

ZAPORIZHZHIA STATE MEDICAL UNIVERSITY

DEPARTMENT OF HOSPITAL PEDIATRICS

**DISEASES OF BLOOD AND ENDOCRINE SYSTEM IN
CHILDREN**

**(the collection of test tasks on hospital pediatrics for the 5 year
English speaking students of medical faculty)**

ZAPORIZHZHIA

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Zaporizhzhia State Medical University

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Introduction

Pediatrics is one of important clinical disciplines, without deep knowledge of which the forming of modern specialist-physician is not possible.

Pediatrics as educational discipline is based on students knowledge of substantive provisions of anatomy, histology, physiology, physiopathology, pathoanatomy, propedeutics and infectious diseases

Acquisition of thorough knowledges and abilities from paediatrics allows to utilize them for the savation of clinical problems of diagnostics, prophylaxis and treatment of diseases.

Educational and methodical manual “Diseases of blood and endocrine system in children” for the 5 year students of medical faculty compiled in accordance to “Educational profesional programm for high education” after professional direction of „Medicine” , ratified by the Ministries of Education and Health of Ukraine. In developing of materials the long-term experience of pediatrics department of Zaporizhzhia State Medical University and the recommendations of supporting department of pediatrics is used (hospital pediatrics department of National Medical University).

Materials given in the Manual show by itself the guidance on the practical classes leadthrough for the 5 year students of medical faculty studying on speciality of Pediatrics and General Practitioner. Taking into account progressive development of pediatrics, change of requirements to the specialists the given educational methodical Manual will incompletely reflects to the pedagogical and professional necessities, that is why will be perfected and complemented.

TEST CREDIT STRUCTURE OF THE MODULE 3. Diseases of blood and endocrine system in children.

Topic	Lectures	Pract. classes	Ind. prep.	Individual work
<i>Semantic module 10. Diseases of blood in children.</i>				
1. Scarce anemias in children	2	4	2	
2. Hemoblastoses in children	2	4	2	
3. Hemorrhagic diseases in children	2	4	2	
<i>Semantic module 11. Diseases of endocrine system in children</i>				
4. Diabetes mellitus in children	2	4	2	
5. Thyroid diseases in children	2	4	2	
6. Diseases of hypothalamus pituitary system in children		4	2	

Implementation of individual work			4
Final module control		4	6
Total: ECTS credits - 2,0; hours - 60	10	28	22

TIMETABLE OF LECTURES. MODULE 3

Diseases of blood and endocrine system in children

#	Topic	Hours
1	Anemias in children	2
2	Hemoblastoses in children	2
3	Hemorrhagic diseases in children	2
4	Diabetes mellitus in children	2
5	Thyroid diseases in children	2
	Total	10

TIMETABLE OF PRACTICAL CLASSES. MODULE 3.

Diseases of blood and endocrine system in children

#	Topic	Hours
1	Scarce anemias in children	4
2	Hemoblastoses in children	4
3	Hemorrhagic diseases in children	4
4	Diabetes mellitus in children	4
5	Thyroid diseases in children	4
6	Diseases of hypothalamus, pituitary and sexual glands in children.	4
7	Final module control	4
	Total	28

THEMATIC PLAN OF STUDENTS INDIVIDUAL PREPARATION (SIP). MODULE

3.

Diseases of blood and endocrine system in children

№ п/п	Kind of SIP	Hours	Control types
1	Preparation to practical classes	12	Current control on the practical classes.
2	Working of themes which are not included in the plan of		Final module control

	audience classes.		
3	SIP:case reports analysis, preparing of referates and the performancies in clinical meetings.	4	Final module control. Current control on the practical classes.
4	Preparing to the final module control	6	Підсумковий модульний контроль
	Total	20	Final module control.

**POINTS DISTRIBUTION FOR THE ASSESMENT OF STUDENTS
PERFORMANCIES. MODULE 3.МОДУЛЬ 3.**

Diseases of blood and endocrine system in children

.Module 3 (volume of the estimated activity)	Maximal points are possible
<i>Semantic module 10.</i>	
Topic 1. Scarce anemias in children	20
Topic 2. Hemoblastoses in children	20
Topic 3. Hemorrhagic diseases in children	20
<i>Semantic module 11</i>	
Topic 1. Diabetes mellitus in children	20
Topic 2. Thyroid diseases in children	20
Topic 3. Diseases of hypothalamus, pituitary and sexual glands in children.	20
Current performance in total	120
Final module control	80
SUMMARISED MODULE POINTS	200

Note: In mastering the topic after traditional system points gives to the a student as follows: «5» - 20 points, «4» - 16 points, «3» - 12 points, «2» - 0 points.. Maximal amount of points for current educational performance of student are 120.

A student is allowed to pass the final module control in terms of him performance according to the requirements of tutorial and in case of obtaining no less than 72 points for current performance during the practical classes. (12 x 6).

Final module control is setting off to the student if he get not less than 50 of 80 points.

Theme. Iron-, protein-and vitamin scarce anaemias.

Study time: 4 hours

Etiology, pathogenesis, classification, clinical presentation, diagnostics, differential diagnostics, treatment, prophylaxis. Emergency in haemorrhage. Prognosis.

I. Actuality of the theme.

Anemia is a frequent laboratory abnormality in children. As many as 20 percent of children in the United States and 80 percent of children in developing countries will be anemic at some point by the age of 18 years.

Childhood anaemia poses a major public health issue leading to an increased risk of child mortality, as well as the negative consequences of iron deficiency anaemia on cognitive and physical development.³ The United Nations General Assembly set a goal at its special session on children in 2003 to reduce the prevalence of anaemia by one third by 2010. Anemia (uh-NEE-me-eh) is a condition in which your blood has a lower than normal number of red blood cells. This condition also can occur if your red blood cells don't contain enough hemoglobin (HEE-muh-glow-bin). Hemoglobin is an iron-rich protein that gives blood its red color. This protein helps red blood cells to carry oxygen from the lungs to the rest of the body. If you have anemia, your body doesn't get enough oxygen-rich blood. As a result, you may feel tired and have other symptoms. With severe or long-lasting anemia, the lack of oxygen in the blood can damage the heart, brain, and other organs of the body. Very severe anemia may even cause death. Red blood cells are disc-shaped and look like doughnuts without holes in the center. They carry oxygen and remove carbon dioxide (a waste product) from your body. These cells are made in the bone marrow—a sponge-like tissue inside the bones. Red blood cells live for about 120 days in the bloodstream and then die. White blood cells and platelets (PLATE-lets) are also made in the bone marrow. White blood cells help to fight infection. Platelets stick together to seal small cuts or breaks on the blood vessel walls and stop bleeding. With some types of anemia, you may have low numbers of all three types of blood cells. Anemia has three main causes: blood loss, lack of red blood cell production, or high rates of red blood cell destruction. These causes may be due to a number of diseases, conditions, or other factors. Many types of anemia can be mild, short term, and easily treated. Some types can even be prevented with a healthy diet. Other types can be treated with dietary supplements. However, certain types of anemia may be severe, long lasting, and life threatening if not diagnosed and treated.

Concrete purposes:

1. To determine the etiological and pathogenetic factors in iron-, protein-and vitamin scarce anaemias in children .
2. To classify and analyze the typical clinical manifestation of iron-, protein-and vitamin scarce anaemias in children.
3. To make the plan of investigation and analyse the information about laboratory and instrumental data of iron-, protein-and vitamin scarce anaemias in children.
4. To demonstrate skills of treatment, rehabilitation and prophylaxis in iron-, protein-and vitamin scarce anaemias in children .

5. To diagnose and render an urgent help in haemorrhage.
6. To conduct differential diagnostics of iron-,protein-and vitamin scarce anaemias in children and make a preliminary diagnosis.
7. To determine the prognosis for life in iron-, protein-and vitamin scarce anaemias in children.
8. To demonstrate the skills of medical specialist moral and deontological principles and principles of professional subordination in pediatrics.

II. Classes (pointing of planned mastering level)

1. A student must know (to familiarize): $\alpha 1$
 - About the place of iron-,protein-and vitamin scarce anaemias in the structure of haematology system diseases in children, widespread in different age-dependent and ethnic groups;
 - About statistical information in relation to morbidity, frequencies of complications, lethality, the nearest and remote prognosis in patients with iron , protein-and vitamin scarce anaemias ;
 - About history of scientific study and payment of domestic scientists;

2. A student must know : $\alpha 2$

- etiology of iron-,protein-and vitamin scarce anaemias in children;
- key links of iron-,protein-and vitamin scarce anaemias pathogenesis ;
- clinical classification of iron-,protein-and vitamin scarce anaemias;
- the classic clinical manifestation of iron-,protein-and vitamin scarce anaemias;
- laboratory diagnosis iron-,protein-and vitamin scarce anaemias;
- laboratory and instrumental diagnosis of iron-, protein-and vitamin scarce anaemias;
- complications of iron-,protein-and vitamin scarce anaemias in children;
- the treatment principles of iron-,protein-and vitamin scarce anaemias in children;

3. A student must seize: $\alpha 3$

By skills:

- collection of complaints and anamnesis of disease;
- examination of patients with iron-,protein-and vitamin scarce anaemias and revealing the main symptoms and syndromes.
- formulating and substantiating the preliminary diagnosis;
- determinat a laboratory and instrumental examination plan of patients investigation (with obedience of diagnostics standards);

By the abilities:

- interpreting the results of laboratory and instrumental investigations.
- conducting a differential diagnosis among different kinds of anaemias ;
- giving recommendations in relation to the patient regimen and diet with anaemias- taking into account the stage of disease, severity of the state and concomitant pathology;
- completing the treatment plan for anaemias according to standards taking into account the stage of disease, complications and concomitant pathology.
- rendering the first aid in extreme situations and exigent states.

III. Aims of personality development (educative aims):

- A student must learn to adhere to the rules of behaviour and principles of medical etiquette and deontology near a bed ridden patient with iron-, protein- and vitamin scarce anaemias -to try hands on ability to set a psychological contact with a patient and his family;
- to master a sense of professional responsibility for a timely , adequate and skilled medicare.

IV. Interdisciplinary integration:

Subject	To know	To be able
1. Previous (providing)		
Anatomy	Structure of blood in children	To determine the location of thyroid projection and palpation, of local lymphatic nodes.
Physiology	Physiology of hematopoietic system in newborns, normative indices of laboratory and instrumental investigational methods and their assesment.	To asses laboratory data and instrumental investigational methods.
Pathologic physiology	Key links of the pathogenesis of hematopoietic system	
Pathologic anatomy	Morphological features of the hematopoietic system in newborns development depending of disease stage.	To analyze and interpret the information about clinical examination and about additional methods of investigation
Pharmacology	Pharmacokinetics and pharmacodynamics, the side effects of preparations.	To prescribe: age dependent and individual patient's characteristics of treatment to identify the stage of disease and establish an individual prescription to take with the correct dosage. To be able to make a prescription.
Propedeutical pediatrics.	The basic stages and methods for the clinical examination of patients.	To collect complaints and anamnesis vitae et morbid- to find out the basic risk factors in disease of hematopoietic system , be able to conduct patient's examination, to reveal the clinical signs of anaemias and to be able to interpret the data

		about additional methods of investigation.
2. Followings (provided)		
Hospital pediatrics.	Clinical signs of anaemias, differential diagnosis and treatment tactics.	To reveal the clinical signs of anaemias and complications, to conduct differential diagnosis, be able to prescribe treatment.

Methodical materials for the class basic stage supporting

The questions for the control of the primary knowledge level of abilities and skills:

1. What is the function of erythrocytes?
2. How can we use erythrocytic index ?
3. What is general for anaemias?
4. What is the typical triad for hemolytic anemia?
5. What are the specific clinical signs of hypoplastic anaemias?
6. What are the typical syndromes for iron-deficiency anemia?
7. What are the laboratory findings of pernicious anemia ?
8. To explain the ferrotherapy in children with iron-deficiency anemia.
9. To explain the pathogenesis of hereditary spherocytosis.
10. To explain the pathogenesis of anemia praematarorum.
11. What is the prophylaxis of iron-deficiency anemia in children?
12. To explain the clinical manifestation of Diamond-Blackfan syndrome.

Primary tests

1. The amount of iron in blood serum in chronic iron scarce anemia:

- A. <30,4 mcmol/l
- B. < 15 mcmol/l
- C. <22 mcmol/l
- D. < 12,5 mcmol/l
- E. <20 mcmol/l

2. Daily requirement for iron in children of an early age:

- A. 2 mg
- B. 10 mg
- C. 50 mg
- D. 20 mg
- E. 1mg

3. Daily requirement for iron in teenagers:

- A. 100 mg.
- B. 10 mg.

- C. 15-20 mg.
- D. 2 mg.
- E. 40 mg

4. What kind of iron is better absorbed?

- A. Iron chloride.
- B. 3-valent.
- C. Iron in a complex with proteins.
- D. . 2 valent.
- E. Iron in a complex with polivitamines

.5. Who is a juvenile chlorosis inherent to the most often?

- A. Newborns
- B. Young men of 16 years old.
- C. Young women of 15 years old.
- D. Girls of 15-20 years old.
- E Children of early age

6. What is the cell –color index in juvenile chlorosis?

- A. 0,44-0,5
- B. . 1,1-1,3
- C. 0,82-1,65
- D. 0,7-0,85
- E. More than 1,3

7. What is the basic prominent feature of a peripheral blood in newborns?

- A. Lymphopenia
- B. Neutrophile leukocytosis .
- C . Anemia.
- D. Erythrocytosis
- E. Lymphocytosis

8. Is there a possibility of iron deficiency correction with a diet?

- A. No
- B. Yes.
- C. Possible with the help of animal parentage products.
- D. Possible with the help of using vegetable products.
- E. Possible with the help both of vegetable and animal parentage products

9. Are hemotransfusions indicated in iron scarce anemia?

- A. No.
- B. Yes.
- C. Indicated in hemoglobin amount lower than 100 g/l.
- D. Indicated in a hemoglobin amount lower than 90g/l
- E. Indicated in hemoglobin amount lower than 80 g/l.

10. In what age the Diamond-Blackfan syndrome develops the most frequently?

- A. 1-2
- B. 2-6
- C. 8
- D. 10-15
- E. The first month of life

11. What is the diameter of erythrocytes in the case of macrocytic [megalocytic] anemia ?

- A. 7,2-8,3 mkm
- B. 10-12 mkm
- C. 7-12 mkm
- D. >7 mkm
- E. <12 mkm

12. What is the name of erythrocytes with thorns?

- A. macrocyt
- B. stomatocyt
- C. spherocyt
- D. acanth(r)ocyte
- E. microcyt

13. What is the cell –color index in B12 deficiency anaemia ?

- A. 0,44-0,5
- B. 0,7-0,85
- C. 0,82-1,05
- D. >1,05
- E. <0,44

Answers: 1-B, 2-A, 3-D, 4-C, 5-A, 6-A, 7-B, 8-C, 9-D,A, 10-A, 11-B, 12-E, 13-B.

Typical situational tasks of 2 level

Task 1.

A child of 4 years old is hospitalized with complaints to be pale. On examination the liver and spleen are not enlarged, congenital anomalies (dysmorphic facies). Laboratory findings : macrocytic anaemia with elevated levels of folic acid and vitamin B12, elevated fetal hemoglobin (Hb F) and increased expression of "i" antigen. Bone marrow culture shows markedly reduced numbers of colony-forming units—erythrocyte (CFU-E) and BFU-E.

1. What is the preliminary diagnosis?
2. At what age is a disease most often begins?
3. Key basic diagnostic criteria.

4. Prescribe treatment.
5. Prognosis

Standard of answer. Task 1.

1. Diamond-Blackfan syndrome
2. This rare condition usually becomes symptomatic in early infancy.
3. frequently with pallor in the neonatal period, but may first be noted later in childhood. About 50% of children are diagnosed by 2 mo of age, and 75% by 6 mo. The most characteristic features are macrocytic anemia, reticulocytopenia, and a deficiency or absence of red blood cell (RBC) precursors in an otherwise normally cellular bone marrow.
4. Corticosteroid therapy is frequently beneficial if begun early, with three fourths of patients responding initially. The mechanism of its effect is unknown. Prednisone in three or four divided doses totaling 2 mg/kg/24 hr is used as an initial trial. This dose should then be doubled, used on alternate days, and tapered still further while maintaining the hemoglobin level at 10 g/dL or above. In some patients, very small amounts of prednisone, as low as 2.5 mg, may be sufficient to sustain adequate erythropoiesis.
5. The outlook is best in those who respond to corticosteroid therapy. About one half of the patients are long-term responders. The liver and spleen enlarge, and secondary hypersplenism with leukopenia and thrombocytopenia may occur in children who are not chelated adequately or in those with chronic hepatitis acquired from transfusions. The complications of chronic transfusions are similar to those seen in β -thalassemia major, and prevention and treatment of iron overload should be equally aggressive in both groups of transfused patients .

Task 2.

A girl of 11 years old firstly hospitalized in the department with complains of weakness, irritability, anorexia. The tongue is smooth, red, and painful. Neurologic manifestations include ataxia, paresthesias, hyporeflexia. The anemia is macrocytic, with prominent macro-ovalocytosis of the RBCs. Serum vitamin B12 levels are <100 pg/mL. Concentrations of serum iron and serum folic acid are normal.

1. What is the preliminary diagnosis?
2. At what age is a disease most often begins?
3. Key basic diagnostic criteria.
4. Prescribe treatment.
5. Prognosis

Standard of answer. Task 2.

1. JUVENILE PERNICIOUS ANEMIA
2. The symptoms of juvenile pernicious anemia become prominent at 9 mo to 11 yr of age.
3. The anemia is macrocytic, with prominent macro-ovalocytosis of the RBCs. The neutrophils may be large and hypersegmented. In advanced cases neutropenia and thrombocytopenia, simulating aplastic anemia or leukemia, are seen. Serum

vitamin B12 levels are <100 pg/mL. Concentrations of serum iron and serum folic acid are normal or elevated. Serum LDH activity is markedly increased. Moderate elevations (2–3 mg/dL) of serum bilirubin levels may be seen. Excessive excretion of methylmalonic acid in the urine (normal amount, 0–3.5 mg/24 hr) is a reliable and sensitive index of vitamin B12 deficiency.

