

Ministry of health care of Ukraine
Highest state scientific institution of the Ukraine
«Ukrainian medical stomatological academy»

"Approved"

at a meeting of the Department of Experimental
and Clinical Pharmacology with Clinical
Immunology and Allergology

Head of the department

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" _____ " _____ 2017 Pr. № _____

**Methodical guidance for students' self-directed
work when preparing for practical session**

Academic discipline	Clinical Immunology and Allergology
Module №1	Clinical Immunology and Allergology
Semantic module №1	Immunological status. Immunodeficiency diseases and immune-pathology
Topic 4	Natural innate immunodeficiency diseases. Age immunology.
Year of study	5
Faculty	medical

1. Relevance of theme:

Immunodeficiency states (IDS immunodeficiencies (IDA), immune deficiency (ID - it changes in the immune system result in violation of state maturation, differentiation, functional activity and the number of cells That participate in the immune response. IDS have a hereditary basis (primary CID and to form a different life periods exposed immunotoxins , Viruses, and other factors (secondary IDS The probability exists in each case of chronic or recurrent microbial-inflammatory , Purulent, parasitic or fungal processes, so IDS study It is relevant and necessary.

2. Learning objectives:

1. Learn the principles of diagnosis and differential diagnosis of primary IDS, Principles of treatment and prevention of IDS .
2. To master the basic principles immunodiagnostics, tactics and approaches to the doctor Combined treatment of T - and B - associates immunodeficiencies.
3. Identify the main features of IDS due to impaired phagocytic immunity deficiency and complement proteins .
4. Learn the etiology and pathogenesis of immune disorders in AIDS.
5. Learn the principles of AIDS prevention.
6. To develop creative abilities in the process of clinical, theoretical, Laboratory studies of patients with primary and secondary immunodeficiency .

3. Basic knowledge, skills necessary for studying the subject (interdisciplinary integration)

The name of the previous disciplines	These skills
Anatomy	Know the structure of the thymus, lymph nodes, plaques, spleen, bone marrow. Spend of examination of patients.
Normal physiology	To know the functioning of the central and peripheral organs of the immune system. Take hold of the basics of clinical and laboratory studies.
Biochemistry	Learn the basics of biochemical laboratory studies. Action and effect of cytokines of different groups of biologically active substances.
Microbiology and Virology	Know the immune response, diagnosis of bacterial and viral infections. Take hold of the basics of special microbiological methods.
Therapy	Know the pathogenesis and clinical manifestations of allergic diseases and secondary immunodeficiencies. To be able to collect immunological and allergic history.
Phthisiatry	To master the mechanisms of the immune response cell dependence, course and clinical manifestations of tuberculosis.
Infectious diseases	Learn the pathogenesis, clinical manifestations of bacterial and viral infections d. To be able to diagnose bacterial and viral infections.

4. Tasks for independent work during preparation for classes.

4.1. List of basic terms, parameters, characteristics that must learn art udent while preparing for the class:

Term	definition
Immune deficiency and its status	a group of diseases which are based on immunopathology
Infection syndrome	a generic term that includes a host of infectious and inflammatory diseases of different etiology (bacterial, viral, fungal, and complementous, parasitic, etc.) and various localization, which is allocated a number of features
Allergic syndrome	immune pathological condition as pathogenetic basis of the clinical manifestations of allergic diseases.
Lymphoproliferative	immune pathological condition characterized by a decrease in

(Cancer Syndrome)	antitumor resistance of the organism and the development of cancer.
Auto immune syndrome	immune pathological condition associated with disturbed self-tolerance mechanisms to the antigens of the body. Clinically manifests itself auto immune diseases auto immune component during the inflammatory process.

4.2. Theoretical questions for the class:

1. Basic principles of classification of immunodeficiency.
2. Natural Born combined immunodeficiencies and immunodeficiencies B- and T-cell links: mechanisms of development, especially the clinical course, and treatment of immunodiagnostics.
3. Natural Born immunodeficiencies phagocytic link of the immune system and the complement system: the development of mechanisms, especially the clinical course, and treatment of immunodiagnostics.
4. The concept of acquired immunodeficiency. The causes, clinical features, immunodiagnostics and immunotherapy.
5. Clinical and laboratory diagnostic criteria, principles of treatment of HIV/AIDS.

