

Ministry of Health of Ukraine
Bogomolets National Medical University

METHODOLOGICAL RECOMMENDATIONS

for practical classes for students

Academic discipline: OK25. Propaedeutics of pediatrics including nursing practice, basic medical skills in the pediatric department

Area of knowledge: 22 "Healthcare"

Specialty: 222 "Medicine"

Pediatrics Department № 2

Approved at the meeting of the department dated **August 27, 2026**, protocol №1

Considered and approved: Central Medical Committee on Pediatric Disciplines

dated August 28, 2026 protocol №1

Topic: Anatomical and physiological features, general semiotics of diseases of the immune system in children

1. Purpose: formation of professional competencies to achieve program learning outcomes in anatomical and physiological features, methods of physical examination and semiotics of diseases of the immune system in children, namely:

student acquisition of knowledge on:

- anatomical and physiological features of the immune system in children;
- general semiotics of diseases of the immune system in children;
- main symptoms and syndromes of damage to the immune system in children, methods of clinical examination in children;
- instrumental and laboratory examination methods for assessing the state of the child's immune system.

student acquisition of skills on:

- evaluation of the results of instrumental and laboratory examination methods for assessing the state of the child's digestive system;
- analysis of the main symptoms and syndromes of damage to the child's digestive system.

2. Competencies:

- compilation of a diagnostic algorithm in children with pathology of the immune system;
- interpretation of the results of instrumental and laboratory examination methods of children with pathology of the immune system.

Stage	Stage description *	Levels of mastery *	Time (min)
Preparatory stage	Organizational measures Setting learning goals, motivating students	*	10
Main stage	Test control on the topic of the lesson, checking and announcing the results. - theoretical survey; - demonstration of practical skills, explaining the most important points regarding the semiotics of diseases of the immune system in children; - students' work on acquiring skills in clinical examination of children with pathology of the immune system;	** *, **, ***	20 55

	- students' acquisition of skills in assessing changes in the state of the immune system in various diseases in children. Solving problems on the topic of the lesson.	****	20
Summary	Analysis and evaluation of the results of students' work. Announcement of the topic of the next lesson, an indicative map for independent work with literature.	*	10
	Total	2.5. academic hours	115

Equipment – textbook, computer, mannequin, phonendoscope, spatula.

Lesson plan and organizational structure

* Familiarization, **reproductive, *** reconstructive, **** creative levels of learning.

3. Topic content. Features of the immune system in children.

!!! Tip: before studying the topic, remember the ontogenesis, anatomy and physiology of the immune system.

- The formation of the immune system begins in the early stages of embryonic development and is genetically determined. The immune status of a newborn depends on:
 - genetic characteristics of the parents;
 - the course of pregnancy and childbirth;
 - environmental conditions.

Feature: organs and cells of the immune system are formed gradually, at different times, and their functional maturity by the time of birth is not the same.

Ontogenesis of the immune system

Immune System Organs and Immunocompetent Cells	Gestation period
Gestation Period Primary stem cell first begins to function	A) Yolk sac at 3rd week B) Liver -3-6 weeks C) Spleen at 3 months Термін рестації
Bone marrow	It is laid at the end of the 3rd month, but only from the 5th month does bone marrow hematopoiesis begin.
Thymus	is formed at 6 weeks Begins to function at 12 weeks.
Lymph nodes: a) peripheral	3 – months 5 – months

b)gastrointestinal tract (mesentery)	
Macrophages	4-week
NK	6-week
TI	11 weeks.
Basic formation of immune system organs, differentiation of T lymphocytes (helpers, suppressors), functional maturity of B lymphocytes	From the 20th week
Ig synthesis СИНТЕЗ Ig	12 weeks
HLA system	6 weeks
Complement system	20 weeks
Beginning of IgA synthesis	30 weeks

It is important to remember "4–6–11–20–30"

2. The main stages of the formation of the immune system. The stem cells from which the cells of the immune system are formed appear sequentially: **yolk sac** (3rd week) → **liver** (6th week) → **spleen** (3–5th month) → **bone marrow** (after 5th month).

3. Formation of the organs of the immune system

- The thymus ensures the maturation and differentiation of T-lymphocytes. starting with its laying (6th week), the formation of Hassall's bodies (12th week), the appearance of thymocytes (14th week) and the completion of the formation of the thymus and the differentiation of T-lymphocytes (20th week).

