co-administration with hormones of a 10-14-day rate of penicillin or its analogs remains in force. In the presence of the multiple and often becoming aggravated infection centers the course of penicillin therapy is extended, and according to indications, patients already in a hospital are transferred to treatment by Bicillinum.

Along with bright positive effect of steroid hormones many authors note their side effect. Clinical signs of undesirable influence of corticosteroids are various and are expressed by tranzitorny increase in arterial blood pressure, an excess adiposity, a hypertrichosis, disturbance of a menstrual cycle, changes of skin (dryness, eels, a nevus pigmentosus, etc.), functions of nervous system and digestive tract, etc.

In therapy of primary rheumatic carditis with defeat of the valve device and also the RL long forms so far widely use drugs of a quinolinic row: delagil, plaquenil, etc.

In the last two decades in connection with the changed RL course at its treatment use only non-steroidal anti-inflammatory drugs more and more widely. One of the first in this disease began to apply indometacin.

In the last decade at RL are widely used sodium diclofenac (Voltarenum, betaren, etc.). Appoint at the rate of 2-3 mg/kg of body weight a day. A course of treatment – 1-1.5 months. Brufenum (ibuprofen) apply at RL at children in a daily dose 600-800 mg. for a long time.

Researches of therapeutic activity of various antirheumatic means allowed to establish that non-steroidal anti-inflammatory drugs at RL at children unlike adults considerably concede to hormonal therapy, especially in the presence of a carditis of various degree of manifestation. However this question is widely discussed.

At the severe forms of a disease proceeding with a pancarditis and a polyserositis the pulse therapy can be used by Methylprednisolonum.

The second important stage of complex recovery treatment of sick RL still should be considered local rheumatologic sanatorium. Its task is in that by use of the corresponding medical and motive mode and a number of therapeutic and preventive actions to achieve final subsiding of activity and full compensation of rheumatic process and also recovery of functional capacity of a cardiovascular system.

The third component of rehabilitation therapy is dispensary observation for the children who had a rheumatic carditis. It includes regular survey of the patient, continuation (in need of) anti-inflammatory treatment, purpose of all-improving actions, the tempering procedures, the dosed physical education, performing secondary prevention of a recurrence of a disease.

Prevention.

Being guided by theoretical concepts of emergence of RL, long experience of rheumatologists, the existing methodical instructions and also WHO recommendations (1989), the program of prevention of RL and fight against a disease recurrence, i.e. primary and secondary prevention was developed.

Primary prevention still covers two stages:

1. *Measures of the general plan.*
2. *Fight against a streptococcal infection.*

1. The measures of the general plan providing strengthening of health of children and teenagers:

- ensuring correct physical development of the child;
- hardening since the first months of life;
- the good vitaminized nutrition;
- maximum use of fresh air;
- rational physical education and sport;

- fight against density in dwellings, schools, child care facilities;
- performance of a wide complex of sanitary and hygienic rules.

2. Measures against a streptococcal infection:

- early diagnostics;
- the correct treatment of the upper respiratory tract infections caused by group A streptococcus.

The drama growth of streptococcal infections noted from the middle of the 80th years in the developed countries causes big concern in view of the possible growth of RL and RPS caused by it. Therefore the active measures including a number of the directions, first of all treatment of streptococcal infections are necessary.

Penicillin remains highly effective drug of the choice in a streptococcal infection (tonsillitis, exacerbation of an adenoid disease, scarlet fever). It is appointed on 750,000 PIECES/days to preschool children, by 1 000 000 – 1 500 000 Piece/days to patients of school age within 10-14 days or 5 first days with the subsequent administration of Bicillinum-5 in a dose of 750 000 - 1 50 000 PIECES/days twice with an interval of 5 days. At oral administration the dose of phenoxymethylpenicillin and its analogs makes: Oxacillinum, ampicillin – 500,000 - 750,000 PIECES/days to children of preschool age and 1 000 000 – 1 500 000 Piece/days of school.

According to WHO recommendations (1989) effective drug in treatment of a nasopharyngeal infection is oral acid resisting penicillin – phenoxymethylpenicillin (Ospenum) in the same doses. Optimum drug from this group should be considered amoxicillin today.

For the patients having the allergy accepted by an alternative erythromycin, a semi-synthetic makrolidny antibiotic azithromycin (sumamed) and roksitromitsin are.

Other antibiotics of a broad spectrum of activity, for example cephalosporins, can also effectively eliminirovat a streptococcus of group A of upper airways.

Development and questions of anti-streptococcal immunization, antistreptococcal vaccine still are in a discussion stage. The achievements in the field of molecular biology which allowed to have valuable additional information on group A streptococci give promise on creation of effective vaccine in the near future.

The new direction of primary prevention of RL is development of methods of forecasting of a disease. First of all, the concept of risk factors which is a basis for forecasting is developed

The secondary prevention directed to prevention of a recurrence and progressing of a disease at the children who transferred RL consists in regular administration of Bicillinum (penicillin of the prolonged action) which high efficiency is proved by long-term researches of domestic and foreign authors.

Optimum should consider the year-round prevention which is carried out monthly. It is appointed to all children who transferred RL within the next 5 years. It is connected with the fact that the greatest number of a recurrence fall on the first 5 years after the previous attack. However duration of secondary prevention, making not less than 5 years, for the children who had arthritis or a chorea without damage of heart is defined individually for each patient. So, the patient who transferred primary or returnable RL with damage of heart, especially with symptoms of the formed or created heart disease, year-round prevention has to be carried out up to achievement of 18-year age, and if necessary longer.

Year-round prevention carry out by means of Bicillinum-5 in a dose 1,500,000 Pieces once in 4 weeks to children of school age and teenagers. to preschool children Bicillinum-5 are entered in a dose of 750,000 Pieces of 1 times into 2 weeks.

Under WHO recommendations (1989), to the patients subject to high risk of recuring of rheumatic process, it is necessary to enter Bicillinum-5 1 time into 3 weeks in a dose of 1,500,000 Pieces to school students and 1 time in 10 days on 750,000 Pieces – children of preschool age. As perspective antibiotics of the prolonged action are considered also a benzatina benzillenitsillin – **retarpen** and new benzylpenicillin of a benzatin – **ekstentsillin** which well proved for secondary prevention of RL at the adult contingent of patients.

Secondary prevention can be carried out by daily reception of antibiotics (penicillin and its analogs) inside.

Besides the measures of secondary prevention stated above, at accession of acute respiratory infections, tonsillitis, pharyngitis, after tonsilectomies and other surgeries performing the current prevention (10-day rate of penicillin) is recommended to sick RL.

Scheme of inspection of the patient

When collecting the anamnesis to pay attention on:

- the patient's family tree (presence of rheumatic, streptococcal, allergic diseases in family and at the patient's relatives);
- features of the immune system (frequency of viral, bacterial infections, their course, the performed therapy and reactions to it, inoculations and inoculative reaction);
- allergic mood;
- chronic centers of an infection (antritis, tonsillitis, otitis, caries of teeth). Efficiency of treatment;
- domestic conditions (density), day regimen (stay in the fresh air);
- complaints, development of a disease, duration of separate symptoms (fever, its character, joint syndrome, etc.)

At objective survey to pay attention on:

- indicators of physical development, their dynamics;
- condition of nervous system (change of mentality, behavior, character, condition of cranial nerves, tendon jerks, focal symptomatology), vegetative disturbances;
- damage of heart (carditis, cardiosclerosis: calculation of pulse, measurement of the ABP, auskultativny and percussion data). Diagnosis of a circulatory unefficiency according to indications (use of functional trials);
- sizes of a liver, spleen (both manifestation of a visceral syndrome, and circulatory unefficiency), lymph nodes;
- condition of the musculoskeletal system: survey, a palpation, volume of passive and active movements, the number of the joints involved in process, existence of contractures, ankiloz, periartikulyarny changes, muscles (polymyalgias, a miositis, consolidation), skins (color, elasticity, dryness, turgor, rashes – character, duration).

At assessment of paraclinic methods of a research to pay attention on:

- in blood test on a leukocytosis, increase SOE, a disproteinemia, a hyperglobulinemia, hyper alpha-2-globulinemiyu, seromucoids, DFA, credits of streptococcal antibodies.
- the analysis of urine (a daily proteinuria, test on Nechiporenko, Addis-Kakovsky, across Zimnitsky, clearance of endogenous creatinine);
- X-ray inspection of bodies of a thorax (heart sizes, expansion of borders, symptoms of a pericarditis), joints (osteoporosis, ossifluence, destruction of a cartilage, ankiloza, thickening and flattening of an articulate bag);
- tool grafichesike researches: The ECG – disturbance of a rhythm, conductivity, change of teeth; FKG – easing, fragmentation and deformation of complexes, the sir - or protosystolic noise on a top, pericardial rub; EKHOKG – disturbance of sokratitelny ability of heart, a condition of the valve device, the sizes of cardial cavities and large vessels, etc.
- survey of the oculist, ENT specialist, neuropathologist, etc. experts.

Tasks for independent preparation:

1. Solve situational problems.
2. Make tasks of test control on a subject.
3. Examine and describe the patient with rheumatism. Leave the plan of inspection.
4. Write prescriptions in workbooks:
   a) Bicillinum 5
   b) azithromycin
   c) penicillin
   d) diclofenac
   e) Prednisolonum

Situational tasks.

Task No. 1

The boy I., 11 years, came to department by gravity.

From the anamnesis it is known that had scarlet fever 2.5 months ago (a typiform, moderate severity). Received antibacterial therapy. In a month it is written out in school. Then began to note changes of handwriting, the boy became restless, the progress at

school decreased, the tearfulness appeared. Soon mother began to notice at the boy of twitching of facial muscles, inaccuracy of movements when clothing and at meal time. Periodically temperature to subfebrile figures increased, the catarrhal phenomena were not. Saw a doctor, blood test in which changes are not revealed was made. The diagnosis was made: "Flu, asthenic syndrome". Received Oxacillinum within 7 days, without effect. Neurologic disorders accrued: grimacing manifestations amplified, the boy could not put on independently, the help sometimes was required at food, the tearfulness and irritability in this connection the patient was hospitalized remained.

At receipt serious condition. The boy is whining, irritable, quickly is tired, the speech skandirovannost, inexact performance of coordination tests, a hypomyotonia, grimacing is noted. In lungs vesicular breath sound, there are no rattles. The area of heart is visually not changed. Heart borders: right – on the right edge of a breast, upper – on the III edge, left – on 1 cm of a knutra from the average and clavicular line. Cardiac sounds are moderately muffled, not rough systolic noise on a top is listened, not carried out, in an ortostaza its intensity decreases. The soft stomach, is available to a palpation, the liver, a spleen are not increased.

Questions:

1. Prove and formulate the preliminary diagnosis.
2. What examinations should be performed to the patient.
3. Make the treatment plan.

Task No. 2

The boy V., 1 years 2 months, came to department with complaints to a loss of appetite, vomiting, loss of body weight, damp cough.

From the anamnesis it is known that till 1 year the child developed according to age, goes independently from 10 months. At the age of 11.5 months had the acute respiratory disease which was followed by the catarrhal phenomena and an abdominal syndrome (an abdominal pain, a liquid chair), subfebrile temperature was noted. The specified changes remained within 7 days.

In 2-3 weeks after recovery the parents noted that the child began to be tired quickly at physical activity during the games, an asthma was noted. The state gradually worsened:

periodically there were symptoms of concern and damp cough at night, vomiting, the appetite worsened, the boy lost in weight, the pallor of integuments attracted attention. Temperature did not increase. The state is regarded by the local pediatrician as manifestation of an iron deficiency anemia, the child is directed to hospitalization for inspection.

At receipt serious condition, the appetite is reduced, inactive. Integuments, pharynx light pink. Respiration rate 44 1 minute, in lungs single damp rattles in lower parts are listened. Area of heart: visually – a small warm left-side hump, palpatorno – an apical beat diffuse, the area is it about 8 cm2, is percussion – limits of relative warm dullness: right – on the right edge of a breast, left – on the front axillary line, upper – the II mezhreberye, auskultativno – ChSS - 140 beats/min, cardiac sounds are muffled, more the I tone on a top, is listened not rough timbre the systolic noise occupying 1/3 systoles, connected with the I tone in the same place. A soft stomach, a liver + 6 cmon the right average and clavicular line, a spleen + 1 cm. Free, painless urination.

General blood test*:* Hb - 110 g/l. Ayr - 4, l xl012/l, Leyk - 5.0xl09/l, p.b. - 2%, with - 56%, l - 40%, m - 2%, SOE - 10 mm/hour.

ECG: a low voltage of the QRS complexes in standard leads, sinus tachycardia to 140 in a minute, the alpha makes a corner -5 °. Signs of an overload of the left auricle and left ventricle. Negative teeth of T in I, II, aVL, V5, V6 assignments, RV5 <RV6.

Thorax X-ray analysis in a direct projection: the pulmonary drawing is strengthened. KTI - 60%.

EKHOKG: increase in a cavity of a left ventricle and left auricle, ejection fraction is 40%.

Task:

1. Prove and formulate the diagnosis.
2. Estimate the provided results of inspection.
3. What else examinations would you like to perform to the child?
4. Carry out the differential diagnosis.
5. Make the treatment plan of this child.

Task No. 3

Sick R., 9 years, came to a hospital with complaints to long subfebrile condition, weakness and fatigue, small appetite.

Anamnesis of a disease: these complaints appeared after removal of carious tooth 4 weeks ago. Parents did not see a doctor, conducted treatment independently febrifuges. However fever remained, the weakness and deterioration in health increased in this connection, the child was hospitalized.

Anamnesis of life: the girl were born from the first normally proceeding pregnancy, births in time, did not lag behind in physical and psychomotor development, at the age of 1 month the systolic noise with punctum maximum in the III-IV mezhreberye to the left of a breast was listened. After inspection the defect of an interventricular partition of the small sizes located in a hymenoid part subaortalno is diagnosed. Further the health of the girl remained good, symptoms of heart failure were not observed, did not receive treatment.

At receipt a condition of the patient heavy, it is very pale, sluggish, an asthma at rest to 28 in a minute is noted. In lungs vesicular breath sound, there are no rattles. The area of heart is visually not changed. At a palpation the apical beat diffuse and strengthened, is located in the IV-V mezhreberye on 2 cmof a knaruzha from the left average and clavicular line. In area III-IV of a mezhreberye the systolic trembling, diastolic trembling in the II-III mezhreberye to the left of a breast is defined at the left. Heart borders at percussion: right – on the right edge of a breast, upper – in the II mezhreberye, left – on 2 cmof a knaruzha from the average and clavicular line. At auscultation: in the III-IV mezhreberye to the left of a breast the systolic noise connected with the I tone and occupying 3/4 systoles is listened rough, the scraping timbre; noise is carried out practically it is necessary all area of heart. In the II-III mezhreberye to the left of a breast the protodiastolic noise which is carried out along the left edge of a breast is listened. In the II mezhreberye at the left – accent of the II tone. Heart rate is 100 beats/min. ABP of 115/40 mm Hg. The soft stomach, is available to a deep palpation, the liver acts on 3 cmfrom under edge of a costal arch on the right average and clavicular line.

General blood test: Ayr - 4, lxl012/l, Hb - 105 g/l, Leyk. – 12.0x109/l, p.b. - 7%, with - 37%, e - 3%, l - 50% of m - 3%, SOE - 40 mm/hour.

General analysis of urine: specific weight-1018, protein - 0.05 ‰, leukocytes - 2-3 in p/z, erythrocytes - are absent.

ECG: sinus tachycardia, normal provision of an electrical axis of heart, signs of an overload of the right and left ventricles.

Task:

1. Prove and formulate the preliminary diagnosis.
2. Call the main clinical criteria of this disease including not provided at the patient.
3. What else examinations does this patient need to perform? What their expected results?
4. Make the treatment plan of the patient.

Class in a subject:

"JUVENILE PSEUDORHEUMATISM"

I. Scientific and methodical justification of a subject:

According to WHO data, diffusion diseases of connective tissue occupy one of the leading places in structure of incidence adults, every 10th disabled person has this pathology. Diseases of this group quite often begin at children's and youthful age, are difficult for early diagnostics, proceed heavier, than at adults. Early diagnosis of these diseases and the corresponding therapy have basic value for the forecast of a disease and the patient's life.

II. Purpose of activity of students.

The student has to know:

- main diagnostic criteria and features of a course of the juvenile pseudorheumatism (JP);
- principles of diagnosis of YuRA;
- treatment and preventive actions at YuRA.

The student has to be able:

- to resolve diagnostics issues on the leading clinical and laboratory syndromes;
- to define a degree of activity of autoimmune process, the individual forecast;
- to prove the diagnosis;

- to make the treatment plan;
- to plan preventive actions, to define the forecast.

III. Content of training:

1. Main concepts of pathogenesis of diffusion diseases of connective tissue.
2. Nosological forms of diffusion diseases of connective tissue.
3. Kliniko-morfologichesky changes in diffusion diseases of connective tissue.
4. The leading clinical and laboratory syndromes in diffusion diseases of connective tissue.
5. Principles of therapy and prevention of diffusion diseases of connective tissue.
6. Differential diagnosis of inflammatory diseases of joints (rheumatic, YuRA, infectious and allergic).
7. "Big" and "small" diagnostic criteria of a juvenile pseudorheumatism.
8. Working group of the YuRA clinical forms at children's age.
9. Principles of treatment of YuRA.
10. Features of a course of diffusion diseases of connective tissue.
11. Principle of treatment of these diseases and prevention.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.

3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.
4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.
5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.
6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.
7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.
8. Lectures on pediatrics.
9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. The contributing factors in development of rheumatic diseases (rheumatism, hard currency, YuRA) at children.
2. State the main concepts of pathogenesis of diffusion diseases of connective tissue.
3. Give definition and call nosological forms of diffusion diseases of connective tissue.

VII. List of questions for check of final level of knowledge:

1. Call kliniko-morphological changes in a scleroderma, a system lupus erythematosus, a dermatomyositis, a nodular periarteritis.
2. Determine a degree of activity of autoimmune process in diffusion diseases of connective tissue by the leading clinical and laboratory syndromes.
3. List differential and diagnostic criteria of inflammatory diseases of joints (rheumatic, YuRA, infectious and allergic).
4. Call features of a course of diffusion diseases of connective tissue at children's age.
5. Define medical tactics in diseases of connective tissue and a measure of their prevention.
6. Call "big" and "small" diagnostic criteria of a juvenile pseudorheumatism.
7. List the basic principles of therapy of YuRA at children.

Information block.

Juvenile Pseudorheumatism (JP)

represents the chronic inflammatory disease of joints with not clear etiology and difficult pathogenesis leading to gradual destruction of joints and which is combined at a number of patients with the significant extraarticular manifestations.

Etiology and pathogenesis.

YuRA belongs to the multifactorial, poligenno inherited diseases in which development take place not only environmental and infectious, but also hereditary factors, including immunogenetic. Among sick YuRA authentically more often than in population, A1, A28, B27, B40, DR5 antigens meet.

Clinical picture.

Clinical manifestations of YuRA are diverse and depend on the different reasons: age and sex of the child, factors which provoked a disease, the nature of further development of process.

In most cases the clinical picture of a disease is defined by damage of joints. The beginning of a disease can be slow, hardly noticeable, from appearance of weak pain and a swelling in any one joint, is more often knee or talocrural. Later 2-3 weeks or 1-2 months the pathological process develops in other symmetric joint. This symmetry of defeat is typical for YuRA. Besides pain and restriction of mobility of joints children test a general malaise, weakness, they reduce body weight, there is subfebrile condition, SOE increases to 20-25 mm/h. Further, in the absence of timely diagnostics and adequate therapy, other joints – radiocarpal, elbow, maxillary and temporal, small joints of a brush and foot, neck joints also can gradually be involved in process. At damage of 2-4 joints it is accepted to speak about an oligoarthritis. Involvement in process of 5 joints also more demonstrates development of polyarthritis. Such subacute course of process is usually observed at children 5 years, mainly at school students are more senior. At the children who got sick with YuRA at early age (1-4 years) the acute onset of a disease which is followed by fever, the profound arthralgias with involvement in process at once of 4 joints and more, including small joints of a brush and tables is more often observed. Against the background of fever they quite often have polymorphic enanthesis, increase in peripheral lymph nodes, liver and spleen. Patients cease to move, hold the forced position providing

the smallest morbidity at rest. This forced situation with time is fixed thanks to muscular contractures, and the child loses an opportunity to move and serve himself.

Acute onset of YuRA sometimes is not followed by distinct damage of joints. Can be the leading clinical manifestations persistent intermittent fever with temperature rises in the morning, persistent polymorphic rash and the profound arthralgias, mainly in large joints – knee, talocrural, coxofemoral. At the same time in blood tests the high neutrophylic leukocytosis decides on stab shift, significant increase in SOE (up to 60 mm/h). The lack of distinct changes in joints considerably complicates diagnosis of a disease. Quite seldom YuRA proceeds in the form of monoarthritis – most often a knee joint. Usually it is the persistent synovitis at girls of the first years of life which is followed by damage of eyes – a uveitis. At children of 1-2 years the diagnosis of a uveitis is considerably complicated due to the lack of complaints at the child that quite often leads to late diagnostics – at a stage of considerable loss of sight or even development of a blindness. Considering, consistently clinical picture YuRA, it is necessary to stop, first of all, on features of an articulate syndrome as main manifestation of a disease.

Extraarticular changes are usually observed at the severe YuRA system forms.

Damages of heart at YuRA are not a rarity. Most often changes in heart are observed at children with acute onset of a disease in combination with other extraarticular changes, such as fever, rash, hyperadenosis, a liver, a spleen. At this category of patients in 40-43% of cases (according to kliniko-tool researches) myocarditis comes to light, diffusion is more often. It has very typical clinical picture: the patient has a cyanosis of lips and a Crocq's disease, takhi-, bradycardia, increase in the sizes of heart, its diffuse pulsation, a priglushennost of tones, soft systolic noise on a top is more rare. Data of tool methods – the ECG and EhoKG – confirm the clinical diagnosis of myocarditis. Signs of disturbance of blood circulation appear in hard cases: the liver increases, pastosity of shins appears. It should be noted that myocarditis quickly responds to complex treatment. Endocardium is involved in process extremely seldom with what almost total absence of valve heart diseases at patients is connected with YuRA.

Approximately in 50% of cases the damage of a myocardium is combined with pericardiac changes. However the rheumatoid pericarditis can proceed separately or against the background of a polyserositis – pleurisy, serous peritonitis. Such option of a

pericarditis is quite often observed at children of preschool age. Against the background of adequate therapy (usually within 7-10 days) distinct positive dynamics with a further favorable outcome is noted.

Extremely seldom at children the coronaritisis phenomena are observed, and, as a rule, this pathology is found also only at a tool research. Small branches of coronary arteries are involved in process. At defeat of larger vessels, clinical symptoms appear: pain behind a breast, sensation of fear. The coronaritisis, as a rule, is one of symptoms of a generalized vasculitis and the general malignant course of process.