4.3. Practical questions for the class:

1. Detecting signs of irritation of the immune system according to leykogram.
2. Know the basic principles of purpose Immunotropic therapy in the treatment of immune complex diseases.
3. Assign immunotropic therapy in treatment of infectious diseases.
4. To be able to determine the prognosis, conduct primary and secondary immunization with immunodependent diseases.
5. Evaluate the effectiveness of immunotherapy assigned dynamically.
6. Conduct immunoglobulin replacement therapy drugs.
7. The ability to independently conduct curation a patient with clinical writing history.

Content topics

Medical history includes The following sections and features:

- Title page previous and clinical diagnosis;
- Complaints, medical history, personal history, where the student must pay special attention to previous Possible signs of immunosuppression and its causes;
- When carrying out an objective review you need to pay special attention to the investigation of lymphotropic system symptoms of metabolic disorders;
- The formulation of previous the diagnosis is necessary to allocate the basic clinical and immunological syndrome on the basis of which develops Survey plan (taking into account the latest achievements of laboratory immunology two nosological forms are determined for the differential diagnosis;
- In the section, "Etiology. Pathogenesis" special attention you need to pay on the immunological aspects of these issues;
- The disclosure Treatment, prevention, prognosis the disease must be considered modern achievements Clinical Immunology;
- In the history of the disease is necessary to bring two diaries progress of the disease with the mapping of the dynamics of laboratory examinations, treatment characteristics.

With the obsession theme for adoption

Innate immunodeficient STATUS

Immunodeficiency state (IDS – a group of diseases, the foundation with is immunopathology.

Immunological status manifest themselves major clinical syndromes:

- infectious disease;
- immunoproliferative/Oncology;
- allergic;

- Hashimoto.

Infection Syndrome - a generic term that includes infectious-inflammatory diseases of different etiology (bacterial, viral, fungal, mycoplasma parasitic, etc.) and various localization, which is allocated a number of features:

- recurrence of acute infection;
- protracted course of disease;
- pronounced tendency to generalize infection;
- a high risk of chronic diseases with frequent exacerbations and the progressive nature of the course of the pathological process;
- quick accession conditionally pathogenic microflora;
- Home the role of mixed infections in the formation of inflammatory process;
- unusual pathogens;
- atypical forms of disease;
- heavy course of the disease;
- opportunistic infections;
- resistance standard therapy (a combination of antibiotics, the need intravenous introduction of antibiotics, their long-term use and frequent change no etiological recovery after repeated treatments.

Allergic syndrome - a immunopathological state As clinical pathogenetic basis manifestations of allergic diseases. Violation of immunity in the form of change differentiation processes immunoregulatory T-lymphocytes, the E overproduction, reduce the isolation of IgA, determine the immune profile of patients with atopic and often predetermined by genetic factors. The clinical manifestations of allergy syndrome is an allergic disease.

Lymphoproliferative th / Cancer Syndrome - is immunopathological state Which is characterized by a decrease in antitumor resistance of the organism and the development of cancer.

Auto immune syndrome - a immunopathological state Connected with disturbed self-tolerance mechanisms to antigens of the body. Clinically discovers themselves autoimmune diseases or autoimmune component during the inflammatory process.

IDS can detect themselves as insulated E syndromes and their combinations of those. There are two big Group immunodeficiencies - Primary (innate and secondary (acquired).

Primary CID - A congenital disorders of the immune system, which characterizes the early implementation of clinical immunopathology . Most primary CID are hereditary conditions . The predominant type of inheritance is autosomal recessive, with many of the classic forms of primary IDS inherited graft X-linked, so in the structure primary IDS up to 80% up boys. The clinical manifestation of primary IDS It begins with the expansion of antigen load in early childhood. In this clinical picture of primary immunodeficiency is determined by the level of experience of the immune system, that is a specific syndrome and constituent factors: living conditions, the state of the local Immunity, heredity, concomitant disease states from other organs and systems, adaptive capabilities of the organism, the timely diagnosis of immunodeficiency Status and curative interventions.

