FEDERAL STATE BUDGETARY EDUCATIONAL INSTITUTION

OF HIGHER EDUCATION
«BASHKIR STATE MEDICAL UNIVERSITY»
OF THE MINISTRY OF HEALTHCARE OF RUSSIAN FEDERATION

DEPARTMENT REPRODUCTIVE HUMAN HEALTH

WITH COURCE OF IMMUNOLOGY

APPROVED by
Head of the department

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Methodical recommendations For professors
to the practice session on the topic:
«Primary immunodeficiencies»

Discipline: Clinical Immunology
Specialty: 31.05.01. «General education»
Course 4
Semester7
Hours: 4

Ufa - 2021

Methodological instructions for students for practical lessons in the discipline "Clinical Immunology " were developed by the faculty of the department in accordance with the work program of the academic discipline (Ufa, 2021), the curriculum (2021) and taking into account the requirements of the Federal State Educational Standard of Higher Education 3 ++ according to specialty 31.05.01 General education (M., 2020).

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1. The theme and its relevance: "Primary immunodeficiencies»

Immunodeficiency, or immunological failure, is a disruption of the immune response, accompanied by the development of recurrent infections and tumors. Primary (congenital) immunodeficiencies, or primary immunological failure, are caused by defects in the genes of immune system cells (IS) and appear in the first months and years of life. They are often fatal.

Primary immunodeficiencies are accompanied by the following clinical symptoms and diseases.

1. Recurrent infections that are not treatable.
2. Persistent opportunistic infections, destructive pneumonia, abscesses.
3. Developmental lag in infants combined with infections and diarrhea.
4. Lymphadenopathy, hepatosplenomegaly, arthritis, scleroderma, dermatomyositis and other autoimmune diseases.
5. Hypoplasia of lymphoid tissue.

Primary immunodeficiencies are associated with genetic defects of one or more components of the immune system, namely: cellular and humoral immunity, phagocytosis, complementary.

Primary immunodeficiencies are relatively rare diseases, the frequency of them is 1 case per 25,000-100,000 people. The exception is selective immunodeficiency Of IgA, which is found at a frequency of 1 case per 500-1000 people.

The severity of humoral immunodeficiency conditions, i.e. antibody formation disorders, depends on the level of genetic defects in lymphocytes. Examples: x-chromosome agammaglobulinemia (Bruton disease), selective IgA deficiency, selective deficiency of the subclasses Of IgG, hyper-IgM syndrome, general variable immunodeficiency, transient hypogammaglobulinemia in newborns.

Primary immunodeficiencies caused by defects in T lymphocytes genes are characterized by a more severe clinical condition than in antibody defects. The insufficiency of T-l manifests itself to varying degrees: from severe combined immunodeficiency (in which T-lymphocytes do not function) to less severe (when the functions of these cells are partially preserved): X-clutched heavy combined immunodeficiencies, severe combined immunodeficiencies with autosomal recessive inheritance mechanism.

Unlike patients with heavy combined immunodeficiencies, patients with combined forms of IDS retain minimal activity of T-lymphocytes.

The most well-known are the following variants of combined immunodeficiencies: ataxia-teleangiectasia (Louis-Bar syndrome), Wiscott-Aldrich syndrome (immunodeficiency, accompanied by thrombocytopenia and eczema), Di Georgie syndrome, impaired expression Grade I and II.

The most well-known variants of the deficit of the phagocytosis system are: chronic granulomatosis disease, Chediac-Higashi syndrome, deficiency of leukocyte adhesion molecules, myelperoxideidase deficiency.

Deficiency of the system of complement and their receptors on the cells leads to the development of various pathological conditions, as complementary proteins are involved in the withdrawal of immune complexes and microbial bodies (hereditary angioneurotic) from circulating blood swelling).

2. Learning purpose: mastering knowledge about the concept of Immunity, types. Factors of non-specific resistance, knowledge of pathogenesis of each type of reaction and clinical manifestations.

To form professional competencies, the student must know:

- anatomical and physiological features of the formation of organs and systems that play a role in the formation of primary immunodeficiency;
- methods of diagnosis of different types of primary immunodeficiency;
- features of clinical manifestations of different types of primary immunodeficiency.

To form professional competencies, the student must be able to:

- collect anamnesis, determine the patient's examination plan for organs and systems;
- determine the plan of additional examination of the patient;
- evaluate the results of clinical and laboratory-instrumental data;
- formulate a diagnosis in accordance with the modern classification;
- assign a treatment plan;
- master the following competencies: GC 1, GC 6, GPC 5, PC 1, PC 5_.