Pulmonary changes at YuRA meet considerably less than cordial, in the form of the isolated damage of a pleura, fusion of sine, plevroperikardialny commissures, the combination of exudative pleurisy to changes in pulmonary fabric – a pneumonitis meets less often. As a rule, it is unilateral process, but maybe bilateral. At the patient the asthma sharply accrues, appear persuasive cough, dry in the beginning, and then damp, abundance of small-bubbling and crepitant rattles in lower parts of lungs, more often on the right. These pulmonary and pleural changes are followed by other symptoms of activity of YuRA – fever, rash, involvement in process of a reticulohistocytosis system (increase in peripheral lymph nodes, a liver, a spleen). Radiological during this period strengthening of the vascular and interstitial drawing of lungs of spotty and cellular character, a "muddy", or "milk" background, sometimes passing ochagovopodobny shadows comes to light.

Damage of kidneys at YuRA is shown by usually uric syndrome, and depending on structure of a deposit, presence or lack of protein, duration of reception of NPVP and basic drugs the treatment it can be different. According to morphologists, it is possible to allocate three types of damage of kidneys:

- glomerulonephritis,
- interstitial nephrite,
- amyloidosis.

The last is the most frequent and terrible complication of YuRA which is on the first place among the possible reasons of a lethal outcome. At YuRA the amyloidosis can develop at children at any age, and not always at the long course of the disease. This terrible complication develops mainly at patients with system forms of a disease – at Steel and Visslera-Fankoni's syndromes.

Eyes are quite often involved in pathological process in a pseudorheumatism.

Clinically is followed by mild reddening of a conjunctiva of eyes, a photophobia. As a rule, at the same time is available oligo- or monoarthritis. At biomicroscopy during this period the vasodilatation of a conjunctiva, small and average precipitated calcium superphosphates, quickly formed pigmentary synechias, change of a form of a pupil are noted. Functions of an eye during an acute period are reduced, but are restored when stopping process. More frequent and heavy sign of rheumatoid damage of eyes is the chronic front iridocyclitis. It is characterized by sluggish, almost asymptomatic course. The diagnosis is made sometimes at routine inspection. As a rule, rough changes of eyes and the significant decrease in visual functions come to light: the iris is atrophied, its pupillary edge is displaced, the pigmentary border is often destroyed. At 35% of children almost full fusion of a pupil, a partial or full cataract, an opacity of the vitreous body are noted.

The triad of symptoms is considered the most characteristic of rheumatoid damage of eyes: a slow uveitis, tape-like dystrophy of a cornea and the complicated cataract. These changes can appear already at early stages of a disease. Damage of muscles is observed at 70-75% of the children suffering from YuRA. Most of authors agree that muscular atrophies are secondary and connected with dysfunction of joints. As a rule, the muscular atrophy is most distinct more proximally than a sore joint, is symmetric and especially pronounced at generalized process with considerable decrease in functional capacity of joints. Less often the miositis which is shown myalgias meets.

The hyperadenosis is observed at children with YuRA quite often, especially in an acute period of a disease and at system forms of a disease. In sizes all peripheral lymph nodes can increase, but the adenopathy of axillary and inguinal groups prevails more often.

The splenomegaly meets much less often, mainly at Steel's syndrome. Clinically the spleen is palpated on 2-3 see belowa costal arch, edge its usually elastic, at a favorable course of process it quickly decreases in sizes.

At the majority of sick YuRA the temperature rises of a body to subfebrile figures are observed in an acute period of a disease and are short. Higher and persistent fever is typical for system forms of a disease – Steel and Visslera-Fankoni's syndromes

(allergosepticheskу option). At Steel's syndrome temperature increases more often in the evening, but no more than 38-38.5 °C, and sometimes is subfebrile. The feverish period proceeds 2-3 weeks and is stopped against the background of adequate therapy. The highest and persistent fever is noted at an allergosepticheskу syndrome. Temperature rise is usually observed early morning hours (from 5 to 7 h), quite often is followed by a strong fever. Temperature reaches 39-40 °C, sticks to 4-5 h, then decreases, being followed pouring then. In the evening temperature increases seldom.

Rash. Skin rashes at sick YuRA, as a rule, are observed at system forms of a disease. At Steel's syndrome the rash usually punctulate, is more rare spotty and papular, is localized on a breast, a stomach, sometimes on an extensor surface of upper and lower extremities. Brightness and abundance of rash sometimes match temperature increase. At height of activity of process it can remain 1-3 weeks and gradually die away against the background of therapy. Absolutely other is rash at an allergosepticheskу syndrome. Usually it arises against the background of sharp temperature rise and is very different: from scarlatiniform and korepodobny to the "linear" rashes, typical for this syndrome, presenting themselves an accumulation of erythematic rash in the form of the lines from 1 to 3-4 cm long which are localized usually on the internal surface of hips and the side surfaces of a thorax. Despite abundance and brightness, rash is never followed by an itching that distinguishes it from allergic, in particular medicamentous rash. In process of falling of fever (to 12-13 h day) it dies away, but does not disappear completely. Long, persistent rash, as a rule, accompanies severe forms of YuRA and is predictively an adverse symptom.

Lag in growth and decrease in body weight – frequent manifestations of YuRA which expressiveness is directly proportional to severity of a disease. So, at mainly articulate form of a disease with multiple damage of joints the lag of growth of bones in length is observed, at the same time, the child got sick earlier, the this dependence is more significant. There is a direct local connection between damage of joints and a bone. However growth of children with the YuRA system forms most intensively slows down.

Data of laboratory researches. Laboratory indicators at YuRA most often reflect existence and expressiveness of inflammatory process and therefore are not specific. The exception represents detection of the rheumatoid factor (RF). Unfortunately, at children

the rheumatoid factor is found no more than at 15-20% of patients, and, according to us, is twice more often in synovial fluid, than in blood serum.

General blood test. One of the most important indicators is the increase in SOE substantially depending on a disease form. So, at mono - and SOE oligoarthritis seldom reaches high figures (30-35 mm/h), in polyarthritis and the YuRA system forms increases to 50-60 mm/h. Level of leukocytes also depends on a disease form. In a leukocytosis oligoarthritis in general can not be or the number of leukocytes does not exceed 8-10Õ109/l. More considerable shifts in analyses of peripheral blood are observed at system forms. Quickly progressing hypochromia anemia in a moderate leukocytosis is characteristic of Steel's syndrome, and even leukopenias with increase in SOE up to 40-50 mm/h. The profound leukocytosis to 20-30Õ109/l with a neutrocytosis to 70-80% and stab (10-12%) shift is typical for an allergosepticheskу syndrome. SOE can reach 50-60 mm/h, quantity of thrombocytes – 400-500 thousand. It should be noted that the size SOE and activity of process not always directly correlate among themselves. At a number of patients against the background of therapy the inflammatory phenomena in joints practically disappear, there are no signs of visceral defeats, and the SOE level continues to remain rather high that, apparently, indicates incomplete remission of process.

Serumal immunoglobulins. As a rule, in active phase YuRA at patients the level of serumal immunoglobulins, and at some patients of all fractions increases (And, M, G).

The S-reactive Protein (SRP) is also one of nonspecific indicators of activity of rheumatoid process, and fluctuation reflect its degree; increase in the SRB level comes to light at 60-70% of patients. In recent years the origin of SRB is in more detail studied. It is developed by hepatocytes under the influence of interleukin-6 (IL-6) which synthesis is induced by interleukin-1 (IL-1) and a factor of necrosis of tumors an alpha (ΦНО-α). It is considered that the similar origin has also amyloid protein.

Antinuclear factor (ANF) representing an antibody to an integral kernel of a cell use usually as screening the test at a system lupus erythematosus. At sick YuRA it is defined rather seldom and testifies in this case in favor of weight of process. However there is a group of patients with frequent presence of ANF - it is girls aged up to 4 years from mono - or the oligoarthritis which is followed by development of a uveitis. Detection of ANF at them testifies to a high risk of development of damage of eyes.

Radiological data at YuRA. In total distinguish 4 stages of radiological changes:

The I stage – osteoporosis, mainly epiphyseal;

The II stage – osteoporosis and initial cartilaginous destruction, narrowing of an articulate crack;

The III stage – the significant destruction of a cartilage and bone, bone erosion;

The IV stage – symptoms of the III stage and an ankiloza.

The radiological method gives full and most exact information on a condition of the musculoskeletal system of the patient. It is necessary to consider that at the same patient the different joints have various radiological changes.

However at assessment of weight take the maximum changes of any joint into account.

Diagnostic criteria of YuRA.

In practice, for diagnostics of YuRA, use diagnostic criteria of the American Rheumatologic Association (ARA):

1. *the beginning of a disease to 16-year age;*
2. *the damage of one or more joints which is characterized by a swelling/exudate or having at least two of the following signs: function restriction, morbidity at a palpation, increase in local temperature;*
3. *duration of articulate changes is not less than 6 weeks;*
4. *exception of all other rheumatic diseases.*

It should be noted that diagnosis of a certain YuRA by criteria of MACAW requires all 4 signs.

Клинико-анатомическая характеристика заболевания	Клинико-иммунологическая характеристика	Течение болезни	Степень активности процесса	Рентгенологическая стадия артрита	Функциональная способность больного
Преимущественно суставная форма: • полиартрит • олигоартрит (2–3 сустава) • моноартрит	ЮРА–РФ+	Быстро прогрессирующее	I, II, III степень	I – околосуставной остеопороз	I – способность к самообслуживанию сохранена
Суставно-висцеральная форма: • с ограниченными висцеритами • синдром Стилла • аллергосептический синдром (Висслера–Фанкони) *Ревматоидный артрит с поражением глаз*	ЮРА–РФ–	Медленно прогрессирующее	Ремиссия	II – остеопороз, хрящевая деструкция, сужение суставной щели III – костные эрозии IV – анкилоз	II – способность к самообслуживанию частично утрачена III – способность к самообслуживанию утрачена полностью

Steel's syndrome according to literature meets rather seldom, in only 12-16% of all cases of YuRA at children, mainly preschool age. Of it it is characteristic rather sharp, manifest, than the subacute beginning, and, at the earliest stages of the disease joints are involved in process – polyarthritis from the first days of a disease is one of the leading symptoms at Steel's syndrome. Extraarticular manifestations, first of all fever are characteristic of this option of YuRA, is more often febrile, sometimes reaching 39-40 °C, proceeding, as a rule, no more than 3-4 weeks. In process of progressing of an articulate syndrome the fever decreases, but the separate temperature rises which are quite often followed by skin rashes can remain. Rash at Steel's syndrome more often punctulate, reminding scarlatinal, is more rare korepodobny, unstable, disappearing together with fever.

During a disease at children the general dystrophy quickly develops. The increased peripheral lymph nodes sometimes reaching the hazelnut sizes, especially in axillary and inguinal areas are sharply designated. The liver and a spleen increase in sizes. Heart at Steel's syndrome suffers at 75% of patients, mainly a myocardium, at 20% – a pericardium, at 10% the aortitis is formed. It should be noted that at this form of a disease the secondary amyloidosis (up to 20%) with primary damage of kidneys most often develops. At the same time at patients within 2-3 years the selection proteinuria can remain, and on the 4-5th year it becomes a constant. A process course at Steel's syndrome, as a rule, quickly progressing. The high humoral activity – SOE to 60 mm/h, increase in all classes of immunoglobulins is noted (it is more G), increase in SRB. The big tendency to **a leukopenia, than to a leukocytosis** is noted, hypochromia anemia quickly accrues. On the 2-3rd year of a disease, destructive changes of a cartilage, bone erosion, in some cases – the ankiloza of ossicles of a wrist defined radiological can already develop. Steel's syndrome, as a rule, it is long the current process. The invalidization at it reaches 25-30%.

The Allergosepticheskу syndrome (**Visslera-Fankoni's syndrome**) significantly differs from Steel's syndrome, first of all, in the delayed articulate manifestations that considerably complicates early diagnostics. As a rule, large joints participate in process: knee, talocrural, often coxofemoral and very seldom joints of brushes and feet. Destructive changes pretty fast develop in them: the cartilage razvolokneniye, bone erosion, and this process quickly progresses, leading to considerable destruction, for example, femur heads.

For an allergosepticheskiy syndrome svoystven and special nature of fever: it usually develops early morning hours, starting with 5-6 h, after a strong fever. Fever duration – 3-4 h, then comes the gektichesky temperature drop which is followed pouring then. The beginning of fever often matches appearance of skin rashes. Rash at an allergosepticheskiy syndrome has extremely persistent character, and, just as fever can last for months. It is localized on a trunk and extensor surfaces of extremities, it is usually polymorphic, a group accumulation on the side surfaces of a breast, on hips is observed. The so-called "linear rash" presenting an accumulation of small elements of rash in the form of lines 1-2 cm long is very typical.

Considerably also the blood picture differs from Steel's syndrome. From the very beginning of a disease at Visslera-Fankoni's syndrome the high neutrophylic leukocytosis with stab shift is observed (a leukopenia at Steel's syndrome!) up to 30000-40000 leukocytes, from them 15-20% – stab.

From visceral defeats at allergosepticheskiy option are observed a lymphadenopathy, gepato-, the splenomegaly is more rare, myocarditis quite often develops and mioperikardit, considerably making heavier a clinical picture of a disease. The course of the disease fast-progressing at 10-12% of patients is formed a secondary amyloidosis, most often kidneys. At late diagnostics and inadequate therapy the allergosepticheskiy option of YuRA gives big percent of an invalidization of patients mainly owing to damage of hip joints.

Treatment of YuRA represents a difficult task and demands special knowledge for the choice of the right direction of therapy depending on a form and the nature of a course of the disease. However the basic principle anyway remains: use of high-speed non-steroidal anti-inflammatory drugs (NPVP) and basic, or drugs of long action. A specific place in treatment of YuRA is held by the glucocorticoids applied now mainly at system forms of a disease.

Non-steroidal anti-inflammatory drugs. Such drugs as indometacin (derivative an indole) and Voltarenum became widespread (derivative phenylacetic acid). Indometacin and Voltarenum appoint to children in a dose of 2-3 mg/kg a day, i.e. from 25 to 100 mg/days, depending on the body weight of patients, to long term (within months and years) therefore, considering side effects, especially ulcerogenic impact on a mucous

membrane of a stomach, constant endoscopic control and also use of cytoprotectors are necessary.

To children appoint Brufenum at the rate of 30-40 mg/kg a day within several months. Naproxenum (Naprosinum) is comparable on the therapeutic effect with Brufenum, but in comparison with Brufenum has longer period of removal from plasma – 10-17 h. The children's form is issued in tablets on 250 mg, – in the form of syrup.

The basic drugs operating slowly unlike high-speed NPVP, more influence immunological processes, slow down articulate destruction, influence remission terms even after cancellation because of their cumulative properties. The quinolinic derivatives which are most often used at children, a methotrexate, cyclosporine A, Sulfasalazinum enter into this group. At adults often use the gold salts which were not widespread so in practice of pediatricians because of considerable side effects. For the same reasons at children also such basic drug as d-Penicillaminum is not used.

Delagil is appointed to children of 1 times a day (to night) on 4 mg/kg, but by no more than 250 mg in time or after a meal. Plaquenil (gtsdroksikhlorokhin) is appointed also once at the rate of 8 mg/kg a day, but by no more than 400 mg. Long-term practice of use of quinolinic drugs for children with not severe slowly progressing YuRA severe forms showed their efficiency and safety at long use.

One of the best immunosuppressants, including at treatment of severe forms of YuRA, the methotrexate used in many rheumatic diseases is considered now, appoint once a week from 2.5 to 7.5-10 mg., depending on a body surface of the child, to teenagers as much as possible on 15 mg.

It is necessary to carry cyclosporine A widely known in transplantology to the new immunosuppressive drugs appointed at treatment of YuRA. Prescribing of cyclosporine A is shown to sick heavy YuRA with a generalized articulate syndrome or its system forms. Cyclosporine A dose – from 2.5 to 4 mg/kg a day in 2 receptions. Careful control of function of kidneys as one of serious side effects of drug is accumulation of slags, increase in level of creatinine is necessary. Control has to be exercised 1 time in 10 days.

One of immunosuppressants now in use is Sulfasalazinum (salazosulfatsyaridin). It is known that this drug with success is used at treatment of ulcer colitis and has the significant antibacterial activity in relation to indestinal flora. In a pseudorheumatism the

positive effect occurs in 2-3 months from an initiation of treatment. The adult appoint 0.5 g 3-4 times a day, to children depending on body weight – from 0.5 to 1.5 g a day.

Glucocorticoids. Now at treatment of YuRA use generally two drugs – Prednisolonum and Methylprednisolonum (Methypredum, solyumedrol), having both anti-inflammatory, and immunosuppressive properties. At all the advantages the glucocorticoids have so obvious negative side effects that their use at YuRA is limited in recent years the heaviest options of a disease. They are appointed at system forms – Steel's syndromes and allergosepticheskу, in the form of courses the pulse therapy is more often in recent years. At severe forms of YuRA appoint Prednisolonum inside in a dose of 0.5-1 mg/kg a day. This dose is accepted during no more than 2-3 weeks and behind that reduce it on 1/4, 1/8 tablets of 1 times in 4-5 days to a maintenance dose of 5 mg/days. Substantially this phenomenon can be prevented simultaneous introduction of glucocorticoids vnutrisustavno. At the same time use such drugs as Methypredum, solomedrol and diprospan (phosphate dinatrium betamethasone + betamethasone dilropionat). Drugs enter into all painful joints in a dose from 0.2 to 1 ml. depending on the joint size. The frequency rate of introduction to one joint usually does not exceed 2-3 times.

Intravenous human immunoglobulin. In recent years in treatment of the YuRA system forms with acute onset, high fever, rash, a neutrophylic leukocytosis successfully use intravenous immunoglobulin – VVIG (sandoglobulin, intraglobin F, a pentaglobin, octadin). The dose of the entered VVIG makes 0.4-0.5 g/kg on a course. One course of therapy consists of triple administration of immunoglobulin every other day. At the corresponding indications the course of treatment is repeated monthly or with an interval of 3-6 months.

Antitsitokinovy therapy. Among a wide range of the "pro-inflammatory" mediators which are taking part in pathogenesis of RA, the special attention is drawn to a necrosis factor опухоли-α (ФНО-α) which is considered as the main cytokine defining development of synovial inflammation and osteoklastoposredovanny bone destruction in arthritises. Therefore now ФНО-α is the major target for so-called *antitsitokinovy therapy* of RA. It formed the basis for development of the FNO-α inhibitors blocking biological activity of this cytokine in circulation and at the cellular level. In the world pharmaceutical

market there are 3 drugs which are blockers ФНО-α: infliksimab, adalimumab and etanertsept.

Local therapy, i.e. impact directly on joints, plays an important role in the general treatment of YuRA. So, intra articulate introduction of corticosteroids – Methylprednisolonum, a solyumedrol, a diprospan is widely used. At topical administration of corticosteroids the fast and significant effect occurs: pain disappears, the mobility of a joint is restored, especially in an exudative synovitis when at the time of a puncture a significant amount of synovial fluid is removed. The amount of the administered drugs depends on the size of a joint and age of the child. Into small joints enter 0.1-0.2 ml of drug, into larger to 1 ml. The same joint is punktirut once a week, by in total no more than 2-3 punctures. Duration of effect is various – from several weeks to several months.

During stay of patients in a hospital and sanatorium it is also possible to administer the drugs by means of ultrasound, to apply low-energy laser radiation.

At decrease in the general and local activity of process the massage and physiotherapy exercises are recommended.

Tasks for independent preparation:

1. Examine the patient with YuRA, using the scheme below. Describe the changes in a workbook revealed by you.
2. Solve situational problems.
3. Make questions for test control on a subject.

Scheme of inspection of the patient

When collecting the anamnesis to pay attention on:

- the patient's family tree (presence of rheumatic, streptococcal, allergic diseases in family and at the patient's relatives);
- features of the immune system (frequency of viral, bacterial infections, their course, the performed therapy and reactions to it, inoculations and inoculative reaction);
- allergic mood;

- chronic centers of an infection (antritis, tonsillitis, otitis, caries of teeth). Efficiency of treatment;
- domestic conditions (density), day regimen (stay in the fresh air);
- complaints, development of a disease, duration of separate symptoms (fever, its character, morning constraint, joint syndrome, etc.).

At objective survey to pay attention on:

- indicators of physical development, their dynamics;
- condition of nervous system (change of mentality, behavior, character, condition of cranial nerves, tendon jerks, focal symptomatology), vegetative disturbances;
- damage of heart (carditis, cardiosclerosis: calculation of pulse, measurement of the ABP, auskultativny and percussion data). Diagnosis of a circulatory unefficiency according to indications (use of functional trials);
- damage of kidneys (glomerulonephritis): to estimate a diuresis, hypostases, changes of a cardiovascular system;
- damage of lungs (alveolites, pulmonary vasculites, the progressing pneumofibrosis);
- sizes of a liver, spleen (both manifestation of a visceral syndrome, and circulatory unefficiency), lymph nodes;
- condition of a digestive tract;
- condition of the musculoskeletal system: survey, a palpation, volume of passive and active movements, the number of the joints involved in process, existence of contractures, ankiloz, periartikulyarny changes, muscles (polymyalgias, a miositis, consolidation), skins (color, elasticity, dryness, turg, a rash – character, duration).

At assessment of paraclinic methods of a research to pay attention on:

- in blood test: on a leukocytosis, increase SOE, a disproteinemia, a hyperglobulinemia, hyper alpha-2-globulinemiyu, existence of lupoid cells (LE) and a rheumatoid factor, the maintenance of SRB, seromucoids, DFA, credits of streptococcal antibodies. Maintenance of ALT and nuclear heating plant, cholesterol, bilirubin, urea, potassium, sodium, sedimentary reactions, amylase content.

- the analysis of urine (a daily proteinuria, test on Nechiporenko, Addis-Kakovsky, across Zimnitsky, clearance of endogenous creatinine);
- X-ray inspection of bodies of a thorax (heart sizes, expansion of borders, symptoms of a pericarditis), joints (osteoporosis, ossifluence, destruction of a cartilage, ankiloza, thickening and flattening of an articulate bag);
- tool grafichesike researches: The ECG – disturbance of a rhythm, conductivity, change of teeth; FKG – easing, fragmentation and deformation of complexes, the sir - or protosystolic noise on a top, pericardial rub; EKHOKG – disturbance of sokratitelny ability of heart, a condition of the valve device, the sizes of cardial cavities and large vessels, etc.
- survey of the oculist, ENT specialist, neuropathologist, etc. experts.

Situational tasks.

Task No. 1

Sick O., 13 years, came to department repeatedly for performing complex therapy.

From the anamnesis it is known that the disease began at 3-year age when after the postponed flu the boy began to limp – as it appeared at survey, because of damage of a knee joint. The joint was spherical shape, hot to the touch, restriction of movements was noted. Further involvement of other joints in pathological process was noted. Almost constantly the child received non-steroidal anti-inflammatory drugs, on this background the periods of remission lasting up to 10-12 months were noted, however the disease gradually progressed. During the periods of aggravation of the patient showed complaints to morning constraint.