The classification of primary IDS They use the term syndrome. The name of the syndrome foundation take a particular value - determining a defect, eg , Hyper-Ig M syndrome. Bright clinical signs such as ataxia telangiectasia . The etiologic factor, such as Staphylococcus aureus syndrome; names of the authors who first described the syndrome, and the patient's name, as an example with first described the syndrome ohm, syndrome Wiskott-Oldricha, Job's syndrome

Classification of primary IDS (Stephanie D., JE Veltishchev in 1996)

I. Lack of humoral immunity (System B-lymphocytes).

1. X-linked agammaglobulinemia.
2. Disgama globulinemia:
 - a) the total, variable gipogamaglobulinemia;
 - b) selective deficit IgA;
 - c) immunoglobulin deficiency IGG, IGA, with an increase in the synthesis of IgM - hyper IgM syndrome.
 - g) subclass deficiency IGG (absence of IgG1, IgG2, IgG3, IgG4 IgM levels with increasing

or without it.

II. Lack of cellular immune responses (T-cell system).

1. Lymphocytic dysgenesis (Nezelofa syndrome, a French type of IDS)
2. Thymic hypoplasia and parathyroid glands (Di George syndrome).

III. Combined IDS (heavy combined immune deficiency - VKIN)

1. Reticular dysgenesis.
2. Hereditary alymphocytosis, S lymphocytic (Swiss type IDS).
3. Syndrome "bare" lymphocytes.
4. IDS with thymoma.
5. Viskott- Syndrome Aboulafia.

IV. Violation of interleukins in the system cells and cooperative in immune response.

V. IDS in hereditary anomalies exchange.

1. Lack of ADA.
2. Lack purinnucleated.

VI. Insufficient system complement . Lack of phagocytosis.

1. Violation of chemotaxis , Migration and degranulation.
 - a) X syndrome Chediak-IGASOM;
 - b) hyper IgE - syndrome.
2. Violation of the processes of digestion (killing microbes:
 - a) septic granulomatosis;
 - b) lipohomoglystiasis;
 - c) fermentopathy neutrophilic granulocytes: myeloperoxidase deficiency , NADH oxidase, glutathione peroxidase, Glucose-6-phosphate dehydrogenase .
3. Defects opsonization and absorption:
 - a) defects opsonization;
 - b) deficit taftsin;
 - c) the absence of membrane glycoproteins LAF-1, Gp 150, 95, Mac-1 and others.

VIII. Pathology of local immunity.

IX. Small (minor) compensated or immune system abnormalities. Transient (transitional) forms IDS.

Syndrome in primary IDS It differs from the classical concept of the syndrome as a specific symptom . We can not always provide a specific clinical manifestations of the syndrome. For example, selective IgA deficiency clinically may take the form of infectious syndrome (localization - the skin and mucosa), allergic, autoimmune or . At the same time, can not detect clinically themselves with adequate compensation for the defect due to other factors of local immunity selective IgA deficiency . Therefore, in detecting primary IDS We can not advance to forecast the health and life of the patient. Early diagnosis of primary IDS It makes it possible to predict the resistance to decline a certain range of microorganisms and using the preventive and therapeutic measures to prevent the formation of foci of chronic infection of generalized heavy and other infections.

Primary CID - 10 warning signs:

1. Frequent otitis disease (no less than 6-8 times during one year).
2. Several confirmed heavy sinusitis (at least 4-6 times a year).
3. More two confirmed pneumonia.
4. Recurrent deep skin abscesses or internal organs.
5. The need for long-term therapy with antibiotics to kill the infection (up to two months or longer).
6. The need for intravenous antibiotics to kill the infection.
7. At least two deep infections such as meningitis, osteomyelitis Sepsis.
8. Backlog babies in height and weight.
9. Persistent thrush or fungal skin seems to age no more 1 of the year.
10. The presence of relatives of the primary IDA, an early death from severe infections or one of the above symptoms.

If a child is marked more than one of these signs Then immunodeficiency high accuracy.

Frequency IDS in the population, according to different researchers, ambiguous, due to the lack of common programs and methodological limitations. The possibility of laboratory errors. These data are based on generally not immunological and pathological symptoms that are discovered at dead children. Among the different forms of IDS, often (50-75%) of the whole antibodies marked deficits in 20% of cases in combination with immunopathological failure, 10% - cellular immunity, 18% - lack of phagocytosis, 2% - the complement system defects. In some primary IDS, a number of options.

Lack of humoral (B-cell system) - This is a CID with an overwhelming shortage of B-mediated immunity - humoral immune responses.