4. Formation of the cells of the immune system **Macrophages** appear on the 4th week. At the 6th week, the first cells with the function of natural killers (NK) are detected in the liver **T-lymphocytes**—appear at the 11th week, at the 12th week—they are already capable of a functional response, at the 20th week—differentiation of T-lymphocytes into the main functional subpopulations occurs.

!!! T-lymphocytes are formed before their functional capabilities fully mature. Maturation of **B-lymphocytes** begins in the liver. At the 8th week—B-lymphocytes with surface immunoglobulin receptors appear; at the 9th–10th week—IgM receptors; at the 11th–12th week—IgA receptors. At the 20th week, B-lymphocytes acquire the ability to transform into plasma cells and synthesize immunoglobulins. IgA synthesis begins at the 30th week.

Important to remember: Yolk sac → liver → spleen → bone marrow

Thymus → T-lymphocytes

Liver → B-lymphocytes → plasma cells → immunoglobulins

Clinical significance The immaturity of individual components of the immune system in the fetus and newborn explains the age-related features of the immune response and the increased susceptibility of young children to certain infections, as well as understanding the nature of the occurrence of congenital immunodeficiency states.

Features of the fetal immune system

1. **Phagocytosis** is present, but it is not effective enough.
2. **The complement system** is not active enough. At birth: C3 and C4 $\approx$ 50% of the adult level; C8, C9 and MAK $\approx$ 10%. Complement reaches adult levels at approximately 3 months of age.
3. **T-cell and NK** defenses are functionally immature: T-cells can proliferate, but their ability to destroy infected cells is still weak. The above features make the fetus particularly vulnerable to intracellular infections (rubella, herpes, chickenpox, hepatitis viruses; bacteria: Listeria; toxoplasma)
4. **Own synthesis of IgM and IgA** is very low The fetus is already able to synthesize immunoglobulins, but their number is small. At birth: IgM $\approx$ 8–10% of the adult level; IgA $\approx$ 0.4% of the adult level. ***Very important for diagnosis: IgM and IgA do not pass through the placenta.*** Therefore, if they are elevated in the blood of the newborn, it means that they were produced by the fetus itself, this may be a sign of intrauterine infection (if in umbilical cord blood: IgM > 0.2 g/l IgA > 0.05 g/l)
5. The fetus's own synthesis of **IgG** begins around the 15th–20th week, but it is small. However, the level of IgG in the newborn may be even higher than in the mother, because IgG passes through the placenta from the mother to the fetus. This transport occurs especially actively in the last weeks of pregnancy.

!!! A premature baby has a lower level of IgG (less time to receive maternal IgG).

The main value of IgG. Maternal IgG provides the baby with passive protection against infections even before birth and in the first months of life. 6. Interferon and amniotic fluid The fetus, placenta and amniotic fluid contain a lot of α -interferon. but it mainly regulates the development of the immune system, and does not provide a powerful antiviral response. The actual production of a sufficient amount of interferon becomes possible only after 1 year of life. Amniotic fluid contains substances with antibacterial action: lysozyme; transferrin; IgG, which protect the fetus from bacteria.

!!!!It is worth remembering

- IgM + IgA in the newborn $\rightarrow$ own antibodies $\rightarrow$ possible marker of intrauterine infection.
- IgG in the newborn $\rightarrow$ mainly maternal antibodies $\rightarrow$ passive protection.
- Prematurely born $\rightarrow$ less transplacental IgG $\rightarrow$ weaker passive protection.

Features of the functioning of the immune system in newborns.

The immune system of a newborn is already working, but is still functionally immature. After birth, the child first comes into mass contact with antigens through the skin, respiratory tract and especially the intestines. This stimulates the rapid maturation of the lymphoid tissue and the immune system.

1. **Lymphoid tissue** develops rapidly after birth:
 - enlargement of lymph nodes, especially mesenteric;

- active formation of lymphocytes;
- appearance of a large number of plasma cells;
- beginning of active production of antibodies.

Already during the first week of life, the number of lymphocytes in the blood increases significantly, the 1st physiological crossover of the leukocyte formula occurs. Physiological lymphocytosis persists until 4–6 years, when the 2nd crossover occurs.