At receipt serious condition, the defiguration and a swelling of interphalangeal, radiocarpal, elbow joints, restriction of movements in the right hip joint is noted. In lungs there are no rattles. Heart borders: right – on the right edge of a breast, upper – on the III edge, left – on 1 cm of a knutra from the left average and clavicular line. Rhythmical cardiac sounds, sonorous, there are no noise.

Questions:

1. Formulate and prove the preliminary diagnosis.
2. Make the plan of inspection of the patient.

3. Carry out differential diagnostics.

4. Appoint treatment.

Task No. 2

The boy Z., 13 years, arrived on inspection with complaints to a polyarthralgia within the last 4 months, long subfebrile condition, increased fatigue.

Anamnesis of a disease: the beginning of this disease is connected with the postponed SARS proceeding with high fever. Already against the background of the remaining subfebrile condition the boy had a rest in the summer in the Crimea then the specified complaints amplified.

From the anamnesis of life it is known that to the real disease the child grew and developed normally, had 2-3 times a year the catarrhal diseases proceeding rather not hard.

At receipt moderately severe state. Patient of the correct constitution, satisfactory food. Pale integuments. The elements which are faintly painted erythematic deskvamatoznye on a face, mainly on cheeks and a nose bridge are noted. There are changes of joints in the form of a swelling and moderate morbidity of radiocarpal, elbow and ankle joints. Axillary, back cervical and kubitalny lymph nodes are moderately increased. In lungs a percussion sound pulmonary, vesicular breath sound. Limits of relative warm dullness: right – on the right edge of a breast, upper – on the III edge, left on 1 cmof a knutra from the left average and clavicular line. Cardiac sounds are a little muffled, rhythmical, there are no noise. A soft stomach, painless, the liver and a spleen are not increased. The chair issued urination is not broken.

General blood test: Ayr – 4,2 x 10 12/l, Nv – 100 g/l, Blood clot. – 90 x 10 9/l, Leyk. – 105 x 10 9/l, p.b. – 2%, with – 62%, e – 2%, l – 31%, m – 3%, SOE – 50 mm/hour.

General analysis of urine: ud. weight – 1012, protein – 0.33 ‰, leukocytes – 3-4 in p/z, erythrocytes – 20-25 in p/z,

Biochemical analysis of blood: crude protein – 83 g/l, albumine – 46%, globulins: α1 – 5%, α2 – 12%, β- 5%, γ- 32%, seromucoid – 0.8, ALT – 32 Pieces/l, nuclear heating plant – 25 Pieces/l, urea – 4.5 mmol/l, creatinine – 98 mmol/l.

Zimnitsky's test: ud. weight – 1006-1014, a day diuresis – 320 ml, night – 460 ml.

Clearance on creatinine – 80 ml/min.

Questions:

1. Prove the preliminary diagnosis.
2. What additional examinations does the patient need to perform?
3. Call the principles of treatment of this disease.

Class in a subject:
"DIFFUSION DISEASES OF CONNECTIVE TISSUE"

I. Scientific and methodical justification of a subject:

According to WHO data, diffusion diseases of connective tissue occupy one of the leading places in structure of incidence adults, every 10th disabled person has this pathology. Diseases of this group quite often begin at children's and youthful age, are difficult for early diagnostics, proceed heavier, than at adults. Early diagnosis of these diseases and the corresponding therapy have basic value for the forecast of a disease and the patient's life.

II. Purpose of activity of students.

The student has to know:

- the main kliniko-morphological manifestations in a scleroderma (with prevalence of processes of fibrosing), a system lupus erythematosus (destructive and nuclear pathology), a nodular periarteritis (defeat of large vessels), a dermatomyositis (damage of muscles);
- principles of diagnosis of diffusion diseases of connective tissue and YuRA;
- treatment and preventive actions in these diseases.

The student has to be able:

- to resolve diagnostics issues on the leading clinical and laboratory syndromes;
- to define a degree of activity of autoimmune process, the individual forecast;
- to prove the diagnosis;
- to make the treatment plan;
- to plan preventive actions, to define the forecast.

III. Content of training:

1. Main concepts of pathogenesis of diffusion diseases of connective tissue.
2. Nosological forms of diffusion diseases of connective tissue.
3. Kliniko-morfologichesky changes in diffusion diseases of connective tissue.
4. The leading clinical and laboratory syndromes in diffusion diseases of connective tissue.
5. Principles of therapy and prevention of diffusion diseases of connective tissue.
6. Differential diagnosis of inflammatory diseases of joints (rheumatic, YuRA, infectious and allergic).
7. Features of a course of diffusion diseases of connective tissue.
8. Principle of treatment of these diseases and prevention.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.
3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.
4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.
5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.
6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.
8. Lectures on pediatrics.
9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. The contributing factors in development of rheumatic diseases (rheumatism, hard currency, YuRA) at children.
2. State the main concepts of pathogenesis of diffusion diseases of connective tissue.
3. Give definition and call nosological forms of diffusion diseases of connective tissue.

VII. List of questions for check of final level of knowledge:

1. Call kliniko-morphological changes in a scleroderma, a system lupus erythematosus, a dermatomyositis, a nodular periarteritis.
2. Determine a degree of activity of autoimmune process in diffusion diseases of connective tissue by the leading clinical and laboratory syndromes.
3. List differential and diagnostic criteria of inflammatory diseases of joints (rheumatic, YuRA, infectious and allergic).
4. Call features of a course of diffusion diseases of connective tissue at children's age.
5. Define medical tactics in diseases of connective tissue and a measure of their prevention.

Information block.

Former name of this group of diseases of "collagenose". It combines a number of nosological forms of which systemic lesion of connective tissue and vessels of autoimmune genesis owing to immune complex deposition, polymorphism of a clinical picture and the progressing nature of a course are characteristic. According to modern classifications of rheumatic diseases, this subclass included a system lupus erythematosus, a scleroderma, a dermatomyositis, a diffusion fascitis, rheumatic polymyalgia, a recurrent

polychondritis and a panniculitis, Shegren's syndrome, the mixed disease of connective tissue.

Etiology and pathogenesis. Finally are not specified. The etiological role of persistent viral infection is supposed: clumsy and korepodobny viruses, retroviruses, etc., implementing the action against the background of multifactorial type of predisposition and also immunogenetic factors connected with the 6th couple of chromosomes. In a basis pato- and morfo-genesis humoral and cellular immunopathological processes lie.

The prevailing mechanism of damage is T dysregulation - and V-cellular factors of immunity in the presence in blood of excess content of antigen including autoimmune origin. The formed antigen-antibody complexes activate the coagulant system of blood, bodies and fabrics are postponed on a basal membrane of vessels, krovosnabzhayushchy (kidneys, synovial, serous membranes, a brain, etc.); the lizosomalny enzymes released at the same time from the englobing cells promote defeat deepening. The complement fixed by cell-bound immune complexes and also sensibilized small lymphocytes have cytotoxic effect. Defeat of vessels at the level of a microcirculation is the cornerstone of system damage of connective tissue and parenchymatous bodies. Localization of defeat, primary and characteristic of each disease of this group, is defined by implementation of an effector phase of the immune response in a target organ.

The nosological specificity is inherent to each disease along with community of pathogenetic and morphological features. Distinctive feature of a system lupus erythematosus is the sensitization to nucleinic connections of virus or nuclear origin therefore anti-nucleinic and antinuclear antibodies are formed; the damaging part is assigned to an antinuclear factor – to the antibodies directed against the main components of a kernel of a cell. Hyperproduction of antibodies to native (two-spiral) DNA is especially characteristic; the complexes formed with their participation are responsible, in particular, for development of lyupus-nephrite and also vasculitis in other bodies.

The originality of a morphogenesis of a system scleroderma consists in strengthening of a kollagenoobrazovaniye and disturbance of microcirculation in combination with peculiar almost acellular inflammation. The central role in development of fibroblastichesky process, fast forming of a sclerosis, a hyalinosis of fabrics belongs to the increased function of fibroblasts and other kollagenoobrazuyushchy cells. As a result

of excess formation of soluble forms of collagen, injuries of an endothelium of vessels develop microcirculator disturbances. Steady progressing of process is caused by autoimmune reactions to collagen in view of its excess education.

At patients with a dermatomyositis the sensitization of lymphocytes to antigens of muscle tissue is revealed and the role of cell-bound immune complexes in development of vasculites in muscles is shown.

At children owing to anatomo-physiological features all diffusion diseases of connective tissue proceed heavier, than at adults. Acute, rapid development with fast forming of polysystem pathological process with the significant and raspostranenny vascular reactions, an exudative component of inflammation at high probability of a recurrence and further progressing is peculiar to these diseases. Girls, mainly younger school and prepubertatny age get sick more often.

SYSTEM LUPUS ERYTHEMATOSUS. The most serious illness from this group. Distinguish sharp, subacute and chronic options of a course.

Clinical picture. Manifestations are diverse and are observed in various combinations. Leaders are a joint and muscular syndrome (at 80-0% of patients), damage of skin (at 85% of patients), feverish reactions of various type and duration. Often heart is surprised: the pericarditis, Liebman-Saks's endocarditis, myocarditis, are more rare a pancarditis. Pleuropulmonary changes develop at 40-80% of children (pleurisy, a pulmonitis, etc.). Approximately a half of patients has neurologic symptoms: epileptiform spasms, various motive and sensitive disturbances, mental disorders. Damage of kidneys, mainly like a diffusion glomerulonephritis with a nephrotic component is typical. Lupoid nephrite quite often defines the forecast of a disease. The moderate lymphadenopathy comes to light almost at all patients; increase in a liver and spleen – less frequent symptom (at 15-30% of children).

The joint and muscular syndrome is characterized by damage of joints, sinews, tendinous vaginas of muscles. Muscle pains can be very severe, feign a dermatomyositis, at a palpation of a muscle are painful, are condensed with places, are noted a diffusion atrophy them, decrease in animal force up to a myasthenia. The arthralgia of various degree of manifestation migrating. Arthritis proceeds sharply, subacutely, seldom chronically. Acute arthritis reminds rheumatic on "volatility" of defeat; subacute is similar

to rheumatoid with a characteristic defiguration of joints, a muscular atrophy, constraint in the mornings and restriction of functions. Most often proximal interphalangeal and metacarpophalangeal joints, knee and ankle joints are surprised. Manifestations of a disease can keep long time, but then undergo full involution. Incomplete dislocations are possible; ankiloza are formed seldom; a large amount of exudate in joints is observed also infrequently. Multiple and symmetric damage of joints, tendency to involution and recuring are characteristic. Radiological changes of joints: spotty or diffusion osteoporosis of epiphyseal areas without signs of osteoarticular destruction. In synovial fluid the low protein content and cellular elements decides on prevalence degenerative with the phenomena of nuclear disintegration; sometimes lupoid cells and an antinuclear factor are found.

The joint and muscular syndrome proceeds wavy; its aggravation is combined most often with defeat of serous covers and skins, development of heavy viscerites (lupoid nephrite, tserebrovaskulit) the joint syndrome dies away.

At some patients the aseptic subchondral osteonecrosis mainly in the field of a femur head develops, is more rare in knee and shoulder joints. Clinically the affected joint pain is defined, at localization in a hip joint – restriction of rotation and assignment, lameness. Radiological the picture characteristic of various stages of aseptic necrosis comes to light. Recovery of bone structure or does not happen, or it proceeds very slowly that is promoted by long-term treatment by glucocorticoids.

The unusual polymorphism and various localization are inherent to skin rashes. More often they are shown by an erythema and hypostasis, infiltration, a hyperkeratosis with tendency to formation of necroses with the subsequent pigmentation and scarring. "Butterfly", pathognomonic for a system lupus erythematosus, on a face in a bridge of the nose and cheeks meets approximately at 1/3 patients. The cheilitis, the vascular phenomena (capillarites, livedo, Reynaud's syndrome, hemorrhages in skin, mucous membranes), trophic changes of skin, nails, a hair loss are quite often observed. At survey of an eyeground, hemorrhages, a vasomotor spasm, the velvet-like whitish centers come to light; the uveitis, an episcleritis are less characteristic.

Treatment. At treatment consider course option, a degree of activity and weights of the visceral phenomena. The central place is taken by glucocorticoids. The general

duration of a course is individual – from several months to several years. Therapy by Prednisolonum is combined with quinolinic derivatives (delagil, plaquenil).

The patient with diffusion damage of kidneys, heavy gemopatiya appoint cytostatic means – Azathioprinum (imuran) or cyclophosphamide in a dose of 1-3 mg/kg a day in combination with Prednisolonum which dose it is possible against the background of cytostatic means to lower.

Intensive care is performed at patients with the profound immunocomplex pathology at a nephrotic syndrome, a heavy tserebrovaskulit, cytopenias, a system vasculitis. It is carried out with the help pulse therapy in a combination with Cyclophosphanum and extracorporal methods – a plasma exchange and a karbogemoperfuziya.

DERMATOMYOSITIS. Disease which is followed by systemic lesion of skeletal and unstriated muscles, skin and internals. Distinguish primary and secondary, or paraneoplastic, the dermatomyositis connected with tumoral process. The last meets at children extremely seldom. Primary dermatomyositis at children's age differs in two features: frequent development of vasculites in an initial stage and as their investigation — calcification of fabrics in later phase and also association with carriage of HLA-B8 and DR-3 that indicates participation of genetic factors. The tendency to giperergichesky vascular reactions, a perversion of enzymatic and protein metabolism are observed.

There is no standard classification of a dermatomyositis. On the nature of a course at children allocate sharp, subacute and primary and chronic options. They are distinguished by sharpness, weight, degree of systemacity of clinical manifestations and rates of progressing.

Clinical picture. Differs in a mnogosindromnost, muscles and almost with the same frequency skin most often are surprised. Generally cross-striped muscles are surprised, it is rare — unstriated muscles. The miositis at height of activity of process has mainly symmetric raspostranenny character. Muscles of a shoulder and pelvic girdle become pasty consistence or dense, increase in volume, are painful at a palpation. Muscle pains constant or passing, various intensity. Muscle weakness to a full obezdvizhennost of the patient is profound. Mimic and chewing muscles suffer. In the most adverse cases respiratory and pharyngeal muscles are surprised.

Skin manifestations are various. The purple-lilac (heliotropic) erythema and hypostases of various localization are typical. They most often are located in paraorbital area in the form of glasses or a half mask, can extend to adjacent sites, including a hairy part of the head and also to a trunk and an extremity, including to a dorsum of metacarpophalangeal and proximal interphalangeal joints, sometimes in combination with a Quincke's disease. Also papular, bullous elements, a purpura, teleangiectasias, a hyperkeratosis, a poikiloderma, an alopecia, livedo, vascular staza, a red border meet at the basis of nails and so forth. At a heavy course small or extensive necroses of cover fabrics of various localization are formed. At 50% of children the heavy raspostranenny calcification of muscles and hypodermic cellulose, at fistulas, some with education, develops.

Almost at all patients the damage of mucous membranes of a mouth, upper airways, conjunctivas, vaginas is observed (ulcers, hemorrhages, hyperaemia, puffiness, trophic changes and so forth).

At a part of patients the joint syndrome – an arthralgia and (or) arthritis, sometimes a pseudoarthritis – the seeming damage of joints at the expense of hypostasis of periartikulyarny fabrics or a contracture in connection with fibrous changes of muscles develops; arthritis usually passing like polyarthritis, with involvement in process symmetric radiocarpal, knee, talocrural and interphalangeal joints without the expressed exudative component of inflammation.

Visceral manifestations are caused by defeat of the respiratory device (pulmonary insufficiency in connection with involvement in process of respiratory muscles and a diaphragm, vascular and interstitial pneumonia, pleurisy); cardiovascular system (mainly myocarditis, myocardial dystrophy); nervous system (mainly peripheral and vegetative); digestive tract (dysphagy, abdominal pain, gastroenterocolitis, ulcer process). Damage of kidneys comes to light infrequently, has passing character. Actively current process is always followed by subfebrile body temperature, high temperature reaction is inherent to acute onset and high activity of a disease. Are characteristic loss of body weight — increase of dystrophy and trophic disorders, endocrine pathology.

SYSTEM SCLERODERMA. A system sclerosis of connective tissue with primary damage of skin. Allocate three stages of skin changes: hypostasis, induration and atrophy.

The first stage proceeds imperceptibly, in an induration stage the skin and hypodermic cellulose become dense, accept cartilaginous consistence, skin cold, does not gather pleated, has a wax shade, is pigmented. In an atrophy stage the skin becomes thinner, sites of pigmentation alternate with depigmented, ossifluence of nail phalanxes, an ulceration of finger-tips, Reynaud's syndrome develop. Damage of skin is combined with damage of joints, bones and muscles, constraint, contractures of muscles, false ankyloses develop. At widespread process the physical activity of the child is limited. On the roentgenogram of joints the osteoporosis, narrowing of an articulate crack, an uzuration of cartilages, incomplete dislocations, ossifluence of nail phalanxes, sometimes – deposits of calcium in periartikulyarny fabrics come to light. In an articulate exudate which manages to be received very seldom, – the changes similar to those in a pseudorheumatism.

Damage of internals meets often, but mainly the erased symptomatology: fibrosis of a myocardium, endocardium, it is rare — a pericarditis; the basal pneumosclerosis, is more rare – pleurisy; dysphagy, dispepsichesky disorders; focal nephrite, it is rare – a true sklerodermichesky kidney with violently developing renal failure.

Tasks for independent preparation:

1. Examine the patient with hard currency, using the scheme below. Describe the changes in a workbook revealed by you.
2. Solve situational problems.
3. Make questions for test control on a subject.

Scheme of inspection of the patient

When collecting the anamnesis to pay attention on:

- the patient's family tree (presence of rheumatic, streptococcal, allergic diseases in family and at the patient's relatives);
- features of the immune system (frequency of viral, bacterial infections, their course, the performed therapy and reactions to it, inoculations and inoculative reaction);
- allergic mood;
- chronic centers of an infection (antritis, tonsillitis, otitis, caries of teeth). Efficiency of treatment;

- domestic conditions (density), day regimen (stay in the fresh air);
- complaints, development of a disease, duration of separate symptoms (fever, its character, morning constraint, joint syndrome, etc.).

At objective survey to pay attention on:

- indicators of physical development, their dynamics;
- condition of nervous system (change of mentality, behavior, character, condition of cranial nerves, tendon jerks, focal symptomatology), vegetative disturbances;
- damage of heart (carditis, cardiosclerosis: calculation of pulse, measurement of the ABP, auskultativny and percussion data). Diagnosis of a circulatory unefficiency according to indications (use of functional trials);
- damage of kidneys (glomerulonephritis): to estimate a diuresis, hypostases, changes of a cardiovascular system;
- damage of lungs (alveolites, pulmonary vasculites, the progressing pneumofibrosis);
- sizes of a liver, spleen (both manifestation of a visceral syndrome, and circulatory unefficiency), lymph nodes;
- condition of a digestive tract;
- condition of the musculoskeletal system: survey, a palpation, volume of passive and active movements, the number of the joints involved in process, existence of contractures, ankiloz, periartikulyarny changes, muscles (polymyalgias, a miositis, consolidation), skins (color, elasticity, dryness, turg, a rash – character, duration).

At assessment of paraclinic methods of a research to pay attention on:

- in blood test: on a leukocytosis, increase SOE, a disproteinemia, a hyperglobulinemia, hyper alpha-2-globulinemiyu, existence of lupoid cells (LE) and a rheumatoid factor, the maintenance of SRB, seromucoids, DFA, credits of streptococcal antibodies. Maintenance of ALT and nuclear heating plant, cholesterol, bilirubin, urea, potassium, sodium, sedimentary reactions, amylase content.
- the analysis of urine (a daily proteinuria, test on Nechiporenko, Addis-Kakovsky, across Zimnitsky, clearance of endogenous creatinine);

- X-ray inspection of bodies of a thorax (heart sizes, expansion of borders, symptoms of a pericarditis), joints (osteoporosis, ossifluence, destruction of a cartilage, ankiloza, thickening and flattening of an articulate bag);
- tool grafichesike researches: The ECG – disturbance of a rhythm, conductivity, change of teeth; FKG – easing, fragmentation and deformation of complexes, the sir - or protosystolic noise on a top, pericardial rub; EKHOKG – disturbance of sokratitelny ability of heart, a condition of the valve device, the sizes of cardial cavities and large vessels, etc.
- survey of the oculist, ENT specialist, neuropathologist, etc. experts.

Class in a subject:

"LEUKOSES"

I. Scientific and methodical justification of a subject.

Acute leukoses belong to the serious hematologic illness widespread in the basic in children's age. Lately the expressed tendency to growth of number of hemoblastoses at children is noted. Early diagnostics, timely use of modern schemes of therapy now considerably improved the forecast of acute leukoses.

Therefore, knowledge of clinical options and the cornerstone pathological changes in an organism, their timely recognition, the correct organization of adequate medical care are a condition of decrease in lethality, preservation of conditions of the correct growth and development of children, and then and working capacity and lifetime of adults.

II. Purpose of activity of students on occupation:

The student has to know:

- modern theories of an etiology of acute leukoses;
- pathogenesis of acute leukoses;
- main pathomorphologic changes in marrow and other bodies and fabrics;
- clinical manifestations of an acute leukosis;
- classification of leukoses (clinical and morphological);
- changes of peripheral blood and miyelogramma in an acute leukosis;

- the differential diagnosis of an acute leukosis with chronic leukoses and leukemoid tests;
- the contributing factors to development of hemorrhagic diseases;
- the main pathophysiological mechanisms of disturbances in the system of a hemostasis;
- key laboratory indicators of a system of a hemostasis;
- main nosological forms of hemorrhagic diseases (hemorrhagic vasculitis, trombotsitopatiya, thrombocytopenia, hemophilia);
- complications of hemorrhagic diseases;
- IDCS: etiopathogenesis, clinical laboratory diagnostics, medical tactics;
- emergency aid in hemorrhagic diseases;
- principles of modern pathogenetic therapy of clinical options of hemorrhagic diseases;
- principles of rehabilitation of hemorrhagic diseases, landmark medical examination.

The student has to be able:

- to collect the purposeful anamnesis;
- to perform objective examination of the patient, to allocate disease symptoms;
- to make the plan of necessary additional inspection, to estimate the received results;
- to carry out the differential diagnosis of basic diseases of blood;
- to prove the final diagnosis;
- to make the plan of treatment;
- to write the prescription on the main medicines;
- to make the plan of dispensary observation and rehabilitation of the patient with blood diseases.

III. Content of training.

1. Physiology, pathophysiology of a system of a hemopoiesis.
2. Objective research of the patient and semiotics of diseases of blood.
3. Main methods of laboratory diagnosis of diseases of a system of blood.

4. Acute leukosis at children (an etiology, pathogenesis, classification, diagnostics, diff. diagnostics).

5. The principles of landmark therapy, rehabilitation and dispensary observation of acute leukoses at children.

6. The differentiated approach in treatment of a hemorrhagic syndrome in blood diseases at children.

7. Dispensary observation and rehabilitation of children with leukoses.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.