4. A prompt hematologic response follows parenteral administration of vitamin B12 (1 mg), usually with reticulocytosis in 2–4 days, unless there is concurrent inflammatory disease. The physiologic requirement for vitamin B12 is 1–5 μ g/24 hr, and hematologic responses have been observed with these small doses, indicating that administration of a minidose may be used as a therapeutic test when the diagnosis of vitamin B12 deficiency is in doubt. If there is evidence of neurologic involvement, 1 mg should be injected intramuscularly daily for at least 2 wk. Maintenance therapy is necessary throughout the patient's life; monthly intramuscular administration of 1 mg of vitamin B12 is sufficient. Oral therapy may succeed because of mucosal diffusion with high doses, but it is not generally advisable due to uncertainty of absorption.

5. prognosis - a full recovery with a balanced diet

Task 3.

A girl of 10 years old firstly hospitalized in the department with complaints of pagophagia. On examination tachycardia and cardiac dilatation occur, and systolic murmurs are present. The spleen is enlarged. Laboratory findings: microcytosis with increasing deficiency the RBCs, hypochromia, poikilocytosis, and increased red cell distribution width (RDW). Reticulocytes are moderately elevated. White blood cell counts are normal. The bone marrow is hypercellular, with erythroid hyperplasia. Leukocytes and megakaryocytes are normal.

1. What is the preliminary diagnosis?
2. At what age is a disease most often begins?
3. Key basic diagnostic criteria.
4. Prescribe treatment.
5. Prognosis

Standard of answer . Task 3.

1. Iron-deficiency anemia

2. 2-3 month, juvenile period

3. Normal ranges are age dependent, and decreased levels accompany iron deficiency. Next, there is a decrease in serum iron (also age dependent), the iron-binding capacity of the serum increases, and the percent saturation falls below normal (also varies with age). When the availability of iron becomes rate limiting for hemoglobin synthesis, a moderate accumulation of heme precursors, free erythrocyte protoporphyrins (FEP), results. As the deficiency progresses, the red blood cells (RBCs) become smaller than normal and their hemoglobin content decreases. The morphologic characteristics of RBCs are best quantified by the determination of mean corpuscular hemoglobin (MCH) and mean corpuscular volume (MCV). With increasing deficiency the RBCs become deformed and mis-shapen and present

characteristic microcytosis, hypochromia, poikilocytosis, and increased red cell distribution width (RDW)

4. Oral administration of simple ferrous salts (sulfate, gluconate, fumarate) provides inexpensive and satisfactory therapy. There is no evidence that addition of any trace metal, vitamin, or other hematinic substance significantly increases the response to simple ferrous salts. For routine clinical use the physician should be familiar with an inexpensive preparation of one of the simple ferrous compounds. The therapeutic dose should be calculated in terms of elemental iron; ferrous sulfate is 20% elemental iron by weight. A daily total of 6 mg/kg of elemental iron in three divided doses provides an optimal amount of iron for the stimulated bone marrow to use. Better absorption may result when medicinal iron is given between meals. Intolerance to oral iron is uncommon. A parenteral iron preparation (iron dextran) is an effective form of iron and is usually safe when given in a properly calculated dose, but the response to parenteral iron is no more rapid or complete than that obtained with proper oral administration of iron, unless malabsorption is present.

5. Prognosis of the disease - a full recovery with a balanced diet

Task 4.

A preterm black male infant was found to be jaundiced 12 h after birth. At 36 h of age, his serum bilirubin was 18 mg/dL, hemoglobin concentration was 12.5 g/dL, and reticulocyte count 9%. Many nucleated red cells and some spherocytes were seen in the peripheral blood smear.

1. What is the preliminary diagnosis?
2. Etiology?
3. Key basic diagnostic criteria.
4. Prescribe treatment.
5. Which of the following should the differential diagnosis include?

Standard of answer.Task 4.

1. Hereditary spherocytosis

2. The most common molecular defect is an abnormality of spectrin, which is a major component of the cytoskeleton responsible for red cell shape. A recessive defect has been described in α -spectrin; dominant defects in β -spectrin and in protein 3; and dominant and recessive defects in ankyrin. A deficiency in spectrin, protein 3, or ankyrin results in uncoupling in the "vertical" interactions of the lipid bilayer skeleton and the loss of membrane microvesicles).

3. Spherocytosis can be seen in hyperthermia, hereditary spherocytosis, G6PD deficiency, or ABO incompatibility. Hyperbilirubinemia has been associated with black preterm infants with G6PD deficiency. The blood smear of the affected infant usually reveals nucleated red cells, spherocytes, poikilocytes, "blister" cells, and fragmented cells. Neonatal hyperbilirubinemia occurs in about 50% of patients with hereditary spherocytosis. Spherocytosis occurs in ABO incompatibility but not in Rh incompatibility. The hemolytic manifestations of ABO incompatibility and hereditary spherocytosis are very similar. The blood types of the mother and of the infant should be determined along with the results of a direct Coombs test of the infant and the

presence or absence of a family history of hemolytic disease (spherocytosis). Sickle cell disease would not be expected to cause problems in newborns due to the protection by fetal hemoglobin.

4. Since the spherocytes in hereditary spherocytosis are destroyed almost exclusively in the spleen, splenectomy eliminates most of the hemolysis associated with this disorder. Vaccines for encapsulated organisms such as pneumococcus, meningococcus, and *Haemophilus influenzae* should be administered prior to splenectomy, and prophylactic penicillin (age ≤ 5 yr: 125 mg/12 hr; age >5 yr through adulthood: 250 mg/12 hr) administered thereafter. Postsplenectomy thrombocytosis is commonly observed but needs no treatment and usually resolves spontaneously.

5. The major alternative consideration when large numbers of spherocytes are seen on the blood film is immune hemolysis. Isoimmune hemolytic disease of the newborn, particularly due to ABO incompatibility, mimics hereditary spherocytosis. The detection of antibody on the infant's red cells using a direct Coombs test should establish the diagnosis of immune hemolysis. Other autoimmune hemolytic anemias also are characterized by spherocytes, and there may be evidence of a previously normal hemoglobin, hematocrit, and reticulocyte count. Rare causes of spherocytosis include thermal injury, clostridia septicemia with exotoxemia, and Wilson disease, each of which may present with a transient hemolytic anemia.

Task 5.

1. On a routine-screening complete blood count, a 1-year-old is noted to have a microcytic anemia. A follow-up hemoglobin electrophoresis demonstrates an increased concentration of hemoglobin A₂.

1. What is the preliminary diagnosis?
2. Which of the following ethnic groups is the lowest incidence in?
3. Key basic diagnostic criteria.
4. Prescribe treatment.
5. Prognosis

Standard of answer. Task 5.

1. β -thalassemia trait
2. Their main distribution includes areas bordering the Mediterranean Sea, much of Africa, the Middle East, the Indian subcontinent, and Southeast Asia.
3. The concentration of hemoglobin A₂ is β -thalassemia trait. In severe iron deficiency, hemoglobin A₂ may be increased or decreased. In mild-to-moderate iron deficiency, the level of hemoglobin A₂ is normal. The level is also normal in sickle cell anemia, chronic systemic illness, and lead poisoning.
4. Transfusions are given on a regular basis to maintain the hemoglobin level above 10 g/dL. This "hypertransfusion" regimen has striking clinical benefits; it permits normal activity with comfort, prevents progressive marrow expansion and cosmetic problems associated with facial bone changes, and minimizes cardiac dilatation and osteoporosis. Transfusions of 15–20 mL/kg of

packed cells are usually necessary every 4–5 wk. Cross-matching should be performed to forestall alloimmunization and prevent transfusion reactions. A sustained high blood level of deferoxamine is needed for adequate iron excretion. The drug is administered subcutaneously over an 8- to 12-hr period using a small portable pump (during sleep), 5 or 6 nights/wk. Patients who adhere to this regimen can maintain serum ferritin levels of lower than 1,000 ng/mL, which is well below the toxic range.

5. Bone marrow transplantation is curative in these patients and has been performed with increasing success, even in patients who have been transfused extensively. This procedure, however, carries considerable risks of morbidity and mortality and generally can only be used for patients who have nonaffected histocompatible siblings.

Methodical materials for the class

A professional algorithm of patients management implementation (reference chart) for the practical skills and abilities forming .

№	Task	Sequence of implementation	Remarks and warnings related to self-control
1	To conduct patient's examination for anaemias.	1.To conduct complaints and disease's anamnesis gathering. 2.To gather thoroughly the patient's life anamnesis. 3.To conduct examination of the patient. 4.To investigate cardiovascular system of the patient (palpation, percussion).	To pay attention to features of disease course , underlying factors, concomitant diseases etc. To establish the risk factors which can cause the development of disease. To assess patient general condition, position in bed, color and humidity of skin and mucouse, presence of neck veins and extermities swelling. To pay regard to pulse rhythm, it's tension and size on both hands, apex shove, its properties, margins of absolute and relative cardiac dullness, its changes, HR(tachi-or bradycardia, extrasystole),BP.

		<p>5.To conduct heart and main vessels auscultation.</p> <p>6.To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the system of digestion.</p> <p>9. To conduct examination and palpation of thyroid gland and local lymphatic nodes.</p>	<p>To pay regard to heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To pay attention to features of percussion and auscultation in children of different age.</p>
2	To formulate the preliminary diagnosis.	<p>1.To formulate the preliminary diagnosis</p> <p>2.To substantiate all the components of preliminary diagnosis based on complaints,anamnesis, and examinations.</p>	To formulate the preliminary diagnosis of anaemias and substantiate each component of it.
3	To evaluate the parameters of additional laboratory investigations.	<p>1.To evaluate the blood count data.</p> <p>2. To evaluate the biochemistry data.</p> <p>3.To evaluate the blood hormonal profile.</p>	<p>To pay attention to the signs of anemia, reticulocyte count iron, ferritin and total iron binding capacity levels, leucocytosis, changing of formula, blood film, red cell enzyme studies, folate, vitamin B12 levels , elevation of sedimentation rate.</p> <p>To pay attention to cholesterol, lipids and glucose levels.</p>
4	To undersrtand the data of additional and laboratory	To undersrtand the data of bone marrow	

	investigation.	puncture.	
5.	To conduct differential diagnosis.	<p>1.Consistently to find the common signs in complaints,life and disease anamnesis, data of examination, data of laboratory and instrumental investiagtions in patient and in similar states.</p> <p>2.To find differences between complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumenttal methods of research and in similar nosology.</p> <p>3.On the basis of the differences found to exclude similar diseases from the list of possible diagnoses.</p> <p>4. To conduct differential diagnostics according to the above mentioned algorithm among all the nosologies having the similar signs, among other kinds of anaemias</p> <p>5.Taking into account the impossibility to exclude the diagnosis of anaemias from the list of credible diagnoses to draw a conclusion about the probability of such a diagnosis.</p>	Special attention must be paid to differential diagnosis among the leucaemias
6	To formulate the final clininical diagnosis.	1.To formulate the final clininical diagnosis.	Basing on modern classification of

		2. Basing on the preliminary diagnosis, additional investigations data, conducted differential diagnosis, substantiate all elements of the final clinical diagnosis.	anaemias, formulate the diagnosis, complications of disease and the presence of concomitant diseases.
7	To prescribe treatment for patients.	1. To prescribe no medicinal treatment 2. To prescribe medicinal treatment.	Specify the regimen and detailed diet according to a disease. Taking into account the age, severity of patient's state, the stage of disease, the presence of complications and concomitant pathology, to prescribe modern medicinal treatment in accordance with the standards of anaemias therapy.

The material for the control of the secondary level of abilities and skills:

The secondary tests

1. A girl of 10 years old, is admitted to hospital. Laboratory findings: macrocytic anaemia, serum vitamin B12 levels are 80 pg/mL. Concentrations of serum iron and serum folic acid are elevated.

What is the preliminary diagnosis?

- A. Iron deficiency
- B. vitamin B12 deficiency
- C. vitamin B12 hypervitaminosis
- D. thalassemia
- E. Minkowsky-Shauffard disease

2. A 5-year-old previously well boy develops pallor, dark urine, and jaundice. There has been no apparent exposure to a jaundiced person or to any toxins. He is taking trimethoprim-sulfamethoxazole for otitis media. You consider the possibility of a hemolytic crisis caused by glucose-6-phosphate dehydrogenase (G6PD) deficiency.

- A. African American

- B. Greek
- C. Chinese
- D. Middle Eastern

3. A 11-year-old boy with signs of malnutrition consulted a doctor about smell and taste distortion, angular stomatitis. Objectively: marked blue sclerae. The patient was diagnosed with iron deficiency anemia. What is the dominating clinical syndrome?

- A. Haemolytic
- B. Anaemic
- C. Myelodysplastic
- D. Sideropenic
- E. Haemologic

4. A 17-year-old patient complains of nasal haemorrhages, multiple bruises on the anterior surface of the trunk and extremities, sudden weakness. In blood: Hb- 74 g/l, reticulocytes - 16%, RBCs - $2,5 \cdot 10^{12}/l$, platelets – $30 \cdot 10^9/l$, ESR- 25 mm/h. What is the most effective measure for the treatment of thrombocytopenia?

- A. Splenectomy
- B. Iron preparations
- C. Cytostatics
- D. Hemotransfusion
- E. Vitamin B₁₂

5. A 2950-g black baby boy is born at home at term. On arrival at the hospital, he appears pale, but the physical examination is otherwise normal. Laboratory studies reveal the following: mother's blood type A, Rhpositive; baby's blood type O, Rh-positive; hematocrit 38%; reticulocyte count 5%. Which of the following is the most likely cause of anemia?

- A. Fetomaternal transfusion
- B. ABO incompatibility
- C. Physiologic anemia of the newborn
- D. Sickle cell anemia

6. What are the blood parameters in the drepanocytic anemia?

- A. drepanocytosis
- B. elliptocytosis
- C. target cell anemia
- D. acanth(r)ocytosis
- E. spherocytosis

7. In what kind of anaemias can we find Heinz's bodies?

- A. Iron deficiency
- B. vitamin B12 deficiency

- C. Diamond-Blackfan syndrome
- D. thalassemia
- E. Minkowsky-Shauffard disease

8. What are the blood parameters in the Diamond-Blackfan syndrome?

- A. Microspherocytosis, anemia, reticulocytosis.
- B. Anisocytosis, decreasing of erythrocytes, thrombocytopenia.
- C. Makroanisocytosis, poicilocytosis of erythrocytes, decreasing of hemoglobin.
- D. Normocytosis, decreasing of erythrocytes.
- E. Macrocytosis, elevated fetal hemoglobin (Hb F), thrombocytosis

9. The Etiology of hemolytic anemia

- A. snake venom
- B. sulfonamide
- C. blood transfusion
- D. becoming too cold
- E. all listed above

10. The stimulator of eritropoesis....

- A. vasopressin
- B. aldosterone
- C. insulin
- D. thyroxin
- E. all listed above

Answers: 1-B, 2-A, 3-D, 4-C, 5-A, 6-A, 7-B, 8-C, 9-D, 10-A

Materials of the medical support for the students independent training: a reference chart for organization of students independent work with educational literature.

Tasks	Instructions
To study the etiology and pathogenesis of iron-, protein- and vitamin scarce anaemias in children	To select the key links of anaemias pathogenesis.
To study the clinical manifestations of iron-, protein- and vitamin scarce anaemias in children.	To establish the symptoms and gather it to clinical syndromes which enable to make the credible diagnosis of anaemias.
To study diagnostic criteria of anaemias.	To make the structural plan of disease
To study the additional methods of	To work out a plan of patient's

research (laboratory, instrumental)	examination.
To study the changes in additional investigational methods which are pathognomonic for anaemias.	To enumerate the basic diagnostic criteria of anaemias according to the data of additional investigational methods.
To conduct differential diagnostics, to establish a final diagnosis	To substantiate the basic components of diagnosis in accordance with the modern classification, and to conduct a differential diagnosis.
To prescribe the individual holiatry to patient with anaemias. To be able to render the first aid in haemorrhage for children.	To make the prescribing chart specifying the regimen, diet, medicinal treatment, taking into account the age, severity of patient state, stage of disease, presence of complications and concomitant diseases.

Basic literature:

1. Nelson textbook 18th Edition by Robert M. Kliegman, MD, Richard E. Behrman, MD, Hal B. Jenson, MD and Bonita F. Stanton, MD. SAUNDERS. 2007. – 3200 p.
2. Essential pediatrics. 6th Edition Revised and Enlarged, O.P. Ghai, Piyush Gupta, V.K. Paul. Published by Dr. Ghai Delhi-92, 719 p.
3. Майданник В.Г. Педиатрия. Учебник (2-е издание, испр. и доп.) - Харьков: Фолио, 2005. - 1125 с.
4. Schafermeyer RW: Pediatric abdominal emergencies. In: Tininalli JE et al (eds). *Emergency Medicine*. McGraw-Hill, New York, 2000, p. 846.

Informational resources:

1. <http://www.medscape.org/>
2. <http://www.netmedicos.com/>
3. <http://www.sciencedaily.com/>
4. <http://www.ncbi.nlm.nih.gov/omim>
5. <http://www.hugenavigator.net/HuGENavigator/home.do>
6. <http://kroktest.org.ua/kroki/krok-2/foreign-tests/base-foreign-students-englishstep-2>
7. <http://www.ncbi.nlm.nih.gov/pubmed>
8. <http://pediatrics.aappublications.org/>
9. <http://www.jpeds.com/>
10. <http://www.springer.com/medicine/pediatrics/journal/431>
11. <http://www.angelfire.com/in/pedscapes/>
12. <http://www.mdlinx.com/pediatrics/news.cfm>

Theme: Leukemias in children.

Leukemias and lymphadenomas in children: etiology, pathogenesis, classification, diagnostics, differential diagnostics among other diseases of the blood system and diseases with hyperplastic syndrome, treatment. The first aid in hemorrhagic syndrome and in syndromes of prelum. Prognosis.

I. Actuality of the theme.

Leukemias are the most common childhood cancers, accounting for about 33% of pediatric malignancies. Acute lymphoblastic leukemia (ALL) represents about 75% of all cases, with a peak at the age of 4 yr. Acute myeloid leukemia (AML) accounts for about 20% of leukemias, with an incidence that is stable from birth through the age of 10 yr, increasing slightly during adolescence. Most of the remaining leukemias are the chronic myeloid form; chronic lymphocytic leukemia is rarely seen in children. The overall annual incidence of leukemia is 42.1 per million white children and 24.3 per million black children. The difference is due mainly to the lower incidence of ALL among black children. General clinical features of the leukemias are similar because all involve and severe disruption of bone marrow function. Specific clinical and laboratory features differ, however there is marked variability in responses to therapy and in prognosis.