2. **The number of neutrophils** and their phagocytic activity in a newborn is reduced, which causes weaker protection against bacteria. This is especially true for pustular infections
3. **Macrophages** have a reduced ability to migrate to the site of infection, absorb and digest microorganisms. They are especially insufficiently destroyed by *Haemophilus influenzae* and *Klebsiella*. Therefore, newborns are particularly susceptible to sepsis and meningitis caused by these pathogens.
4. **The inflammatory response** is not sufficiently expressed. Monocytes of newborns produce less IL-1 (stimulates the synthesis of CRP), IL-6 (participates in the stimulation of antibody formation) and TNF- α - an important mediator of inflammation.

**!!!!Clinical significance: in a newborn, infection can occur without high fever.
That is, the absence of severe fever does not exclude severe infection!!!!**

5. Antitoxic protection is weak There are not enough factors that neutralize bacterial toxins. Therefore, newborns are particularly sensitive to endotoxins and may have severe intoxication. Cellular immunity
6. T-lymphocytes are still immature. In newborns, the ability of T lymphocytes to form an effective cytotoxic response is reduced. This is especially true for infections caused by: streptococci. pneumococci. Therefore, the risk of severe bacterial infections is high.
7. Memory T cells are almost absent In a newborn: memory T cells $\approx$ 5%, in an adult $\approx$ 40–50%. Memory T cells are needed for a rapid response upon repeated encounter with an antigen. Therefore, the immune response of a newborn to infection and vaccination is not yet sufficiently formed. Repeated doses of vaccines are necessary to form stable immunity.
8. T and B lymphocytes do not interact effectively enough In newborns, T lymphocytes produce less of some cytokines, and the interaction of T and B cells is not effective enough. B lymphocytes produce mainly antibodies that bind less strongly to the antigen. The response to viruses, gram-negative bacteria, intracellular pathogens, and some parasitic infections is especially weak. Infection can quickly become generalized (sepsis, meningitis, etc.). Humoral immunity
9. The synthesis of immunoglobulins is low, especially: secretory IgA Its deficiency leads to easier penetration of microorganisms through the mucous membranes.
10. IgM - the first line of defense against bacteria IgM is synthesized one of the first during the immune response: agglutinates antigens well; complement, especially important in the fight against gram-negative bacteria. But the newborn's own IgM is not yet enough. Therefore, the risk of gram-negative sepsis is high. Particularly dangerous: *Salmonella*, *Klebsiella* and other gram-negative bacteria.
11. The level of IgG in the newborn is sufficient The newborn receives maternal IgG through the placenta and the child has passive protection against those infections to which the mother has antibodies. Protection is especially important in the first 2–3 months of life. Maternal Ig G does not protect sufficiently against all bacteria and viruses. Therefore, transplacental

antibodies do not provide complete protection, for example, from staphylococci, Escherichia coli, measles, etc.

12. The level of IgG₂ is insufficient. This subclass is important for the response to polysaccharide antigens (pneumococcus, meningococcus). Therefore, newborns are especially sensitive to these bacteria.
13. The number of natural killers (NK) in newborns is reduced, which causes weak antiviral and antitumor protection;
14. 14. The immunity of the newborn has a “suppressor” orientation Despite the fact that newborns formally have more helpers, functionally the immune system of the newborn has a suppressor orientation: in addition, the suppressor function is performed not only by CD8 cells, but also by immature T-lymphocytes, NK cells, and other regulatory mechanisms. This is a protective mechanism aimed at regulating (inhibiting) excessive inflammatory and autoimmune reactions in case of sudden contact with a huge number of new antigens immediately after birth. Therefore, at the beginning of life, the immune system is somewhat "restrained"
15. The skin and mucous membranes are a weak barrier, immature. Together with insufficient phagocytosis, this increases the risk of pustular skin infections. Therefore, careful care of the skin and mucous membranes is very important for the newborn.
16. Complement The activity of complement in newborns is approximately 50% of that in adults.
17. **Lysozyme** is one of the most effective factors Among the factors of the innate immunity of the newborn, lysozyme is of great importance. It damages the cell wall of bacteria.
18. **The thymus (retrosternal gland)** is the central organ of T-cell immunity. After birth, it grows actively: at birth, its mass is 10–15 g, puberty 30–40 g. After puberty → it gradually decreases (age-related involution of the thymus), but in older age its function does not disappear completely.