2. Educational medical documentation (case histories, laboratory researches, roentgenograms).

3. Technical means of training.

4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.

2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.

3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.

4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.

5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.

6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. Physiology, pathophysiology of a system of a hemopoiesis.
2. Objective research of the patient and semiotics of diseases of blood.
3. Main methods of laboratory diagnosis of diseases of a system of blood.
4. Physiology and pathology of a system of a hemostasis at children.
5. Classification of hemorrhagic diseases.

VII. List of questions for check of final level of knowledge:

1. What theories of the reasons of development of an acute leukosis in children do you know?
2. Call the main pathogenetic mechanisms in development of an acute leukosis.
3. What classification of acute leukoses at children is used in modern hematology? By what principle the classification is carried out?
4. What modern methods of a research are used at diagnosis of acute leukoses?
5. With what diseases it is necessary to differentiate acute leukoses at children?
6. What risk factors contribute to development of a leukosis in children?
7. What form of an acute leukosis meets at children more often others?
8. At what age the rise in incidence of an acute lymphoblastoid leukosis at children is noted?
9. What periods allocate in clinic of an acute leukosis at children?
10. What complications can arise in an acute leukosis at children.
11. What is "landmark" therapy of acute leukoses meant definition? Call the basic principles of landmark therapy of acute leukoses at children.
12. Call the basic principles of rehabilitation and dispensary observation in acute leukoses at children.

Information block.

Leukoses – the general name of the malignant tumors arising from the haematogenic cells. 75% of patients – children with the acute lymphoblastoid leukosis (ALL), 15-20% –

with an acute not lymphoblastoid leukosis (OnLL), 1-3% – with the chronic myeloid leukosis (CML), the others – with not identified options of the acute leukosis (AL).

Etiology:

a) ionizing radiation;

b) influence of chemicals;

c) virus theory;

d) genetic predisposition;

e) role of oncogenes and mutations.

Classification:

OLL – 3 morphological options (depending on the lymphoblast sizes); OnLL – 7 options:

M1 is miyeloblastny, maturing is absent,

Sq.m is miyeloblastny, semimaturation,

The m3 is promiyelotsitarny,

M4 is miyelomonoblastny,

M5 is monoblast,

M6 – an erythremic myelosis,

M7 is megakarioblastny.

At HML allocate "adult" type, "juvenile" type and also blast crisis. The congenital leukosis is described as the OL special form.

Clinic.

Combination *of anemic* and *intoxication* symptom complex (pallor of mucous membranes and skin with a gray and icteric shade, slackness, weakness, subfebrile condition), *proliferative* (increase in peripheral, mediastinal or belly lymph nodes, a gepatosplenomegaliya, tumoral formation of an eye-socket, flat bones of a skull, etc.) and *hemorrhagic* syndromes (skin hemorrhages, nasal bleedings, etc.). Quite often already at the first clinical identification of OL – clinical and radiological signs of damage of skeletal system, brain and its covers (headaches, nausea, vomiting, disturbances of consciousness, damage of cranial nerves – are more often the VI couples, parapareses, paralyzes, spasms, etc.), testicles, leukemic infiltrates on skin.

***Clinical stages**:*

a) period of initial manifestations;

b) the period of the developed clinical manifestations;

c) incomplete and full clinical laboratory remission;

d) disease recurrence;

e) end-stage.

At many children one or two characteristic syndromes appear in 4-6 weeks prior to development of an acute leukosis that allows to make the diagnosis correctly.

At OnLL the proliferative syndrome is seldom shown, it is frequent – anemic, intoxication, hemorrhagic syndromes, infections.

Datas of laboratory.

In analyses of peripheral blood find blast cells, anemia, thrombocytopenia in sick OL (but at 10% of sick children of aberrations in analyses is not present). In a miyelogramma – oppression of an erythrogenesis, thrombocytopoiesis, abundance of blast elements (total number of blasts more than 30%). Cytochemical researches are conducted in specialized hematology units for differential diagnostics of OLL. Search of specific markers by means of marked monoclones is necessary for identification of options of OLL.

Treatment.

At OL the therapy is divided into the periods and is carried out in specialized (hematologic) department: induction of remission, consolidation, the supporting treatment by periodic courses of active therapy (in particular, prevention of damages of central nervous system). The therapy supporting remission includes: 6 Mercaptopurinum – 50 mg/sq.m a day, inside in 3 receptions daily; the methotrexate – to 50 mg/sq.m is each 8 weeks intravenously; Prednisolonum – 2 mg/kg/days inside within 4 weeks, every 8-16 weeks.

New methods of treatment of sick OL concern various aspects of bone marrow transplantation, stem cells that is especially important for sick OnLL which often in the course of treatment have a marrow aplasia. Transplantation can be singenny, allogenic and autologous.

Diet. The sick acute leukosis recommends high-calorific with one-and-a-half, in comparison with age norms, amount of proteins, vitaminized, rich with mineral substances (a table 10a). When assigning glucocorticoids the diet is enriched with products, with the increased content of salts of potassium and calcium.

Dispensary observation is carried out by the hematologist of the specialized center and the district doctor. Once in 2 weeks do blood test. At maintenance therapy of a cytostatics appoint once a week, before it it is necessary to count number of leukocytes. If them less than 1.0Õ109/l, then cytostatics do not appoint, and give the medicines promoting increase in number of leukocytes and only at increase in number of leukocytes to 1.5Õ109/l resume cytostatic therapy. Do to Miyelogramm before and after each course of a reinduktion which is conducted in a hospital.

Change of climatic conditions is not shown. The child is exempted from preventive inoculations, exercises. He should be preserved against physical activities, mental traumas, cooling, accidental infections. House training is recommended.

The forecast depends on activity of the supporting antirecurrent therapy, quality of dispensary observation and the applied khimiopreparat, a possibility of the selection of the donor compatible on HLA, for bone marrow transplantation or stem cells, the accumulating infectious complications and also individual sensitivity of the patient to side effects of the applied khimiopreparat, existence of a gene of multiple resistance at it to drugs.

Usually a recurrence of OL comes in the first 2 years after achievement of the first remission. It is considered that if the first remission continues 3-4 years, then it is possible to ask about the therapy termination, however it is necessary to speak about recovery of the child very carefully.

The scheme of inspection of the patient with an acute leukosis.

When collecting the anamnesis to pay attention on:

- genetic predisposition;
- the short-term or repeating hemorrhages, anemias, metabolism diseases, general diseases, etc.;
- professional harm of parents;
- defects of food in family;
- growth and development during various age periods;
- infectious diseases, their frequency, weight;

- reaction of the haematogenic system (hemorrhagic manifestations, anemias, leukemoid tests), reaction of lymphatic system (repeatability, duration);
- the first symptoms of a disease – their character, dynamics.

At objective inspection to pay attention on:

- weight of a state;
- integuments and mucous (color, shade, hemorrhagic manifestations);
- skeletal system: morbidity in various departments (spontaneous, at movements, at a palpation, etc.);
- lymphatic system: careful survey and characteristic of peripheral lymph nodes;
- palpation of a liver and spleen – the sizes, morbidity;
- survey on bodies and systems;
- to make the plan of additional researches;
- to compare data of objective survey with indicators of additional researches.

When reading laboratory analyses to pay attention on:

- characteristic of an erythrocyte system (quantity of erythrocytes, reticulocytes, Hb;
- condition of a leukocytic formula (quantity of leukocytes, morphological characteristic);
- existence of young and undifferentiated forms;
- leukemic "failure";
- to miyelogramm.

Test tasks.

1. Primary place of formation of leucemic cells is:

a) lymph nodes

b) Central nervous system

c) marrow

d) lymphoid bodies

2. Acute and chronic leukoses differ from each other:

a) degree of a differentiation of tumor cells

b) sharpness of clinical manifestations

c) disease duration

3. The diagnosis of an acute leukosis does not raise doubts at:

a) emergence of blast cells in a gemogramma

b) anemic and hemorrhagic syndromes

c) oppression of all sprouts of a hemopoiesis in marrow

d) total blastoz in marrow

4. Quantity of blast cells in marrow, necessary for statement

diagnosis of a leukosis:

a) total blastoz

b)> 30%

c)> 20%

d)> 5%

5. At an extramedullary recurrence of an acute lymphoblastoid leukosis are surprised:

a) lymph nodes

b) Central nervous system

c) liver

d) testicles

e) marrow

6. Induction of remission of an acute leukosis includes use:

a) glucocorticoids

b) polychemotherapy

c) radiation therapy

d) glucocorticoids + polychemotherapy

e) glucocorticoids + polychemotherapy + radiation therapy

Situational tasks.

Task No. 1.

The boy R., 12 years, came to department with complaints to weakness, nausea, vomiting, temperature increase, onychalgias.

From the anamnesis it is known that within the last 3 months the boy began to be tired quickly, the appetite decreased. 2 weeks ago parents noticed that the child turned pale. The real aggravation of symptoms was noted 10 days ago when temperature up to 39.3 °C increased, submaxillary lymph nodes increased. In out-patient blood test the hyperleukocytosis up to 200 x l09/l is revealed.

At receipt condition of the child heavy. Intoxication symptoms are pronounced. Integuments and visible mucous pale, on extremities numerous ecchymomas. Submaxillary, cervical lymph nodes by the sizes up to 1.5 cm, mobile, painless are palpated; axillary, inguinal lymph nodes up to 1.0 cm in the diameter. In lungs the vesicular breath sound, in lower parts is weakened on the right, rattles are not listened. Clear, rhythmical cardiac sounds. A soft, painless stomach at a palpation. The liver + 4.0 cm, a spleen + 2.0 cm is lower than edge of a costal arch. The stiff neck, a positive Kernig's sign is noted.

General blood test*:* N - 86 g/l, Ayr – Z,2 h 1012/l, Blood clot - single, Leyk – 208 x 109/l, blasts - 76%, p.b. - 1%, with - 4%, l - 19%, SOE - 64 mm/hour.

Miyelogramma: marrow giperplazirovan, blasts - 96%, a neutrophylic sprout - 3%, an erythroidal sprout - 1%, megacaryocytes - are not found.

Task:

1. Make the preliminary diagnosis.
2. What methods of a research does this child need to carry out still?
3. Call the main stages of treatment of this disease.
4. What was the reason of development of neurologic symptomatology?

Class in a subject:

"DISEASES WHICH ARE FOLLOWED BY BLEEDING"

I. Scientific and methodical justification of a subject.

Hemorrhagic diseases are widespread at children. Having begun at children's age, they quite often accompany the patient during all subsequent life. Doctors of all profiles should face these diseases, their complications or consequences. Therefore, knowledge of clinical options and the cornerstone pathological changes in an organism, their timely recognition, the correct organization of adequate medical care are a condition of decrease in lethality in these diseases, preservation of conditions of the correct growth and development of children, and then and working capacity and lifetime of adults.

II. Purpose of activity of students on occupation:

The student has to know:

- the contributing factors to development of hemorrhagic diseases;
- the main pathophysiological mechanisms of disturbances in the system of a hemostasis;
- key laboratory indicators of a system of a hemostasis;
- main nosological forms of hemorrhagic diseases (hemorrhagic vasculitis, trombotsitopatiya, thrombocytopenia, hemophilia);
- complications of hemorrhagic diseases;
- IDCS: etiopathogenesis, clinical laboratory diagnostics, medical tactics;
- emergency aid in hemorrhagic diseases;
- principles of modern pathogenetic therapy of clinical options of hemorrhagic diseases;
- principles of rehabilitation of hemorrhagic diseases, landmark medical examination.

The student has to be able:

- to collect the purposeful anamnesis;
- to perform objective examination of the patient, to allocate disease symptoms;
- to make the plan of necessary additional inspection, to estimate the received results;
- to carry out the differential diagnosis of basic diseases of blood;
- to prove the final diagnosis;
- to make the plan of treatment;

- to write the prescription on the main medicines;
- to make the plan of dispensary observation and rehabilitation of the patient with blood diseases.

III. Content of training.

1. Physiology, pathophysiology of a system of a hemopoiesis.
2. Objective research of the patient and semiotics of diseases of blood.
3. Main methods of laboratory diagnosis of diseases of a system of blood.
4. Classification of hemorrhagic diseases.
5. Hemorrhagic vasculitis (etiology, pathogenesis, classification, clinical and laboratory diagnostics, principles of therapy, complication).
6. Idiopathic Werlhof's disease (etiology, pathogenesis, classification, diagnostics, therapy).
7. Hemophilia (etiopathogenesis, classification, clinical manifestations, laboratory diagnostics, complications, treatment).
8. The differentiated approach in treatment of a hemorrhagic syndrome in blood diseases at children.
9. Dispensary observation and rehabilitation of children with hemorrhagic diathesis.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.
3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.

4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.

5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.

6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. Physiology, pathophysiology of a system of a hemopoiesis.
2. Objective research of the patient and semiotics of diseases of blood.
3. Main methods of laboratory diagnosis of diseases of a system of blood.
4. Physiology and pathology of a system of a hemostasis at children.
5. Classification of hemorrhagic diseases.

VII. List of questions for check of final level of knowledge:

1. Give characteristic to the main etiopatogeneticheskу mechanisms of a hemorrhagic vasculitis at children.
2. What principle is the basis for modern classification of a hemorrhagic vasculitis?
3. What main clinical manifestations of a hemorrhagic vasculitis at children? What methods of laboratory diagnostics are used at production of this diagnosis?
4. Call possible complications of a hemorrhagic vasculitis.
5. Call the basic principles of therapy of a hemorrhagic vasculitis at children.
6. Give definition to the concept "idiopathic Werlhof's disease". Give main types of trombotsitopeniye, the reasons for their development.
7. Describe the main clinical manifestations of a hemorrhagic syndrome in thrombocytopenia.
8. What methods of a research can be used for diagnosis "An idiopathic Werlhof's disease"?
9. What modern methods of treatment of a Werlhof's disease do you know?

10. Call etiopatogeneticheskу mechanisms of development of a hemorrhagic syndrome in hemophilia.

11. What data of the anamnesis and clinical manifestation are inherent for the patient with hemophilia?

12. What laboratory diagnostics is necessary for diagnosis "Hemophilia"?

13. What complications can arise in hemophilia?

14. Call the basic principles of therapy, prevention, dispensary observation and rehabilitation of hemophilia at the present stage.

Information block.

HEMOPHILIA

– the hereditary disease caused by deficit or molecular anomalies of one of plasma blood-coagulation factors.

Distinguish: hemophilia And, the classical form connected with deficit of the VIII factor (70-78% of patients of the total number of patients with hemophilia), a Cristmas disease, Kristmas's disease, at deficit of the IX factor (6-13%); hemophilia With, Rosenthal's disease, at deficit of the XI factor (1-2%).

Cristmas disease frequency the different countries fluctuates from 6.6 to 18 cases on 100 thousand male inhabitants.

Hemophilias And yes In are inherited on the recessive, linked to X-chromosome type in this connection only men are ill. Daughters, inheriting X-chromosome from healthy mother and X-chromosome from the father hemophiliac, without having clinical manifestations of a disease, are conductors of hemophilia. The sons who were born from such marriage are healthy and further do not transmit a disease to posterity. Theoretically a half of sons of conductresses of hemophilia can receive abnormal X-chromosome from mother and be born patients, also as well as at a half of the been born daughters the carriage of hemophilia is possible.

Hemophilia With is inherited autosomally, persons of both sexes are ill.

The Cristmas disease to family can sporadically arise, being a consequence of natural mutations of genes in X-chromosome that makes about 28% of hemophilia And yes 9% of a Cristmas disease.

Pathogenesis.

Bleeding arises in connection with disturbance of blood clotting owing to a lack of factors of VIII, IX or X.

Clinic.

Clinically separate forms of hemophilia proceed almost equally, being shown by a hemorrhagic syndrome with gematomny type of bleeding which expressiveness depends on a form of hemophilia and degree of deficiency of the corresponding pro-coagulant. Definition of a form of hemophilia is possible only on the basis of special laboratory researches.

Depending on degree of deficiency of responsible pro-coagulant, allocate:

1) a severe form of a disease with the level of pro-coagulant it is lower than 1-3%;

2) a moderately severe form – with the level of pro-coagulant from 3 to 5%;

3) an easy form – with the level of pro-coagulant from 6 to 15%, but with possibility of heavy bleedings in injuries and the operations performed without sufficient replacement therapy;

4) a latent form – with the level of pro-coagulant it is higher than 15%.

The course of hemophilia is characterized by the periods of the increased bleeding which are replaced by intervals of relative clinical wellbeing.

The disease comes to light, as a rule, at children's age. At easy forms of hemophilia the disease can be shown at youthful and later age (20-23 years). At severe forms of hemophilia some newborns have extensive cephalohematomas, late bleedings from an umbilical cord, hypodermic and intracutaneous, is rare – intracraneal hemorrhages, bleedings during the cutting or a rupture of a bridle of language. At many children hemorrhages are not observed while they do not begin to creep and go. By 1.5-2-m years, bruises on a forehead, extremities, massive intermuscular hematomas on buttocks, gingival bleedings are characteristic in traumatization toys, eruption zu6ov, from a mucous membrane of a throat at shout, crying, cough, menacing with asphyxia. Intramuscular injections, inoculations can also cause bleeding. After three-year age, in connection with increase in physical activity of the child, damages of joints which begin to prevail further join and take the first in frequency place among other symptoms of a disease.

Developing of bleedings at the disturbances of integrity of skin continuing from several hours to many days obviously not adequate to injury force is characteristic. In the postoperative period of surgical interventions, after extraction of tooth the recurrence of bleedings delayed on time several hours can be celebrated.

In case of compression by a hematoma of nervous trunks of extremities, sinews, vessels contractures, paralyzes, an atrophy of muscles, necroses of fabrics develop. The hematomas of submaxillary area and a neck squeezing airways are dangerous.

Bleedings from urinary tract at younger children's age are very rare, but the frequency and intensity of a hamaturia (a gross hematuria in combination with the dysuric phenomena) increases over the years. Bleedings from a digestive tract, first of all, diffusion capillary are also rare. More often there are gastrointestinal bleedings at patients with a Cristmas disease a combination to a peptic ulcer of a stomach, a 12-perstny gut leading to a sharp anemization.

One of the most typical symptoms of hemophilia – damage of joints (hemarthrosis). Defeat knee, talocrural, elbow, is more rare – shoulder, hip, radiocarpal and small joints of brushes and feet is usually connected with an injury which in most cases happens to the insignificant and inadequate size of hemorrhage, or arises spontaneously. Damage of joints takes place a certain staging in classical cases: the hemarthrosis – gemartrit – an anchylosis.

The acute hemarthrosis is characterized by sharp joint pain, increase it in volume, a dermahemia and temperature increase over it. In an extensive hemarthrosis the general state worsens, body temperature increases, SOE accrues and the neutrophylic leukocytosis in blood can develop. At the correct timely treatment the first acute hemarthrosis can resolve completely in 2-3 weeks, without leaving significant changes. Repeated hemorrhages cause secondary inflammatory processes in joint tissues with the subsequent development chronic hemorrhagic - a destructive osteoarthrosis with the significant permanent deformation of a joint, an anchylosis, an atrophy of muscles of extremities and the corresponding radiological changes (osteoporosis, narrowing of articulate cracks, deformation of the articulate ends, metafizarny cross strips of a sclerosis, incomplete dislocations, intra articulate fractures), leading the patient to an invalidization which degree, in many respects defines social suitability of the person.

In diagnosis of hemophilia the family anamnesis is of great importance. However almost at 1/3 patients it negative, and the disease can be suspected on the basis of gematomny type of bleeding, the long delayed bleedings, damages of joints.

Datas of laboratory.

The general blood test at patients during remission usually normal. After plentiful bleeding or in the presence of extensive hematomas the posthemorrhagic anemia of varying severity, a neutrophylic leukocytosis, sometimes – the raised SOE and the increased quantity of thrombocytes is defined. The bleeding time is normal. Retraction of a clot is also not broken. Increase in duration of fibrillation according to Li-Whyte (norm of 5-7 minutes), a calcium clotting time of plasma, heparin time, decrease in consumption of a prothrombin in the course of fibrillation (norm of 80-100%) is characteristic.

The final diagnosis is possible at assessment of high-quality, and quantitative maintenance of separate pro-coagulants that it allows to establish a form of hemophilia and degree of its weight.

Complications in hemophilia appear in process of increase of duration of a disease and, as a rule, are caused by immune mechanisms. This development of a so-called inhibitory form of the hemophilia caused by emergence in blood of sick antibodies anticoagulants – inhibitors VIII or IX of factors which inactivate the anti-hemophilic haemo drugs applied in therapy, appearance of hemolytic anemia, thrombocytopenia, a leukopenia, a glomerulonephritis with an amyloidosis of kidneys and an outcome in HNP. At transfusion hematotherapy there is a danger of transfer of a viral hepatitis, AIDS.

Treatment of hemophilia pathogenetic – introduction to an organism of sick missing blood-coagulation factors in quantity adequate to a specific case for the purpose of a bleeding stop, treatment of consequences of bleedings (a hemarthrosis, hematomas and so forth) and prevention of bleedings.

In hemophilia And VIII, recombinant drugs of a factor VIII apply the cleaned concentrates of a factor of VIII, a high cleaning concentrate of a pork factor. As a last resort, cryoprecipitate use is possible.

In a Cristmas disease such drugs are the cleaned concentrates of a factor of IX, drugs of a recombinant factor IX, concentrates of factors of a prothrombin complex (PPSB).

At inhibitory forms of hemophilia use a concentrate of factors of a prothrombin complex, the recombinant activated factor of VII, Prednisolonum, immunodepressants, desmopressin, also the plasma exchange is effective.

Two programs of treatment of patients with hemophilia are applied: systematic transfusion treatment (at severe forms of a disease) and periodic symptomatic transfusion treatment during the first hours after hemorrhage or an extensive injury.

At a pre-hospital stage haemo static drugs of nonspecific action are used: lagokhilus intoxicating (infusion of 5%, 10% inside on 1 tablespoon, 3-6 times a day; 10% alcohol tincture – on 1 tsp in 1/2 glasses of water of 3-5 times a day; tablets on 0.2 g); an epsilon - aminocapronic acid inside on 1 g, each 4 hours, in/in 100 ml of 5% of solution; Haemophobinum (3% solution inside on 1 tablespoon, 2-3 times a day or for tampons); an absorbable gelatin sponge with thrombin.

Prevention of exacerbations of a disease includes the correct information of parents and the child on the nature of a disease, its complications, consequences, etc.; creation of the guarding mode, since early age, the maximum restriction of traumatic situations, games, sports (swimming is shown), physical education classes, use for protection of joints of the foam guards sewed in clothes; vocational guidance of the patient (brainwork); prevention of infectious and other diseases. Preventive inoculations are resolved (except high doses of gamma-globulin). The place of a prick is recommended to be pressed a finger for several minutes.

In need of treatment of associated diseases of medicine are appointed only inside or intravenously. Such drugs as acetylsalicylic acid, pyrazolon and indolovy derivatives, Brufenum, Euphyllinum, a papaverine, furosemide, nitrofurans, high doses of penicillin and its semi-synthetic analogs, etc. are contraindicated.