Agammaglobulinemia Engaged with X-linked (Bruton's disease) is characterized by a significant reduction in the content of Ig in blood, severe hypoplasia, lack of maturation of the body's cells, plasmatic number while preserving reactions of cellular immunity. It is a hereditary disease-linked X chromosome, which affects only boys. It is believed that the pathogenesis of the disease is associated with the lack of B-lymphocyte precursors. The fact that bursectomy of chickens is an exact model of the disease. Significantly reduced maintenance of IGG, IGA and serum IGM, AT after immunization are not formed. Bacterial infections occur the first year of life. The lymph nodes show typical changes that are characterized by the presence of only a T-dependent paracortical area. Cells plasmatic number never seen. The children of the first years of life in the thymus occur late appearance of fatty metamorphosis of the accumulation of fat in the cells of the cortical areas. Children usually suffer severe bacterial lesions of the respiratory and digestive tract, purulent meningitis, sepsis. Despite intensive antibiotic therapy, the prognosis is unfavorable. Death occurs in the first months or first years of life. Agammaglobulinemia often combined with Hodgkin's disease, malignant lymphoma, autoimmune diseases.

Dysgammaglobulinemia. Near hypogammaglobulinemia, which is peculiar to the deficit of all major classes of Ig, happens to form a selective deficiency of one of the classes of Ig. They can be combined with a combined deficiency. Most often it is a selective deficit of IgA, in second place - IgM deficiency, then a selective deficit of secretory IgA, and very rarely - IGG. The heterogeneous group consists of impaired ability of B-lymphocytes to transform into plasma cells. Leading clinical symptoms are chronic bacterial infections of the respiratory, digestive tract, skin lesions. The content of B-lymphocytes in the peripheral blood is not altered, but they can be functionally inferior. With this reduced helper T lymphocyte activity.

Lack of cellular immune responses. Many hereditary diseases (syndromes) are manifested primarily as disorders of T-link of the immune system. Individuals with these disorders are affected by viral and fungal diseases, their complications occur after vaccination with BCG. It is a serious course of herpes, chicken pox. In patients with reduced content of lymphocytes in the peripheral blood and the thymus-dependent areas and organs of the immune system.

Nezelof Syndrome (French type of IDS, alymphocytosis) - a hereditary disease in which there are no cellular responses to them, no protection. Inherited by type. Manifestations of the disease are marked on the first days of life. The syndrome manifests delay in the child's development, protracted septic processes with the cells in the lungs, skin. In peripheral blood - reduced content of lymphocytes, the ratio of CD4⁺/CD8⁺ does not change significantly, suppressed RBTL, the level of all classes maintained in the normal range, the ability to formation of joint-stock company does not change. Histologically show malnutrition of thymus, there are very few lymphocytes in the lymph nodes near the T-dependent areas. The treatment - a bone marrow transplant.

Hypoplasia of the thymus gland and parathyroid glands (Di George syndrome and, agenesis of the thymus, pharyngeal recess syndrome) - a congenital selective defect in cellular immunity. It is characterized by agenesis or hypoplasia of the large thymus and parathyroid glands, the aortic arch, the formation of facial structure violations. The etiology of this disease is not defined. Do not rule out that the pathogenesis associated with the action of environmental agent environment during the critical period of the formation of the W-TU gill pockets of the parathyroid glands, the thymus, the aortic arch (6 - 8 weeks embryogenesis). Clinically note hypocalcemia with an attack of tetany. Death comes suddenly, in the early days or the first two years of life. For this syndrome characteristic flaws in facial structure: hyperthyroidism, antimongoloid shape of the eyes,

a small mouth, mikrogia, low placed ears. There are also esophageal atresia, hypothyroidism. In lymphoid tissue - expressive devastation T-dependent areas. In place of the particles of the thymus - fibrous connective tissue or tissue. Infectious diseases are often caused by a viral or bacterial and gram-negative flora. Children are killed or serious infectious diseases, or congenital defects

Combined IDS. Severe combined IDS (Swiss type of immune deficiency, limfopenichnaya agammaglobulinemia, thymic alymphoplasia, congenital dneplasia thymus) is characterized by impaired cellular and humoral immunity. Inherited by recessive manner. It is believed that the cause of the defect is a lymphoid stem cells. Clinically unstable leuko- and lymphopenia. Quickly changed infectious diseases that lead to death on the 6-8 th month of life. In the early days of life on the skin appear root rash, thrush in the mouth membrane, signs of skin candidiasis. Children are very susceptible to viral infections. The considerable blood lymphopenia (less than $1 \times 10^9/L$), reduced content of T-lymphocytes, an increase of B-lymphocytes.