3. Materials for self-preparation to master this topic:

Self-training questions:

1. The concept of primary and secondary immunodeficiency.
2. Classification of primary immunodeficiencies.
3. Issues of diagnosis of primary immunodeficiencies.
4. Defects in humoral immunity (Bruton's agammaglobulinemia, hyper-IgM syndrome, selective IgA deficiency, severe combined immunodeficiency).
5. Defects in cellular immunity (Wiskott-Aldrich syndromes, ataxia-telangiectasias, DiGeorge, hyper-IgE syndrome).
6. Defects in phagocytosis (chronic granulomatous disease, leukocyte adhesion defect).
7. Complement defects, hereditary angioedema.

4. Type of lesson: practical lesson
5. Duration: 4 hours
6. Equipment: computer, projector
7. The content of the lesson.
Technological map of the lesson:

№	Stages of classes and their content	Time in min.	Used visual aids, methodological manuals, etc.	Location of classes	Purpose and nature of the activity	
					of the student's activity	The purpose and nature of the teacher's activity
1	2	3	4	5	6	7
1	Organizational stage	5				
2	Control of students' initial knowledge using test tasks	40	Textbook for classroom and extracurricular work of students Tests.	Study room	Assimilation of theoretical material. Solving typical tasks using tests	Control of the initial level of knowledge.
3	Familiarization of students with the content of the lesson	15	Training tables, slides.			
4	Independent work of students under the guidance of a teacher:	105	Training tests, training situational tasks.		Consolidation of knowledge on the topic, self-examination of the level of assimilation of the material	Control over the work of students.
5	Control of the final level of knowledge and skills on the topic	15	Situational tasks that control test tasks.	Study room		Summing up the lesson. Checking the test results, the level of assimilation of the lesson topic
6	Homework assignment	5				

7.1. Control of the initial level of knowledge and skills. Self-control assignments: students' decision on individual sets of test assignments on the topic

Task 1. Inbound Testing. Choose the correct answers

1. IMMUNE STATUS IS DETERMINED AS QUANTITY AND FUNCTIONAL ACTIVITY
1) T-cells
2) B-cells
3) phagocytes
4) blood coagulation factors
5) indicators of the system of nonspecific resistance

2. HUMORAL IMMUNE RESPONSE IS PROVIDED
1) B-lymphocytes
2) T-lymphocytes
3) mast cells
4) monocytes
5) neurosecretory cells

3. IMMUNE REACTION AS A MULTISTAGE PROCESS INCLUDES
1) antigen recognition
2) release of glucocorticoids
3) production of cytokines
4) the formation of antibodies
5) synthesis of chemokines

4. ANTIGEN TO LYMPHOCYTES IS PRESENTED
1) erythrocytes
2) plasma cells
3) macrophages
4) platelets
5) mast cells

5. PHAGOCYTOSIS IS CARRIED OUT
1) monocyte-macrophage cells
2) hepatocytes
3) B-lymphocytes
4) interferons
5) T-lymphocytes

6. IMMUNITY IS
1) phagocytosis
2) protection from genetically alien living bodies and substances
3) functional activity of T- and B-lymphocytes
4) the number of natural killers
5) the concentration of immunoglobulins

7. IMMUNOLOGICAL DIAGNOSIS INCLUDES
1) collection of immunological history
2) physical examination
3) laboratory immunological studies
4) formulation of the immunological diagnosis
5) Ultrasound of internal organs

8. INDICATIONS FOR ASSESSMENT OF IMMUNE STATUS

1) autoimmune pathology, allergic diseases
2) pathology of pregnancy
3) secondary immunological deficiency
4) control of lipid-lowering therapy
5) genetic defects of the immune system

9. A SPECIFIC IMMUNE RESPONSE IS PROVIDED
1) T-lymphocytes
2) neutrophils
3) platelets
4) basophils
5) eosinophils

10. NON-SPECIFIC PROTECTION IS CARRIED OUT
1) T-helpers
2) class G immunoglobulins
3) macrophages
4) T-killers
5) B-lymphocytes

11. MECHANICAL FACTOR OF NON-SPECIFIC RESISTANCE IS
1) intact surface of the skin and mucous membranes
2) motor activity of smooth muscles
3) function of epithelial cilia
4) lysozyme
5) interferons

12. MARKER OF IMMUNODEFICIENCY IS
1) lymphadenopathy
2) shortness of breath
3) weight loss
4) infection, including opportunistic
5) prolonged fever