Even small surgeries at patients with hemophilia are carried out with obligatory preparation in the form of replacement therapy.

Prevention of hemophilia as diseases assumes establishment of carriage of a pathological gene and prenatal diagnosis of hemophilia And by means of the family analysis of polymorphism of lengths of restrictive fragments of DNA (PDRF the analysis).

AUTOIMMUNE WERLHOF'S DISEASE.

– a disease in which hemorrhagic syndrome is connected with formation of autoantibodies against:

a) antigens of thrombocytes,

b) against antigens of thrombocytes and megacaryocytes,

c) antigens of megacaryocytes,

d) against antigen, the general for thrombocytes and erythrocytes or thrombocytes, erythrocytes and leukocytes.

The immune cytolysis of thrombocytes can be primary when the reason of an autoaggression does not manage to be established also symptomatic – secondary (heteroimmune to thrombocytopenia), caused by destruction of thrombocytes the antibodies which arose in an organism in response to change of an antigenic structure of thrombocytes.

Most often at children's age the idiopathic Werlhof's disease (IWD) which makes 47% among all Werlhof's diseases meets.

The disease occurs in children of all age groups, since chest age (is more often in 3-6 years); in the pubertal period the disease frequency at girls is 2-3 times higher, than at boys.

Etiology.

Can be the previous factors of development of ITP postponed in 2-3 weeks prior to a disease sharp respiratory virus, bacterial infections, use with the medical purpose of medicines, preventive inoculations are more rare (AKDS, protivokorevy, administration of gamma-globulin, etc.). It is not possible to establish a proximate cause of development of ITP more often. However the listed above factors need to be considered rather as pathogenetic.

Pathogenesis.

Immune genesis of ITP is conventional. Thrombocytopenia is caused by the increased destruction of thrombocytes, mainly in a spleen and in the peripheral blood channel, under the influence of the antibodies which are formed, mainly, a splenic pool of lymphocytes. At the same time the products and quantity of megacaryocytes and, respectively, thrombocytes in an inert brain increase in comparison with norm by 2.5-5 times.

It is supposed that autoimmune mechanisms participate in pathogenesis of ITP therefore autoantibodies are developed against own not changed antigen of thrombocytes that defines clinical features of ITP accepting a chronic course. At the same time the therapeutic effect can be gained only when using the immunosuppressive means suppressing an autoaggression or during removal of immunocompetent body – spleens.

In pathogenesis of bleeding at ITP, along with thrombocytopenia, functional features of thrombocytes, their participation in a hemostasis with angiotrofichesky (permeability of a vascular wall) and adhesive and aggregation (formation of a platelet stopper) matter functions.

Clinical picture.

In a course allocate the sharp and chronic ITP forms (lasting purpura more than 6 months) with the periods of aggravation, clinical and kliniko-hematologic remission. At patients with a chronic course of ITP consider the number of a recurrence (with rare, with a frequent recurrence, continuously recurrent course). Distinguish the ITP easy forms at which there is only a skin hemorrhagic syndrome, there are no bleedings. At medium-weight forms bleeding is significant moderately, the number of thrombocytes fluctuates in limits 50-100Õ109/l. At patients with severe forms of ITP the long or plentiful bleedings leading to a heavy anemization are noted, the number of thrombocytes of peripheral blood makes less than 30Õ109/l. However, thrombocytopenia degree not always corresponds to expressiveness of bleeding.

The ITP chronic form most often develops at children of 7-10 years. As a rule, in the anamnesis it is difficult to reveal any previous factor. The gradual beginning at rather satisfactory condition of the patient and normal temperature is characteristic of the ITP chronic form, in the absence of symptoms of intoxication. In the period of crisis in clinical picture ITP on the first in frequency place there is a skin hemorrhagic syndrome – a purpura in the form of polymorphic rashes – ecchymomas from small to the considerable sizes (more than 10 cmin the diameter) and punctate petekhialny rash. Hemorrhages are located asymmetrically on skin of a trunk, extremities, persons, except for a hairy part of the head, palms and feet. Their polikhromnost is characteristic. At 50% of patients of a hemorrhage are localized on a mucous membrane of an oral cavity, tonsils, a back wall of a throat, a soft and hard palate. Hemorrhages in a sclera are sometimes noted.

Hemorrhages usually appear spontaneously, is more often at night. Positive signs of bandage and pinch are characteristic.

The second in frequency symptom at ITP are bleedings which in chronic option of a disease, as a rule, are combined with a skin hemorrhagic syndrome. Persistent, plentiful nasal bleedings, bleedings from a mucous membrane of a mouth, tonsils, a back wall of a throat are most typical. Less often gastrointestinal, renal bleedings meet. At 10% of sick ITP cerebral hemorrhages which can lead to a lethal outcome are noted. Girls the first symptom of a disease can have plentiful long periods.

Datas of laboratory.

In peripheral blood the level of thrombocytes, sometimes is reduced to their total disappearance (norm 150-400Õ109/l). The quantity of erythrocytes and level of hemoglobin is normal, or posthemorrhagic anemia (anisocytosis, a reticulocytosis, a poikilocytosis, a hypochromia of erythrocytes) is noted, the number of leukocytes is not changed. At a research of a gemostaziogramma the lengthening of a bleeding time to 30 minutes and more by the Dyyuka method (is noted at norm within 2-5 minutes), reduction of retraction of a blood clot (less than 75%). Blood clotting according to Li-Whyte, as a rule, normal – 7-10 minutes. The maintenance of plasma blood-coagulation factors is normal. The increased or normal number of megacaryocytes – 0.05-0.15Õ109/l is characteristic of marrow at ITP, it is a lot of young forms, thrombocytes are not found or them very little because of fast receipt them in blood. After splenectomy the quantity of megacaryocytes decreases to norm that needs to be regarded as positive effect of therapy.

Principles of treatment.

In the period of crisis the hospitalization in a specialized hospital is shown. The high bed rest is recommended before recovery of the minimum physiological level of thrombocytes. Both at sharp, and at the ITP chronic form apply:

1) intravenous infusions of immunoglobulin (gamimmun, gamma-globulin) in a dose of 400 mg/kg within 5 days that suppresses immune mechanisms of a course of a disease. This type of therapy can be a splenectomy alternative and also be used at the ITP forms refractory to treatment by corticosteroids, immunodepressants, to splenectomy;

2) corticosteroids (Prednisolonum – 2 mg/kg/days), according to indications in a look pulse therapy;

3) at inefficiency of hormonal means and splenectomy apply immunodepressants (cyclophosphamide, Azathioprinum, vinblastine, etc.), a synthetic androgen danazol;

4) the plasma exchange at ITP leads to almost total disappearance of antithrombocytic antibodies and the CEC from blood of the patient and to the antibodyformation termination;

5) splenectomy is shown only at inefficiency of therapy by immunoglobulin and drugs of steroid hormones at the ITP chronic form and also in sharp cases when it is impossible to stop heavy bleedings, life-threatening patients or the cerebral hemorrhages interfaced to threat. The efficiency of a method is 75-90%;

6) the symptomatic therapy directed to increase in resistance of a vascular wall, improvement of functional properties of thrombocytes and performed with the haemo static purpose: vitamins C, P, A, calcium pantothenate, aminocapronic acid, thrombin, absorbable gelatin sponge, cryotherapy, Adroxonum, Dicynonum, phytotherapy (nettle, dogrose, corn stigmas, water pepper, yarrow);

7) in the profound anemia the transfusion of the washed erythrocytes is possible.

The medicines breaking aggregation properties of thrombocytes are contraindicated (aspirin, Butadionum, indolovy derivatives, etc.).

The scheme of inspection of the patient with a hemorrhagic disease.

When collecting the anamnesis to pay attention on:

- diseases of the haematogenic system among the immediate family (bleeding, hemorrhage, a condition of their emergence, communication with food, diseases);
- family living conditions (food, unhealthy work conditions);
- incidence of a SARS (frequency, weight, reaction of the haematogenic system, appearance of hemorrhages);
- time emergence of the first hemorrhagic signs, their character, variability, duration, weight, repeatability, communication with other diseases.

At objective inspection to pay attention on:

- weight of a state;
- assessment of initial development – by the standard methods (compliance of the child I will increase on physical and psychological indicators);

- integuments, mucous (color, vascular drawing), hemorrhagic rashes (quantity, arrangement, size, depth, dynamics, coloring), hemorrhages;
- bone and muscular systems: morbidity in various departments spontaneous, in an injury (adequacy to an injury), localization, existence or lack of palpatorny changes;
- to perform system inspection with emphasis on nervous system (the general neurologic status for identification of secondary deviations), a cardiovascular system, the sizes of a liver and spleen, survey of a chair (identification of impurity of blood);
- to allocate defeat syndromes on systems;
- to carry out the differential diagnosis of hemorrhagic rash.

At assessment of laboratory results to pay attention on:

- morphological composition of peripheral blood in dynamics;
- standard hemorrhagic complex (quantity of thrombocytes, coagulation time, bleeding time), key indicators of a coagulant system of blood, biochemical blood test (bilirubin free, hematocrit, hepatic tests, prothrombin tests).

Test tasks on a subject.

1. Change is characteristic of an idiopathic Werlhof's disease:
   a) bleeding time
   b) coagulation time
   c) and that and another
   d) neither that, nor another
2. At an idiopathic Werlhof's disease bleeding type:
   a) gematomny
   b) vaskulitno-purple
   c) petekhialno-spotty
   d) mixed
   e) angiomatous
3. At an idiopathic Werlhof's disease thrombocytopenia caused:
   a) insufficiency of formation of thrombocytes
   b) the increased destruction of thrombocytes

c) redistribution of thrombocytes

4. At an idiopathic Werlhof's disease in a miyelogramma it is characteristic:

a) oppression of a megakariotsitarny sprout

b) normal number of megacaryocytes

c) irritation of a megakariotsitarny sprout

5. Of a hemorrhagic syndrome at ITP it is characteristic:

a) symmetry of rashes

b) polymorphism of rashes

c) polikhromnost of rashes

d) asymmetry of rashes

e) existence of favourite localization

e) lack of favourite localization

6. Treatment of a hemorrhagic syndrome at ITP is carried out:

a) transfusion of the trombokoncentrat

b) introduction of missing factors of coagulation

c) Dicynonum

d) Vikasolum

e) heparin

e) Prednisolonum

g) antiagregant

h) α-interferon drugs

7. In the general blood test in a hemorrhagic vasculitis it is characteristic:

a) anemia

b) thrombocytopenia

c) thrombocytosis

d) neutrophylic leukocytosis

e) the accelerated SOE

8. Of a hemorrhagic syndrome at GV it is characteristic:

a) presence of an itching

b) existence of favourite localization

c) tendency of elements to merge

d) asymmetry of rashes

e) existence of petechias

e) nasal bleedings

9. The drugs which are applied to treatment of a hemorrhagic vasculitis:

a) Dicynonum

b) Prednisolonum

c) heparin

d) antiagregant

e) concentrates of blood-coagulation factors

e) trombokoncentrat

10. In hemophilia a bleeding time:

a) it is extended

b) does not change

c) it is shortened

11. Mode of inheritance in hemophilia:

a) linked to X-chromosome

b) autosomal and dominant

c) autosomal and recessive

12. Clinical manifestations of hemophilia And are connected with deficit of a factor:

a) VIII

b) IX

c) X

d) XI

13. In what mechanism of coagulation changes in hemophilia are noted?

a) in external

b) in internal

c) both in external and in internal

14. In a Cristmas disease to a koagulogramma change of tests is characteristic:

a) autokoagulyatsionny

b) the activated partial tromboplastinovy time

c) tromboplastinovy time

d) thrombin time

e) plasma calcium clotting time

e) emergence of products of degradation of fibrin

15. At treatment of hemophilia And are applied:

a) native plasma

b) Prednisolonum

c) Dicynonum

d) trombokoncentrat

e) cryoprecipitate

e) concentrate of a factor of VIII

g) Vikasolum

Situational tasks

Task No. 1

The boy of 6 years, arrived with complaints to temperature, rash on hands and legs.

The disease began sharply, temperature to 38.5ºæ rose, on skin of hands and legs the plentiful hemorrhagic rash developed. For the second day the boy was hospitalized.

State at receipt heavy, it is sluggish, pale. On skin of an extensor surface of hands, legs, buttocks the plentiful spotty and papular symmetrically located hemorrhagic rash. The left eyelid is edematous, an eye is closed. Knee, ankle joints are hydropic and sharply painful. All these phenomena kept within 3 days. For the 5th day of a disease the new wave of hemorrhagic rash and at the same time paroxysmal abdominal pain, vomiting appeared. The boy refused food. An abdominal pain kept 4 days, the morbidity at a palpation was localized around a navel, in the right ileal area, feigning an appendicular syndrome. From the 20th day the diseases of new aggravations were not.

Blood test: Ayr. – 3.4 x 1012/l, Nv – 112 g/l, Leyk. – 6.6 x 109/l (during attacks of an abdominal pain a leukocytosis 18.2 x 109/l), blood clot. – 408 x 109/l, a bleeding time – 3 min. 12 sec., a blood clotting time: the beginning – 1 min. 20 sec., the end – 4 min. 10 sec.

The analysis of urine – without pathology.

Questions:

1. Make the diagnosis, prove it.
2. What surgical complications can develop in this disease?
3. Appoint treatment.

Task No. 2

The girl of 12 years is brought in a hospital with the plentiful uterine bleeding which arose for the first time.

The last 6 months notes frequent, plentiful bleedings, education after microtraumas of hemorrhages of different size, a form and coloring.

Serious condition. Pale. On skin multiple asymmetric hemorrhages (from small to extensive). Several sites of hemorrhage on a mucous membrane of a mouth.

Peripheral lymph nodes, liver and spleen are not increased. ABP of 100/55 mm Hg.

Blood test: Ayr. - 3.0 x 1012/l, Nv – 90 g/l, Tsv. pok. – 0.9, reticulocytes – 10%, Leyk. – 11.2 x 109/l, Blood clot. – 32 x 109/l, e/f – 6%, basophiles – 3%, p.b. – 12%, with / I am 55%, limf – 20%, monocytes – 4%, SOE – 12 mm/h.

Questions:

1. Your diagnosis? Prove your assumptions.
2. What research most informatively for confirmation of the diagnosis?
3. Appoint treatment to this patient.

Task No. 3

The boy of 6 years came to reception of children's hospital with the diagnosis "Rheumatism".

State at moderately severe receipt. Subnutrition. Pale. On skin of the right shoulder an ecchymoma with a diameter of 5 cm. Peripheral lymph nodes are slightly increased, mainly in submaxillary area. Carious teeth are partially debrided. Cardiac sounds are slightly muffled, functional systolic noise. The liver and a spleen are not increased. The neurologic status without features. The right knee joint of spherical shape, the movement in it are limited and sharply painful.

From the anamnesis it is known that the child from early age suffers from the increased bleeding: after slight injuries, hematomas on a trunk and extremities are noted.

At a puncture of the right knee joint gemolizirovanny blood is received.

Blood test: eritr. – 3.8 x 1012/l, NV – 110 g/l, leyk. – 6.5 x 109/l, SOE – 23 mm/h, blood clot – 200 x 109/l.

Blood clotting according to Li-Whyte – 18 min. A calcium clotting time – 450 min. Addition of fresh donor plasma normalized a calcium clotting time, addition is long the stored plasma the calcium clotting time did not change.

Questions:

1. Make the preliminary diagnosis. Prove it.
2. What additional researches need to be conducted?
3. What treatment does this patient need to appoint?

Class in a subject:

"DISEASES OF KIDNEYS AT CHILDREN"

I. Scientific and methodical justification of a subject:

The glomerulonephritis and pyelonephritis have the clinical and pathogenetic features at children. Studying pathology of kidneys in age aspect will promote the best understanding and assimilation of clinic, the principles of treatment and prevention of these diseases.

II. Purpose of activity of students.

The student has to know:

- anatomo-physiological features of an urinary system at children;
- features of an etiology, pathogenesis and patomorfologiya of diseases of kidneys at children;
- additional methods of a research in pathology of kidneys;
- features of clinic of a glomerulonephritis and pyelonephritis at children's age;
- classification of diseases of kidneys at children;
- the reasons promoting a chronic course of a glomerulonephritis and pyelonephritis;
- the principles of treatment and dietotherapy in diseases of kidneys at children;
- features of rehabilitation of children with these diseases.

The student has to be able:

- to collect the anamnesis (genealogical, social) and to mark out risk factors of developing pathology of kidneys;
- to examine the patient with a disease of an urinary system;
- to make the plan of laboratory, X-ray inspections and to estimate their results;
- to make the developed clinical diagnosis on classification;
- to carry out the differential diagnosis with diseases with similar clinical symptomatology;
- to appoint a diet, pathogenetic and emergency treatment by the patient taking into account a form of a disease and degree of a renal failure, to write the prescription on medications;
- to make the plan of further rehabilitation of patients.

III. Content of training:

1. Glomerulonephritis (etiology, patomorfologiya, clinic).
2. Main laboratory symptoms of a glomerulonephritis.
3. Complications of an acute glomerulonephritis.
4. OPN in an acute glomerulonephritis.
5. Principles of treatment of patients with an acute glomerulonephritis (diet, medicamentous therapy).
6. Dispensary observation of the children who had an acute glomerulonephritis.
7. Chronic glomerulonephritis (etiology, forms, stages, morphological changes, laboratory signs).
8. Principles of a dietotherapy and drug treatment, dispensary observation of patients with a chronic glomerulonephritis.
9. Etiology, pathogenesis, morphological changes in pyelonephritis.
10. Clinical, laboratory diagnosis of pyelonephritis.
11. Treatment, prevention, dispensary observation of patients with pyelonephritis.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches,

roentgenograms).

3. Technical means of training.

4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.

2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.

3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.

4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.

5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.

6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge:

1. Anatomo-fiziologichesky features of an urinary system at children.

2. What complaints are shown by patients with pathology of an urinary path?

3. What changes can be revealed at external inspection of such patients?

4. What additional research is appointed to children with pathology of kidneys? What changes at the same time can be revealed?

5. What factors, contributing to development of pathology of an urinary path do you know?

VII. List of questions for check of final level of knowledge:

1. Call etiological factors and pathomorphologic features of a glomerulonephritis at children.
2. List the main clinical and laboratory signs of a glomerulonephritis.
3. What main clinical and laboratory signs of nephritic and nephrotic syndromes?
4. Call complications of an acute glomerulonephritis.
5. List the main symptoms of OPN in an acute glomerulonephritis.
6. Call the principles of treatment of patients with an acute glomerulonephritis (diet, medicamentous therapy).
7. What main objectives are solved at dispensary observation of the children who had an acute glomerulonephritis.
8. Call forms and stages of a chronic glomerulonephritis.
9. What morphological changes and laboratory signs are characteristic in a chronic glomerulonephritis?
10. Call the principles of a dietotherapy and drug treatment, dispensary observation of patients with a chronic glomerulonephritis.
11. What etiological factors, contribute to the development of pyelonephritis in children?
12. Call the main links of pathogenesis and morphological changes in pyelonephritis.
13. What researches does the patient need to conduct for confirmation of the diagnosis of pyelonephritis?
14. Characterize the principles of treatment-and-prophylactic actions and dispensary observation of patients with pyelonephritis.

Information block

ACUTE GLOMERULONEPHRITIS (OGN)

– the acute, cyclically developing after an infection disease of kidneys of infectious and allergic (immunoallergic) genesis which is clinically shown various syndromes (hypostases, a hypertension, a hamaturia).

In classification of OGN one of four clinical syndromes defines its such forms:

1) nephritic (moderate hypostases, hypertensia, an uric syndrome), being the classical or typical beginning of this GN form;

2) nephrotic (prevalence of hypostases, the profound proteinuria – more than 3 gr / days);

3) the isolated uric syndrome (without hypostases and hypertensia);

4) mixed (a nephrotic syndrome with a hamaturia and hypertensia).

On activity of process allocate: period of initial manifestations, period of involution and transition to chronic GN.

The condition of kidneys is estimated: without renal failure, with a renal failure and OPN.

At a nephritic syndrome the hypostases and hypertensia are profound unsharply; a constant symptom is the hamaturia. Hypostases develop in the field of a century (can be absent), the face becomes pastose, skin – sharply pale. The ABP moderately raises, usually by 8-10th day from an onset of the illness it is normalized. The expressiveness of a hamaturia is different. Low it is at this form and a proteinuria (less than 3 gr / days) to which fast decrease is peculiar.

OGN in the form of a nephrotic syndrome meets mainly at children of preschool age.

Clinic: widespread hypostases (up to ascites); urine changes (a high proteinuria, a cylindruria – hyaline, granular and epithelial). The hamaturia is not characteristic of this GN form. Hypertensia is absent or short-term. In blood find a hypoproteinemia, cholesterol content in plasma increases. The general pallor of integuments is observed.

OGN with a nephrotic syndrome, a hamaturia and hypertensia is observed at children of advanced age (12-15 years), on the nature of a course and an outcome is the most severe form of OGN.

Complaints**:** severe headache, asthma.

Clinic: persistent hypertensia, hypostases are less profound, than at a nephrotic form, but the edematous period more long; proteinuria, hamaturia. In an acute period of a disease the oliguria is observed. There is a danger of development of OPN. A constant symptom is the proteinuria, it can be not really massive, but long. The hamaturia or a mikrogemauriya is noted. In an urocheras hyaline, granular cylinders are found. In blood – a hypoproteinemia, a hypercholesterolemia.

The forecast of a disease in the presence of this syndrome always serious.

Chronic GN is characterized by a long recurrent or continuous course. It is formed usually by the end of the 1st year of a disease of OGN at its adverse course. Lead to synchronization: disturbances of the dietary and motive modes, stratification of various infectious diseases, etc.

On the nature of clinical manifestations, allocate three forms of chronic GN:

1) nephrotic;

2) mixed;

3) gematurichesky.

Signs of a nephrotic syndrome are characteristic of *a nephrotic form*: hypostases of various degree of manifestation, proteinuria, hypoproteinemia, гипер-α-глобулинемия$_2$, hypoalbuminemia, hypercholesterolemia and lipidemiya. The urocheras is normal, hypertensia is absent. At most of patients the high-selection proteinuria and normal function of kidneys is observed. Some children can have a restriction of glomerular filtration (50-70 ml/min.). The course of a nephrotic form of chronic GN is more often recurrent.

The mixed form of chronic GN is characterized by signs of a nephrotic syndrome, a hamaturia with hypertensia. Long hypertensia is followed by a hypertrophy of a left ventricle, sometimes with development of acute cardiovascular insufficiency and distsirkulyatorny (vascular) encephalopathy. The proteinuria exceeds 2.5 gr. in day, non-selective, moderate restriction glomerular filtration, early appears and usually disturbance of canalicular functions progresses, anemia, a course of the disease – progressing is observed.

The Gematurichesky form is shown only by an uric syndrome in the form of a hamaturia without proteinuria or in combination with a small proteinuria, hypostases and hypertensia are absent, functions of kidneys are kept the long period.