Concrete purposes:

1. To determine the etiologic and pathogenetic factors in diffuse leukemias and lymphadenomas in children.
2. To classify and analyse the typical clinical manifestation of leukemias and lymphadenomas in children.
3. To determine the features of leukemias and lymphadenomas in children and put the initial clinical diagnosis.
4. To make the plan of examination and analyse the information about laboratory and instrumental data in the classic course of leukemias and lymphadenomas in children.
5. To demonstrate skills of treatment, rehabilitation and prophylaxis of leukemias and lymphadenomas in children.
6. To diagnose and render an urgent help in cell lysis crisis in children.
7. To conduct differential diagnostics among leukemias and lymphadenomas and put the final diagnosis.
8. To determine the prognosis for life in leukemia and lymphadenoma in children.
9. To demonstrate the skills of medical specialist's moral and deontological principles and principles of professional subordination in pediatrics.

II. Classes (pointing of planned mastering level)

1. A student must have a notion (to familiarize): $\alpha 1$

- About the place of leukemia in the structure of blood system's diseases in children, dependent on different age and ethnic groups;
- About statistical information in relation to morbidity, frequency of complications, lethality, immediate and long-term prognosis for patients;
- About the history of scientific studying and the contribution of domestic scientists;

2. A student must know (to master): $\alpha 2$

- etiology of leukemia;
- key links of leukemia's pathogenesis;
- citochemical and immunologic classification of leukemias;
- classical clinical manifestation of leukemia;
- classical clinical manifestation of lymphadenoma;
- laboratory and instrumental diagnosis of leukemia;
- complications of leukemia and lymphadenoma;
- treatment principles of leukemias and lymphadenomas in children;

3. A student must master: $\alpha 3$

Skills:

- Complaints and anamnesis taking;
- Examination of patient with leukemia and revealing the main symptoms and syndromes.
- To formulate and substantiate the initial diagnosis;
- Determination of laboratory and instrumental examination, to make the plan of patient's investigation (according to diagnostics' standards).

Abilities:

- To interpret the results of laboratory and instrumental tests;
- To conduct differential diagnostics with a mielodysplastic syndrome, lymphogranulomatosis, non-Hodjkin lymphadenomas, and other clinical states which are accompanied by the increase of lymphatic nodes, increase of temperature, bleeding and other signs of disease;
- To give recommendations in relation to the patient's regimen and diet in leukemia and lymphadenoma, taking into account the stage of disease, severity of the state and the concomitant pathology;
- Taking into account the stage of disease to specify the severity of the state and concomitant pathology;

- To complete the treatment plan in leukemia and lymphadenomas according to the standards taking into account the stage of the disease, complications and concomitant pathology;
- To render the first aid in extreme situations and exigent states.

III. Aims of personality development (educative aims):

- A student must learn to adhere to the rules of behaviour and principles of medical etiquette and deontology near a patient's bed;
- Be able to set a psychological contact with a patient and his family;
- To master the sense of professional responsibility for a timeliness and adequacy of skilled medicare.

IV. Interdisciplinary integration:

Subject	To know	To be able
1. Previous (providing)		
Anatomy	Structure of the human's haemopoetic and lymphatic systems	
Histology	Structure of haemopoetic organs, their morphological features during the process of maturation	
Physiology	Physiology of haemopoetic organs and normal process of bone marrow maturation, normative indices of laboratory and instrumental methods and their value	To interpret the data of laboratory and instrumental investigational methods.
Pathologic physiology	Key links of leukomogenesis	
Pathologic anatomy	Morphological features of leukemias developing depend on the stage of the process	To analyse and interpret the information of clinical examination and about additional methods of investigation
Pharmacology	Pharmacokinetics and pharmacodynamics, preparations side effects	To prescribe age-dependent treatment of patient, taking into account individual

	(antibiotics, chemotherapy drugs, etc.), used in the treatment of patients with leukemias	features and period of disease, to establish the individual regimen of taking the preparations and their dosage. To prescribe recipes.
Propedeutical pediatrics.	Basic stages and methods of patient's clinical examination	To take complaints, anamnesis vitae et morbi, to find out the basic risk factors of leukemia, to conduct patient's examination, to reveal the clinical signs of leukemia, to interpret the data on additional methods of investigation.
Sternal puncture	Normative indices of sternal puncture	To interpret the information of sternal puncture
3. Intradiscipline integration		
Lymphogranulomatosis	Clinical signs of lymphogranulomatosis	To reveal the characteristic clinical signs of lymphogranulomatosis and differential diagnostics of the signs of leukemia
Non-Hodgkin disease	The signs of Non – Hodgkin disease	To reveal the main clinical signs of Non –Hodgkin disease and differential diagnostics among the signs of leukemia
Trombocitopenia	The signs of trombocitopenia	To reveal the characteristic clinical signs of trombocitopenia and differential diagnostics of the signs of leukemia.

Methodical materials for the class basic stage supporting

A professional algorithm of patient's management (reference chart) for the practical skills and abilities forming.

No	Task	Sequence of implementation	Remarks and warnings related to self-control
1	To conduct an	1.To conduct the complaints and	To pay attention to the

	examination of a patient with leukemia	<p>disease's anamnesis taking.</p> <p>1.To conduct the complaints and disease's anamnesis taking.</p> <p>3.To conduct examination of the patient.</p> <p>4.To examine lymphatic nodes system of the patient (palpation).</p>	<p>features of disease course, underlying factors, concomitant diseases etc.</p> <p>To establish the risk factors of the disease occurrence.</p> <p>To assess patient's general condition, position in bed, color and wetness of skin and mucouses, presence of neck veins and extremities swelling.</p> <p>To pay regard to the rhythm of pulse, it tension and size on both hands, apex shove, its properties, margins of absolute and relative cardiac dullness, its changes, HR (tachi or bradycardia, extrasystole), BP.</p>
		<p>5.To conduct condition of heart and main vessels auscultation.</p> <p>6.To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the digestion system.</p>	<p>To pay regard to heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To pay attention to the features of percussion and auscultation in different aged children.</p>
2	To formulate the initial diagnosis.	<p>1.To formulate the initial diagnosis</p> <p>2.To substantiate all the components of initial diagnosis, based on complaints,anamnesis, and examinations.</p>	<p>Taking the classification as a starting point to formulate the initial diagnosis of leukemia and to substantiate each component of it.</p>
3	To evaluate the parameters of	<p>1.To evaluate the blood count data.</p>	<p>To pay attention to the presence of leucocyto-</p>

	additional laboratory investigations.	2.To interpret the data of CSF.	sis, shifting of formula, increasing of SR, presence of blasts. To pay attention to the presence of blasts and their morphology, features of red stem in SP. To pay attention to agent and sensitiveness to antibiotics.
4	To understand the data of additional and laboratory investigation.	To understand the chest X-Ray data, SP, ECG, and ultrasound.	To pay special attention to the signs of pneumonia, lungs infiltration, to additional formations in lungs etc., ECG signs, hepatosplenome by ultrasound.
5	To conduct differential diagnosis.	1.Consistently to find the common signs in complaints,life and disease anamnesis, data of the examination, data of laboratory and instrumental examination of the patient and in similar states. 2.To find out the differences between complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumental methods of research and in similar nosology. 3.On the basis of the differences to found out similar diseases from the list of probable diagnoses. 4. To conduct differential diagnostics according to the mentioned algorithm among all the nosologies which have the similar signs. 5.Taking into account the impossibility to exclude the diagnosis of leukemia from the list of probable diagnoses to draw a conclusion about the highest degree of probability of such diagnosis.	Special attention must be payed to differential diagnosis among the lymphogranulomatosis, non-Hodgkin disease and hemorrhagic syndromes.
6	To formulate the	1. To formulate the final clinical	Taking as a starting

	concluding clinical diagnosis.	diagnosis. 2. Taking as a starting point the provisional diagnosis, additional investigations data, conducted differential diagnosis to substantiate all the elements of concluding clinical diagnosis.	point the classification of leukemia to formulate a provisional diagnosis, complications of the disease and the presence of concomitant diseases.
7	To prescribe treatment for patient.	1.To prescribe non medicinal treatment. 2.To prescribe the medicinal treatment	Expressly to specify the regimen and detailed diet according to the disease. Taking into account the age, severity of the patient's state, the stage of the disease, the presence of complications and concomitant pathology, to prescribe modern medicinal treatment in accordance with the standards of leukemia's therapy.

VII. Material for the control and medical providing of the class.

A question for initial level control of knowledge, skills and abilities:

Definition of hemoblastoses.

1. To determine an etiology of acute and chronic leukemias in children.
2. To specify the key links of leukogenesis.
3. Basic statements of modern cytomorphologic and immunologic classification of leukemias.
4. To call the facilitating factors for leukemias origin.
5. To call the typical clinical manifestation of acute and chronic leucemia in children.
6. To compose the plan of laboratory and instrumental investigation of the patient.
7. To specify the most typical complications of leukoses in children.
8. To specify the principle of acute leucoses therapy. BFM protocols.
9. To specify the main groups of chemopreparations in the treatment of children with leukemia.
10. Prognosis and sensitivity to chemotherapy based on the most typical chromosome aberrations in acute and chronic leukemias in children.

The first level tests

1. To the child sick with acute lymphoblast leukemia, for correction of anemia the hemotransfusion with packed red cells was performed. What laboratory investigations should be done necessarily after a hemotransfusion?

- A. Coombs test, functional liver tests.
- B. Determine hemoglobin, RBC, urine tests.
- C. Proteinogramme, coagulogramme.
- D. Electrolytes in a blood and urine.
- E. Urinary acid of blood, acid and alkaline condition of blood.

2. The family of a child having been diagnosed with acute lymphoblastic leukemia applied for information about the child's prognosis. Which of the following included is a poor prognostic sign?

- a. White blood cell count at diagnosis of less than $50 \times 10^9/l$
- b. Hyperdiploidy with more than 50 chromosomes
- c. Presence of a mediastinal mass
- d. Age between 1 and 10 years
- e. Early pre-B-cell variety of the disease

3. . A boy of 12 years old, has been admitted to the clinic with complaints on short wind, cough, increasing of body temperature up to $37, 0C$. He's been falling sick during 3 months. On thorax X-ray a "pipe"-like mediastinal shadow with presence of polycyclic contours has been revealed. What preliminary diagnosis is the most probable?

- A. Leukemia
- B Tubercular bronchadenitis
- C Lungs cancer
- D Sarcoidosis
- E Lymphogranulomatosis

4. A boy of 4 years old, has been admitted with complaints of fever, itch, raised sweating, enlarged cervical and axillary lymph nodes. On examination of the patient the lymph nodes conglomerate (like potatoes in the bag) in the left site of the neck and in the right axillary area are palpated. The liver and lien are enlarged. What are the most typical signs of lymphogranulomatosis in the patient?

- A. Splenomegaly
- B. Itch
- C Conglomerate of lymphonoduses
- D Hepatomegalia
- E Fever

5. A girl of 12 years old during 6 months has been complaining of growing thin, labored respiration and dry cough. On X-ray of the thorax there are considerably enlarged mediastinal lymph nodes. Mantoux test is negative. Hemogramme: Hb-90 g/l, erythrocytes. - $2,9 \times 10^{12}/l$, thrombocytes - $94 \times 10^9/l$, leucocytes - $12 \times 10^9/l$, relating to stab neutrophiles - 12 %, segmented - 70 %, blood sedimentation rate is 18 mm / hour. What is the prime test for establishing of diagnosis?

- A. Histological investigation of mediastinal lymphonoduses
- B Morphological investigation of bone marrow
- C Computer tomography of abdominal cavity
- D Spirographic tests with Metacholine and Salbutamololum
- E Thermometry in each 3 hours during a week

6. A girl of 6 years old, was taken to hematology department in a serious state: a high fever, all groups of lymph nodes are enlarged, hemorrhagic syndrome, hepatosplenomegalia. Blood count: RBC - $2,0 \times 10^9/L$, HB - 84 g/l, CI: 0,75, WBC- $24,0 \times 10^9/L$, eosinophiles.-3 %, relating to stab neutrophile -1 %, segmented neutrofiles.-16 %, lymphocytes-75 %, monocytes -5 %, a thrombocytes.- $150 \times 10^9/l$, ESR-56 mm/h . In a myelogram the blasts is 92 %. Which of the listed parameters

plays the main role for the establishing of a diagnosis?

- A. Blastosis in a myelogram
- B. Leukocytosis
- C. Lymphocytosis
- D. Thrombocytopenia
- E. Anemia

7. A child of 4 years old. During the last 4 months asthenia, dermal hemorrhages admitted. Nasal bleedings, paleness, hyperthermia. On the routine blood analysis : haemoglobin - 45 g/L, erythrocytes - $1,2 \times 10^9$ /L, a color index - 0,9, leucocytes $1,5 \times 10^9$ /L, relating to stab neutrophile - 1 %, segmented - 25 %, eosinocytes - 1 %, lymphocytes - 5 %, monocytes - 4 %, blood sedimentation rate 50 mm / h, thrombocytes - 40×10^9 /L. What is the preliminary diagnosis?

- A. Iron deficiency anemia
- B. Aplastic anemia
- C. B12 - deficiency anemiamaculae
- D. Hemolytic anemia
- E. Acute leukosis

8. An 8-year-old child being treated with a combination of chemotherapy agents develops very red, inflammed sores in the mouth and esophagus. He has difficulty at eating and drinking food and liquids. Which of the following

antineoplastic agents is the most likely etiology?

- A. Cephasoline
- B. Prednisone
- C. Dexametasone
- D. Antifungal drugs
- E. Methotrexate

9. What are the most distinctive features of peripheral blood and bone marrow changes that allow to suspect a myelodysplastic syndrome?

- A. WBC increasing, normal or decreased elements in bone marrow.
- B. PLTC increasing and increased bone marrow elements.
- C. WBC increasing with simultaneously decreased thrombocytes and erythrocytes and the signs of elements differentiation impairment in the bone marrow.
- D. Decreasing of blood elements (RBC, WBC, PLTC) in simultaneously significant decreasing of bone marrow elements
- E. RBC, WBC, PLTC decreasing and simultaneously increasing bone marrow elements and the signs of impaired bone marrow elements differentiation.

10. In what types of acute lymphoblastic leukemia leucemides on the skin is the most common sign?

- A. Myeloblast
- B. Lymphoblast
- C. Monoblast.
- D. Promyelocytic.
- E. Megacariocytic.

Answers: 1-b, 2-c, 3-e, 4-c, 5-a , 6-c, 7-b, 8-e, 9-a, 10-a.

Typical situational tasks of 2 level

Task -1

Patient D., 7 years, transferred to children department from ophthalmologic department, where he had stayed because of lacrimal sacculus phlegmon.

On examination: general condition is mild, skin pallor, some hemorrhages on the upper extremities. On cheek mucosae there is a hemorrhagic rash. Enlarged lymph nodes palpated (up to 2 cm in diameter): subscullar, retrocervical, supraclavicular, subinguinal, inguinal are painful and elastic in consistency

The lung breathing is unchanged. Pulse rate is 74 per min. BP 144 to 90 mm Hg. Tongue is clear. Hemorrhages on cheek mucous and palate. Gums are pale. Throat is red. Glands are uneven, enlarged. Liver and spleen are unchanged.

Blood count: Er. $2.1 \times 10^{12}/l$, Hb 74 g/l, CI 1, polichromathophilia, anisocytosis, poicilocytosys, reticulocytosys 30%, leuc. $4.5 \times 10^9/l$, bands: 6%, s: 10%, lymph..80%, blasts 4%, tr. $33 \times 10^9/l$, SR 55 mm. per hour. Urine count is unchanged.

1. Establish the diagnosis.
2. What additional investigational methods are needed?
3. How to explain the mucosal hemorrhages in this patient?
4. Evaluate the blood count
5. Does lacrimal saccus phlegmone related to basic condition in this patient?

Standard of answer . Task 1.

1. Acute leucosis.
2. Bone marrow punction.
3. Mucosal hemorrhages could be explained by the megakaryocytic stem cell depression caused by the bone marrow blast transformation.
4. Blood count demonstrates severe anemia, elevated reticulocytes, thrombocytopenia and presence of blast cells, increased sedimentation rate.
5. Lacrimal saccus phlegmone like other unusual opportunistic infections could be seen because of depressed immunity in leukemic patient.

Task-2

Boy U. was admitted to the clinic with complaints on enlargement of subjaw and parauricular lymphatic nodes, increased body temperature. He has been feeling seek during a month, was treated in the outpatient department for the diagnosis of

epidemic parotitis without any effect. Besides, the hemorrhages on skin and pallor appeared. In the outpatient department the blood count was taken where the leucocytes consists of $3,7 \times 10^9/l$, lymphocytes were 90 %, SR 70 mm/h.

From the anamnesis it was reported: from the first pregnancy, normal development. In admission to the hospital the general state is mild, pallor, ecchymomas on the extremities, enlargement of cervical and subjaw lymphnodes up to 2-3 cm in diameter, dyspnea, weakened breathing on lungs auscultation, no rales. On heart auscultation the systolic murmur over the fifth site was heard with mild intensity. Liver is 4 cm under the costal arch, spleen is 3 cm, dense and painless.

Blood count: erh. $2.56 \times 10^{12}/l$, Hb 60 g/l, anisocytosis. Leucocytes: $3,5 \times 10^9/l$, e: 1%, bands: 5%, s: 9%, lymph 11%, mon:1%, lymphoblasts 73 %, SR 2 mm/h.

1. What disease could be suggested?
2. What are the additional investigations?
3. What test will confirm the clinical diagnosis?
4. How to explain heart disorders in this patient?
5. How to explain the initial non specific signs of diseases and their unsuccessful treatment?

Standard of answer . Task 2.

1. Acute lymphoblast leukemia
2. Bone marrow puncture.
3. The presence of lymphoblasts, anemia in the hemogramme, lymphadenopathy, hepatolienal syndrome.
4. Heart disorders like murmurs is secondary by severe anemia and decreased blood viscosity with consequent blood flow turbulence.
- 5, It could be explained by preleukemic stage of the disease.