In some patients the number of T-lymphocytes in the normal range, but they are functionally immature. Reduced maintenance Ig all classes. At postmortem examination lymphoid tissue elsewhere in the state expressive hypoplasia. The thymus consists of retikuloepiteliya, which prevents the formation of thymus cells, lymphocytes little division in the cortical and medullary zone there.

Immunodeficiency syndrome ataxia-telangiectasia (Louis-Bar syndrome) - a deficiency of cellular and partially humoral immunity, which is combined with progressive cerebellar ataxia and okolobulbarnymi teleangiectaze. Inherited by avtosomnotetive type. It is caused by a defect of the final differentiation of T lymphocytes associated with impaired thymic epithelium, in particular, from de defects in DNA repair. It is believed that the progressive loss of CNS and endocrine system depends on the autoimmune process of thymus defects. Immunological study of patients there GTS and shows lymphocyte defect in the RBT of PHA and specific AG. In peripheral blood - lymphopenia, and eosinophilia. Ataxia develops after 4 years of life and progresses, telangiectasia occur in the first year of life. Observed skin lesions, retardation in the physical development and do not develop secondary sexual characteristics. Often there is the development of malignant tumors. At postmortem study - a significant hypoplasia mainly T-dependent areas. In the thymus - fatty metamorphosis do not age-matched. The cerebellum - atrophy of the cortex, the expansion of the IV ventricle.

The lack of the complement system. It is believed that this group of diseases - the most frequent hereditary anomaly of human proteins. The complement system includes over 20 proteins. It occurs hereditary defects of one or more components of the classical (C1-C9) and the alternative pathways (properdin et al.). As a result of gene mutation occurs protein synthesis or produced a protein that has no activity. Most of the defects inherited codominant (Table).DV Stephanie and YE Veltishchev (1996) believe that the failure of the complement system should be provided in such clinical situations:

1. Autoimmune diseases (systemic lupus erythematosus, anaphylactoid purpura, dermatomyositis, chronic glomerulonephritis), if there is no typical clinical and laboratory features.
2. Recurrent pyogenic infections that can occur with a deficit of various factors of the classical and alternative pathways of complement activation. Clinical manifestations are similar to the IDS, but do not have the appropriate laboratory confirmation.

Recurrent meningococcus and gonococcal infections (meningitis, arthritis), are associated with defects in the final components of the complement system.

Hereditary defects in the complement system (JR Firehand, RP Johnston, 1998)

Insufficient - of components	Types inheritance	Disease	infection
Clq	definitely not	Systemic lupus erythematosus, in Ascula, membranoprolifera-tive th glomerulonephritis	Recurrent bacterial, fungal; dermatitis, meningitis
Clq -ingibitor	House inantny (D)	angioedema	
SIs		Systemic lupus erythematosus	

C2	Autosomal-codominant (DCE)	Systemic lupus erythematosus, chronic glomerulonephritis, a disease Shsileyna-Henoch, der matomiozit, trombocitopenic purpura	Septicaemia (usually Streptococcus pneumoniae), meningitis, pneumonia
NW		Membranoproliferativ - ny glomerulonephritis, systemic lupus erythematosus, vasculitis	Heavy generalizing bacterial
C4		Systemic lupus erythematosus, Henoch disease - Henoch, Sergena syndrome	Bacteremia, meningitis
C5		Systemic lupus erythematosus	Disseminative gonococci, stump-coccal; pyoderma, meningitis
C6		Systemic lupus erythematosus, membranoproliferat glomerulonefrit, Sergena syndrome	
C7		Systemic lupus erythematosus, scleroderma, ankylosing spondylitis, rheumatoid arthritis	Disseminative gonococci , stump-coccal ; pioder mia, meningitis
C8	System ink-valued erythematosus	Systemic lupus erythematosus	
C9 Factor D properdin			Tendency to meningococcal meningitis. Relapse Virus sinusitis, bronchitis, bronhoectaziz. Tendency to meningococcal vomu meningitis
SZL-receptor	AKD	Systemic lupus erythematosus	