Depending on expressiveness of symptoms, allocate the next periods: 1) aggravations;

2) partial remission;

3) clinical laboratory remission at which there are no all symptoms of a disease.

At chronic GN it is possible to tell about recovery in case of preservation of full clinical laboratory remission within 5 years and more.

Chronic GN can proceed without renal failure with disturbance of their main functions and chronic kidney disease.

Treatment of the acute, quickly progressing and chronic glomerulonephritis has to be complex (the mode, a rational diet, medicamentous therapy, sanatorium treatment) and individual.

1. The bed rest makes 2-3 weeks.

2. Diet**.** In an active stage table salt is excluded or limited, proteinaceous loading at preservation of a sufficient kalorazh and liquid decreases. The saltless diet is recommended at oligoanuriya, hypostases and hypertensia. Liquid is appointed, on a daily urine of previous day taking into account extrarenal losses. The child has to receive during a day of not less than 500 ml. liquids (taking into account meal). The amount of crude protein is 50-55 gr. in day; Caloric content remains due to increase in carbohydrates and fats.

At a convergence of hypostases the enrichment of a diet with potassium is reasonable.

3. Antibiotics (a penicillinic row, macroleads, tsealosporina of the II-III generation) are appointed in middle-aged doses. In the presence of symptoms of a renal failure, oligoanuriya of a dose of antibiotics decrease up to 1/-1/3 daily doses as their cumulation and emergence of by-effects is possible. Antibiotic treatment duration 3-4 weeks, each 8-10 days it is necessary to change an antibiotic. Nefrotoksichny antibiotics are contraindicated: gentamycin, tetracycline, sisomicin, Kanamycinum, etc.

4. The drugs improving rheological properties of blood – dezagregant (curantyl, trental), anticoagulants (heparin)

5. Glucocorticoids (Prednisolonum, Methylprednisolonum, dexamethasone).

The most widespread scheme – daily prescribing of drug within 1.5-2 months with the subsequent transition to faltering therapy. The full daily dose of Prednisolonum is equal to 1.5-2 mg/kg is appointed in 3 inclusion in the first half of day. The dose of drug decreases each 5-7 days by 2.5-5 mg. Faltering treatment is carried out with gradual increase in a

break from 1 to 4 days (drug – a break receives 4 days 3 days of the patient). The maintenance dose is given in one step, in the morning.

6. Cytostatics (Chlorbutinum, Cyclophosphanum, Azathioprinum) are applied at treatment gormonoustoychivy the GN forms.

7. At an edematous syndrome diuretic drugs – saluretics are shown: hydrochlorothiazide, furosemide (lasixum), Acidum etacrynicum (Uregitum), Clopamidum, Triamterenum, Triampur compositum, Aldactonum.

Besides saluretics, with the diuretic purpose it is possible to use osmodiuretik: albumine in/in 0.5-1.0 g/kg, 10-20% Mannitolum solution on 200-300 ml a day (at the rate of 1 gr. substances on 1 kg. body weights), 20% solution of glucose of 10-15 ml/kg, in/in struyno, reopoliglyukin 10-15 ml/kg.

8. At hyper azotemic syndromes the infusional therapy and alkaline drink, use of a lespeflan inside on 5-15 ml, 3-4 times a day (increase in a dose is possible) 3-4 weeks are also necessary or lespenefrit inside 3-4 teaspoons a day or 2-3 times, in oil on 1 ampoule, a day 3-4 weeks. Apply at azotemias Unithiolum and enterosorbents (enterosgel, Polyphepanum, a smekta) in age doses.

9. At Hypertensinum a syndrome appoint drugs: atenolol, enalapril, ramiprit, captopril, kapoten. Early use of antihypertensives against the background of the medical and guarding mode, sharp restriction of table salt in a diet gives the chance to prevent development of a renal eclampsia.

10. The immunoallergic component of genesis of a glomerulonephritis dictates need of use of antihistaminic drugs (Diazolinum, tavegil, etc.) for age doses courses for 3-4 weeks.

11. Sick with the GN gematurichesky forms appoint Ascorutinum, Vikasolum, Dicynonum, aminocapronic acid in age doses.

12. Non-steroidal anti-inflammatory drugs (NPVP) are inhibitors of prostaglandins and stabilizers of cellular membranes, suppress action of a histamine, serotonin, bradykinin on fabric, possess antitromboticheskу action, immunosuppressive, anticomplementary and fibrinolytic activity. NPVP are used in the standard doses, courses of 4-6 weeks (Voltarenum (Ortophenum), diclofenac-sodium, indometacin, an ibuprofen (Brufenum)).

13. Membrane stabilizing and antioxidant therapy: dimefosfon, lipoic acid, an aloe, magnesium oxide, Essentiale, karsit, Unithiolum, Aevitum, vitamin B, oxygenotherapy).

Dispensary observation.

Frequency of surveys: the pediatrician of 1 times in 2 weeks within 3 months, further 1 r / month within the first year of observation, then 1 times in 3 months within 3 years, further 2 times a year. The nephrologist examines the child within 3 months, further 1 time in 3 months within the first year of observation, further 1 time in 6 months within 3 years, further once a year once a month. The stomatologist, the ophthalmologist and the ENT specialist examine the child 2 times a year, other specialists in indications.

Dispensary observation continues 5 years at all forms of sharp GN and completely normalized blood and urine count tests. Patients with chronically GN are observed before transfer to adult policlinic.

PYELONEPHRITIS

Pyelonephritis (PN) – a nonspecific microbic and inflammatory disease of kidneys with primary defeat of a pyelocaliceal system and tubulointerstitsialny tissue of kidneys.

Etiology: bacterial infection: colibacillus, proteas, staphylococcus, enterococci, mycoplasmas, chlamydias, viruses (adenoviruses, enteroviruses, viruses of herpes and parainfluenza), klebsiyella, pyocyanic stick, streptococcus. The etiological role of mushrooms increased.

Pathogenesis: development of PN is possible in the presence of the virulent and rather massive infection capable to overcome a threshold of individual body resistance.

Major factors of risk: genetic predisposition, immaturity and disturbance of a differentiation of renal fabric; disturbance of an intra renal blood-groove and lymph flow; disturbance of normal outflow of urine; diffusion or segmentary hypoplasia of muscles of an ureter; decrease in the general and local immunological protection; the factors leading to tubulointerstitsialny changes (crystalluria, infections, disturbance of a homeostasis, influence of medicines); bacterial allergy.

In development of secondary PN significance is attached to congenital anomalies of the kidneys and uric ways leading to disturbance of a passage of urine and disorder of a hemodynamics of a kidney, primary secondary tubulopatiya; to dizmetabolichesky nefropatiya, vesicoureteral and to other refluxes.

As primary PN understand not obstructive process, as secondary – obstructive. Sharp PN is characteristic of primary option of a disease and, as a rule, comes to an end in 4-6 weeks with full clinical laboratory remission (80-90%). Chronic PN is diagnosed at preservation of a disease more than 6 months from its beginning or at nalichiya for this period not less than two-three recurrence.

Clinic of sharp Mon.

At children of the first year of life the urinogenny way of spread of an infection prevails. PN is shown by high temperature of a body, the dyspepsia phenomena, chair disorder, proceeds behind a mask of intestinal toxicosis or a SARS with prevalence of neurologic symptomatology (spasms, meningeal symptoms). Such course of the disease is connected with tendency of children of early age to generalization of inflammatory process. Children are uneasy, especially at urination. Urination happens in the small portions, a weak stream, to the long bladder emptying which is followed by stomach muscle tension – suprapubic tension. At babies the disease is shown more often at 4-5-month age.

The beginning of sharp PN at children of advanced age is shown by high fever and disturbance of the general state. Complaints: headache, weakness, fatigue. The stomach at a palpation painful, especially on the course of ureters, is noted the muscle tension of an abdominal wall, a back pain, Pasternatsky's symptom – positive. There are "shadows" around eyes, pastosity a century less often – shins, the phenomenon of a dysuria and dyspepsia. There are a nocturia, enuresis. Muddy urine, contains in it a considerable deposit and flakes, reaction – acid. A characteristic bacteriuria – over 100,000 microbic bodies in 1 ml of urine. In urine, irrespective of age of the child, prevail a neutrophylic leukocyturia, up to a pyuria, protein within 1-2% contains (in daily urine – over 100 mg.), single fresh erythrocytes, hyaline cylinders, a renal epithelium are defined. The relative density of urine is not broken. The hyperazotemia does not arise. Some delay of secretory and excretory function of the canalicular device of nephron is noted.

In blood the neutrophylic leukocytosis, the increased SOE are defined.

On ultrasonography in the acute stage of inflammation – increase in a parenchyma of a kidney, expansion of a pelvis. Increase in the sizes of a kidney happens at the expense

of interstitial hypostasis. The increased pyramids of a medulla and a reinforced renal sine are visible.

Allocate the following main syndromes of sharp PN:

- syndrome of the general intoxication,
- gastrointestinal,
- urological,
- painful,
- dysuric,
- uric.

Clinic of chronic PN depend on a form (primary or secondary), stages of a disease, a course of process, extent of distribution, one - or bilateral damage of kidneys.

Clinical picture (main signs):

- intoxication (a hyperthermia without the catarrhal phenomena, a headache, weakness, a loss of appetite, "shadows" under eyes);
- to abdominal pain and/or waist;
- morbidity at a palpation of area of kidneys, ureteric points.
- morbidity at effleurage on a waist;
- urination disturbance.

PN course options:

• acute manifest onset;

• gradual emergence of symptoms of a disease;

• latent oligosymptomatic course.

Cardinal symptoms of a disease:

- anamnestic data (a dysuria, a pollakiuria, pain and tension in a side, fever, hyper pyrexia attacks without local symptoms);
- morbidity at a palpation of kidneys;
- disturbance of concentration ability of kidneys;
- bacteriuria;
- pathological uric deposit;

– radiological changes, according to excretory, retrograde urography, a selection angiography and a tsistouretrografiya.

Obligatory clinical laboratory methods:

– clinical inspection;
– assessment of physical development;
– measurement of arterial blood pressure;
– complete blood count test (a leukocytosis, a neutrocytosis with shift to the left, increase in SOE)
– clinical analysis of urine (leukocyturia over 10 leukocytes under review);
– urine crops – a bacteriuria, bacterial number are higher than 100,000 microbic bodies in 1 ml. wet);
– Zimnitsky's test – disturbance of a rhythm of a mocheotdeleniye, there can be disturbances of ability to concentrate and dissolve urine;
– research daily excretion of salts (oxalates, urates, calcium, phosphates).

Obligatory special diagnostic methods:

– ultrasound examination of kidneys and bladder with assessment of situation, the sizes, forms and structures of bodies;
– miktsionny tsistouretrografiya;
– excretory urography.

Treatment.

1. Mode:

a) bed – in the presence of intoxication, a hyperthermia, a fever, a pain syndrome, the profound dysuric disorders. Duration is 3-5 days.

b) ward – at normalization of temperature, reduction or elimination of extrarenal symptoms, improvement of an uric deposit – 3-5 days.

c) the general – at elimination of extrarenal symptoms and considerable improvement of an uric deposit.

2. Diet: at extrarenal manifestations appoint a table with moderate restriction of protein (1.5 g/kg) and chloride sodium (2-3 gr. in day). Additional intake of liquid: up to 7

years of 500-700 ml.; 7-10 years – 700-1000 ml.; 10 years – 1.0-1.5 lare more senior. – cranberry or cowberry drink, dry apples and pears decoction. Depending on urine reaction, alternation of the proteinaceous (acidifying) and vegetable (alkalinizing) food is recommended each 3-5 days. It creates unfavorable conditions for growth of bacteria.

3. Antibacterial therapy:

At extrarenal signs and high paraclinic activity (increase in SOE is higher than 30 mm/h, a leukocyturia more than 100 leukocytes in p/zr, a bacteriuria over 100,000 microbic bodies in 1 ml.) before receiving the answer of crops of urine to sensitivity appoint semi-synthetic penicillin in combination with uroseptiky (furagin). Duration of therapy is 10-14 days.

At establishment of gram-negative flora (colibacillus, enterococci, klebsiyella, salmonellas, proteas, a pyocyanic stick) it is necessary to appoint:

• Cefazolin (Kefzolum) of 20-40 mg/kg a day, in 2 introductions;

• Cefalexin of 25-50 mg/kg a day, in 4 introductions;

• Tsefuroksy 30-100 mg/kg a day, in 3 introductions;

• Tsefaklor of 20 mg/kg a day, in 3 introductions;

• Tsefotaksy (klaforan) 50-100 mg/kg a day, in 2-4 introductions;

• Tseftrizoksy (epotsellin) 1.0-2.0 mg a day, in 2-4 introductions;

• 2 times a day drive (roksitromitsin) 1/2 tablets.

In a recurrent infection and existence of the mixed flora the combination of two antibiotics is admissible or it is better – an antibiotic with antibacterial drug:

• the 5-nok (nitroxoline) renders anti-inflammatory and antisclerous effect, 5-10 mg/kg a day, in 4 receptions, orally, after a meal.

• Gramurinum – is effective against a protea, colibacillus, golden staphylococcus. Children are from 2nd to 12 years old and the adult on 2 tablets (0.5 gr.) 3 times a day, orally, after a meal. A course of treatment from 7-10 days to 2-4 weeks.

• Palinum – in multi-infection 1 capsule 2 times a day orally, after a meal.

• Bactrimum (Biseptolum) – to children is 2-5 years on 2 tablets (120 mg.) 2 times a day, 6-12 years 4 tablets (120 mg.) – orally, to food. Formation of crystals of drug in tubules of nephrons is possible therefore carry out prevention.

• Nolicin – on 0.5 tablets, 2 times a day, orally, after a meal.

• Norbaktin – on 1/3-1/2-1 tablet, 2 times a day, orally, after a meal.

• Nitrofuran drugs (furagin, furadonin) 5-8 mg/kg of body weight a day, in 3-4 receptions, orally, after a meal. Are appointed more rare as give numerous side effects. A course of treatment of 7-10 days, if necessary repeat after a 10-15-day break.

Treatment by some antibiotics continues within 4 weeks with drug change each 8-10 days. At a negative two-triple bacteriuria the treatment is continued by one of antibacterial drugs the same cycles (8-10 days), all – up to 4-6 weeks.

After 4-6 weeks of continuous treatment continue treatment on an outpatient basis or in sanatorium by a faltering course of 10-12 days in month) 3 months. Every time drugs change, phytotherapy is in parallel appointed. At secondary PN, during aggravation, treatment is conducted similarly sharp, then the faltering course proceeds up to 6 months.

4. Detoksikatsionny therapy: it is carried out at children with a severe form of a disease. Appoint in/in by drop infusion 10% glucose solution (10 ml/kg), reopoliglyukin (at the rate of 10-20 ml/kg day) and at the end of injection – lasixum (furosemide) in a dose of 1 ml/kg. Natural juice, compotes, kissels, dogrose broths, mineral waters are inside shown (Truskavets, Borjomi, Yessentuki, etc.).

5. Pathogenetic and symptomatic therapy. The desensibilizing drugs (Diazolinum, tavegil, Suprastinum, Phencarolum, Pipolphenum) in age doses, a continuous course 3-4 weeks, during the active period of a disease are appointed.

6. Anti-spastic therapy: it is recommended at a pain syndrome (Nospanum in a dose of 1 mg/kg a day; Baralginum, maxidin); duration – 3-5 days. Use of Euphyllinum in an age dose in / is shown century.

7. In the presence of an immunodeficiency immunomodulators under control of an immunogramma are appointed.

8. Activators of intracellular exchange and membrane stabilizing means – vitamins A, groups B, E, ATP, cocarboxylase, 2.5% solution of a xydibackground, 15% solution of a dimefosfon, Unithiolum in an age dose, courses 3-4 weeks (2-3 times a year at chronic PN). At dizmetabolichesky disturbances use Essentiale, magnesium oxide within 2-3 weeks.

Dispensary observation.

Surveys of experts: the pediatrician – 1 time in 1-2 months, the nephrologist 2 r / year.

Analysis of urine 1 r / month, An. wet across Nechiporenko 1 time in 3 months, crops of urine on flora of 1 times in 3 months.

Duration of dispensary observation is 6 months.

Tasks for independent work:

1. Solve situational problems and tasks of test control.
2. Examine the patient with pathology of an urinary path, describe the revealed changes in state of his health.
3. Write out:
   - klaritromitsin
   - furagin
   - Bactrimum
   - veroshpiron
   - delagil

Independent work of students.

Scheme of inspection of the patient:

When collecting the anamnesis to pay attention on:

- existence at the child of the chronic centers of a streptococcal infection;
- diseases preceding development of a glomerulonephritis or pyelonephritis;
- beginning of a disease and sequence of manifestation of symptoms of a disease;
- the treatment spent at home.

At the general survey to pay attention on:

- color of integuments;
- presence of hypostases, their localization, degree of manifestation;
- ABP;
- condition of a cardiovascular system;
- condition of other bodies and systems (survey of a pharynx, stomach palpation).

At assessment of paraclinic methods of a research to pay attention on:

- analysis of urine (protein content, erythrocytes, leukocytes, cylinders);
- bacteriuria degree;
- crops of urine on flora;
- Zimnitsky's test;
- clearance of endogenous creatinine;
- complete blood count test;
- biochemical analysis of blood (protein content, protein fractions, cholesterol, β-lipoproteids, urea, residual nitrogen, creatinine);
- data of ultrasonography of kidneys (form, size, arrangement, condition of a pyelocaliceal system, existence of concrements); given to excretory urography, a miktsionny tsistografiya.

Situational tasks.

Task No. 1

The patient is 7 years old, came to clinic for the 3rd day of a disease with complaints to a headache, puffiness of the face, shins, emergence of urine in the form of "meat slops".

The child from the first pregnancy proceeding with toxicosis of the first half, the first births in time. Weight at the birth 3150 gr., length is 50 cm, Assessment on a scale Apgar 8/8 points. Prematurity without features. On breastfeeding up to 7 months, it is imparted on age. From 5 years stays on the dispensary registry in connection with an adenoid disease, frequent SARS. The genealogical anamnesis is not burdened.

The real disease began in 2 weeks after the postponed tonsillitis. At receipt a moderately severe state. Skin and visible mucous usual coloring, clean, is noted puffiness of the person, pastosity of shins and feet. The pharynx is unsharply hyperemic, tonsils of the II-III degree, are loosened, without imposings. Cardiopulmonary activity satisfactory, ABP of 130/85 mm Hg. The stomach of a usual form, soft, is available to a deep palpation in all departments, painless. A liver at edge of a costal arch. Kidneys are not palpated, Pasternatsky's symptom negative on both sides. Daily urine of 300-400 ml., urine of red color.

General blood test*:* Ayr - 4.3x 1012/l, Nv - 125 g/l, Leyk - 12.3Õ109/l, p.b. - 5%. with - 60%, e - 5%, l - 24%. m - 6%, SOE - 20 mm/hour.

General analysis of urine: quantity - 70.0 ml, color - red, transparency - incomplete, reaction - alkaline, relative density - 1023, an epithelium - 1-2 in p/z, the erythrocytes - changed cover all fields of vision, leukocytes - 2-3 in p/z, cylinders - granular 3-4 in p/z, protein - 0.9 ‰.

Biochemical analysis of blood*:* crude protein - 65 g/l, albumine - 53%, an alpha 1 globulins - 3%, alfa2-globulins - 17%, beta globulins - 12%, gamma-globulins - 15%, urea - 17.2 mmol/l, creatinine - 1.87 mmol/l, potassium - 5.21 mmol/l, sodium - 141.1 mmol/l, cholesterol - 6.0 mmol/l, a beta - lipoproteids - 2.0 g/l.

Biochemical analysis of urine*:* protein - 600 mg/days (norm - to 200), phosphorus - 21 mmol/days (norm - till 19-32), calcium - 3.6 mmol/days (norm 1.5-4), creatinine - 2.5 mmol/days (norm - 2.5-15), ammonia - 28 mmol/days (norm - 30-65), titratsionny acidity - 40 mmol/days (norm - 48-62), oxalates - 44 mg/days (norm - to 17).

Ultrasonography: a liver, a gall bladder, a pancreas, a spleen without pathology. Kidneys are bred usually, the sizes are not increased, the parenchyma is not changed. ChLS has the ordinary building.

Task:

1. Formulate the developed clinical diagnosis.
2. Make the plan of laboratory and tool inspection.
3. What methods should be used for specification of a functional condition of kidneys?
4. Appoint necessary treatment.
5. Duration of dispensary observation for the patient?

Task No. 2

The girl of 9 years, came to department concerning pains in lumbar area, the speeded-up urination.

The child from the first pregnancy proceeding with toxicosis of the first half. Childbirth on the 38th week. Weight at the birth 3500 gr., length of 52 cm. The period of a neonatality proceeded without features. From children's infections had chicken pox, a rubella. A SARS - it is frequent. Allergoanamnez is not burdened.

The disease was preceded by overcooling. Next day there was a headache, an adynamia, abdominal pain and lumbar area at the left, temperature increased up to 39 °C.

The catarrhal phenomena were not noted. During the next 4 days continued to be in a fever highly, the pollakiuria was observed, urine was muddy.

At receipt in a hospital a moderately severe state. Pale integuments, hypostases were not observed, the body temperature of 38 °C. Pasternatsky's symptom positive on both sides, is more at the left. The palpation of a left kidney is painful. The speeded-up urination is noted.

General blood test: Ayr - 4.5xl012/l, Nv - 140 g/l, Leyk – 10.5kh 109/l, p.b. - 10%, with - 60%, l - 22%, m - 8%. SOE - 28 mm/hour.

General analysis of urine: reaction is neutral, protein - 0.09 ‰, leukocytes - entirely all fields of vision, erythrocytes-1 in p/z, salts - oxalates. - it is a lot of bacterium.

Biochemical analysis of blood*:* crude protein - 72.0 g/l, SRB +++, seromucoid - 0.3, urea - 4.3 mmol/l.

Ultrasonography of kidneys*:* kidneys are located correctly, left - 107x42x13 mm., right - 94x37x13 mm. The echo signal from a collective system is changed on both sides, it is more at the left, is expanded. Suspicion on doubling of a left kidney.

Urine crops*:* colibacillus in number of 100,000 microbic bodies/ml is sowed.

Task:

1. Your diagnosis? Justification of the diagnosis.
2. Describe an etiology and pathogenesis of this disease.
3. Make the plan of further inspection of the child.
4. Specify additional methods of a research for specification of the diagnosis.
5. List functional methods of a research of kidneys.
6. What medical tactics of maintaining child?

Test tasks.