Task-3

A patient, 4 years, old, had symptoms of an acute disease: fever, relapse vomiting. Next 2-3 days a considerable general weakness appeared. He was

admitted to the hospital with a diagnosis of Botkin's disease to infectious department and after examination and blood count taking was transferred to children department. From the anamnesis it was reported that the child was born healthy, in the past he hadn't been ill. The parents are healthy.

On examination: the general condition is grave, the skin is pale with a rather yellow tint, scleres are subicteric. Pereferic lymphonodes are palpated up to the kidney bean dimensions, dense, painless. Pulse is 76 beats per minute, rhythmic, filled satisfactory, Heart margins are normal, on auscultation the systoloic murmur over all the sites of auscultation. Liver is 2 cm under the costal arch, painless and soft. Spleen is 4-5 cm under the costal arch, soft and painless. Body temperature is 37.8- 39.8 C. Blood count: erh.0.98*10x12/l, HB 28 g/ l, L: 3.8*10x9/l, neutrophilosis without shifting in formula. In the blood smear there is a big amount of normoerhythroblasts. In blood data Tr.12*10x9/l, reticulocytes 22%, General bilirubin is 102 mcmol/l, indirect bilirubin 96 mcm/l. Urine is darkly brown color with big sediment of urates. Urobilin test is very positive.

1. Establish preliminary diagnosis.
2. What additional methods of investigation are necessary?
3. How to explain the urates has detected in urine?
4. Asses the bilirubin level.
5. With what conditions it should be differentiate?

Standard of answer . Task 3.

- 1.Hemolytic anemia.
2. Osmotic resistance of erhythrocytes.
3. The presence of urates in urine caused by increased cell metabolism is seen in hemolysis.
4. Patient have indirect hyperbilirubibemia caused by increased destruction of erhythrocytes.
5. Acute leukemia, hypoplastic anemia, Estren- Damesek anemia, acute hepatitis,

Task-4

Patient G., 6 years, was admitted to the hospital with complaints of stomach-ache, general weakness, bad appetite. Stomach-ache and general weakness had appeared 2 months ago, and the appetite disappeared then. To these phenomena pallor and fever were added. In the hospital the state of the child is severe, the child is flaccid, the paleness progresses: peripheral lymph nodes are enlarged, insignificant pain on patting the thorax, single hemorrhages. The spleen is considerably enlarged (lower pole palpated near the pelvis) and dense in consistency.

The liver is 4 cm under the edge of costal arc. Blood count: $er.2.5 \cdot 10^{12}/l$, Leuc: $44 \cdot 10^9/l$, 77% of blasts are the myelocytes, 1% are the bands, 1% are segmented cells, 2% are lymphocytes, 19% are normoblasts -2:100, SR - 18 mm/h

1. To establish the initial diagnosis.
2. Why can't given the data allow to establish the final diagnosis?
3. Work out a plan of examination.
4. How to explain the stomach ache in his patient.
5. With which conditions it should be differentiate?

Standard of answer . Task 4.

1. Acute leukemia
2. Because, there is a requirement for the determination of blasts type.
3. Bone marrow puncture, clotting tests, ultrasound of the heart, spleen and hepatobiliary system, chest X-ray.
4. Stomach aches in this patient caused by enlargement of intestinal lymph nodes.
5. Hypoplastic anemia, hemolytic anemia, chronic gastritis/

The questions for the control of secondary knowledge level of abilities and skills:

1. Definiton of hemoblastoses.
2. To determ an etiology of acute and cronic leukemias in children.
3. To specify the key links of leukomogenesis.
4. Basic statements of modern cytomorphologic and immunologic classification of leukomias.
5. To define the facilitating factors for leukemias origin.
6. To define the typical clinical manifestation of acute and chronic leucemia in children.
7. To compose the plan of laboratory and instrumental investigation of leukemic child patient.
8. To specify most typical complication of leukoses in children.
9. To specify the principle of acute leucoses therapy. BFM protocols.
10. To specify the main groups of chemoreparations in treatment children with leukemia.
11. Prognosis and sensitivity to chemotherapy based on the most typical chromosome abberations in acute and chronic leukemias in children.

VII. Materials of the medical support for the students independent training: a reference chart for organization of students independent work with educational literature.

Tasks	Instructions
To study the etiology and pathogenesis of leukemia and lymphadenoma in children. To be able to detect the risk group for the severity of leukemia.	To enumerate basic etiologic factors, select the key links of leukemia, pathogenesis.
To study clinical manifestations of leukemias and lymphadenomas in children.	To establish the symptoms and gather it to clinical syndromes which enable to put the credible diagnosis of leukemia.

To study diagnostic criteria of leukemia.	To make the flow diagram of the disease
To study the additional methods of research (laboratory, instrumental)	To work out a plan of patient's investigation.
To study the changes in additional investigational methods which are pathognomonic for leukemias.	To enumerate the basic diagnostic criteria of leukemia according to the data of additional investigational methods.
To conduct differential diagnostics, to establish a final diagnosis	To substantiate the basic components of diagnosis in accordance with the modern classification, and to conduct a differential diagnosis.
To prescribe the individual poliatry to patient with the leukemia. To be able to render the first aid in cell lysis crisis for children.	To make the prescribing chart specifying the regimen, diet, medicinal treatment, taking into account the age, severity of patient's state, the stage of the disease, the presence of complications and concomitant diseases.

The second level tests

1. The otherwise healthy 17-year-old complains of swollen glands in his neck and groin for the last 6 months and an increasing cough for the previous 2 weeks. He also reports some fevers, especially at night, and possibly some weight loss. On examination, you notice that he has nontender cervical, supraclavicular, axillary, and inguinal nodes, no hepatosplenomegaly, and otherwise looks to be fairly healthy. Which of the following would be the appropriate next step?

- a. Chest radiograph

- b. Complete and differential blood counts
- c. Trial of antituberculous drugs
- d. Urine tests
- e. Cat-scratch titers

2. A child of 8 years old. Increasing paleness, weakness, hemorrhagic eruption on the skin have appeared. On bone marrow puncture the depression of hemopoiesis was marked. What basic method of therapy is indicated for this case?

- A. Antibiotics + hemotransfusion
- B. Splenectomy
- C. Haemotransfusion + cytotoxic agents
- D. Cytotoxic agents + bone marrow transplantation
- E. Corticosteroids +bone marrow transplantation

3. Most favourable variant of acute lymphoblast leucemia:

- A. O-cell
- B. T-cell
- C. Pre-B-cell.
- D. Variant with translocations (9;22)
- E. B-cell

4. The complete remission criteria in acute leucemia:

- A. Absence of complete remission in acute leucosis.
- B. Satisfactory general condition and normal blood count.
- C. Normal blood count, in bone marrow smear not more than 5% of blasts, normal liquor count.
- D. Normal blood count, spleen enlargement.
- E. Normal blood count, diminished spleen dimensions.

5. From what age is prophylactic cranial irradiation conducted for children in

acute leucosis?:

- A. After 1 year old
- B. After 3 years old
- C. After 10 years old.
- D. After 5 years old
- E. In 1 year old.

6. Concept of “hybrid” leucosis is:

- A. The presence on the blast cells of lymphoid and myeloid markers lines at the same time.
- B. Two clones of blasts belong to myeloid line only (myeloblasts and monoblasts).
- C. Two clones of blasts belong only to lymphoid line but to different cell lines (T- and B- lymphocytes).
- D. The presence of the tumor substrate cells of chronic and acute leucoses at the same time.
- E. Two clones of lymphoid or myeloid blasts at different stage of differentiation belong to one cell line.

7. What are the hematologic signs of 2 stage (stable) true polycytemia.

- A. Hepato-, -splenomegaly, -myelofibrosis.
- B. Hepato-, -splenomegaly, hemorrhagic syndrome.
- C. Hepato-, -splenomegaly, thromboses, hemorrhages, hyperplasia of all stems in the bone marrow.
- D. Anemia, leucocytosis, thrombocytopenia.
- E. All answers are correct.

8. Prophylaxis of neuroleucemia is conducted by intralumbar administration of the following preparations:

- A. Metotrexat 10,0 mg/m²; citosar 20-30 mg/m²; dexamethason 4 mg/m².
- B. Metotrexat 12,5 mg/m²; citosar 20-30 mg/m²; dexamethason 4 mg/m².
- C. Metotrexat 5,0 mg/m²; citosar 20-30 mg/m²; dexamethason 4 mg/m².
- D. Metotrexat 30,0 mg/m²; citosar 20-30 mg/m²; dexamethason 4 mg/m².
- E. Metotrexat 50 mg/m², ; citosar 20-30 mg/m²; dexamethason 4 mg/m².

9. Prophylaxis of neuroleucemia in children's ALL includes:

- A. Cranial irradiation.
- B. High doses of citosar in combination with average high doses of metothrexat
- C. Intrathecal and i.v. metothrexat administration in combination with prophylactic cranial irradiation.
- D. High doses of metothrexat in combination with high doses of citosar.
- E. High doses of metothrexat in combination with prophylactic cranial irradiation.

10. The complete remission criteria in acute leucemia:

- A. Absence of complete remission in acute leucosis.
- B. Satisfactory general condition and normal blood count.
- C. Normal blood count, in bone marrow smear not more than 5% of blasts, normal liquor count.
- D. Normal blood count, spleen enlargement.
- E. Normal blood count, diminished spleen dimensions.

Answers: 1-a, 2-e, 3-b, 4-c, 5-c, 6-a, 7-c, 8-b, 9-e, 10-b

Basic literature:

1. Eds. Greer JP, et al. Wintrobe's clinical hematology. Volume 2. 12th edition. 2009. Lippincott Williams and Wilkins
2. CYPALL. Guidelines for treatment of children and young persons with acute lymphoblastic leukaemia and lymphoblastic lymphoma (Interim Guidelines 3). 2011.

3. Cardiff University. AML17: Working parties on leukaemia in adults and children in acute myeloid leukaemia or high risk myelodysplastic syndrome. Version 7.1. 2011.
4. Neville K, Steuber P. Clinical assessment of the child with suspected cancer. www.uptodate.com (accessed November 2012).
5. [Seif AE](#); Pediatric leukemia predisposition syndromes: clues to understanding leukemogenesis. *Cancer Genet.* 2011 May;204(5):227-44. doi: 10.1016/j.cancergen.2011.04.005.
6. [Rubnitz JE, Inaba H](#); Childhood acute myeloid leukaemia. *Br J Haematol.* 2012 Nov;159(3):259-76. doi: 10.1111/bjh.12040. Epub 2012 Sep 12.
7. [Long term follow up of survivors of childhood cancer](#); Scottish Intercollegiate Guidelines Network - SIGN (Mar 2013)
8. [Grigoropoulos NF, Petter R, Van 't Veer MB, et al](#); Leukaemia update. Part 2: managing patients with leukaemia in the community. *BMJ.* 2013 Apr 9;346:f1932. doi: 10.1136/bmj.f1932.

Theme: Haemorrhagic diseases in children. Haemophilia, thrombopathy, thrombopenia. Etiology. Pathogenesis. Classification. Diagnostics. Differential diagnostics between haemorrhagic diseases in children. Treatment. The first aid in case of haemorrhage. Prognosis.

The amount of studying hours – 4 academic hours.

I. Actuality of the theme. Haemorrhagic syndrome can manifest itself as an independent disease or can be a manifestation of other pathology. A course of haemorrhage is damage in haemostasis, which can be primary in case of congenital haemorrhagic diseases and secondary in case of complications. Bleeding cessation occurs in compliance with interaction of three haemostasis links: vascular, platelets and coagulatory. Isolated or combined breaking in one or several hemostasis links can lead to haemorrhagic syndrome.

II. Classes (pointing planned mastering level with)

1. A student must have conception (to familiarize): $\alpha 1$

- the place of haemorrhagic diseases in the structure of diseases in children;
- statistical information in relation to morbidity, frequencies of complications, lethality, the nearest and remote prognosis;
- the history of scientific studying and assessments of domestic scientists.

2. A student must know (master): $\alpha 2$

- risk factors of manifestation and pathogenesis of haemorrhagic diseases in children;
- modern scheme of coagulation and anticoagulative systems, thrombocyte haemostasis;
- key links of haemorrhagic diseases' pathogenesis;
- clinical classification of haemorrhagic diseases in children;
- classic clinical manifestation of haemophilia;
- classic clinical manifestation of thrombopenia;
- classic clinical manifestation of thrombopathy;
- laboratory diagnosis of haemophilia;
- laboratory and instrumental diagnostics of haemorrhagic diseases;
- complications of haemorrhagic diseases in children;
- treatment principles of haemorrhagic diseases in children;
- preventive measures of haemorrhagic diseases in children, rehabilitation methods of patients and their dispensary observation.

3. A student must master: $\alpha 3$

Skills:

- Collection of complaints and anamnesis morbi;
- Examination of patient with haemorrhagic disease and revealing the main symptoms and syndromes;
- To formulate and substantiate preliminary diagnosis;
- Determination of laboratory and instrumental plan of patient's examination (according to diagnostics standards);
- Giving the first aid in case of acute bleeding, haemorrhagic shock in children;
- To realize life prognosis of a patient with haemophilia, thrombopathy and thrombopenia.

Abilities:

- to interpret the result of laboratory and instrumental tests;
- to conduct differential diagnosis among haemophilias, thrombopenias and thrombopathias;
- to conduct differential diagnosis among diseases with bleeding;
- to give recommendations in relation to the regimen and diet of a patient with haemorrhagic disease, according to the stage of disease, severity of the state and concomitant pathology;
- to complete the treatment plan in haemorrhagic disease according to standards taking into account the stage of disease, complications and concomitant pathology;
- to render the first aid in extreme situation and exigent states.

III. Aims of personality development (educative aims):

- A student must adhere rules of behaviour and principles of medical etiquette and deontology, to develop bedside manner;
- to set a psychological contact with patient and his family;
- to master the sense of professional responsibility for a timely and adequate medicare.

Interdisciplinary integration:

Subject	To know	To be able
1. Previous (providing)		
Physiology and pathologic physiology	Coagulating factors of blood system, platelets haemostasis. Modern scheme of blood clotting.	To determine etiologic factors of possible bleeding.
Pharmacology	Pharmacokinetics and pharmacodynamics, the side effects of hemostyptic therapy.	To prescribe treatment dependent on age and patient's individual features, period of disease, to establish individual regimen of taking the preparations and its

		dosage. To prescribe recipes.
Propedeutical pediatrics.	Clinical and laboratory diagnostic methods of haemorrhagic syndromes. Principles of accomplishment of platelet count, bleeding time, prothrombin time, and activated partial thromboplastin time (APTT), the tourniquet test, whole blood clotting time, prothrombin consumption time, and thromboplastin generation test. Basic stages and methods of patient's clinical examination.	To collect complaints, anamnesis vitae et morbi, to find out the basic risk factors of haemorrhagic diseases, to conduct patient's examination, to reveal clinical signs of diseases of blood system, to interpret the data of additional investigative methods. To lay out medical report, to evaluate severity of child's condition.
2. Followings (provided)		
Hospital pediatrics.	Etiology, pathogenesis, clinical signs of haemorrhagic disease, differential diagnosis and treatment tactics.	- To reveal clinical signs and complications of haemorrhagic diseases; - to conduct survey design and medical plan; - to conduct differential diagnosis, to be able to prescribe treatment.
3. Interdiscipline integration		
Hypoplastic and aplastic anaemia	Clinical manifestations of transient hypoplastic and aplastic anaemia	To establish specific clinical signs of hypoplastic and aplastic anaemias. To conduct differential diagnosis between haemophilias, thrombopenias and thrombopathias.
Purpura rheumatica	Clinical manifestations of purpura rheumatica	To establish specific clinical signs of purpura rheumatica. To conduct differential diagnosis between haemophilias, thrombopenias and thrombopathias.
Acute leucosis	Clinical manifestations of acute leucosis	To establish specific clinical signs of acute leucosis. To conduct differential diagnosis between haemophilias, thrombopenias and thrombopathias.
DIC-syndrome	Clinical manifestations of DIC-syndrome	To establish specific clinical signs of DIC-syndrome. To conduct differential diagnosis between haemophilias, thrombopenias and

		thrombopathias.
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VI. Plan and organizational structure of classes.

№ п/п	Basic stages of classes, their function and maintenance	Educational aims are in the levels of mastering	Methods of control and studies	Educational materials	Distributing of time in minutes
1	<p>Preparatory stage</p> <p>Organizational measures</p> <p>Raising of educational aims and motivation</p> <p>Control of basic knowledges and skills level:</p> <p>1. Etiology of haemophilias, thrombopenias and thrombopathias in children;</p> <p>2. Key links of haemorrhagic diseases pathogenesis;</p> <p>3. Clinical classification of haemorrhagic diseases;</p> <p>4. Features of clinic and diagnostic of different haemorrhagic diseases in children.</p> <p>5. Laboratory and instrumental diagnosis of haemorrhagic diseases in children;</p> <p>6. Differential diagnostic of haemorrhagic diseases in children;</p> <p>6. Complication of haemophilias, thrombopenias and thrombopathias in children;</p> <p>7. Treatment principles of haemophilias, thrombopenias and thrombopathias in children;</p> <p>8. First aid in case of acute haemorrhage or haemorrhagic shock in children;</p> <p>9. Prophylaxis of haemorrhagic diseases in children;</p> <p>10. Rehabilitation of</p>	<p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p>	<p>Individual oral questioning</p> <p>Test control of the second level</p> <p>Individual oral questioning</p> <p>Typical situational task of 2 level</p> <p>Typical situational task of 2 level</p> <p>Typical situational task of 2 level</p> <p>Test control of 2 level</p> <p>Typical situational task of 2 level</p>	<p>p. II «Educational aims»</p> <p>p. I «Actuality of theme»</p> <p>Second level tests the table «classification of haemorrhagic diseases»</p> <p>Structurally logical chart of haemorrhagic diseases</p> <p>Typical situational task 2 level</p> <p>Typical situational task of 2 level</p> <p>Tests of 2 level</p> <p>Typical situational tasks of 2 level</p> <p>Kit of medicines.</p>	<p>3 min.</p> <p>12 min.</p> <p>20 min.</p>

	children with haemorrhagic syndrome.				
2	<p>Basic stage of professional skills and abilities forming:</p> <p>1.To conduct the patient management with haemorrhagic diseases, to take complaints and anamnesis.</p> <p>2.To conduct the pateint examination, to detect main symptoms and syndromes of haemorrhagic disease.</p> <p>3.To formulate and substantiate the preliminary diagnosis</p> <p>4.To compose the plan of patients laboratory and instrumental investigation.</p> <p>5.To interpret the results of laboratory and instrumental investigation.</p> <p>6.To conduct differential diagnosis among clinical conditions accompanied by blood systems changes.</p> <p>7.To give the recom-mendations for regimen and diet of patient.</p> <p>8.To compose the plan of treatment of patient with haemorrhagic dise-ase according to the sta-ge of disease and the presence of complications.</p> <p>9.To be able to render the first aid in extreme situations</p>	<p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p> <p>α3</p>	<p>Practical professional training</p> <p>Practical professional training</p> <p>Practical professional training</p> <p>Practical professional training</p> <p>Practical professional training.</p> <p>Tests and the third level control. The third level test control.</p> <p>The practical pro-fessional training is in solving of non standard clinical situations.</p> <p>The third level test control.</p> <p>Practical profess-ional training.</p> <p>The third level test control.</p> <p>Practical profess-ional training.</p> <p>The practical professional training in solving of non typical clinical situations. The third level test control. Practical professional</p>	<p>Algyrthmes for forming practical skills. Patients. Case history.</p> <p>A reference chart for forming of professional abilities. Case history.</p> <p>A reference chart for forming of professional abilities.Situatio nal typical tasks of third level.The third level tests. Prescribing chart.</p> <p>The third level Non-typical situational tasks. Treatment algo-rythm for the haemorrhagic diseases patients. The third level non-typical situational tasks. The first aid algyrthm in case of haemorrhage or haemorrhagic shock.</p>	115 min.

			training in solving of non typical clinical situations.		
3	Concluding stage. Control and correction of professional abilities and skills. Working out the totals of class. Home work (basic and additional literature on the topic)		Analysis of clinical work performances Solving of non typical tasks and the third level tests. Estimation of clinical work.	Clinical work performances The third level Non-typical situational tasks. A reference chart for independent work with literature	30 min.