Accidental transformation of the thymus - temporary reduction of the thymus under the influence of severe stress:

- severe infection;
- glucocorticoids;
- cytostatics;
- starvation;
- radiation;
- other strong stress factors.

At the same time, the number of lymphocytes in the thymus cortex decreases - the organ decreases, its function temporarily decreases.

The main difference:

age-related involution - physiological and gradual; accidental transformation - pathological, but reversible - after eliminating the cause, the thymus can restore its size and function.

Thymomegaly - enlargement of the thymus. An enlarged thymus does not necessarily mean an increase in its function and can occur due to adipose and connective tissue, rather than functional tissue.

Thymus hormones.

- thymosin → stimulates the maturation of lymphocytes and the function of T-helpers;
- thymopoietin → immunomodulation;

- thymulin → participates in the differentiation of T-lymphocytes. The main function of the thymus: maturation + selection of T-lymphocytes, removal of T-cells that incorrectly recognize their own tissues - this helps prevent autoimmune reactions.

Critical periods of development of immunobiological reactivity in children.

There are 5 main critical periods:

- 1. newborn period**
- 2. 3–6 months;**
- 3. 2nd year of life;**
- 4. 4–6 years;**
- 5. adolescence.**

1- period-period of newborn period (see above).

2nd period -3-6 months of life.

1. The suppressive orientation of immune reactions is preserved.
2. Passive immunity is weakened due to the decay of maternal IgG.
3. The primary immunological response is still carried out at the expense of IgM, which does not leave immunological memory, as a result of which whooping cough, chickenpox and other diseases. have an atypical course. There may be repeated diseases.
4. There is still an insufficient amount of IgA, so children often get sick with RS viruses, pneumonias, secondary ARVI.
5. In children of the first months of life, antibodies to some intestinal infections (salmonellosis, escherichiosis, dysentery) are formed weakly and can be more severe and without a sufficient antibody response.
6. Primary or secondary immunodeficiency states (IDS) may appear. That is why this period is called "physiological transient (temporary) hypogammaglobulinemia" - that is, the level of own immunoglobulins is temporarily insufficient.

3rd period - 2nd year of life.

1. An important restructuring of the immune system occurs - the suppressor orientation changes to the helper one. The immune system becomes more active - there is a greater tendency to immune complex reactions (vasculitis, hemolytic-uremic syndrome and other immune complex conditions. At the same time, autoimmune diseases are still rare.
2. **Humoral immunity** increases B-lymphocytes become more sensitive to T-lymphocyte signals - antibodies are formed more actively. The level of IgG reaches 80% of the adult level; IgA - 40%. It is especially important that IgA begins to grow actively - the mucous membranes are better protected, so it becomes more difficult for allergens to penetrate through the mucous membranes. Transient allergic manifestations may decrease.
3. **Not enough IgG₂ and IgG** - the child remains more sensitive to pneumococcus;, Haemophilus influenzae.
4. **Lymphoid tissue** reacts very actively During infection, the lymph nodes and tonsils increase greatly. This is called hyperplasia of the lymphoid tissue, and the increase can persist for a long time after recovery.

!!!!Therefore, increased lymph nodes or tonsils in children of this age **do not** necessarily mean **chronic pathology** - it may be a manifestation of an active immune response.

5. Local immunity is not yet mature enough The protection of the mucous membranes remains especially weak: the child is easily infected through the respiratory tract. In addition, at this age, there are more contacts with the environment, the incidence of ARVI, bronchitis, pneumonia increases.

- 4th critical period - 4–6 years**
1. This is the period of the immune system approaching the adult type. The 2nd "crossover" of the leukocyte formula occurs - the number of neutrophils increases, lymphocytes decrease, their level approximately equalizes. After that, neutrophils predominate in the blood, as in adults.
 2. T-helpers and T-suppressors approach the adult ratio The immune system becomes more mature. Conditions appear for the development of autoimmune diseases in genetically predisposed children.
 3. Immunoglobulins are actively maturing. IgM and IgG reach adult levels. IgA is still somewhat lower. At the same time, IgE increases significantly, which is associated with a high frequency of atopic diseases.
 4. A full-fledged immunological memory is formed The immune system is already able to form a memory at the first contact, and to respond quickly at repeated contact. But the mucous membranes are not yet mature enough: the permeability of the mucous membranes for microorganisms and allergens is still increased. Therefore, atopic, parasitic, and some autoimmune diseases are often found at 4–6 years of age. Late immunodeficiencies and chronic diseases may also appear.