For primary IDS most characteristic is the infectious syndrome. The most typical localization of lesions is broncho-pulmonary system lororgany, gastrointestinal tract, skin, are associated with increased antigenic load on them. In addition, the compound may infectious syndrome allergic, autoimmune; infectious syndrome with lymphoproliferative / oncology. For example age specific mortality from tumors in groups of primary IDA exceed 10-200 times the expected figure for the general population. The greatest number of tumors observed in patients from ataksieleangiectazy and Wiskott-Aldrich syndrome. With this type of tumors in patients with primary tumors of different BPI that are observed in the population. This usually limforeticular malignancies. Some of these features are reliable clonal proliferation which is associated with infection of Epstein-barr virus. In severe combined immune deficiency (SCID) recorded 50-fold increase in the frequency of gastrointestinal malignant tumors and an increase of approximately 300 times the frequency of lymphoma in women. Today described a significant number of autoimmune slaughtering eases associated with IDA. Among them, pernicious anemia, autoimmune hemolytic anemia, idiopathic trombocitopenic purpura, CAS, tiroidit, Sjogren's syndrome, chronic active hepatitis, myasthenia gravis.

The prognosis of primary IDA defined set of data, with one of the important points have

early diagnosis. The diagnosis of primary IDS can be tiring build only when a special examination, with the use of immunoassays, genetic and molecular analysis techniques. This requires consultation and examination by a clinical immunologist.

Materials for students' self-directed work

A. Tests to verify the initial level of knowledge:

1. Physiology crosshairs blood counts in children occur:

1. The end of the first month of age
2. in the first week of age
3. 8 to 10 years
4. 3 to 4 years

2. For central authorities the human immune system are:

1. Spleen
2. Lymph nodes
3. Bone marrow
4. Minnengitis
5. None of the above

3. Which statements regarding the thymus is wrong:

1. The highest activity is observed in childhood
2. It is the central body of T-lymphocytes
3. Is the main site of synthesis of antibodies
4. In the thymus , and there is the development of antigen lymphocytes

4. For the B-lymphocytes are not peculiar:

1. The ability to recognize the antigen
2. The ability to phagocytosis
3. The cellular cytotoxicity
4. Standalone operation

5. Which statements regarding the bone marrow are wrong:

1. Is the place of antigen-activated lymphocytes
2. It is the central body for B-lymphocytes
3. The highest activity is observed in childhood
4. Combines the features of central and peripheral organs of the immune system

6. Agammoglobul and him , and I caused the Swiss type:

- a) deficiency of complement system;
- b) T cell deficiency;
- c) B cell deficiency;
- g) barrel deficient cells.

7. The main etiologic manifestations of humoral immunodeficiency:

- a) The intracellular pathogen
- b) Simple
- c) extracellular bacteria
- d) Fungal generalizing ir ovannaya flora

8. Girl in 1 g . 3 months. It is in the hospital to determine the cause of periodic attacks clone and tothich their seizures and frequent SARS. Born in due course. The treatment with vitamin D. In the hospital has been vaccinated, BCG - no sign of skin. Vaccination accompanied entsefal measles and partly a reaction. What is the most reliable diagnosis?

A spazmofilii

B hypoparathyroidism

C meningoencephalitis

D Thymic aplasia tgland (D syndrome and -George)

E Agammaglobulinemia, coupling with the X chromosome (X-linked agammaglobulinemia)

9. The child was born on the 8th month of pregnancy. She found: microcephaly, cataracts, heart defect. Having a child in the second month of pregnancy was sick: he was short-lived rise in

temperature to 37.5°C, swollen lymph nodes small rash on the face, trunk and extremities, which took place without residual effects. What is the most likely previous diagnosis of the child and the mother?

- A Rubella
- B cytomegalovirus infection
- C Herpetic infection
- D chlamydia infection
- E toxoplasmosis

10. The baby was born 1.5 years, body mass 3100, length 51 cm . I breastfeed. After the introduction of complementary foods (oatmeal) has ceased to be added to the mass, were emptying with an unpleasant odor in large quantities. Objectively: the phenomenon of malnutrition . II of Art, pale skin, big belly. What is the most reliable diagnosis?