Questions for elementary level of knowledges control.

1. To determine the concept of haemorrhagic syndrome in children.
2. Modern scheme of clotting, anticoagulative system, thrombocyte haemostasis.
3. What are haemophilia's, thrombopenia's and thrombopathia's etiology and pathogenesis?
4. What are the clinical manifestations of haemophilia, thrombopenia and thrombopathia in children?
5. What are the main diagnostic principles of haemophilia, thrombopenia and thrombopathia in children? To conduct differential diagnostic of haemorrhagic syndrome in children.
6. What is the first aid in case of acute bleeding or haemorrhagic shock?
7. To prescribe treatment, prophylactic and rehabilitations measures in children with haemophilia, thrombopenia and thrombopathia.

Methodical materials for the class basic stage supporting

Primary control tests

1. Newborn boy from the first pregnancy and first delivery. Mother's blood type is O(I) Rh(+), child's is A(II) Rh(+). An icterus increases progrediently after 2 day of life. Liver +4 cm. Bilirubin of blood on the 3-d day of life is 250 mcMol/l, unconjugated is 240 mcMol/l. Direct Coumbs-test is low positive, Hb 160-160 g/l, RBC - $4,5 \times 10^{12}/l$, Ht 0,55. What the most reliable diagnosis?
 - A. Physiologic jaundice
 - B. ABO-conflict
 - C. Jaundice of mother milk
 - D. Conjugated icterus
 - E. Fetal hepatitis

2. Child in age of 9 days, was born in a term with weight 3000g. Apgar score is 8-9 points. From the first day admitted an icterus of skin, liver +3,5cm. Color of urine and feces are not changed. Hb 130 g/l, reticulocytes is 4,6%, common bilirubin is 300 mcMol/l, indirect fraction is - 288 mcMol/l, transaminases: AlAT - 0,28, AsAT - 0,26. During medical treatment the state of child became better, intensity of icterus diminished. Up to 9 day of life icterus acquired greenish color, urine became dark, feces white. In ultrasound cholic channels and gall-bladder scanned clear. Establish the diagnosis:

- A. Biliary atresia
- B. Fetal hepatitis
- C. Intrahepatic cholestasis
- D. Jaundice of Crigler-Najjar
- E. Physiologic jaundice

3. A new-born boy from a mother with the complicated obstetric anamnesis, from third pregnancy, first delivery. At birth a skin is rose. Hb 160 g/l, RBC - 4,5 G/l. Bilirubin of blood from the umbilical vein is 60 mcMol/l. Blood type of mother is O(I) Rh(-), of the child is O(I) Rh(+). The icterus of skin appeared after 8 hours; bilirubin of blood is 116 mcMol/l, unconjugated. Define the tactic of medical treatment.

- A. Exchange blood transfusion
- B. Light-therapy
- C. Enterosorbents
- D. Light-therapy + liquid infusion
- E. Membranstabilizing preparation.

4. In the child of one week an icterus has appeared in the first days and increased due to indirect fraction of bilirubin. Hepatic enzymes are normal. Blood type of mother is A(II) Rh -positive, of the child is 0(I) Rh- positive. What pathology is more likely in this case?

- A. Biliary atresia
- B. Fetal hepatitis
- C. Conjugated icterus.
- D. Hemolytic disease of newborns

E. Crigler-Najjar syndrome

5. In newborn child with hemolytic disease induced by Rh – conflict the blood type is O (I) Rh (+), in mother is (II) Rh(-). What blood must be poured during the operation of exchange blood transfusion?

- A. (II) Rh (-)
- B. A(I) Rh (-)
- C. A (II) Rh(+)
- D. O (I) Rh (+)
- E. B(III) Rh(-)

6. In newborn boy from the first pregnancy in age of one day there was an icterus. Common bilirubin in blood serum is 148 $\mu\text{mol/l}$, indirect bilirubin is 126 $\mu\text{mol/l}$. Coumbs test is positive. A mother has blood type O(I) Rh(-). What is more likely causes the jaundice?

- A. Biliary atresia
- B. The rhesus conflict
- C. Physiological jaundice
- D. ABO-incompatibility
- E. Fetal hepatitis

7. Girl after delivery have following clinical data: icterus, pallor, splenohepatomegalia. Blood type is B(III) Rh(+); Hb in blood 150 g/l, RBC is $4,2 \cdot 10^{12}/\text{l}$, reticulocytes 9 %. Bilirubin of blood is 58 $\mu\text{mol/l}$, unconjugated. Mother's blood is B(III) Rh(-), titer of anti- Rh-antibodies during pregnancy are 1:127; 1:254. What test more reliable will help to define the tactic of treatment?

- A. Routine blood test
- B. Clinical supervision
- C. Increasing of bilirubin per hour
- D. Proteinogramme
- E. Level of hepatospecific enzymes

8. A girl 2-nd day. Pale, hemorrhages on a skin, general edema: liver +6cm. Apgar score is 6 points. Mother's blood is O(I) Rh(-), child's is O(I) Rh(+). The Hb in umbilical cord blood 70 g/l, RBC $1,5 \cdot 10^{12}/\text{l}$, reticulocytes 15%. Bilirubin at birth is

59 $\mu\text{mol/l}$, unconjugated. Woman has abortions in her anamnesis. What the most reliable diagnosis?

- A. Sepsis of newborns
- B. B.Congenital leucosis
- C. Rhesus conflict, edematic form
- D. Fetal hepatitis
- E. Hereditary hemolytic anemia

9. Boy of 3 days. In the end of first day of life an icterus of skin has appeared, a liver was enlarged to 3 cm. Child is enough active, reflexes and muscular tone are not changed. Bilirubin of blood $175 \mu\text{mol/l}$, unconjugated, Hb 150 g/l, RBC $4,7 \times 10^{12}/\text{l}$, Ht 0,5. Define the tactic of medical treatment.

- A. Hemotransfusion
- B. Light-therapy
- C. Exchange blood transfusion
- D. Extracorporal hemosorbtion
- E. Hemotransfusion + membranestabilizing preparations

10. In girl 2 days of life an icterus appeared at the end of the first day. In clinical examination an- icterus of skin and sclera admitted. A live under edge of costal arc on 4 cm, spleen on 2 cm. Mother's blood type is- (0) the Rh+ child's II Rh(+). In routine blood test the reticulocytosis 15 ‰, RBC $2,8 \times 10^{12}/\text{l}$, hemoglobin 120 g/l, bilirubin of umbilical blood is $78 \mu\text{mol/l}$, after 8 hour is $190 \mu\text{mol/l}$. Choose the method of medical treatment:

- A. Exchange blood transfusion
- B. Prescribing of phenobarbital
- C. Light -therapy
- D. Liquid infusion
- E. Intragastral dropping linfusion

Answers: 1-B, 2-C, 3-A, 4-D, 5-B, 6-D, 7-C, 8-C, 9-B, 10-A.

Typical situational tasks of 2 level

Task 1

Both the mother and baby have O-positive blood. The baby's direct serum bilirubin is 20 mcMol/L, with a repeat total serum bilirubin of 118 mMol/L. Urine bilirubin is positive. The mother's white count is 13,000/L with a differential of 53% polymorphonuclear cells, 46% lymphocytes, and 6% monocytes. The hemoglobin is 170 g/L, and the platelet count is 278,000/L. Reticulocyte count is 1,5%. The peripheral smear does not show fragments or abnormal cell shapes. Blood cultures are pending in the laboratory. Liver enzymes and liver ultrasound are normal. G6PD levels and osmotic fragility testing are normal.

1. The most likely diagnosis in this infant is...
2. What the mechanism of it?
3. What the term of appearing?
4. What the criteria for diagnosis?
5. Provide differential diagnosis.

Standard of answer . Task 1.

1. Physiologic jaundice.
2. The development of jaundice in a healthy full-term baby may be considered the result of a normal physiologic process if the time of onset and duration of the jaundice and the pattern of serially determined serum concentrations of bilirubin are in conformity with currently accepted safe criteria.

3. Physiologic jaundice becomes apparent on the second or third day of life, peaks to levels no higher than about 12 mg/dL on the fourth or fifth day, and disappears by the end of the week. The rate of rise is less than 5 mg/dL per 24 h and levels of conjugated bilirubin do not exceed about 1 mg/dL. Concern about neonatal jaundice relates to the risk of the neurotoxic effects of unconjugated bilirubin. The precise level and duration of exposure necessary to produce toxic effects are not known, but bilirubin encephalopathy, or kernicterus, is rare in term infants whose bilirubin level is kept below 18 to 20 mg/dL. Certain risk factors affecting premature or sick newborns increase their susceptibility to kernicterus at much lower levels of bilirubin.

4. The diagnosis of physiologic jaundice is made by excluding other causes of hyperbilirubinemia by means of history, physical examination, and laboratory

determinations. Jaundice appearing in the first 24 h is usually a feature of hemolytic states and is accompanied by an indirect hyperbilirubinemia, reticulocytosis, and evidence of red-cell destruction on smear.

5. In the absence of blood group or Rh incompatibility, congenital hemolytic states (e.g., spherocytic anemia) or G6PD deficiency should be considered. With infection, hemolytic and hepatotoxic factors are reflected in the increased levels of both direct and indirect bilirubin. Studies should include maternal and infant Rh types and blood groups and Coombs tests to detect blood group or Rh incompatibility and sensitization. Measurements of total and direct bilirubin concentrations help to determine the level of production of bilirubin and the presence of conjugated hyperbilirubinemia. Hematocrit and reticulocyte count provide information as to the degree of hemolysis and anemia, and a complete blood count screens for the possibility of sepsis and the need for cultures. Examination of the blood smear is useful in differentiating common hemolytic disorders. Except for determinations of total and direct bilirubin, tests of liver function are not particularly helpful in establishing the cause of early-onset jaundice. Transient elevations of transaminases (AST and ALT) related to the trauma of delivery and to hypoxia have been noted. Biliary atresia and neonatal hepatitis can be accompanied by elevated levels of transaminase but characteristically present as chronic cholestatic jaundice with mixed hyperbilirubinemia after the first week of life.

Task 2.

A newborn was born at term, it was his mother's 1-st pregnancy. The jaundice was revealed on the 2-nd day of life, then it progressed. The adynamia, vomiting and hepatomegaly were presented. The indirect bilirubin level was 280 $\mu\text{mol/L}$, the direct bilirubin level - 6 $\mu\text{mol/L}$, Hb- 145 g/L. Mother's blood group - O(I), Rh+, child's blood group - A(II), Rh+.

1. Make a diagnosis.
2. What the pathogenesis of disease?
3. What the term of clinical manifestation appearing?
4. What the typical blood changes?
5. What the changes in mother organism are you waiting for?

Standard of answer . Task 2.

1. Hemolytic disease of newborn (ABO incompatibility), icteric type.
2. Hemolysis is primarily extravascular, although intravascular hemolysis that is not induced by complement also occurs.
3. Infants sometimes develop anemia, reticulocytosis, and hyperbilirubinemia within the first 24 hours of life.
4. The hallmark of ABO hemolytic disease is the presence of microspherocytes on the peripheral blood smear. In Rh hemolytic disease, on the other hand, microspherocytes are rarely noted. The direct antiglobulin test should be at least weakly positive for anti-A or anti-B; however, because of the sparse distribution of antigenic sites on a newborn's red cells, ABO hemolytic disease may be present even without a positive result on the direct antiglobulin test.
5. The maternal serum should have high titers of IgG directed against A or B. In the absence of clinical hemolytic disease, laboratory evidence of erythrocyte sensitization should not be considered isoimmune hemolytic disease.

Task 3.

You are speaking to a couple who are expecting their first baby in about 2 weeks. They are concerned about the safety of childhood immunizations and also about “unnecessary” medications given to newborns in the hospital. They ask about the purpose of the routine administration of intramuscular vitamin K. You explain to them about hemorrhagic disease of the newborn, stating that the untreated baby can manifest.

1. What the risk in newborn?
2. What the prophylactic steps of disease?
3. Describe clinical manifestations of it?
4. What the diagnostic?
5. Prescribe the treatment.

Standard of answer . Task 3.

1. A prolonged prothrombin time and a risk of serious hemorrhage in the days following delivery.
2. Prophylactically to newborn infants is associated with a decline in the levels of vitamin K-dependent coagulation factors.
3. In less than 1% of infants (but especially those fed human breast milk), the levels reached are low enough to produce classic hemorrhagic manifestations on the

second to seventh day of life. These manifestations include melena, hematuria, and bleeding from the circumcision; intracranial hemorrhage and hypovolemic shock are serious complications.

4. Diagnosis of this condition is indicated by a prolonged prothrombin time, which reflects inadequate concentrations of factors II, VII, IX, and X.

5. Replacement therapy with vitamin K.

Methodical materials to support basic stage class.

Professional algorithm of patient's management for practical skills and abilities forming.

№	Task	Sequence of implementation	Remarks and warnings related to self-control
1	To conduct examination of patient with haemorrhagic disease.	1.To conduct gathering of complaints and disease anamnesis. 2.To gather thoroughly the patient's life anamnesis. 3.To conduct patient's examination. 4.To investigate cardiovascular system of the patient (palpation, percussion).	Pay attention to features of disease course, underlying factors, concomitant diseases etc. To establish the availability of risk factors which facilitate disease occurrence. To assess patient's general condition, position in bed, color and humidity of skin and mucose, presence of petechias, bruises, haematomas on it, presence of neck veins and extremities swelling. To pay regard for rhythm of pulse, it's tension on both hands, apex shove, it's properties, margins of absolute and relative cardiac dullness, it's changes, HR(tachi-or bradycardia, extrasystole),BP, presence of bleedings from mocoses, nasal bleedings, melena, hemarthrones, hematomas, bronchial

			hemorrhage, and so on.
		<p>5.To conduct auscultation of the heart and the main vessels.</p> <p>6.To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the system of digestion.</p>	<p>To pay regard for heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To focus attention on features of percussion and auscultation of different age children.</p>
2	To formulate the preliminary diagnosis.	<p>1.To formulate the preliminary diagnosis</p> <p>2.To substantiate all components of preliminary diagnosis based on complaints,anamnesis, and examinations.</p>	To formulate preliminary diagnosis of haemorrhagic disease and substantiate each component of it, based on modern classification
3	To evaluate the parameters of additional laboratory tests.	<p>1.To evaluate the blood count data.</p> <p>2. To evaluate the biochemistry data.</p> <p>3.To evaluate the platelet count, bleeding time, prothrombin time, and activated partial thromboplastin time (APTT), tourniquet test, whole blood clotting time, prothrombin consumption time, and thromboplastin generation test.</p> <p>4.To evaluate the data of instrumental investigation.</p>	<p>To pay attention to platelet count, bleeding time, prothrombin time, and activated partial thromboplastin time (APTT), tourniquet test, whole blood clotting time, prothrombin consumption time, and thromboplastin generation test, cholesterol, lipids and glucose levels.</p> <p>To pay attention to US of internal organs, radiography of chest organs, joints.</p>
4.	To conduct differential diagnosis.	1.To find out common signs in complaints, life and disease anamnesis, the data of examination, the data of laboratory and instrumental tests in patients with similar status.	Special attention must be paid to differential diagnosis among the DIC-syndrome, hypo- and aplastic anaemias, leucosis, haemorrhagic vasculitis.