5th critical period - adolescence Approximately: girls - 12–13 years, boys - 14–15 years
There is a decrease in the mass of lymphoid organs. An increase in the level of sex hormones that affect the functioning of the immune system: IgE decreases and many children have atopic manifestations become less pronounced; individual features of the immune response are finally formed - mainly: a strong or weaker type of immune response is formed. But new risks appear Adolescence is often accompanied by the influence of adverse factors: smoking; alcohol; drugs; other toxic influences. This can contribute to the development of chronic inflammatory diseases, autoimmune diseases, lymphoproliferative diseases.

Remember the sequence: 3–6 months → we lose our mother's antibodies. 2 years → our own antibodies actively mature. 4–6 years → immunity becomes similar to an adult.
Adolescence → hormones rebuild the immune system.

!!!!The less mature the immune system — the more infections. The more active it becomes — the more likely allergies and autoimmune reactions.

Immunodeficiency diseases (IDD)

- Immunodeficiency is a condition in which one or more mechanisms of the immune system do not work effectively enough.
 - ✓ Inborn errors of immunity (IEI), genetically determined disorders of the immune system.
 - ✓ Secondary immune deficiencies, caused by diseases or external factors.

The body becomes more susceptible to:

- frequent infections;

- prolonged course of diseases;
 - repeated relapses;
- chronic infections;
- unusual or opportunistic infections.

The severity of immune deficiency is assessed comprehensively according to clinical manifestations, nature of infections, laboratory and functional indicators, and for congenital defects - according to a specific molecular genetic diagnosis.

Clinical and anamnestic signs on the basis of which it is possible to suspect the presence of immune system dysfunction, and refer children for consultation to a pediatric clinical immunologist

1. Recurrent bacterial infections that have the following features

Frequent or severe bacterial infections

- ≥ 2 purulent otitis or sinusitis per year;
- ≥ 2 pneumonias per year;
- recurrent abscesses and pyodermas;
- repeated severe infections: o meningitis; o osteomyelitis; o sepsis; o abscesses of internal organs;
- chronic urinary tract infections without anatomic cause;
- recurrent stomatitis;
- infections caused by unusual or conditionally pathogenic microorganisms, fungal infections.
- weak effect of repeated courses of antibiotics.

!!!!Frequent + severe + unusual + poorly treated → think about immunodeficiency.

2. Persistent candidiasis Oral thrush after 6 months, if it:

- does not go away;
- often recurs;
- spreads. May be a sign of impaired cellular immunity.

3. Chronic diarrhea (possible immunodeficiency)

Especially if it accompanied by: diarrhea + insufficient weight gain / malabsorption.

4. Unusual course of viral infection

- frequent herpes;
- long-term or severe herpes;
- severe course of chickenpox;
- long-term aphthous stomatitis;
- opportunistic infections.

5. Immunodeficiency in the family

- cases of unexplained death of children in the family;
- repeated severe infections in relatives;
- known primary immunodeficiency;
- other unusual or severe diseases in family members.

These facts may indicate the hereditary nature of the immunodeficiency.

6. Unusual reactions to live vaccines

- vaccine-associated poliomyelitis
- severe reaction after BCG;
- generalized infection after live vaccine.

7. Unusual course of common infections

- repeated chickenpox; • repeated measles;
- unusually severe or prolonged mononucleosis.

An infection that is usually easily tolerated, but in a child has a severe or recurrent course is a reason to check immunity.

8. Very frequent respiratory infections

≥8 episodes of acute respiratory infections per year or:

- severe respiratory tract infections;
- unusually long course;
- recurrent pneumonias;
- chronic bronchopulmonary changes.

Important: frequent acute respiratory infections in themselves do not mean immunodeficiency, especially in children who attend a children's group. The combination of symptoms is important.

9. Other signs that, together with infections, may indicate ID

Developmental disorders:

- poor weight gain;
- growth retardation;
- short stature;
- increased fatigue.