- A Intestinal infection
- B Simple indigestion
- C Cystic fibrosis
- D Inticative
- E worm infestation

11. Annual boy was hospitalized for pneumococ pneumonia . In the history of the child numerous cases of bacterial infections. Four months ago, I had been ill core. An examination of lymphoid tissue underdeveloped. The major classes of immunoglobulins (IgM and IgA) - no, it was found a small amount of IgG, but the phagocytic ability of leukocytes is not broken. What is the correct diagnosis can be made?

- A total variable and pogammaglobulen
- B Syndrome and cattle-Oldrich
- C selective immunoglobulin deficiency
- D Nezelofa syndrome
- E G and pogammaglobulenem and I Bruton

12. In the six-month history of a boy in a variety of frequent infections. Please suffered candidiasis, then he contracted meningitis caused by Haemophilus influenzae, and subsequently developed Pneumocystis I have pneumonia. Taking all this into consideration, we can think about what immunodeficiency?

- A. The failure of B-cells etok differentiate into plasma cells
- B. Failure to CD8 cells to differentiate in the thymus e
- C. Failure stem cells to differentiate into precursors of B and T cells
- D. Failure of neutrophils synthesize enzymes oxygen explosion.

Answers to tests. 12; 2 - 3; 3 -3; 4 -2; -1 5; 6 -; C7; -A 8; -A 9; -A 10; 11 -E; 12 -E. 13 - S.

B. Control of the final level of knowledge:

1. A syndrome , and George is characterized by:

- a) In hypoplasia-dependent nodal zones;
- b) malformations of the second and fourth gill pockets;
- c) hypoplasia, thymus aplasia;
- g) the lack and zogemaglyutin and called on to.

2. By pathological symptoms of illness, Louis -Bar

- a) deficiency of B-lymphocytes, and very low levels of immunoglobulins;
- b) eczema and thrombocytopenia;
- c) ataxia and teleang and project .

3. In a typical manifestation of the syndrome and -Aldrich cattle is:

- a) eczema and tromboditopen and I;
- b) ataxia and teleang and project .

B. lobar pneumonia

4. Violation of neutrophil chemotaxis is typical:

- a) Nezelofa syndrome;
- b) D syndrome and George;

- c) disease Jobs;
- g) syndrome and cattle-Oldrich .

5. For which of the following states of primary immune deficiency characteristic of an extremely high incidence of systemic lupus syndrome vovchanki?

- a) often associated with deficiency of complement component C1 or C2 -C4;
- b) a genetic deficiency and ng and B and C1 of the torus;
- c) Nezelofa syndrome;
- g) H disease and aka-C and quenched.

6. What fragment of the immune system in the first harvest is meningococcal infection?

a) failure on the surface of erythrocytes regulatory molecules which antagonize the effect of lytic complement;

- b) a reduced content properdin;
- c) hereditary deficiency C2-C5 complement fragments;
- d) failure of C3 convertase.

7. Which of the following variants of pathological changes typical of most children's X-linked agamaglobulin ?

a) T-lymphocyte deficiency;

b) the disappearance of lymphocytes from the B and lacorative second zone of the lymph nodes;

c) very low levels of immunoglobulins in normal T-lymphocyte function.

8. What is immunity in the first to suffer when extracellular bacterial) infections, intoxication?

- 1. Natural killer cells
- 2. Macrophages, monocytes
- 3. The components of the complement
- 4. B lymphocytes

9. Baby 8 months. He admitted to hospital with a generalizing fungal infection. In a draft of 2m. Recedivating and purulent infection results of the survey: the spleen and lymph nodes are not palpable; thymus according rentgogram we have not found; hemogram: neutrophil. 95%;

- a) chronic granulomatosis;
- b) X conected agammaglobulinemia;
- c) multiple myeloma
- d) severe combined immune odefitsitny state.

10. Histological examination of the lymph nodes of the child, who suffered from long-term recurrent bacterial infections detected by the absence of the primary and secondary follicular structures. This finding may indicate that:

- a) disease Hodzhkin;
- b) inherent Mr. and pogamaglobulative;
- c) D syndrome and George;
- g) Nezelofa syndrome.

11. When autopsy child (died during the measles epidemic) observed hypoplasia of the para-thyroid gland, thyroid, thymus lack gland. In pathological lymph node study observed the absence of lymphocytes in paracortical a second zone.

- a) X-linked agammaglobulinemia;
- b) D syndrome and George;
- c) disease Hodzhkin;
- g) Chad disease and aka-C and quenched .