		<p>2.To find differences among complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumental methods in similar nosology.</p> <p>3. To find out the differences for excluding similar diseases from the list of probable diagnoses, being based on this algorithm.</p> <p>4. To conduct differential diagnostic among all of nosologies which have the similar signs, among other blood diseases, using this algorithm</p> <p>5.Taking into account the impossibility of excluding the diagnosis of haemorrhagic disease from the list of probable diagnoses to draw conclusion about the probability of such diagnosis.</p>	
6	To formulate the final clinical diagnosis.	<p>1. To formulate the final clinical diagnosis.</p> <p>2. Based on primary diagnosis, additional investigations datas, conducted differential diagnosis to substantiate all elements of concluding clinical diagnosis.</p>	Based on modern classification of haemorrhagic diseases to formulate diagnosis, complications of disease and concomitant diseases.
7	To prescribe treatment for patients.	<p>1.To prescribe non-medicinal treatment</p> <p>2.To prescribe medicinal treatment.</p>	<p>To specify regimen and detalized diet according to the disease.</p> <p>To prescribe modern medicinal treatment in accordance with the</p>

			standards of haemorrhagic diseases therapy, taking into account age, severity of patient state, stage of disease, presence of complications and concomitant pathology,
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Materials of control for conclusive classes stage:

Conclusive test control:

1. In newborn girl diagnosed the hemolytic disease by Rh-factor. The amount of bilirubin is critical. Blood type of child B(III), mother's is - A(II). Exchange blood transfusion is indicated. What selection of donor blood is needed for this purpose?
 - A. Blood type (III), rhesus factor positive
 - B. Blood (III) type, rhesus factor negative
 - C. Blood type (II), rhesus ifactor negative
 - D. Blood type (II), rhesus factor positive
 - E. Blood type O(I), rhesus factor negative

2. For the isoimmune conflict prophylaxis it is needed to administrate for mother an anti-D-Rh immunoglobulin, if following criteria are keeping:
 - A. In mother the Rh (+), antibodies are not present, in newborn Rh (+)
 - B. In mother Rh (+), antibodies are not present, in newborn Rh (-)
 - C. In mother Rh(-), antibodies are not present, in newborn Rh(+)
 - D. In mother Rh (-), antibodies are present, in new-born Rh (+)
 - E. In mother the Rh (-), antibodies are not present, in newborn Rh (-)

3. Hyperbilirubinemia with the rise of direct fraction of bilirubin is observed in newborn patients in following cases:
 - A. Halactosemia
 - B. Perinatal- TORCH- infections
 - C. Sepsis
 - D. Hepatitis
 - E. All above mensioned states are accompanied with the rise of direct fraction.

4. Diagnostics of hemolytic disease caused by Rh-conflict in newborn child with clinical manifestation, but without antibodies in a mother you: will prescribe during pregnancy?
- A. Direct Combs test
 - B. Indirect Combs test
 - C. Direct and indirect Combs tests
 - D. Level of bilirubin in an umbilical cord blood
 - E. Osmotic resistance of erythrocytes
5. In a new-born girl on a 6 day of life the level of bilirubin in blood is 285 $\mu\text{mol/l}$, direct fraction is 94 $\mu\text{mol/l}$. These parameters are reflected:
- A. Hemolysis as a result of glucose-6-phosphat dehydrogenase deficiency
 - B. CMV -infection
 - C. Viral hepatitis B
 - D. Syndrome of an intrahepatic cholestasis
 - E. Everything is correct except for a G-6-PD deficiency
6. Full-term newborn from 2-nd pregnancy was normal. Labors with partial placental detachment. In 14 hours after labors the melena is marked. What diagnostic method can reveal the reason of bleeding?
- A. Barium clister
 - B. The Apt test
 - C. Gastric lavage with solution of sodium chloride 0,9%
 - D. Count of thrombocytes
 - E. Count prothrombin time and partial thromboplastin time
7. Positive Apt test is testifying for presence in the liquid investigated:
- A. Haemoglobin F (blood of newborn)
 - B. Indirect bilirubin
 - C. Hemoglobin A (maternal blood)
 - D. Direct bilirubin
 - E. Meconium
8. A baby boy was born in time, it was his mother's 1st pregnancy. The jaundice was revealed on the 2-nd day of life, then it progressed. The adynamia, vomiting and

hepatomegaly were presented. The indirect bilirubin level was 270 $\mu\text{mol/L}$, the direct bilirubin level - 7 $\mu\text{mol/L}$, Hb- 155 g/L. Mother's blood group - 0(I)Rh⁺, child's blood group - A(II)Rh⁺. Make a diagnosis.

- A. Jaundice due to conjugation disorder
- B. Physiological jaundice
- C. Hemolytic disease of newborn (ABO incompatibility), icteric type
- D. Hemolytic disease of newborn (Rh - incompatibility)
- E. Hepatitis

9. A baby girl was born in time. The adynamia, vomiting and hepatomegaly were presented. The jaundice was revealed on the 2-nd day of life, when it progressed. The indirect bilirubin level was 305 $\mu\text{mol/L}$, the direct bilirubin level - 5 $\mu\text{mol/L}$, Hb- 160 g/L. Mother's blood group - 0(I)Rh⁺, child's blood group - A(II)Rh⁺. Make a diagnosis.

- A. Physiological jaundice
- B. Jaundice due to conjugation disorder
- C. Hemolytic disease of newborn (Rh - incompatibility)
- D. Hemolytic disease of newborn (ABO incompatibility), icteric type
- E. Hepatitis

10. A newborn boy was born in term. Apgar score 7-8 points. Mother's blood is AB(II)Rh(-). Child's is B(III)Rh(+). An icterus appeared in the first day. Common bilirubin is 204 $\mu\text{mol/l}$, indirect fraction is 196 $\mu\text{mol/l}$, direct is 10 $\mu\text{mol/l}$, Hb- 160 g/l, reticulocytes 4%. Liver +4cm. Urine is light, feces are painted. Coumbs test is positive. Establish the diagnosis:

- A. Physiologic jaundice
- B. Hemorrhagic illness of newborns
- C. Fetal hepatitis
- D. Crigler-Nayyar syndrome
- E. Hemolytic disease of newborns

Answers: 1-B, 2-C, 3-E, 4-C, 5-E, 6-B, 7-C, 8-C, 9-D, 10-E.

Materials of the medical support for students' self-preparation: a reference chart for organization of students' independent work with educational literature.

Tasks	Instructions
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To study etiology of haemorrhagic diseases in children.	To enumerate basic etiologic factors, to select the key links of haemorrhagic disease.
To study pathogenesis of haemorrhagic diseases in children.	To separate out the main links of haemorrhagic diseases' pathogenesis.
To study clinical manifestations of haemorrhagic diseases in children.	To establish the symptoms and to gather it into the clinical syndromes which enable to establish the probable diagnosis of haemorrhagic disease.
To study diagnostic criteria of haemorrhagic diseases	To make the flow diagram of disease
To study additional methods of research (laboratory, instrumental)	To work out a plan of patient's investigation.
To study changes in additional investigational methods which are pathognomonic for haemorrhagic diseases.	To enumerate the basic diagnostic criteria of haemorrhagic diseases according to the data of additional investigational methods.
To establish concluding diagnosis	To substantiate the basic components of diagnosis in accordance to modern classification, and to conduct a differential diagnosis.
To prescribe individual holiatry to patient with haemorrhagic disease. To be able to render the first aid in child with bleeding.	To prescribe specific regimen, diet, medicinal treatment, taking into account the age, severity of patient state, the stage of disease, the presence of complications and concomitant diseases.

THE RECOMMENDED LITERATURE

Basic:

1. Nelson textbook of pediatrics, 15th ed. / edited by Richard E. Behrman, Robert M. Kliegman, Ann M. Arvin; senior editor, Waldo E. Nelson, 1996, part XXII, chapter 441, 444-451.

2. Медицина дитинства / За ред. П.С. Мощича. - К.: Здоров'я, 1994.

3. Педиатрия : Учебник для студентов высших медицинских учебных заведений III – IV уровней аккредитации. – 3-е изд./ В.Г.Майданник;– Харьков: Фолио, 2006.-с.805-854.

Theme: Diabetes mellitus in children.

Study time: 4 hours

Classification of diabetes mellitus in children Etiology, pathogenesis, clinical presentation, diagnostics, differential diagnostics, treatment, prophylaxis, prognosis. Insulin therapy. Hypoglycemic, hyperglycemic coma and ketoacidosis: etiology, pathogenesis, classification, clinical presentation, diagnostics, differential diagnostics. Emergency and prophylaxis in comas. Prognosis.

I. Actuality of the theme.

Diabetes mellitus is a group of metabolic disorders of carbohydrate metabolism in which glucose is underutilized, producing hyperglycemia. Each year, approximately 600,000 people are diagnosed with diabetes. The disease is classified into several categories. Type 1 diabetes mellitus, formerly known as insulin-dependent diabetes mellitus or juvenile-onset diabetes mellitus, is caused by autoimmune destruction of the b-cells of the pancreas, rendering the pancreas unable to synthesize and secrete insulin. Type 2 diabetes mellitus, formerly known as non-insulin-dependent diabetes mellitus or adult-onset diabetes, results from a combination of insulin resistance and inadequate insulin secretion. Other types of diabetes are rare. Type 2 is the most common form, accounting for 90–95% of diabetes in developed countries.

In 1992, the costs of diabetes in the US were estimated to be \$98 billion. The mean annual per capita healthcare costs for an individual with diabetes are approximately fourfold higher than those for individuals who do not have diabetes. Similarly, in the United Kingdom, diabetes accounts for roughly 10% of the National Health Service budget (49 billion).

The high costs of diabetes are attributable to care for both acute conditions (such as hypoglycemia and ketoacidosis) and debilitating complications. The latter include both microvascular complications—predominantly retinopathy, nephropathy, and neuropathy; and macrovascular complications, particularly stroke and coronary artery disease. Together these make diabetes the seventh most common cause of death in the developed world.

Concrete purposes:

1. To determine the etiological and pathogenic factors in diabetes mellitus.
2. To classify and analyze the typical clinical manifestation of diabetes mellitus.
3. To make a plan of investigation and analyze the information about laboratory and instrumental data in the classic course of diabetes mellitus in children.
4. To demonstrate skills of treatment, rehabilitation and prophylaxis of diabetes mellitus in children.

5. To diagnose and render urgent help in hyperglycemic, hypoglycemic, hyperosmolar , hyperlactacidemic and ketoacidosis coma patients.

6. To conduct differential diagnostics of diabetes mellitus in child, different kinds of comas and make a preliminary diagnosis.

7. To determine the prognosis for life in diabetes mellitus.

8. To demonstrate both the skills of medical specialists moral and deontological principles and the principles of professional subordination in pediatrics.

II. Classes (pointing of planned mastering level)

1. A student must know (to familiarize): $\alpha 1$

-about the place of diabetes mellitus in the structure of the endocrine system, and diseases in children which are widespread in different age-dependent and ethnic groups;

- about statistical information in relation to morbidity, frequencies of complications, lethality, and the nearest and remote prognosis in patients with diabetes mellitus;

-about the history of scientific study and payment of domestic scientists;

2. A student must know (master): $\alpha 2$

-the etiology of diabetes mellitus type 1 in children

-key links of diabetes mellitus pathogenesis type 1 ;

-clinical classification of diabetes mellitus type 1;

-the classic clinical manifestation of diabetes mellitus type 1;

-laboratory and instrumental diagnosis of diabetes mellitus type 1;

-the long-term complications of diabetes mellitus type 1;

-the acute complications of diabetes mellitus type 1;

-the treatment principles of diabetes mellitus type 1 in children and long-term complications of diabetes mellitus type 1.

3. A student must seize the skills of: $\alpha 3$

-collection of complaints and anamnesis of disease;

-examination of patients with diabetes mellitus and revealing the main symptoms and syndromes.

-formulating and substantiating the preliminary diagnosis;

-determining a laboratory and instrumental examination plan of patient's investigation (with obedience of diagnostics standards);

By the abilities:

- interpreting the results of laboratory and instrumental investigations;

- conducting a differential diagnosis among diabetes insipidus, kidney glucosuria, short-lived glucosuria and hyperglycemia ;

- conducting a differential diagnosis among different kind of comas;

- giving recommendations in relation to the patient regimen and diet with the diseases of diabetes mellitus - taking into account the stage of disease, severity of state and concomitant pathology;

- completing the treatment plan for diabetes mellitus according to the standards taking into account the stage of disease, complications and concomitant pathology.

- rendering first aid in extreme situations such as hyperglycemia, hypoglycemia, hyperosmolar , hyperlactacidemic, and ketoacidosis coma.

III. Aims of personality development (educative aims):

-A student must learn to adhere to the rules of behaviour and principles of medical etiquette and deontology near a bed ridden patient with diabetes mellitus;

-to try hand on ability to set a psychological contact with a patient and his family;

-to master a sense of professional responsibility for timely, adequate and skilled medicare.

IV. Interdisciplinary integration:

Subject	To know	Be able
1. Previous (providing)		
Anatomy	Structure of human endocrine system, of the pancreas , its circulation	
Histology	Structure of island of Langerhans	
Physiology	Normal physiology of the pancreas, normative indices of laboratory and instrumental investigation methods and their assesment.	To asses laboratory data and instrumental investigation methods.
Pathologic physiology	Key links of the pathogenesis of diabetes mellitus type 1	To asses laboratory data and instrumental investigation methods.
Pathologic anatomy	Morphological features of the pancreas, blood vessels, kidneys, organs of sight, and the nervous system - (depending on disease stage.)	To analyze and interpret the information of a clinical examination and additional methods of investigation
Pharmacology	Pharmacokinetics and pharmacodynamics; the side effects of prescriptions (short-acting , intermediate-acting , and long-acting insulin), and angiotensins (type 1	To prescribe: age dependent and individual patient characteristics treatment to identify the stage of disease and establish an individual prescription to take with the

	blockers,etc.)	correct dosage. To be able to make a prescription.
Propedeutical pediatrics.	The basic stages and methods for the clinical examination of patients.	To collect complaints and anamnesis vitae et morbid - to find out the basic risk factors of diabetes mellitus; to be able to conduct a patient examination to reveal the clinical signs of thyroid gland diseases; <i>and</i> to be able to interpret data for additional methods of investigation.
2.Followings(provided)		
Hospital pediatrics.	Clinical signs of diabetes mellitus and its complications and treatment tactics.	To reveal the clinical signs of diabetes mellitus and complications and be able to prescribe treatment.
3. Interdiscipline integration		
Diabetes insipidus in children	The clinical manifestation of diabetes insipidus	To identify specific clinical signs of diabetes insipidus and conduct a differential diagnosis for diabetes mellitus in children.
Renal glucosuria	The clinical manifestation of renal glucosuria	To identify specific clinical signs of glucosuria and conduct a differential diagnosis for diabetes mellitus in children.
Transient glucosuria and hyperglycemia	The clinical manifestation of transient glucosuria and hyperglycemia	To identify specific clinical signs of transient glucosuria and hyperglycemia and conduct a differential diagnosis for diabetes mellitus in children.
Renal glucosuria	The clinical manifestation of Renal glucosuria	To identify the specific clinical signs of renal glucosuria and hyperglycemia and conduct a differential diagnosis for diabetes mellitus in children.

V. Contents of the theme

Drug Name	Insulin lispro (Humalog)
Description	Onset of action is 10-30 min, peak activity is 1-2 h, and duration of action is 2-4 h.
Adult Dose	0.5-1 U/kg/d SC initially; adjust doses to achieve premeal and bedtime blood glucose levels of 80-140 mg/dL (4-7.5 mMol/L)
Pediatric Dose	0.5-1 U/kg/d SC initially Adjust doses to achieve premeal and bedtime blood glucose levels of: <5 years: 100-200 mg/dL (5.5-10 mMol/L) >5 years: 80-140 mg/dL (4-7.5 mMol/L)
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine isoniazid, diltiazem, diuretics, corticosteroids, thiazide diuretics, thyroid estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin; medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors, alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAOIs, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide, pentamidine, and sulfinpyrazone
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Due to prompt onset of action, administer within 15 min before or immediately after a meal; monitor glucose carefully; dose adjustments may be necessary in renal and hepatic dysfunction

Drug Name	Regular insulin (Humulin R, Novolin R)
Description	Onset of action is 0.25-1 h, peak activity is 1.5-4 h, and duration of action is 5-9 h.
Adult Dose	Adjust to needs
Pediatric Dose	Adjust to needs
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine isoniazid, diltiazem, diuretics, corticosteroids, thiazide diuretics, thyroid estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin; medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors,

	alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAOIs, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide, pentamidine, and sulfinpyrazone
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Dose adjustments may be necessary in renal and hepatic dysfunction

Drug Name	Insulin NPH (Humulin N, Novolin N)
Description	Onset of action is 3-4 h, peak effect is in 8-14 h, and usual duration of action is 16-24 h.
Adult Dose	Adjust to needs
Pediatric Dose	Adjust to needs
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine isoniazid, diltiazem, diuretics, corticosteroids, thiazide diuretics, thyroid estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin; medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors, alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAOIs, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide, pentamidine, and sulfinpyrazone
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Dose adjustments may be necessary in renal and hepatic dysfunction

Drug Name	Protamine zinc (Ultralente)
Description	Onset of action is 2-3 h, peak activity is 4-8 h, and duration of action is 8-16 h.
Adult Dose	Adjust to needs
Pediatric Dose	Adjust to needs
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine isoniazid, diltiazem, diuretics, corticosteroids, thiazide

	diuretics, thyroid estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin; medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors, alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAOIs, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide, pentamidine, and sulfapyrazone
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Dose adjustments may be necessary in renal and hepatic dysfunction

Drug Name	Insulin aspart (NovoLog)
Description	Onset of action is 10-30 min, peak activity is 1-2 h, and duration of action is 3-6 h. Homologous with regular human insulin, with the exception of single substitution of amino acid proline by aspartic acid in position B28. Produced by recombinant DNA technology. Insulin lowers blood glucose levels by stimulating peripheral glucose uptake, especially by skeletal muscle and fat, and by inhibiting hepatic glucose production. Inhibits lipolysis in the adipocyte. Inhibits proteolysis. Enhances protein synthesis. Insulin is the principal hormone required for proper glucose use in normal metabolic processes.
Adult Dose	0.5-1 U/kg/d SC initially; adjust doses to achieve premeal and bedtime blood glucose levels of 80-140 mg/dL (4-7.5 mMol/L)
Pediatric Dose	0.5-1 U/kg/d SC initially Adjust doses to achieve premeal and bedtime blood glucose levels of: <5 years: 100-200 mg/dL (5.5-10 mMol/L)>5 years: 80-140 mg/dL (4-7.5 mMol/L)
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine, isoniazid, diltiazem, diuretics, corticosteroids, thiazide diuretics, thyroid hormone, estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine, phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin Medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors, alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAO inhibitors, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide,

	pentamidine, and sulfinpyrazone
Pregnancy	B - Usually safe but benefits must outweigh the risks.
Precautions	Hyperthyroidism may increase renal clearance of insulin and may need more insulin to treat hyperkalemia; hypothyroidism may delay insulin turnover, requiring less insulin to treat hyperkalemia; due to prompt onset of action, administer within 15 min before or immediately after a meal; monitor glucose carefully; dose adjustments may be necessary in renal and hepatic dysfunction