Unusual skin changes:

- eczema;

alopecia;

- widespread warts;
- unusual rashes.

Unusual combinations symptoms

- ataxia + telangiectasia;
- albinism + photophobia;
- hypoplasia of cartilage and hair.

Lymphoid tissue:

- very few lymph nodes;
- hypoplasia of tonsils;

- thymus abnormalities;
- or, conversely, a significant increase in lymphoid tissue.

10. Laboratory indicators should be evaluated taking into account age and reference limits of the laboratory. Immunological examination helps to determine which part of the immune system may be impaired, but the results are interpreted together with clinical data and age norms. If an antibody defect is suspected, the **level of Ig and specific antibodies is determined**. T-cell defect → **level of lymphocytes**, CD3/CD4/CD8 and functional tests. phagocyte defect → DHR and other functional studies. complement → C3, C4 and functional complement tests. suspected IEI → genetic study by indications.

Modern approach: first clinical phenotype → basic laboratory data → identification of a probable link → specific functional/genetic study.

The most important thing to remember:

1. INFECTIONS: frequent + severe + recurrent + unusual
2. OPPORTUNISTS: candidiasis + unusual microorganisms + severe viral infections
3. VACCINATION: severe reaction to a live vaccine → suspected ID
4. DEVELOPMENT: insufficient growth + weight loss + unusual skin/neurological signs
5. ANALYSES: neutropenia + lymphopenia + low Ig + low CD4

Interpretation of the immunogram

The immunogram cannot be evaluated separately from the clinical picture.

The results of immunological studies should be analyzed taking into account:

- the child's age;
- clinical manifestations;
- frequency and severity of infections;
- type of pathogens;
- response to treatment;
- results of previous studies;
- vaccinations and concomitant diseases.

Immune indicators can temporarily change during acute infection, stress, after vaccination or under the influence of medications, therefore, a single immunogram is only a guideline.

Persistent deviations with corresponding clinical signs are the basis for further examination.

The most informative is the study in dynamics.

!!!The clinic is more important than a separate indicator in the immunogram.

Why can immunogram indicators change?

During infection, immune cells can actively move from the blood to tissues and lymphoid organs. Therefore, a temporary decrease in their number in the peripheral blood does not necessarily mean immunodeficiency.

The results are also influenced by:

- age;
- acute or chronic infection;

- medications;
- nutrition;
- concomitant diseases;
- physiological individual fluctuations.

Therefore, reference values must correspond to age and a specific laboratory.

Examination of a child with suspected immunodeficiency is carried out in 2 stages.

Stage 1 — basic examination:

Complete blood test with leukoformula

- absolute neutrophil count;
- absolute lymphocyte count;
- platelets;
- other cellular changes.

Quantitative immunoglobulins

- IgG;
- IgA;
- IgM;
- if indicated — IgE.

Cellular immunity

- number of T-, B- and NK-cells;
- if necessary — subpopulations of T-cells (CD4, CD8).

Complement

- primarily C3 and C4;
- if indicated — functional tests of the complement system.

Neutrophil function

- special functional tests are prescribed if the clinic allows to suspect a phagocytosis defect.

Stage 2 — specialized examination

Performed according to indications, depending on what defect is suspected.

For example:

- assessment of specific antibodies after vaccination;
- B-lymphocyte function studies;
- T-cell function tests;
- phagocyte function studies;
- complement system studies;
- genetic testing;
- molecular genetic diagnostics.

The main principle is not to “prescribe the entire immunogram”, but to answer a specific clinical question.

Inborn errors of immunity (IEDs)

These are genetically determined disorders of the development or function of the immune system.

Distinguish:

1. Predominantly antibody-deficient disorders
2. Combined immunodeficiencies
3. Deficiencies of cellular immunity
4. Defects of phagocytosis
5. Deficiencies of the complement system
6. Defects of innate immunity
7. Disorders of immune regulation
8. Auto-inflammatory diseases
9. Disorders of the bone marrow/immune system with characteristic syndromes

1. X-linked agammaglobulinemia (Bruton's disease)

Defect of maturation of B-lymphocytes - a sharp decrease in B-cells and all major Ig.

Typical manifestations:

- recurrent bacterial infections;
- otitis;
- sinusitis;
- pneumonia;
- especially after a decrease in maternal IgG.