12. Pregnant R., 26 years old, was admitted to the maternity hospital in a powerful run. This second pregnancy, the first ended in premature birth and stillbirth. After 30 minutes from the start of attempts born alive girl weighing 3600 g, the cry came at once, leather bottom I slightly icteric, determined by an increase in liver and spleen. Litter flaked off and separated after 15 minutes, the weight of the placenta - 800 The mother - Rh-negative blood type 0 (1) of the group, the child - 0 (1) group, Rh-positive. The baby's blood bilirubin adds 64 micromoles \ u1083 , hemoglobin 160 g \ u1083 ?. Which of the diagnoses most likely?

- A** Birthing tumor
- B** Puerperal intracranial injury newborn
- C** Asphyxia severe
- D** Hemolytic disease of the newborn, jaundice-anemic form
- E** Intrauterine infection

13. The boy was born in asphyxia 40 week 6 pathological pregnancy (there was a danger of breaking, preeclampsia of I type II- half), from 3 genera. Mother of 40 years. The child's condition heavy, the weight of 2 kg, there are signs of immaturity, hydrocephalus symptoms. The skin is pale, yellow, akrotose and cianose. Tone deaf heart, rough systolic murmur at all points of auscultation. Abdomen enlarged, liver 3 cm. The urine is full of feces bright. Optometrist found chorioretinitis. Your previous diagnosis?

- A** Congenital heart defect
- B** Hemolytic disease of newborn
- C** Sepsis
- D** nee toxoplasmosis
- E** nee hepatitis

14. The child is 1 year. After the introduction of complementary foods in the draft last month marked loss of appetite, sweeps from the release of a large amount of feces, sometimes vomiting, body temperature normal body weight 7 kg, very pale, swelling in the legs, stomach greatly increased in volume. In coprogramma lot of fatty acids and soaps, has been appointed a gluten-free . What is excluded from the power of this diet?

- A** animal protein
- B** Milk and dairy products
- C** Fruit
- D** Cereals - wheat, oats
- E** Carbohydrate.

Answers to tests 1 -to; 2 -in; 3 -1; 4 -1; 5 th; 6 -to; C7; 8 -4; 9 -r; 10 -b; 11 -b; -A 12; -A 13; 14 - D.

B. Tasks for self-control:

Task 1. Patient M., 8 months. It located in the pulmonary department with a diagnosis of bilateral pneumonia. In the 7th month old fell ill at otitis media, which is very bad for treatable pneumonia and began the process without achieving remission. History: The child from 2 - pregnancy, was born with a weight of 3.8 kg, to the second month of age grew and developed well, suffered an acute viral infection in 4 months of age. Family history: an older sister, 4 years healthy. On the maternal side there is the case of the boy's death in early childhood (from mother's cousin's sister) from sepsis. These additional surveys: Zag. Blood analysis: Air 4,5h1012 / l; HB 135; KP 0.97; A 6,8h109 / l; B 1%; E 3%; Yu 1%; P / I 13%; C / I of 13%; L and M 28%; Mon 11%; 63% T; V1 8%; 40% Ti; 13% T; ESR 24; Ig G 1 g / l; Ig A, M is not defined.

Question: The diagnosis? Tactics of treatment?

Answer. Bruton's syndrome. The mainstay of treatment - Doven on introduction of immunoglobulin preparations

Task 2.

Patient M., 4 years. Often ill with acute respiratory diseases that end, as a rule, purulent bronchitis, from and including. Double ill with pneumonia (2 years 6 months and 3 years 8 months). From history: up to 6 months was not sick. The first SARS had in 6 months of age. In Year 1 went to kindergarden, then every 2 months is SARS, and in time - bronchitis (with bronchospasm).

Immunogramma: A 6,4h109 / l; B 1%; E 8%; P / I 10%; C / I of 50%; A and m 26%; Mon 5%; 60% T; V1 11%; 42% Ti; 18% T; Th / Ts 2, 3. General sputum analysis: gray, purulent, white blood cells in the whole field of view.

Question: Why is the child's illness? What additional immunological surveys necessary to carry out?

Answer: The boy sign immunodeficiency, mainly on the type of humoral as well . In order to clarify the diagnosis should continue immunological examination with obligatory determination

of immunoglobulin levels, indicators of phagocytosis and CD markers.

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