Drug Name	Insulin glargine (Lantus)
Description	Long-acting insulin analogue. Typical onset of action from 1-2 h, duration 20-26 h
Adult Dose	Usually 50% of total daily dose of insulin (0.25-0.5 U/kg); adjust to needs
Pediatric Dose	Licensed age varies between nations (2-6 y); adjust dose as indicated but similar to adult
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Medications that may decrease hypoglycemic effects of insulin include acetazolamide, AIDS antivirals, asparaginase, phenytoin, nicotine, isoniazid, diltiazem, diuretics, corticosteroids, thiazide diuretics, thyroid hormone, estrogens, ethacrynic acid, calcitonin, oral contraceptives, diazoxide, dobutamine, phenothiazines, cyclophosphamide, dextrothyroxine, lithium carbonate, epinephrine, morphine sulfate, and niacin Medications that may increase hypoglycemic effects of insulin include calcium, ACE inhibitors, alcohol, tetracyclines, beta blockers, lithium carbonate, anabolic steroids, pyridoxine, salicylates, MAO inhibitors, mebendazole, sulfonamides, phenylbutazone, chloroquine, clofibrate, fenfluramine, guanethidine, octreotide, pentamidine, and sulfinpyrazone
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Administer at the same time each day; use only if solution is clear and colorless; administer SC only; do not mix with any other insulin or solution; hyperthyroidism may increase renal clearance of insulin and may need more insulin to treat hyperkalemia; hypothyroidism may delay insulin turnover, requiring less insulin; monitor glucose carefully; dose adjustments of insulin may be necessary in patients diagnosed with renal and hepatic dysfunction

Drug Name	Insulin glulisine (Apidra)
Description	<p>Human insulin analog produced by rDNA technology using a nonpathogenic laboratory strain of E coli (K12). Differs from human insulin by replacement of asparagine at B3 position with lysine, and the lysine at the B29 position is replaced by glutamic acid.</p> <p>Insulin regulates glucose metabolism by stimulating peripheral glucose uptake by skeletal muscle and fat, and inhibits hepatic glucose production. Glucose lowering is equipotent to regular human insulin when administered IV. After SC administration, insulin glulisine has more rapid onset and shorter duration of action compared to regular human insulin. Useful to regulate mealtime blood glucose elevation.</p>
Adult Dose	Individualize dose; intended for intermittent SC injection with meals or use by external infusion pump
Pediatric Dose	<p>0.5-1 U/kg/d SC initially;</p> <p>Adjust doses to achieve premeal and bedtime blood glucose levels of:</p> <p><5 years: 100-200 mg/dL (5.5-10 mMol/L)</p> <p>>5 years: 80-140 mg/dL (4-7.5 mMol/L)</p>
Contraindications	Documented hypersensitivity; hypoglycemia
Interactions	Corticosteroids, danazol, diazoxide, diuretics, sympathomimetic agents (eg, epinephrine, albuterol, terbutaline), glucagon, isoniazid, phenothiazines, growth hormone, thyroid hormone, estrogen, progestogens, protease inhibitors, and atypical antipsychotics (eg, olanzapine, clozapine) may increase blood glucose and reduce glucose lowering effect of insulin; oral antidiabetic agents, ACE inhibitors, disopyramide, fibrates, fluoxetine, MAOIs, pentoxifylline, propoxyphene, salicylates, and sulfonamides may decrease blood glucose and cause additive effects to insulin
Pregnancy	C - Safety for use during pregnancy has not been established.
Precautions	Hyperthyroidism may increase renal clearance of insulin and may need more insulin to treat hyperkalemia; hypothyroidism may delay insulin turnover, requiring less insulin to treat hyperkalemia; due to prompt onset of action, administer within 15 min before or immediately after a meal; monitor glucose carefully; dose adjustments may be necessary in renal and hepatic dysfunction

VI. Plan and organizational structure of classes.

№ п/п	Basic stages of classes, their function and maintenance	Educational aims are in the levels of mastering	Methods of control and studies	Educational materials	Distribution of time in minutes
1	Preparatory stage	$\alpha 2$			3 min.
2	Organizational measures of educational aims and motivation	$\alpha 2$		II. II «Educational aims»	12 min.
3	Control of basic knowledge and skill levels:		Individual questioning	II. I «Actuality of theme»	20 min.
	1. Keylinks of pathogenesis of diabetes mellitus type 1 in children	$\alpha 2$	Test control of the second level	Second level tests the table «Pathogenesis of diabetes mellitus type 1 »	
	2. Classification of diabetes mellitus type 1	$\alpha 2$	Individual (oral) questioning	Tests of 2 level	
	3. Classification of complications of diabetes mellitus type 1: long-term and acute	$\alpha 2$	Typical situational task of 2 level	the table «Classification of diabetes mellitus type 1 »	
	4. Laboratory and instrumental diagnosis of diabetes mellitus and complications	$\alpha 2$	Typical situational task of 2 level	Structurally logical chart: long-term and acute complications	
	5 Treatment principles of diabetes mellitus and its complications	$\alpha 2$	Typical situational task of 2 level	Typical situational task of 2 level	
	6. Complications of insulin-therapy	$\alpha 2$	Test control of 2 level	Tests of 2 level	
		$\alpha 2$	Typical situational task of 2 level	Typical situational tasks of 2 level Kit of medicines.	
4	Basic stages of professional skills and abilities forming:				
	1. To conduct the patient's management with diabetes mellitus, to take complaints and anamnesis.	$\alpha 3$	Practical professional training	Patient	115 min.
	2. To conduct the patient's examination and detect the main symptoms and complications with diabetes mellitus.	$\alpha 3$	Practical professional training	Patient	
	3. To formulate and substantiate the preliminary diagnosis	$\alpha 3$	Practical professional training	Case history	
			Practical professional training	A reference chart for the forming of professional abilities. Case	

	4.To compose the plan of patient's laboratory and instrumental investigation.	$\alpha 3$	training	history.	
	5. To interpret the results of laboratory and instrumental investigation.	$\alpha 3$	Practical professional training. Tests and the third level control. The third level test control.	A reference chart for the forming of professional abilities. Situational typical tasks of the third level.The third level tests.	
	6.To conduct differential diagnosis for clinical conditions accompanied by hyperglycemia, polydipsia, glycosuria, polyuria.	$\alpha 3$	The practical professional training is in the solution of non standard clinical situations.	Prescribing chart	
	7.To give recommendations for the regimen and diet of a patient.	$\alpha 3$	The third level test control.Practical professional training. The third level test control.	The third level non typical situational tasks. Treatment algorithm for the diabetes mellitus patients.	
	8.To compose the treatment plan of diabetes mellitus patient's treatment taking into account the stage of disease and the presence of complications.	$\alpha 3$	Practical professional training .	The third level non typical situational tasks. First aid algorithm in diabetes mellitus.	
	9. To be able to render the first aid in extreme situations: hyperglycemic, hypoglycemic, hyperosmolar , hyperlactacidemic, ketoacidosis coma.	$\alpha 3$	The practical professional training on solving of non typical clinical situations.		
5	Concluding stage.		The third level test control.		
6	Control and correction of professional abilities and skills.		Practical professional training on solving of non typical clinical situations.		
7	Working out the totals of class. Home work (basic and additional literature on the topic)		Analysis of clinical work Solution of non typical tasks and the third level tests. Estimation of clinical work.	Clinical work The third level non typical situational tasks. A reference chart for independent work with literature	30 min.

Methodical materials for the class basic stage supporting

The questions for the control of primary knowledge level of abilities and skills:

1. What is the role of genetics in the development of diabetes mellitus type 1?
2. What is the main mechanism in the development of glycosuria in children with diabetes mellitus, type 1 ?
3. What leads to the occurrence of hyperglycemia in children with diabetes mellitus, type 1?

4. How can you estimate the level of glucose with the help of an oral glucose tolerance test (OGTT)?

5. What is the role of glycosylated hemoglobin derivatives (HbA1a, HbA1b, HbA1c) for the diagnosis of diabetes and its complications?

6. What is the role of hyperglycemia in the development of long-term complications (retinopathy, cataracts ,hypertension, progressive renal failure ,early coronary artery disease,peripheral vascular disease ,Neuropathy, both peripheral and autonomic)?

7. Describe the pathogenesis of microalbuminuria in children with diabetes mellitus, type 1.

GFR – glomerular filtration rate

RP – renal perfusion

8. To render the first aid in the case of diabetic ketoacidosis.

9. How many various insulins and mixtures are available for children with diabetes mellitus?

10. Dietary management of diabetes care.

11. To render the first aid in the case of hypoglycemic coma.

12. To conduct a differential diagnosis for hyperglycemic, hypoglycemic, hyperosmolar , hyperlactacidemic, and ketoacidosis coma patients .

Primary tests

1. A boy of 7 years old, was hospitalized with complaints of thirst intensifying and poliuria . During 5 years he has been suffering from diabetes, in anamnesis a diabetic comma three times developed. The level of sugar in the blood is 15,54 mmol/l, in urine it is 5%. By the oculist Retinoangipathy was found. What dose of insulin will be administered to the child?

A. 1,5 U/kg

B.0,25 U/rg

C.0,5 U/kg

D.1,0 U/kg

E.2,0 U/kg

2. A diabetic boy was delivered to the induction centre of child's hospital in severe condition. On examination there was the absence of consciousness . His skin was dry and pale, skin turgor was diminished. Hypotonus of muscles and eyeballs. Lips mucus was dry and in bright red color. Heart tones were hyposthenic, tachycardia, decreased blood pressure, threadlike pulse. Koussmaul type of breathing . Strong smell of acetone midair. Specify the day's dose of insulin for the acute management.

A. 1,5 U/kg

B. 2,5 U/kg

C.2,0 U/kg

D.1,0 U/kg

E.3,0 U/kg

3. Patient O, 13 years old, has been suffering from diabetes for 6 years. Hospitalized to the department in comma. Consciousness was absent, reflexes were low-speed.

Skin and mucous membranes were pale, dry. Breathing was loud. The smell of acetone. Anuria. Laboratory data: blood glucose –is 35 mmol / l, serum potassium is 2,5 mmol / l, sodium is 120 mmol/ l. Blood urea is 9,5 mmol/ l, ketonic bodies in the serum were 7,5 mmol /l. In the urine is +++++, glucose of urine –is 120 mmol /l. What is it necessary for the taking the patient out of coma?

- A. to conduct blood transfusion
- B. to conduct neuroleptanalgesia
- C. to conduct dehydration
- D. to carry out the correction of acid - alkaline equilibrium
- E. to administer hydrocortisone

4. A child of 5 years old was first hospitalized in the department with a diagnosis of I type diabetes, decompensation (ketoacidosis). What is the main mechanism in the development of the disease?

- A. Surplus of glucagon
- B. Insulin insufficiency.
- C. Surplus of somatostatin.
- D. Disturbance of insulin complexing with receptors.
- E. Disturbance of postreceptor mechanism of insulin action.

5. A child of 10 years old was hospitalized with complaints of thirst, intensifying emiction, weight loss, weakness, languor after the clinical and –laboratory examination. The diagnosis of I type diabetes was made, decompensation. How is it possible to explain the symptom of polyuria in the patient ?

- A. Diminished production of thyroid hormone.
- B. Diminished production of antidiuretic hormone.
- C. Increased vasopressin production.
- D. Glucose is selected with urine acts as osmotic diuretic.
- E. Elevated thyroid hormone production.

6. A girl of 12 years old. Since 2-years-old age she has been suffering from diabetes. On examination there was considerably enlarged stomach, liver palpated 6 cm below costal arc, somewhat painful and, dense; Cushing –like type of obesity, nanism. The signs of the sexual maturation are not present. Blood glucose is 17 mmol/l, of urine is 4 mmol/l. In the blood elevated cholesterol, ketonic bodies. What is the preliminary diagnosis?

- A. I type Diabetes, precoma
- B. I type diabetes, Nodding syndrome
- C. I type diabetes, Mauriac syndrome
- D. Cushing disease illness
- E. Cushing syndrome

7. A girl of 12 years old, was delivered to the hospital in the unconscious state. During the last month she has been losing weight, although the appetite is preserved. Stomach-aches, vomiting, appeared. Weakness grew, became put on the brakes. On examination: without consciousness, sharply exhausted, the skin is pale-grey, dry,

hyperemia of cheeks. Breathing is deep, noisy. The smell of acetone. Heart tones are muffled and rhythmic. Pulse 90 b. per minute, blood pressure is 90/50 mmHg. Stomach is falling back. Liver +3,0 cm. What disease can be supposed?

- A. Acetonemic condition.
- B. Coliform infection with neurotoxicosis
- C. Hepatic coma
- D. Diabetic coma
- E. Suprarenal failure

8. A child of 7 years old was hospitalized with complaints on weight loss, thirst, frequent emiction. During the examination in the blood tests the level of sugar on an empty stomach was 14 mmol/l, the level of sugar in urine was 5 g/l. What caused hyperglycemia in the child?

- A. Intensifying of lipolysis .
- B. Intensifying of proteins catabolism.
- C. Decreasing of gluconeogenesis.
- D. Intensifying of glycolysis
- E. Decreasing of glycolysis and intensifying of gluconeogenesis.

9. A child of 7 years old was hospitalized with complaints on thirst, frequent emiction, weight loss (3 kg per month). On examination the level of glucose in the blood on an empty stomach was 19,2 mmol/l. A diagnosis of 1 type diabetes was made. How is it possible to explain the weight loss in the child? .

- A. Intensifying of proteins catabolism and inhibiting of its synthesis.
- B. Decreasing of lipolysis..
- C. Intensifying of gluconeogenesis.
- D. Decreasing of lipolysis. .
- E. Disturbances of basic exchange.

10. A boy of 10 years old, sharp languor, somnolence, thirst, frequent emiction. On examination the smell of acetone from the mouth was marked, on the cheeks bright blush, noisy unsteady breathing, enlarged liver. In the blood tests the level of sugar was 20,5 mmol/l, sugar of urine is 20 g/l, acetone in urine is +++. How is it possible to explain the appearance of acetone in the air and urine? .

- A. By the increased disintegration of ketogenic amino acids and lipids.
- B. By the disturbance of water - electrolyte balance.
- C. By the disturbance of acid - alkaline balance.
- D. By the disturbance of glucose phosphorylating processes.
- E. By the decreasing of glycolysis.

11. A patient of 8 years old, was taken to the hospital without consciousness. A week ago she had a flu, whereupon a girl became sickly, ate badly, mainly milk, tea, grumbled about headache (mother reported). Stomach-aches appeared the day before, repeated vomiting, the girl was sleepy, and lost consciousness to the evening. Consciousness was absent. The skin was dry. The tongue was dry, red. Breathing was deep, noisy, 32/ min., the smell of acetone. Pulse 128 / min., small. Blood pressure -

75/40 mmHg. On palpation the stomach was soft, without pains. Liver + 4 cm, is soft. Body temperature is 36,7 C. Kehr and other symptoms are negative. What is the preliminary diagnosis?

- A. Acute adrenal insufficiency.
- B. Meningitis.
- C. Septic shock.
- D. 1 type diabetes is revealed for the first time, ketoacidic comma.
- E. Hepatitis.

12. Patient K., has been suffering from diabetes for 8 years. Objectively: skin is dry, breathing is noisy, the smell of acetone. What type of comma is it possible to suspect?

- A. ketoacidic
- B. hyperosmotic
- D. lactatacidic
- E. cerebral

13. A boy of 14 years old, complains on thirst, loss of weight, weakness, increased emiction (4 l per day), pain in the legs, itch of the skin. What diagnosis is the most probable?

- A neurogenic polydesum
- B. diabetes
- C. diabetes insipidus
- D. acute nephrite
- E. kidney diabetes

14. A boy of 14 years old, complains on thirst, loss of weight, weakness, increased emiction (4 l per day), pain in the legs, itch of the skin. What is it necessary to prescribe for clarification of the diagnosis?

- A test with xerophagia
- B to determine the level of sugar in the blood
- C. roentgenography of skull
- D. Zimnitski test
- E. to determine the protein loss with urine per day

15. Patient K., was delivered without consciousness. He has been suffering from diabetes for 10 years. During the last week he had gastroenteritis. Objectively: the skin is dry, eyeballs are soft. Breathing is superficial; the smell of acetone is not felt. Blood. pressure is 80/40 mm.Hg What kind of coma is the most probable?

- A hyperosmotic
- B. hyperlactatacidemic
- C. ketoacidic
- D. glucopenia
- E. alcoholic

16. Patient K., was delivered without consciousness. He has been suffering from diabetes during 10 years. During the last week he had gastroenteritis. Objectively: the

skin is dry, eyeballs are soft. Breathing is superficial; the smell of acetone is not felt. Blood pressure is 80/40 mm.Hg What do you expect to get from the results of analyses?

- A.. glycemia is more than 20 mmol/l, acetone in urine is «+++»
- B glycemia is 2,0 mmol/l, acetone in urine is absent
- C. glycemia is than more than 40 mmol/l, acetone in urine is absent
- D. glycemia is up to 10 mmol/l, pH of blood is less than 7,2
- E. glycemia is 5,0 mmol/l, the high level of alcohol in the blood

17. Patient K., 8 years old. He has been suffering from diabetes for 6 years, got insulin 46 U/day. During the last 5 months for compensation of glycemia the dose of insulin was multiplied to 108 U. Glucopenia was not present.

How do you consider such a state?

- A. syndrome of chronic overdose of insulin
- B . resistance to insulin
- C. lability of diabetes course
- D. Kimmelstill –Wilson syndrome
- E. allergy to insulin

18. Patient K., 8 years old. He has been suffering from diabetes for 6 years, got insulin 46 U/day. During the last 5 months for compensation of glycemia the dose of insulin was multiplied to 108 U. Glucopenia was not present.

What is the further tactics of treatment for this patient?

- A, to multiply the dose of insulin
- B. to cancell insulin and to prescribe biguanids.
- C. hyposensibilisation by the small doses of insulin
- D. to increase the amount of carbonhydrates in the diet
- E. to prescribe simple insulin + prednisolon

19. Patient M, without consciousness. The skin is moistured, on the shoulders and thighs there are signs of injections. Breathing is superficial. Blood pressure is 110/170 mmHg. Muscles tones and tendon reflexes are increasded,there are cramps in the extremities. What disease is it possible to think about?

- A.Hyperglycemic comma
- B Glycopenic comma
- C. Hyperosmotic comma
- D. Hyperlactatacid comma
- E. cerebral comma (stroke)

20. Patient K., has been suffering from diabetes for 8 years. During the last year the dose of insulin was diminished to 14 U. In the urine analysis of protein is 1,7 %, sugar 0,8 %, a lot of red corpuscles and cylinders.