Laboratory:

- very low IgG, IgA, IgM;
- sharply reduced or absent CD19/CD20 B-cells;
- T cells are preserved.

2. Selective IgA deficiency

One of the most common primary antibody deficiencies.

Key features:

- very low or absent IgA;
- IgG and IgM are usually preserved;
- B-cell count is usually normal.

May be asymptomatic or manifest with:

- recurrent respiratory tract infections;
- gastrointestinal infections;
- allergic diseases;
- autoimmune diseases.

3. 22q11.2 deletion syndrome (DiGeorge syndrome)

Major triad: Thymus ↓ → T-cell immunity ↓

Parathyroid glands ↓ → hypocalcemia → seizures

Congenital heart defects

Facial changes and other manifestations may be characteristic.

4. Wiskott-Aldrich syndrome

X-linked immunodeficiency.

Classic triad: eczema, thrombocytopenia, recurrent infections.

May also be:

- autoimmune diseases;
- allergies;
- increased risk of malignant diseases.

5. Ataxia-telangiectasia (Louis-Bar syndrome)

Main manifestations: ataxia, telangiectasias, immunodeficiency.

Characteristic:

- progressive cerebellar ataxia;
- telangiectasias;
- recurrent sinopulmonary infections;
- increased risk of malignant diseases.

6. Defects of phagocytosis

- chronic granulomatous disease;
- severe congenital neutropenias;
- cyclic neutropenia.

Chronic granulomatous disease: phagocytes cannot normally form reactive oxygen species: recurrent severe bacterial and fungal infections, granulomas, abscesses.

7. Complement system deficiency. Autoimmune diseases. Congenital angioedema occurs when C3 complement system is insufficient.

Control questions:

1. What are the main features of the functioning of the fetal immune system?
What factors provide its protection against infections?
2. Why are the fetus and newborn particularly sensitive to intracellular infections?
What role do T-cell immunity, NK cells and neutrophils play in this?
3. What role do maternal IgG play in protecting the newborn?
How does their transplacental transfer occur and when is it most intense?
4. What are the main features of the innate immunity of the newborn?
Why can newborns have a weak inflammatory and febrile reaction?
5. How does a child's immune system change during the first years of life? Name the main critical periods of development of immunobiological reactivity and their characteristic features
6. What is physiological transient hypogammaglobulinemia of early childhood? Why does it occur and how can it manifest itself?
7. What clinical signs allow us to suspect immunodeficiency in a child? Name at least 5 "alarm signals".
8. Why can a single immunogram not be the basis for establishing a diagnosis of immunodeficiency? What factors should be taken into account when interpreting it?

9. What basic laboratory tests are included in the initial examination of a child with suspected immunodeficiency? What can be assessed with their help?
10. What main groups of primary immunodeficiencies (innate immunity errors) are distinguished in modern immunology? What links of the immune system are impaired in them?
11. How to suspect according to clinical and laboratory signs: ▯ Bruton's X-linked agammaglobulinemia? ▯ selective IgA deficiency? ▯ DiGeorge syndrome? ▯ Wiskott-Aldrich syndrome? ▯ ataxia-telangiectasia?

Recommended Literature

Basic:

1. Nelson Textbook of Pediatrics. (Robert M. Kliegman et al., 22nd Edition, 2024. Published by Elsevier . V 2. 2222p.
2. Propaedeutic Pediatrics in Syndromes. Boyarchuk O.R., Kinash M.I. Ukrmedknyga.2022. 760 p.
3. “Propaedeutic Pediatrics with Child Care” (T. V. Kapitan, 5th ed., 2021). Vinnytsia. 832p.

Additional:

1. Fundamentals of Pediatrics by Nelson / Karen J. Markdante, Robert M. Kliegman; translation of the 8th English edition in 2 volumes. Scientific editors of the translation V.S. Berezenko, T.V. Pochynok. Kyiv: VSV “Medicine”, -2020.
2. Manual of Propaedeutic Pediatrics/ Lectures from propaedeutic pediatrics. Nikityuk S.O. Ukrmedknyga. English 2020.
3. Fundamentals of immunology. Functions and disorders of the immune system. Abul. K. Abbas. Kyiv. VSV.Medicine. 2020. 336p.

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