1. Test across Zimnitsky allows to estimate:
   a) concentration function of kidneys
   b) filtrational function of kidneys
2. At disturbance of a passage of urine the pyelonephritis is considered:
   a) not obstructive
   b) obstructive

3. In pyelonephritis is surprised:

a) mucous membrane of a bladder

b) blood and lymphatic system of kidneys

c) tubules, pyelocaliceal device and interstitium

d) ball

4. It is possible to speak about synchronization of process at activity of pyelonephritis:

a) more than 3 months.

b) more than 6 months.

c) more than 1 year

5. At high activity of infectious and inflammatory process in kidneys carrying out is possible:

a) urography

b) tsistografiya

d) Ultrasonography of kidneys

e) radio isotope research of kidneys

e) angiography

6. Intake of liquid in pyelonephritis:

a) it is limited

b) raises

7. Are characteristic of pyelonephritis:

a) intoxication symptoms

b) temperature increase

c) urodynias

d) pains in lumbar area

e) hypostases

8. Laboratory changes in pyelonephritis:

a) bacteriuria

b) hamaturia

c) cylindruria

d) proteinuria

e) leukocyturia

e) azotemia

9. In treatment of pyelonephritis are used:

a) diuretic

b) uroseptik

c) hypotensive drugs

d) antibiotics

e) antiagregant

e) glucocorticoids

10. In an acute glomerulonephritis is surprised:

a) interstitial tissue of kidneys

b) ball

c) tubules, pyelocaliceal device and interstitium

d) cortical and marrow of kidneys

e) mucous membrane of a bladder

11. In therapy of an acute glomerulonephritis with a nephrotic syndrome it is applied:

a) hydrocortisone

b) delagil

c) Prednisolonum

d) kapoten

12. Intake of Prednisolonum inside in an acute glomerulonephritis is carried out:

a) evenly within a day

b) mainly in the morning

13. The acute glomerulonephritis with a nephritic syndrome is characterized:

a) moderate hypostases

b) leukocyturia

c) proteinuria

d) azotemia

e) hamaturia

e) lipidemia

g) hypertension

14. An oliguria is called decrease in a diuresis from standard daily age rates:

a) on 1/3

b) on 2/3

c) on 1/2

15. Prednisolonum is appointed at:

a) to a nephrotic form of a chronic glomerulonephritis

b) to a gematurichesky form of a chronic glomerulonephritis

16. The nephrotic syndrome is characterized:

a) profound proteinuria

b) hypoproteinemia

c) hypertensia

d) lipidemia

e) hypostases

e) hamaturia

17. Are characteristic of the mixed form of a chronic glomerulonephritis:

a) hypotonia

b) profound hypostases

c) hypertension

d) insignificant hypostases

e) lipidemia

e) normal level of cholesterol

g) hamaturia

18. Low indicators of specific weight of urine are designated as:

a) hyperstenuria

b) hyposthenuria

c) isosthenuria

d) oliguria

19. The following symptoms are characteristic of cystitis:

a) intoxication

b) hyperthermia

c) urodynias

d) pains in lumbar area

e) leukocyturia

e) bacteriuria

20. At a nephrotic form of a glomerulonephritis the hypostases have the following characteristics:

a) widespread

b) pastosity century and shins

c) "soft"

d) "dense"

Class in a subject:

"DIGESTIVE TRACT DISEASES AT CHILDREN"

I. Scientific and methodical justification of a subject.

Diseases of digestive organs in structure of somatic population morbidity are high on the list. High level of prevalence of gastroenterological diseases and among children is observed. Beginning at children's age, diseases of the digestive system proceed at adults more hard: carry out to development of heavy pathology from a stomach and 12 perstny guts, cholelithiasis, other departments of a gastrobilliarny system also are involved in pathological process. Compliance with it in a task of standards of the broad specialist has to include studying the reasons and conditions of forming of pathology of digestive organs, methods of clinical, laboratory and tool diagnostics, questions of dispensary observation, the principles of continuity of medical care (pediatricians and therapists).

II. Purpose of activity of students on occupation:

The student has to know:

- anatomo-physiological features of a stomach, 12 perstny guts, pancreas at children;
- characteristic of a pain syndrome;
- the principles of functional and special methods of a research in children's gastroenterological practice;
- principles of classification of diseases of a stomach, 12 perstny guts and pancreas;
- diagnostic criteria of diseases of a stomach and 12 perstny guts, pancreas;
- the principles of treatment and dietotherapy in gastroenterological pathology;
- features of dispensary observation for the child with pathology of digestive organs.

The student has to be able:

- to reveal the factors promoting diseases of a stomach, 12 perstny guts and a pancreas at children;
- to examine the patient with diseases of the digestive system, to estimate a pain syndrome, data of the anamnesis;
- to appoint the plan of additional laboratory, tool, X-ray inspection and to estimate their results;
- to make the diagnosis according to the existing classification;
- to appoint a diet and treatment of patients with pathology of digestive system;
- to make the plan of dispensary observation for children with pathology of a stomach, 12 perstny guts and a pancreas.

III. Content of training:

1. The factors contributing to development of gastroenterological pathology.
2. Semiotics of diseases of the digestive system (chronic gastritis, duodenitis, peptic ulcer).
3. Classification of diseases of a digestive tract.
4. Methods of additional inspection of gastroenterological patients.
5. Principles of therapy and prevention of diseases of digestive tract.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.
3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.
4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.
5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.
6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.
7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.
8. Lectures on pediatrics.
9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge.

1. Anatomo-fiziologichesky features of digestive organs at children.
2. Features of a metabolism at children.
3. Composition of normal indestinal flora at children.
4. The contributing factors to development of pathology of digestive organs.

VII. List of questions for check of final level of knowledge.

1. Characteristic of a pain syndrome in diseases of a stomach and a 12-perstny gut.
2. Features of physical inspection of children with diseases of a stomach and a 12-perstny gut.
3. The principles of functional researches in gastroenterological practice.
4. Classification of diseases of a stomach and 12-perstny gut at children.
5. Radiological and tool methods of a research in diseases of a stomach and a 12-perstny gut. Differential diagnostic characters of chronic gastritis, duodenitis, peptic ulcer.
6. The principles of treatment of diseases of a stomach and 12-perstny gut at children.
7. Features of dispensary observation for children with digestive tract diseases.
8. Dietotherapy in diseases of the digestive system.
9. Intestinal dysbiosis at children.

Information block.

CHRONIC GASTRITIS ALSO GASTRODUADENIT

(HG and HGD) – the chronic recurrent phasic inflammation of the mucous membrane of a stomach (MMS) and duodenum (DPK) which is followed by cellular infiltration, disturbance of phisiological regeneration with tendency to progressing and gradual development of an atrophy of the ferruterous device, disorder of secretory, motor, and quite often incretory function of a stomach.

Among etiological factors HG (HGD) associated with an infection of Helicobacter pylori (NR) prevail. HG (HGD) of the allergic nature, reflux often meet at children (associated with the damaging effect of bile).

Most often (80-85%) hyperacid (normoatsidny) HG (HGD) meet, is more rare – gipoatsidny.

Primary obligatory researches at exacerbation of a disease: the general test of blood, urine, the analysis a calla on the occult blood (as necessary), crude protein and protein fractions. Ezofagogastroduodenoskopiya with an aim biopsy SOZH for definition in

bioptata of Nr and a histologic research; intragastralny rn a rn-metriya of ultrasonography of abdominal organs for definition of the accompanying pathology; definition of Nr by a noninvasive method.

Clinic: at *hyperacid* (normoatsidny) HG (HGD) of an abdominal pain on an empty stomach or in 1-1.5 hours after a meal, mainly in epigastric, piloroduodenalny area, heartburn, an eructation air, acid, nausea, constipations.

At *gipoatsidny* HG (HGD) – an early abdominal pain, right after food, weight in epigastric area, reduced appetite, nausea, an eructation food, air, tendency to a diarrhea (unstable chair), a meteorism.

Physical status: At *hyperacid (normoatsidny)* HG (HGD) – morbidity at a palpation in piloroduodenalny area, epigastriums, Mendel's symptom (+, ++), a syndrome of chronic intoxication: weakness, increased fatigue, vegetative disturbances.

At gipoatsidny HG (State Duma) – an asthenic constitution, subnutrition, morbidity at a palpation of an upper and average third of epigastric area, symptoms of chronic intoxication.

Diagnostics. At an endoscopic research allocate HG (HGD): superficial (catarral, erythematic), mixed, subatrophic, nodulyarny (hypertrophic and hyperplastic), erosive, hemorrhagic. SOZH and DPK, motor disturbances define prevalence, activity of inflammatory and destructive changes (a duodeno-gastralny reflux, duodenostaz, insufficiency of the gatekeeper, etc.). At an intragastralny rn of a rn-metriya or fractional gastric sounding) define the stomach sekretoobrazovaniye (raised, normal, lowered), a kislotoobrazovaniye (hyperacidity, the normoatsidnost, a gipoatsidnost) alkalizing function (normal, reduced).

Bioptata SOZH and DPK taken at endoscopic inspection investigate histologically (diagnostics of and the State Duma), define activity of process, the involvement of the ferruterous device, development of a metaplasia, infection of Nr and also for express methods of definition of Nr.

At X-ray inspection reveal signs of inflammation and motor disturbances of a stomach and DPK.

At ultrasonography reveal the accompanying pathology of biliary tract, a pancreas, liver.

Characteristic of treatment: the basic principles of treatment depend on character and form G (State Duma), activity of inflammatory and destructive process, a disease phase.

At aggravation:

1. Solution of a question of treatment conditions (hospital or policlinic).

2. The choice of the motive mode (sparing or usual), use of LFK.

H. Choice of a diet, purpose of dietary food (table No. 1, No. 2, No. 5).

4. Individual selection of complex treatment (taking into account an etiology, the main pathogenetic mechanisms, the leading symptoms).

At (State Duma), associated with Nr-infektsiyey, with the significant activity of process, medicinal therapy there begin with use eradikatsionny (triple or a quadra) therapies on one of the standard schemes. Preference is given to drugs of bismuth (De-nol) as basic, with parallel prescribing of antisekretony drugs.

Modern schemes of treatment of Nr-infektsii at children:

<u>One-week triple therapy with bismuth drug</u>:

1) colloidal subcitrate of bismuth + amoxicillin (roksitromitsin) or klaritromitsin (azithromycin) + furasolidone (makmiror);

<u>One-week triple therapy with blockers of H+/K +-ATFaza</u>:

2) omeprazolum + amoxicillin (roksitromitsin) or klaritromitsin (azithromycin) + furasolidone (makmiror);

<u>One-week kvadroterapiya</u>:

colloidal subcitrate of bismuth + omeprazolum + amoxicillin (roksitromitsin)

or klaritromitsin (azithromycin) + furasolidone (makmiror)

All drugs are appointed 2 times a day (in the morning and in the evening) within 7 days.

After performing eradikatsionny anti-Nr-terapii continue complex treatment of (State Duma), depending on the nature of acid-forming function of a stomach.

At hyperacid (State Duma) – anti-secretory drugs (mainly blockers of N-histamine $_2$receptors: ranitidine or famotidine.

At the second stage of treatment of (State Duma) and also at a reflux gastritis, a syndrome of dyspepsia of diskinetichesky type appoint aluminum - and the

magniysoderzhashchy not soaking up antacids (maaloks, megalofit, gelyusit, alyumag, fosfalyugel, etc.) on 5-15 ml. (or 1/2-1 tablet) in 1.5-2 hours after meal and before going to bed. A basic course of treatment 2 weeks, further – reception of antacids on demand.

With antacids at motor disturbances appoint prokinetics (motilium, koordinaks, peristit, etc.) on 0.005-0.01 gr. 3 times a day to food, 10-14 days, then – on demand.

In parallel, if necessary, cytoprotectors and reparant for a period of 2-3 weeks are appointed (the smekt – on 1/2-1 bag 3 times a day to food, gastrofarm – on 1/2-1 tablet 3 times a day to food, liquiritonum – on 0.05-0.1 gr. 3 times a day to food, Biogastronum, ventrosol, tsitotek, etc.). At spasms and the expressed pain syndrome – spasmolysants (Nospanum, Platyphyllinum, buskopan).

After cancellation of anti-secretory drugs – metabolik – for improvement of a trophicity SOZH (spirulina), vitamin drugs.

At gipoatsidny (State Duma) – stimulators of gastric secretion (juice of cabbage, plantain, abomin, atsidin-pepsin, Plantaglucidum) for a period of 2-4 weeks in combination with cytoprotectors and reparant (4-6 weeks), if necessary – for 7-10 days.

When subsiding aggravation use phytotherapy (courses for 2 weeks), a balneoterapiya (courses for 2 weeks, alternating to phytotreatment), physical therapy (for normalization of secretory and motor function of a stomach, improvements of a trophicity SOZH use an induktoterapiya, KVCh, diadynamotherapy, magnetotherapy, an electrosleep); at gipoatsidny (State Duma) – galvanization of area of a stomach, an electrophoresis with calcium, electrostimulation by DD-currents), a thermotherapy (paraffin, ozokeritovy appliques, etc.).

PEPTIC ULCER (PU)

– the chronic recurrent disease which is characterized by forming of ulcer defect in a stomach and (or) DPK against the background of inflammatory changes SOZH and DPK with involvement in pathological process of other bodies and systems, development of complications, life-threatening the patient. The polyetiological disease which is genetically determined.

The peak of incidence falls on 9-11 years at girls and 12-14 years at boys.

Primary obligatory researches: the general test of blood, urine, blood typing and a

Rhesus factor, the analysis a calla on the occult blood, a proteinogramm, an ezofagogastroduodenoskopiya with an aim biopsy SOZH and DPK and also (in need of a periultserozny zone), carrying out the ureazny test for identification of Nr, a histologic research of bioptat, ultrasonography of abdominal organs for detection of the accompanying pathology of a liver, biliary tract, a pancreas.

be in addition carried out: X-ray inspection, intragastralny rn rn-metriya of an immunogramm, noninvasive methods of identification of Nr.

Note: treatment outcome for aggravation at YaB are always estimated by clinical and endoscopic trials in dynamics.

Anamnesis of YaBZh: an onset of the illness gradual, a course recurrent, with seasonal aggravations; communication with alimentary disturbances.

Anamnesis of YaB DPK: genetic predisposition (to 75% of patients), communication of aggravation with psychoemotional overloads, a course is significant – recurrent, the seasonality is significant at 1/3 patients.

Complaints at YaBZh: pains (aching are more often) behind a xiphoidal shoot and in epigastric area in 0.5-1.5 hours after meal, vomiting, a loss of appetite to anorexia, nausea, heartburn.

Complaints at YaB DPK: intensive (aching and paroxysmal) pains in an upper half of a stomach to food, night, a moyniganovsky rhythm of the pains which are quite often irradiating in a back, in a waist, heartburn, an eructation acid, vomiting (single is more often), tendency to constipations, a headache, emotional lability.

Physical status: YaBZh – palpatorny morbidity, sometimes tension of an abdominal wall in epigastriums.

YaB DPK – the significant local palpatorny and percussion morbidity in a piloroduodenalny zone, a positive molotochkovy symptom, local muscular tension, zones of a skin hyperesthesia of Sacharyin-Hedda, astenovegetativny manifestations.

Complications are observed at 15-20% of patients with YaB, is more often at boys (bleeding – 80%; deformation and a stenosis – 10-11%; perforation – 7-8%; a penetration – 1-1.5%).

Diagnostics: the diagnosis of YaB DPK and its morphological substrate decides only by means of an ezofagogastroduodenoskopiya on an aim biopsy SOZH and DPK.

Determine topography, number, the sizes, a stage of ulcer defect, state existence of complications, expressiveness, option and activity of Nr accompanying the State Duma, contamination SOZH.

X-ray inspection is conducted only in case of impossibility of performance of an endoscopic research. Absolute R-signs of YaB (a symptom of "niche", convergence of folds towards ulcer defect, cicatricial and ulcer deformations) at children's age come to light only at 18-25% of patients. **Characteristic of treatment:** the basic principles of treatment of YaB depend on topography of ulcer defect (DPK or a stomach), the period of a disease, weight of a course, existence of complications, communication with Nr.

At aggravation:

1. The choice of a diet (purpose of dietary food with gradual transition from table No. 1a, 1b, 1 to a table 5), the choice of the motive mode (sparing, LFK).

2. Individual selection of complex treatment taking into account an etiology, the leading pathogenetic mechanisms and kliniko-endoscopic symptom complex.

At Nr-assotsiirovannoy of YaB: treatment is begun with NR eradikation: one of the standard schemes (triple or a kvadroterapiya) within 7 days with the subsequent confirmation of efficiency of an eradikation (not less than 4-6 weeks after the end of antikhelikobakterny therapy) any two methods of verification of Nr is appointed.

In parallel, or at once upon termination of eradikatsionny treatment, anti-secretory therapy (preference is given to selection blockers of N_2 of a histamine of 2-4 generations (groups of ranitidine, famotidine), selection M-cholinolytics (gastrotsepin), by blockers of H+/K+ of ATP-ase (group of omeprazolum, a pantoprazol, a lanzoprazol, a rabeprazol) for a period of 3-4 weeks with gradual cancellation, or purpose of a maintenance dose is appointed (up to 6-8 weeks).

After cancellation of antikhelikobakterny therapy and decrease in anti-secretory drugs for the term of 3-4 weeks appoint: complex antacids (maaloks, Almagelum, gelyusit other) cytoprotectors (the smekt, a sukralfata, a licorice root, synthetic analogs of prostaglandins, dalargin); reparant (sea-buckthorn oil, tykveol, spirulina, ayekol, drugs of propolis, aloe); immunoproofreaders (plant origin).

At disturbance of motility (refluxes, duodenostaz) – prokinetics for 2-3 weeks.

Symptomatic treatment: sedative drugs (persen, new Passitum) on the 1st tablet (1

measured spoon) 2-3 times a day (3 weeks); antistressorny drugs (Sibazonum) in an age dosage for 10-14 days; spasmolysants (Platyphyllinum, Nospanum, buskopan) – parenterally for 10-15 days.

Average course of drug treatment of aggravation of YaB DPK – 4-6 weeks, YaBZh – 6-8 weeks.

CHRONIC PANCREATITIS (CP)

– a polyetiological disease with the phase progressing course, focal or diffusion and degenerative, destructive changes of acinar fabric, exocrine and endocrine function of the pancreas (P).

Primary obligatory researches: complete blood and urine count test, blood sugar (on an empty stomach), amylase and a lipase in blood, a koprogramma, ultrasonography of abdominal organs.

Clinic. The dominating symptom – pain (intensive, aching, sometimes surrounding, it is localized in a middle part of epigastriß area and to the left of a midline, amplifies after a meal, decreases in a prone position on the left side or on a stomach). Long nausea, the pernicious vomiting which is not giving relief. At decrease in vneshnesekretorny pancreatic activity – signs of a maldigestiya (plentiful, foamy, semi-fluid chair and a meteorism). Astenovegetativny syndrome (general weakness, headache, dizziness, sleep disorder). The lack of a pain syndrome at increase of symptoms of vneshnesekretorny insufficiency is characteristic of the HP latent form. At a palpation of a stomach the morbidity in Dezharden's points (PZh head projection), Kacha (PZh body projection) and Mayo-Robsona (projection of tail department of PZh) is defined.

Diagnostics: in peripheral blood – a moderate leukocytosis, it is frequent – stab shift, a lymphocytosis.

Amylase of blood (urine). Increase by 3-5 times from the first day of aggravation and normalization within 3-4 days. An indicator of circulator disorders in PZh.

Lipase of blood (urine). Increase by 2-3 times from 5-7 in the afternoon aggravations. Indicator of weight of defeat of PZh.

Koprogramma – polyexcrements, "a grease look", a fetid smell, a steatorrhea, a creatorrhea.

Ultrasonography – PZh hyper echogenicity, alternation hyper - and hypogene sites, increase in the sizes, change of contours and a shape of gland, expansion of Virsungov Canal.

Characteristic of treatment:

1. The first three days at the significant aggravation – hunger and according to indications – parenteral nutrition. Then – table No. 5 (on Pevznera).

2. Stopping of a pain syndrome: analgetik (Baralginum, analginum), spasmolysants (papaverine hydrochloride; Nospanum in oil; fenikaberan in oil); cholinolytics (buskopan 10 mg. 2-3 times a day after a meal, Platyphyllinum hydrotartrate of 1-2 ml of 5% of solution, subcutaneously), narcotic analgetik (Promedolum of 0.5-1 ml of 2% of solution, in oil).

3. Oppression of functional activity of PZh. Has the strongest overwhelming effect on the PZh function oktreotid (sandostatin) on 25-50-100 mt. 2-3 times a day, in/in or subcutaneously, within 5 days. Dalargin (synthetic analog of opioid peptides) on 1 mg. 2 times a day, in oil. Pancreatic ribonuclease on 1-3 mg/kg, in/in struyno in 20-40 ml. isotonic NaC1 solution. Indirectly oppress secretion of PZh antacids (Almagelum, maaloks, fosfalyugel, etc.) or N_2 gistaminoblokator of the second or third of generations (ranitidine, famotidine).

4. Reduction of an enzymatic toxaemia is carried out at severe forms of HP by parenteral administration of inhibitors of proteolysis (Contrykal, Trasylolum, Gordoxum, zimofen, etc.). The dose is selected depending on degree of a fermentemiya and a condition of the child.

5. For prevention of purulent complications at severe forms of HP appoint antibiotics of a wide range (cephalosporins, macroleads, aminoglycosides).

6. After stopping of a pain syndrome (in 4-6 days) appoint the pancreatic enzymes which are not containing bile (Pancreatinum, mezim-forte, kreon, pan-citrate) on 1 dragee 3 times in time or after a meal.

Duration of hospital treatment is 28-30 days (in the absence of complications).

Tasks for independent preparation:

1. Solve situational problems and test tasks.
2. Examine and describe in a notebook the revealed changes in a condition of the patient with gastrointestinal diseases.
3. Write in a notebook prescriptions on:
   a) mezim-forte
   b) kreon
   c) omeprazolum
   d) renitdin

e)-nol

Scheme of inspection of the patient.

When collecting the anamnesis pay attention on:

- family anamnesis;
- family predisposition;
- prescription of a disease.

Complaints at survey:

- General symptoms: weakness, slackness, increased fatigue, irritability, headaches.
- Abdominal pain.
- Appetite – preservation, decrease.
- Dispeptic disorders:

a) to food, after a meal, are not connected with meal, often, seldom;

b) vomiting (communication with meal, gives relief or not);

c) heartburn (often, seldom, intensity);

d) eructation (air, food, bitter, acid, rotten egg);

e) intestinal dyspepsia – a meteorism, an intestines hyperperistalsis, a diarrhea, constipations, a chair).

- Structure of sensation of pain.
- Localization of pains – the right hypochondrium, the left hypochondrium, area of a stomach, around a navel, the right ileal area, diffuse morbidity on all stomach.
- Irradiation of pains – the right shoulder, a right shoulder-blade, area of heart, the left hypochondrium. Inguinal area, lumbar area, lower extremities.
- Frequency of pains – daily, 1-2 times a week, 1-2 times a month, seldom.
- Duration of pains – of several minutes till 1 o'clock, 1-2 hours, more than 2 hours.
- Nature of an abdominal pain and their intensity: sharp, stupid, aching, pricking, paroxysmal, surrounding, feeling of a raspiraniye after meal.
- Communication with meal – to food, on an empty stomach, after a meal (at once, in 30-40 min., in 1.5-2 hours), are not connected with meal.
- Communication of pain with physical activity: arise or amplify at fast walking, run.