13. THE MAIN OBJECTIVE OF IMMUNE
1) anti-infection protection
2) participation in allergic reactions
3) preservation of antigen structural homeostasis
4) ensuring immunological tolerance
5) synthesis of thyroid hormones

14. CLINICALLY IMMUNOPATOLOGY MANIFESTS
1) chronic infections
2) allergies
3) autoimmune pathology
4) lymphoproliferative diseases
5) chest pain

15. EXAMINATION OF A PATIENT WITH A SUSPECTED IMMUNODEFICIENCY STARTS WITH
1) physical examination
2) collection of immunological history
3) laboratory immunological studies
4) formulation of the immunological diagnosis
5) clinical blood test

16. A SPECIFIC IMMUNE RESPONSE IS PROVIDED
1) lysozyme
2) T-lymphocytes
3) eosinophils
4) B-lymphocytes
5) mast cells

17. HUMORAL FACTORS OF NON-SPECIFIC PROTECTION IS
1) eosinophils
2) dendritic cells
3) macrophages
4) complement system
5) acute phase blood proteins

18. DECREASED IMMUNE SYSTEM FUNCTION UNDERSTANDS
1) primary immunodeficiencies
2) secondary immunological deficiency
3) allergies
4) autoimmune pathology
5) mononucleosis

SITUATIONAL TASKS

SITUATIONAL PROBLEM №1

A 10-year-old child often develops bubble rashes on the lips and around the nose, which occur after hypothermia and prolonged insolation. Rashes are often accompanied by malaise, fever up to 37.10C. Bubbles, holding out for 2-3 days, burst, forming erosion. After healing (after 7-10 days), pigmentation remains on the skin. Physical examination revealed no pathology in organs and systems.
1. Formulate a preliminary immunological diagnosis.
2. What is the clinical syndrome of immunopathology?
3. What stages of immunogenesis are disturbed?
4. Determine the preliminary scope of the immunological examination.
5. Determine the tactics of patient management.

SITATIONAL PROBLEM №2

A 45-year-old man went to the doctor with complaints of episodes of body temperature rise up to 38.00C, skin rashes of a rounded shape on various parts of the body that appeared 2 months ago. The man is homosexual and has had one sexual partner for the last 2 years. Never used injectable drugs.

On examination: the general condition is satisfactory. There were 15 purplish-red, nodular, painless and non-itchy rashes on the trunk. Axillary and inguinal lymph nodes are enlarged up to 2 cm in diameter, elastic on palpation, not soldered to each other and the surrounding tissue, painless. From other organs and systems of pathology is not revealed. No changes were found in the clinical analysis of blood.
1. Formulate a diagnosis and justify the diagnosis
2. Make a plan for an immunological examination.
3. Determine the tactics of patient management.
7.2. Analysis with the teacher of the key questions necessary for the development of the topic of the lesson.
7.3. Presentation by the teacher of the methodology for assessing the state of factors of non-specific protection of the body in the laboratory.
7.4. Independent work of students under the supervision of a teacher (draw in a notebook the stages of phagocytosis, the main schemes of complement activation).
7.5. Control of the final level of assimilation of the topic:
The teacher checks the students ' oral answers to the questions of self-preparation.
Checking the presence of drawings of phagocytosis stages and the main schemes of complement activation in the notebooks.
Materials for monitoring the level of development of the topic:
- a set of test tasks,

- situational tasks.
Place of self-training: study room for independent work of students.
Educational and research work of students on this topic (conducted during school hours): working with the main and additional literature.
The main literature

Serial№	Title	Author(s)	Year, place of publication	Number of copies	
				In library	At the department
1	2	3	4	7	8
	Basic Immunology: Functions and Disorders of the Immune System [Текст] : [учебноеиздание]	**A. K. Abbas, A. H. Lichtman, S. Pillai.**	Elsevier, 2016 – 335 p.	80	0

Additional literature

Serial №	Title	Author(s)	Year, place of publication	Number of copies	
				In library	At the department
1	2	3	4	7	8
•	Lectures in immunology: курс лекций	Maianskii, A. N.	N. Novgorod: Publishing house NSMA, 2004 – 256 p.	40	0
•	**IMMUNOLOGY**	**Khaitov R.M.**	**2008 – 256 c.on-line.**	**access mode:** ЭБС «Консультант студента» http:// www.studmedlib .ru/book/ ISBN978597040 7042.html	unlimited access
•	**Fundamental Immunology.**	**Lippincott Williams & Wilkins**	**2008 –on-line**	**access mode:** Database«LWW Medical Book Collection 2011» http://	unlimited access

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