After exercises, not connected with physical activity.

- Communication with the nature of food: developing of pains after reception – hot dishes, fried, fat, frozen, smoked, cold or carbonated drinks, are not connected with the nature of food.
- Communication of pains with psychological tension (mental traumas, nervousness, fear, etc.).
- Pains pass independently: after intake of milk, soda, drugs, thermal procedures, vomiting.

At an objective research to pay attention on:

- Integuments and mucous – clean, pink coloring, pale. Humidity – norm, dryness or perspiration.
- Intoxication symptoms: a grayish shade of skin, "shadow" under eyes, decrease in elasticity.
- Symptoms of hypovitaminosis: dryness, peeling, cracks in mouth corners, on lips, "perleches".
- Manifestations of exudative diathesis.
- Expansion of venous network – in a thorax, a stomach.
- Peripheral signs.
- Nasal bleedings.
- Hemorrhages on skin.
- Other changes.
- Clean, damp language. Dry, it is imposed with a plaque, "geographical".
- Nature of a plaque: grayish-white, brown, began to smell from a mouth.
- Survey of a stomach: increase in the amount of, it is blown up, muscular tension (in right or left hypochondrium).
- Morbidity at a stomach palpation: in a liver, in a projection of a gall bladder, in epigastriums, around a navel, in a pancreas, on the intestines course (thick, thin, in the Region of a spleen, in the bottom of a stomach).
- Pancreas: morbidity at a palpation in Kass's point, Mayo-Robsona. Condition of intestines

Situational tasks.

Task No. 1

The girl of 11 years, is sick 1 year, complaints to "hungry" pains in epigastriums, appear in the morning on an empty stomach, in 1.5-2 hours after a meal, at night, are stopped meal. Disturb an eructation acid, a chair regular, issued.

Mother of the child has a peptic ulcer of a duodenum, the father has a gastritis, the grandmother in the area of mother has a peptic ulcer of a duodenum. The obstetric and early anamnesis without pathology. Studies at special school of 6 days in a week, 3 times a week are engaged in choreography. On character the introvert.

Survey: height is 148 cm, weight is 34 kg, skin light pink, clean. Stomach: Mendel's syndrome is positive in epigastriums, at a superficial and deep palpation small muscular La Défense and morbidity in epigastriums and piloroduodenalny area, also morbidity in Dezharden's point and Mayo-Robsona. The liver is not increased. On other bodies without pathology.

General blood test: Hb - 128 g/l, C. the item - 0.91, Ayr - 4.2x1012/l; Leyk - 7.2Õ109/l; p.b. - 3%, with / I am 51%, e - 3%, l - 36%, m - 7%, SOE - 6 mm/hour.

General analysis of urine: light yellow color, prozr. half-N; rn - 6.0; density - 1017; there are no squirrels-; sugar - is not present; EDS. C. - 1-2-3 in p/z; leukocytes-2-3 in p/z,

Biochemical analysis of blood: crude protein - 72 g/l, ALT - 19 Pieces/l, nuclear heating plant - 24 Pieces/l, SF - 138 Pieces/l (norm 7-140), amylase - 100 Pieces/l (norm 10-120), thymol turbidity test - 4 pieces, bilirubin - 15 μmol/l, from them svyaz. - 3 μmol/l.

Ezofagogastroduodenoskopiya: mucous a gullet pink, the cardia is closed, In a stomach the muddy slime mucous with focal hyperaemia, in an antruma on walls multiple mixed protrusions. Mucous bulbs duodenum – ochagovo it is hyperemic, hydropic, on a back wall ulcer defect of 0.8x0.6 cm, rounded shape with the hyperemic roller, the bottom is covered with fibrin. The biopsy is taken.

Respiratory ureazny test: positive.

Biopsy test for the NR-infection: positive (++).

Questions:

1. Clinical diagnosis and its justification.

2. Disease etiopathogenesis.

3. Estimate results of the general blood test and whether they correspond to pathology at the child?

4. Modern principles of treatment of this disease.

Test tasks.

2. In chronic gastritis of an abdominal pain:

a) night

b) night and late

c) early

d) depend on localization of gastritis

3. At a chronic gastroduodenit of an abdominal pain:

a) late

b) night

c) early and late

d) late and night

4. An abdominal pain is most characteristic of a peptic ulcer of a 12-perstiy gut:

a) night

b) early and late

c) early and night

d) late and night

5. Drugs possess Antikhelikobakterny action:

a) Almagelum

b) gastrotsepin

c) furasolidone

d) amoxicillin

e) de-Nol

e) Trichopolum

6. At treatment of the diseases associated with Helicobacter pylori it is reasonable to apply a combination of drugs:

a) de-Nol + antibiotic + Trichopolum

b) antacid + de Nol + Trichopolum

c) anti-secretory drugs + antacids + de Nol

d) anti-secretory drugs + de Nol + furasolidone + antibiotic

e) de-Nol + Trichopolum

e) Venter + Trichopolum + antibiotic

7. Of a peptic ulcer of a 12-perstny gut it is characteristic:

a) a normal kislotoobrazovaniye, protective properties of a mucous membrane are reduced

b) the kislotoobrazovaniye is raised, the sekretoobrazovaniye is reduced

c) the kislotoobrazovaniye is reduced, the sekretoobrazovaniye is raised

d) the kislotoobrazovaniye is raised, the sekretoobrazovaniye is raised

8. Clinical signs of bleeding from upper parts of digestive tract are:

a) scarlet blood in a chair

b) weakness

c) girdle pains in a stomach

d) knife-like abdominal pain

e) tar-like chair

e) vomiting "coffee thick"

g) doskoobrazny muscle tension of a stomach

9. Clinical signs of perforation of stomach ulcer and/or a 12-perstny gut are:

a) knife-like pains in anticardium

b) the vomiting which is not giving relief

c) girdle pains

d) liquid chair

e) doskoobrazny muscle tension of a front wall of a stomach

10. Stenozirovaniye of output department of a stomach and/or a 12-perstny gut is shown:

a) eructation

b) the vomiting giving relief

c) expressed meteorism

d) weight loss

e) liquid chair

e) nausea

g) feeling of pressure and completeness in anticardium at once after a meal

11. The pain abdominal syndrome in a peptic ulcer of a 12-perstny gut is characterized:

a) constant character

b) moyninganovsky rhythm

c) chaotic appearance of pain

12. A diagnostic method of gastroduodenal bleeding is:

a) roentgenoscopy of digestive tract with barium

b) survey picture of an abdominal cavity

c) Ultrasonography of an abdominal cavity

d) ezofagogastroduodenoskopiya

13. Complications of a peptic ulcer of a 12-perstny gut are

a) sprue

b) bleeding

c) gullet achalasia

d) penetration

e) perforation

14. For diagnosis of pathology of a stomach and a 12-perstny gut are used:

a) survey picture of an abdominal cavity

b) retrograde pankreatokholangiografiya

c) ezofagogastroduodenoskopiya

d) colonoscopy

21. Specific symptoms of acute pancreatitis at children show

a) vomiting

b) pains in left hypochondrium and/or surrounding

c) gektichesky temperature

d) spotty and papular rash

e) kollaptoidny state

22. Are most informative for diagnostics sharp pancreatitis

a) ultrasonography

b) determination of level of enzymes of a pancreas in blood

c) X-ray inspection

d) koprogramma

23. Biochemical markers of acute pancreatitis are:

a) hyperamilasemia

b) hyperlipasemia

c) disproteinemia

d) decrease in level of standard bicarbonates

e) decrease in level of inhibitor of trypsin

24. Pathogenetic reasonable drugs at treatment of pancreatitis are:

a) streptocides

b) blockers of N-receptors $_2$of a histamine

c) antikholineergichesky drugs

d) sandostatin

e) prokinetics

25. At treatment of a peptic ulcer of a 12-perstiy gut, anti-secretory drugs are used:

a) de-Nol

b) losek

c) ranitidine

d) famotidine

e) fosfalyugel

26. In for the first time the revealed gastritises and a peptic ulcer of a 12-perstny gut associated with a peloric helikobakterioz it is recommended:

a) monotherapy

b) double therapy

c) triple therapy

d) kvadroterapiya

Class in a subject:

"LIVER DISEASES AT CHILDREN"

I. Scientific and methodical justification of a subject.

Diseases of a gepatobiliarny system occupy one of the leading places in structure of pathology of digestive organs children. As a rule, these diseases have continuously recurrent course, often with tendency to progressing of clinical manifestations. Besides functional disturbances of a biliary path, to the real place quite often diagnose such diseases as chronic cholecystitises, cholelithiasis, chronic hepatitis for children. Compliance with it in a task of standards of the broad specialist has to include studying the reasons and conditions of forming of pathology of bodies of a gepatobiliarny system, methods of clinical, laboratory and tool diagnostics, questions of dispensary observation, the principles of continuity of medical care (pediatricians and therapists).

II. Purpose of activity of students on occupation:

The student has to know:

- anatomo-physiological features of a liver and biliary tract;
- the factors promoting a disease of a gepatobilliarny system at children;
- the main symptoms of a disease and gepatobilliarny system at children;
- characteristic of a pain syndrome;
- the principles of functional and special methods of a research in children's gastroenterological practice;
- principles of classification of diseases of a liver and biliary tract;
- diagnostic criteria of diseases of a liver and biliary tract;
- the principles treatment and a dietotherapy in pathology of a liver and biliary tract;;
- features of dispensary observation for the child.

The student has to be able:

- to reveal the factors promoting diseases of a stomach, 12 perstny guts and biliary tract at children;
- to examine the patient with diseases of a gepatobilliarny system, to estimate a pain syndrome, data of the anamnesis;
- to appoint the plan of additional laboratory, tool, X-ray inspection and to estimate their results;

- to make the diagnosis according to the existing classification;
- to appoint a diet and treatment of patients with gepatobilliarny pathology;
- to make the plan of dispensary observation for children with pathology of a liver and biliary tract.

III. Content of training:

1. The factors contributing to development of gepatobiliarny pathology.
2. Semiotics of diseases of bodies of a gepatobiliarny system (chronic hepatitis, chronic cholecystitis, cholelithiasis, dysfunctions of a biliary path).
3. Methods of additional inspection of sick children with chronic diseases of a liver and a biliary path.
4. Classification of diseases of a biliary system.
5. Principles of therapy and prevention of diseases of a gepatobiliarny system.

IV. Educational material security.

1. Visual aids: tables, schemes, multimedia presentations, videos, audiogramma.
2. Educational medical documentation (case histories, laboratory researches, roentgenograms).
3. Technical means of training.
4. Literature.

V. The list of the recommended literature.

1. Children's diseases: the textbook / under the editorship of A.A. Baranov. – M.: GEOTAR-media, 2009. – 1008 pages.
2. Pediatrics: The textbook for medical schools. Under the editorship of N.P. Shabalov. – SPb: SpetsLit, 2006. – 895 pages.
3. Propaedeutics of children's diseases / to N.A. Geppa. – M.: GEOTAR-media, 2009. – 464 pages.
4. N.P. Shabalov. Neonatology: Manual. – M.: MEDpress-inform, 2009.
5. Lectures on pediatrics. A grant for students of medical schools p / an edition M.V. Ehrman. – SPb "Volume", 2001. – 480 pages.

6. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Technique of a research of the child. The study guide for students. – Vladikavkaz, 2011. – 51 pages.

7. Z.D. Kaloyeva, K.M. Dzilikhova, S.K. Karyaeva, etc. Scheme of a case history. The study guide for students. – Vladikavkaz, 2011. – 38 pages.

8. Lectures on pediatrics.

9. Methodical instructions for out-of-class work of students of the 5th course of medical faculty on discipline "Pediatrics".

VI. List of questions for check of initial level of knowledge.

1. Anatomo-fiziologichesky features of digestive organs and zhelchevydelitelny system at children.
2. Features of a metabolism at children.
3. Composition of normal indestinal flora at children.
4. The contributing factors to development of pathology of digestive organs.

VII. List of questions for check of final level of knowledge.

1. Classification of diseases of a bile-excreting system at children.
2. The factors contributing to the development of diseases of biliary tract.
3. Clinical laboratory characteristic of chronic hepatitis at children.
4. Pathogenetic mechanisms of development of cholecystitis and dyskinesia of biliary tract in children.
5. Clinical signs of cholecystitis, dyskinesia of biliary tract on hypo - and to hyperkinetic type.
6. Dietotherapy in diseases of bodies gepatobiliarny sistemyya.

Information block.

DYSKINESIA OF BILIARY TRACT

(ZhVP) – functional disturbances of motility of a gall bladder and/or a tone of the sphincteric device owing to uncoordinated, untimely, insufficient or excessive reduction of a gall bladder and/or the sphincteric device.

Primary obligatory researches: general blood test, general analysis of urine, serumal

cholesterol, alkaline phosphatase, β-lipoproteids, bilirubin the general and fractions; microscopic (fazovokontrastny) and biochemical examination of bile, ultrasonic examination of bodies of a gepatobiliarny system.

Complaints: pain in right hypochondrium, in a navel, paroxysmal (hyperkinetic type) or stupid, aching (hypokinetic type) – after meal, physical or emotional activity; nausea, vomiting, bitterness in a mouth, a loss of appetite, fatigue, emotional lability, dizziness.

Physical status: morbidity at a palpation, in right hypochondrium, in a navel, positive vesical symptoms are possible, at hypokinetic type – increase in the sizes of a liver (soft, mobile, painless, it is quickly reduced after use of holekinetik), it is possible – a distal hyperhidrosis, a pathological dermographism, tendency to an arterial hypertension, functional systolic noise.

Diagnostics:

1. The general blood and urine count test – without features.

2. In hypokinetic type of dyskinesia the moderate biochemical syndrome of a cholestasia is possible (increase in level of serumal cholesterol, β-lipoproteids, cholinesterases, the general bilirubin at the expense of direct fraction).

3. Ultrasonography – lack of indicators of inflammation (consolidation and a thickening of a wall of a gall bladder), reduction of volume of a gall bladder more than for 64% for 60-90 minutes after giving a holekinetik at hyperkinetic type and less than for 34% at hypokinetic type.

4. Ultrasonic multimoment fractional research: a hyper tone of a sphincter of Oddi – lengthening of the 1st and 2nd phases, a hyper tone of a sphincter of Lyutkens – lengthening of the 3rd and 4th phases.

Characteristic of treatment:

1. A diet – table No. 5 on Pevznera.

2. Medicamentous therapy.

A. Hyperkinetic type:

- sedative drugs – infusion of a valerian, sodium bromide, Seduxenum (2-5 mg a day), Relanium (2-8 mg a day); duration of a course and the choice of drug depends on the degree of manifestation of neurologic disorders;
- holespazmolitichesky drugs: Nospanum (1-1.5 mg/kg 3 times a day), Nicospanum

(1 mg/kg 3 times a day), Halidorum (0.05-1 gr. 3 times a day), ditsitet (1 tab. 3 times a day), odeston (1 tab. 3 times a day), spazmopen (1 tab. 2-3 times a day), duration of a course of 10-14 days, if necessary to continue a course, drug needs to be changed;

- choleretics (the drugs stimulating bile synthesis): true (stimulating synthesis of bile acids) – Convaflavinum (1/2-1 tab. 3 times a day), Flaminum (1/3-1 tab. 3 times a day), oksafenamid (0.25-0.5 gr. 3 times a day), febikhol, dekholin, a galstena (5-15 drops 3 times. in day), Cholosasum (1-2 tsps 3 times a day), or the hydrocholeretics (stimulating synthesis of a water component of bile) sodium salicylate. Drugs are appointed a course for 10-14 days;
- physiotherapeutic procedures: thermal procedures (ozokerite, paraffin appliques, a diathermy on area of the right hypochondrium), an inductothermy, an electrophoresis of spasmolysants on area of the right hypochondrium, ultrasound;
- balneoterapiya – mineral waters of a small mineralization and small gas saturation of 3-5 ml/kg of weight on 1 reception, 3 times a day within 1-1.5 months.

B. Hypokinetic type:

- tonic drugs – tincture of a ginseng, Chinese magnolia vine, an eleuterococcus;
- choleretics;
- holekinetik (the drugs stimulating reduction of a gall bladder): cholecystokinin, magnesium sulfate, Pituitrinum P, sorbite (10 ml of 10% of solution of 1 times a day), xylitol, a mannitol, vegetable oils (sunflower, corn, olive), a flax seed;
- prokinetics: motilium (10 mg. 3 times a day), tsizaprid (prepulsid, koordinaks, peristit – 0.2 mg/kg 3 times a day);
- physiotherapeutic procedures: a magnesium sulfate electrophoresis on area of the right hypochondrium, the sinusoidal modulated currents with dirt solution, electrostimulation of a gall bladder;
- balneoterapiya: mineral waters of an average mineralization and average gas saturation (3-5 ml. on 1 kg. masses on reception 3 times a day within 1 month). Duration of hospital treatment – 2 weeks.

Tasks for independent preparation:

1. Solve situational problems and test tasks.
2. Examine and describe in a notebook the revealed changes in a condition of the patient with gastrointestinal diseases.
3. Write in a notebook prescriptions on:
   a) Allocholum
   b) Essentiale
   c) karsit
   d) hofitol
   e) odeston

Scheme of inspection of the patient.

When collecting the anamnesis pay attention on:

- family anamnesis;
- family predisposition;
- prescription of a disease.
-

Complaints at survey:

- General symptoms: weakness, slackness, increased fatigue, irritability, headaches.
- Abdominal pain.
- Appetite – preservation, decrease.
- Dispeptic disorders:

a) to food, after a meal, are not connected with meal, often, seldom;

b) vomiting (communication with meal, gives relief or not);

c) heartburn (often, seldom, intensity);

d) eructation (air, food, bitter, acid, rotten egg);

e) intestinal dyspepsia – a meteorism, an intestines hyperperistalsis, a diarrhea, constipations, a chair).

- Structure of sensation of pain.
- Localization of pains – the right hypochondrium, the left hypochondrium, area of a stomach, around a navel, the right ileal area, diffuse morbidity on all stomach.

- Irradiation of pains – the right shoulder, a right shoulder-blade, area of heart, the left hypochondrium. Inguinal area, lumbar area, lower extremities.
- Frequency of pains – daily, 1-2 times a week, 1-2 times a month, seldom.
- Duration of pains – of several minutes till 1 o'clock, 1-2 hours, more than 2 hours.
- Nature of an abdominal pain and their intensity: sharp, stupid, aching, pricking, paroxysmal, surrounding, feeling of a raspiraniye after meal.
- Communication with meal – to food, on an empty stomach, after a meal (at once, in 30-40 min., in 1.5-2 hours), are not connected with meal.
- Communication of pain with physical activity: arise or amplify at fast walking, run. After exercises, not connected with physical activity.
- Communication with the nature of food: developing of pains after reception – hot dishes, fried, fat, frozen, smoked, cold or carbonated drinks, are not connected with the nature of food.
- Communication of pains with psychological tension (mental traumas, nervousness, fear, etc.).
- Pains pass independently: after intake of milk, soda, drugs, thermal procedures, vomiting.

At an objective research to pay attention on:

- Integuments and mucous – clean, pink coloring, pale. Humidity – norm, dryness or perspiration.
- Intoxication symptoms: a grayish shade of skin, "shadow" under eyes, decrease in elasticity.
- Symptoms of hypovitaminosis: dryness, peeling, cracks in mouth corners, on lips, "perleches".
- Manifestations of exudative diathesis.
- Expansion of venous network – in a thorax, a stomach.
- Peripheral signs.
- Nasal bleedings.
- Hemorrhages on skin.
- Other changes.

- Clean, damp language. Dry, it is imposed with a plaque, "geographical".
- Nature of a plaque: grayish-white, brown, began to smell from a mouth.
- Survey of a stomach: increase in the amount of, it is blown up, muscular tension (in right or left hypochondrium).
- Morbidity at a stomach palpation: in a liver, in a projection of a gall bladder, in epigastriums, around a navel, in a pancreas, on the intestines course (thick, thin, in the Region of a spleen, in the bottom of a stomach).
- Condition of a liver – increase in sizes (the sizes across Kurlov, dense, soft, elastic, painful, painless).
- Condition of biliary tract: Kerr's symptom, Murphy, Ortner's cm, Frenikus-simptom.
- Condition of a spleen: increased, dense, soft, elastic, painful.
- Pancreas: morbidity at a palpation in Kass's point, Mayo-Robsona. Condition of intestines

Situational tasks.

Task No. 1

S.'s belief of 11 years, is sick about a year. Complaints to fervescence, pains in right hypochondrium, amplifying after intake of greasy food, feeling of bitterness in a mouth in the mornings.

Objectively: the state at moderately severe survey, temperature 38.1ºæ, integuments clean, is noted an easy subjikterichnost of scleras. Peripheral lymph nodes (submaxillary, front and zadnesheyny are increased to the 3rd size, mobile, single, painless at a palpation, elastic consistence. In lungs and heart without pathology. The clean pharynx, pink, is a lot of carious teeth. The soft stomach, is blown moderately up, the palpation is painful in right hypochondrium. Kerr, Ortner's positive symptoms, Myussi. The liver and a spleen are not increased. Unstable chair. The diuresis is not broken.

General blood test: Nv –125 g/l, Ayr – 4.7x10 $^{12/l}$, leyk - 12Õ109/l, e e-1, yu-2%, p-10%, with-62%, l-23 of %, sq.m of %, SOE-of 23 mm/hour.

Questions:

1. Preliminary diagnosis.
2. Plan of inspection.

3. Treatment plan.

Test tasks.

1. What of the listed drugs **should not** be applied to stopping of a pain syndrome in bilious colic:

a) atropine

b) papaverine

c) analginum

d) morphine

2. Pains in dyskinesia of biliary tract on hypertensive type have character:

a) surrounding

b) short-term paroxysmal pains in right hypochondrium

c) late pains in an upper half of a stomach

d) constant arching pains in right hypochondrium

e) dull arching aches in paraumbilical area

3. Pains in dyskinesia of biliary tract on hypotonic type have character:

a) surrounding

b) short-term paroxysmal pains in right

c) late pains in an upper half of a stomach

d) constant arching pains in right hypochondrium

4. For final diagnostics of anomalies of a bile-excreting system it is necessary to carry out:

a) Ultrasonography

b) gepatobilistsintigrafiya

c) biochemical research of vesical bile

d) retrograde pankreatokholangiografiya

e) cholecystography

5. In pathogenesis of diseases of biliary tract matter:

a) helikobakterny infection

b) psychological factors

about physical and chemical properties of bile

d) enzymatic insufficiency of a small intestine

e) disturbance of coordinate activity of the sphincteric device

6. Reductions of a gall bladder strengthen:

a) cholecystokinin, gastrin

b) glucagon, calcitonin

c) hypophysis hormones

d) secretin

e) vasoactive intestinal hormone

7. The reasons leading to development of chronic cholecystitis are:

a) disturbance of a diet

b) infectious diseases

c) reflux from a 12-perstny gut in bilious ways

d) food allergy

e) excesses of a gall bladder in the field of a siphon