The indicated signs are the manifestation of:

- A.resistance to insulin
- B. nephrosclerosis
- C. decompensations of diabetes

D. pyelitis

E. syndrome of chronic overdose of insulin

Answers : 1-A, 2-D, 3-D, 4-B, 5-D, 6-B, C, 7-D, 8-D, 9-A, 10-A, 11-D, 12-A, 13-B, 14-B, 15-A,16-C,17-B,E, 18-E,C, 19-C,20-B

Diabetes mellitus in children.

Typical situational tasks of 2 level

1. A boy of 14 years old, from the 12-years-old age has been suffering from diabetes. In the anamnesis hypoglycemic diabetic coma developed, on the eyeground the microaneurysm of vessels, hemorrhages in the retina, pain in the legs, decrease of amplitude on a rheography, level of glucose in the blood is 13,1 mmol/l.

Task Taking modern classification as a basis formulate the diagnosis

Diabetes mellitus in children.

Typical situational tasks of 2 level

2. A boy of 7 years old, was hospitalized with complaints of rashes, weakness, weight loss, he had flu 6 months ago. The level of glucose in the blood is 6,6 mmol/l, oral glucose tolerance test- fasting blood sugar level is 6,16 mmol/l, blood glucose concentration again after 30 min(after glucose) is 7,7 mmol/l, blood glucose concentration again after 60 min is 12,1 mmol/l, blood glucose concentration again after 90 min is 10,54 mmol/l, blood glucose concentration again after 120 min is 11,7 mmol/l.

Task

- To evaluate the oral glucose tolerance test (OGTT)
- What is the initial diagnosis?
- To prescribe treatment
-

Diabetes mellitus in children.

Typical situational tasks of 2 level

3. A girl of 11 years old, from the 9-years-old age has been suffering from diabetes, got insulin 18 U/day. She had a pneumonia 2 weeks ago. The girl was hospitalized with complaints of thirst, stomach-ache, vomiting, loss of consciousness. The smell of acetone. Pulse 120 b. per minute, Blood pressure is 80/45 mmHg . Breathing is deep, noisy.

Task

- What is the preliminary diagnosis?
- To work out a plan of patient investigation.
- To Prescribe treatment

Diabetes mellitus in children.

Typical situational tasks of 2 level

4. A girl of 6 years old from 1-year-old age has been suffering from diabetes, was delivered to the hospital in the unconscious state. On examination the skin is pale-grey, lips mucus is dry and in a bright red color, constriction of pupils, heart tones are hyposthenic, tachycardia, decreased blood pressure, the smell of acetone. Blood glucose is 22,4 mmol/l.

Task

- What is the preliminary diagnosis?
- To render the first aid
- What kind of insulin will most probably be prescribed to the child?

Diabetes mellitus in children.

Typical situational tasks of 2 level

5. A boy of 16 years old. Since 2-years-old age has been suffering from diabetes, got alcohol, hospitalized with complaints of weakness, consciousness,

On examination : the skin is pale and cyanotic , consciousness, sweating.

Breathing is superficial , the smell of alcohol, tachycardia, cramps.

Task

- What is the preliminary diagnosis?
- To render the first aid

Diabetes mellitus in children.

Typical situational tasks of 2 level

6. A patient 8 years old, was taken to the hospital . A week ago had flu. Complains on thirst, loss of weight, weakness, polyuria. On examination: vomiting, the skin is dry, hyperemia of the cheeks. Breathing is deep, noisy. Smell of acetone. Heart tones are muffled and rhythmic. Stomach is falling back.

Task

- What is the preliminary diagnosis?
- To work out a plan of patient investigation.
- To prescribe treatment

Diabetes mellitus in children.

Typical situational tasks of 2 level

7. A patient K., 8 years old. Has been suffering from diabetes during 6 years. On examination his stomach was considerably enlarged, liver palpated 6 cm below the costal arc, somewhat painful, dense; Cushing-like type of obesity, nanism. The signs of the sexual maturation are not present. Blood glucose is 17 mmol/l, urine is 4 mmol/l. In the blood increased cholesterol, ketonic bodies.

Task

- What is the preliminary diagnosis?
- To prescribe treatment

Diabetes mellitus in children.

Typical situational tasks of 2 level

8. A patient M, 12 years old, has been suffering from diabetes during 6 years. Takes insulin of «Aktrapid» 13 U + «Protophan» 14 U. In anamnesis the comatose states relapsed. She is disturbed by the decreasing of sight acuity, weakness and pain in the legs. At ophthalmoscopy on the eyeground the microaneurysm of vessels, hemorrhages in the retina, neovascularisation. Glucose on an empty stomach is 9,7 mmol/l, glucosuria is 2 % (2,5 l), acetone in urine was not discovered.

Task

- What is the preliminary diagnosis?
- What correction of diabetes therapy will you conduct?

Diabetes mellitus in children.

Typical situational tasks of 2 level

9. A child of 10 years old. Has been suffering from diabetes for 10 years. During the last 2 years she had albuminuria, high blood pressure. During the last week had gastroenteritis. Objectively: cramps, the skin is dry, hallucination, eyeballs are soft. Breathing is superficial; the smell of acetone is not felt. Blood pressure is 80/40 mm.Hg. On palpation the stomach is soft, without pains. Hypertonus of muscles.

Task

- What is the preliminary diagnosis?
- What are the laboratory findings
- Prescribe treatment

Diabetes mellitus in children.

Typical situational tasks of 2 level

10. A girl of 12 years old, was delivered to the hospital. During the last month she was losing weight, although the appetite preserved. Stomach-aches, vomiting, appeared. The weakness grew, became put on the brakes. On examination: drowsy, sharply exhausted, the skin pale-grey, dry, hyperemia of cheeks. Breathing is deep, noisy. The smell of acetone. Heart tones are muffled and rhythmic. Pulse 90 b. per minute, Blood pressure is 90/50 mmHg Stomach is falling back. Liver +3,0 cm.

Task

- What disease can be supposed first of all?
- To render the first aid

Diabetes mellitus in children.

Typical situational tasks of 2 level

11. A patient K., 8 years old. Has been suffering from diabetes during 6 years. On examination his stomach was considerably enlarged, liver palpated 6 cm below the costal arc, somewhat painful, dense; Cushing-like type of obesity, nanism. The signs of the sexual maturation are not present. Blood glucose is 17 mmol/l, urine is 4 mmol/l. In the blood increased cholesterol, ketonic bodies.

Task

- What is the preliminary diagnosis?
- To prescribe treatment

Diabetes mellitus in children.

Typical situational tasks of 2 level

12. A patient M, 12 years old, has been suffering from diabetes during 6 years. Takes insulin of «Aktrapid» 13 U + «Protophan» 14 U. In anamnesis the comatose states relapsed. She is disturbed by the decreasing of sight acuity, weakness and pain in the legs. At ophthalmoscopy on the eyeground the microaneurysm of vessels, hemorrhages in the retina, neovascularisation. Glucose on an empty stomach is 9,7 mmol/l, glucosuria is 2% (2,5 l), acetone in urine was not discovered.

Task

- What is the preliminary diagnosis?
- What correction of diabetes therapy will you conduct?

Standard of answer

1. Type 1 diabetes, severe stage, decompensation, diabetic retinopathy 1, peripheral vascular disease.
2. Fasting hyperglycemia and after 120 min. Type 1 diabetes, first diagnosed, decompensation. Injections of short-acting insulin.
3. Ketoacidosis, coma. Blood glucose and urine ketones.
Infusion therapy and multiple injections of short-acting insulin (intravenous infusion of insulin 0.1 U/kg).
4. Ketoacidosis, coma. Infusion therapy and multiple injections of short-acting insulin (intravenous infusion of insulin 0.1 U/kg)
5. Hypoglycemic coma. Manage mild hypoglycemia by giving rapidly absorbed PO carbohydrate or glucose; for a comatose patient, administer an intramuscular injection of the hormone glucagon, which stimulates the release of liver glycogen and releases glucose into the circulation. Where appropriate, an alternative therapy is intravenous glucose (preferably not more than a 10% glucose solution). All treatments for hypoglycemia provide recovery in approximately 10 minutes.
6. Ketoacidosis, coma. Infusion therapy and multiple injections of short-acting insulin (intravenous infusion of insulin 0.1 U/kg).
7. I type diabetes, Mauriac syndrome . Insulin 0.8-1,0 u/kg.
8. Type 1 diabetes, severe stage, decompensation, diabetic retinopathy 111, peripheral vascular disease. Laser coagulation.
9. Hyperosmolar coma.
Hypoglycemia, high level of sodium, high level of chloride, high level of urea, glucosuria. . Infusion therapy and multiple injections of short-acting insulin (intravenous infusion of insulin 0.1 U/kg).
10. Diabetic precoma. Infusion therapy and multiple injections of short-acting insulin (intravenous infusion of insulin 0.1 U/kg).
11. I type diabetes, Mauriac syndrome . Insulin 0.8-1,0 u/kg.
12. Type 1 diabetes, severe stage, decompensation, diabetic retinopathy 111, peripheral vascular disease. Laser coagulation

Methodical materials for the class

A professional algorithm of patients management implementation (reference chart) for the practical skills and abilities forming .

No	Task	Sequence of implementation	Remarks and warnings related to self-control
1	To conduct patient examination for diabetes mellitus	1.To conduct complaints and disease's anamnesis taking.	To pay attention to the features of disease course , underlying factors,

	type 1	<p>2.To take thoroughly the patient's life anamnesis.</p> <p>3.To conduct examination of the patient.</p> <p>4.To investigate cardiovascular system of the patient (palpation, percussion).</p>	<p>concomitant diseases etc.</p> <p>To establish the risk factors which can cause the development of disease.</p> <p>To assess patient's general condition, position in bed, color and humidity of skin and mucose, presence of neck veins and extremities swelling.</p> <p>To pay regard to pulse rhythm, its tension and size on both hands, apex shove, its properties, margins of absolute and relative cardiac dullness, its changes, HR(tachi-or bradycardia, extrasystole), BP.</p>
		<p>5.To conduct heart and main vessels auscultation.</p> <p>6.To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the system of digestion.</p>	<p>To pay regard to heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To pay attention to features of percussion and auscultation in children of different age and stage of compensation.</p> <p>To pay attention to the changes in the case of decompensation and diabetic hepatosis.</p>
2	To formulate the preliminary diagnosis.	<p>1.To formulate the preliminary diagnosis</p> <p>2.To substantiate all the components of preliminary diagnosis taking as a basis complaints, anamnesis, and examinations.</p>	To formulate the preliminary diagnosis of diabetes mellitus and substantiate each component of it.
3	To evaluate the parameters of additional laboratory investigations.	<p>1.To evaluate the blood and urine count data.</p> <p>2. To evaluate the level of</p>	<p>To pay attention to the signs of anemia, leucocytosis, changing of formula, elevation of sedimentation rate, glycosuria, urine ketones.</p> <p>To pay attention to urine</p>

		<p>glycemia and glycosuria, oral glucose tolerance test (OGTT) .</p> <p>3. To evaluate the biochemistry data of blood and urine, renal function tests.</p> <p>4.To evaluate the blood hormonal profile</p>	<p>glucose and blood glucose levels.</p> <p>To pay attention to data of OGTT.</p> <p>To pay attention to cholesterol, lipids, creatinine, glomerular filtration rate , glycated hemoglobin, microalbuminuria.</p> <p>To pay attention to C-peptide changing.</p>
4	To understand the data of additional and laboratory investigation.	To understand the data of thermography, ophthalmoscopy, rheography , vibration sensation	To pay special attention to long-term complications of diabetes mellitus.
5.	To conduct differential diagnosis.	<p>1.Consistently to find the common signs in complaints, life and disease anamnesis, data of examination, data of laboratory and instrumental investigations in patient and in similar states.</p> <p>2.To find differences between complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumental methods of research and in similar nosology.</p> <p>3.On the basis of found the differences to exclude similar diseases from the list of possible diagnoses.</p> <p>4. To conduct differential diagnostics according to the above mentioned algorithm among all the nosologies having the similar signs, among other diseases of thyroid gland</p> <p>5.Taking into account the impossibility to exclude</p>	Special attention must be paid to differential diagnosis among the Diabetes Insipidus, renal glycosuria, transient glucosuria and hyperglycemia, renal glucosuria.

		the diagnosis of diabetes mellitus from the list of credible diagnoses to draw a conclusion about the probability of such a diagnosis.	
6	To formulate the clinical diagnosis.	1.To formulate the final clinical diagnosis. 2.Taking the preliminary diagnosis as a basis, additional investigations data, conducted differential diagnosis, substantiate all the elements of the final clinical diagnosis.	Basing on modern classification of diabetes mellitus, formulate the diagnosis, complications of disease and the presence of concomitant diseases.
7	To prescribe treatment for patients.	1.To prescribe no medicinal treatment 2.To prescribe t medicinal treatment.	To specify the regimen and detalized diet according to a disease. Taking into account the age, severity of patient's state, the stage of disease, the presence of compli-cations and concomitant pathology, to prescribe modern medicinal treatment in accordance to the standards of diabetes mellitus therapy.

The material for the control of the secondary level of abilities and skills:

The secondary tests

1.Patient K., has been suffering from diabetes for 8 years. During the last year the dose of insulin diminished on 14 U. In the analysis of urine protein is 1,7 %, sugar 0,8 %, a lot of red corpuscles, cylinders. The diagnosis of nephrosclerosis was made.

What is the reason of diminishing in insulin necessity?

- A) diminishing of contrainsular hormones activity
- B)diminishing of insulin binding to the proteins
- C)diminishing of insulin disintegration in the kidneys
- D) all indicated reasons
- E) none of the indicated reasons

2. Patient M, 12 years old, has been suffering from diabetes for 6 years. Takes insulin of «Aktrapid» 13 U + «Protophan» 14 U. In anamnesis the comatose states relapsed.

He is disturbed by decrease of sight acuity, weakness and pain in the legs. On ophthalmoscopy on the eyeground the microaneurysm of vessels, hemorrhages in the retina, neovascularisation. Glucose on an empty a stomach is 9,7 mmol/l, glucosuria is 2 % (2,5 l), acetone in urine is not discovered.

Specify the severity and the compensation of diabetes:

- A) mild form, subcompensated
- B) average severity, compensated
- C) severe, subcompensated
- D) mild form, decompensated
- E) severe form, compensated

3. Patient M, 12 years old, has been suffering from diabetes for 6 years. He takes insulin of «Aktrapid» 13 U + «Protophan» 14 U. In anamnesis the comatose states are relapsed. He is disturbed the decrease of sight acuity, weakness and pain in the legs. On ophthalmoscopy on the eyeground the microaneurysm of vessels, hemorrhages in the retina, neovascularisation. Glucose on an empty a stomach is 9,7 mmol/l, glucosuria is 2 % (2,5 l), acetone in urine is not discovered.

What correction of diabetes therapy will you conduct?

- A) to decrease the amount of proteins in day's ration
- B) to include biguanids in the complex of treatment
- C) to the therapy carried out to add äëöëíí
- D) to include the preparations of 1- generation sulphonurea
- E) to include the preparations of 2- generation sulphonurea

4. A child of 7 years old was hospitalized with complaints of thirst, frequent emiction, weight loss (3 kg for a month). On examination the level of glucose in the blood on an empty a stomach is 19,2 mmol/l. The diagnosis of I type diabetes was made. How is it possible to explain the decreasing of child's bodyweight ?

- A) Intensifying of proteins catabolism and inhibition of its synthesis.
- B) Decreasing of lypolysis
- C) Intensifying of gluconeogenesis.
- D) Decreasing of lypolysis.
- E) Decreasing of basic exchange.

5. A girl of 12 years old, since the 2-years-old age has been suffering from diabetes. On examination: considerably enlarged stomach, liver is 6 cm below the costal arc, somewhat painful and dense; Cushing - like type of obesity, nanism. The signs of the sexual maturation are not present. Glucose in the blood is 17 mmol/l, in the urine it is 4 %. In the blood increased content of cholesterol and ketonic bodies.

What is the preliminary diagnosis?

- A) 1n type diabetes, Mauriac syndrome
- B) 1 type diabetes And type, Nobecur
- C) 1 type diabetes, precomma
- D) Cushing disease
- E) Cushing syndrome

6. In the child of 9 years old diabetes was firstly found . How will skin injuries be manifested?

- A) By predilection to the purulent diseases
- B) By depigmentation
- C) By hyperpigmentation.
- D) By development of elephantiasis
- E) By petechias

7. A child of 5 years old was first hospitalized in the department with a diagnosis of I type diabetes, decompensation (ketoacidosis). What is the main mechanism in the development of the disease?

- A) Insulin insufficiency.
- B) Surplus of glucagon.
- C) Surplus of somatostatinum.
- D) Disturbances of insulin binding to the receptors.
- E) Disturbances of postreceptor mechanisms of insulin action.

8. A child of 10 years old was hospitalized with complaints of thirst, increased emiction weight loss, weakness, languor. After clinical and laboratory investigation there was the diagnosis of I type diabetes in the phase of decompensation . How is it possible to explain the symptoms of poliuria in this patient ?

- A) Glucose selected with urine, operates as osmotic diuretic.
- B) Decreased production of antidiuretic hormone.
- C) Increased vasopressin production
- D) Decreased TTH production.
- E) Increased TTH production

9. A boy ill with diabetes was admitted to the induction centre of child's hospital in the severe condition. On examination the absence of consciousness was revealed. The skin is dry, pale, the turgor of skin is decreased. Hypotonus of muscles and decreased tonus of eyeballs. Mucus of lips is dry, in bright red color. Tones of heart are hyposthenic, tachycardia, low blood pressure, threadlike pulse. Breathing of Coumaul. Strong smell of acetone midair. Specify day's dose of insulin for the acute management.

- A) 2,5 U/kg
- B) 1,5 U/kg
- C) 2,0 U/kg
- D) 1,0 U/kg
- E) 3,0 U/kg

10. A boy, 7 years old, was hospitalized with complaints on intensifying of thirst, poliuria. During the last 5 years he was ill with diabetes, in anamnesis a diabetic comma developed three times. The level of glucose in the blood is 15,54 mmol/l, in the urine is 5 %. Retinopathy was found by an oculist. What dose of insulin will most probably be prescribed to the child?

- A) 1,5 U/kg

- B) 0,25 U/kg
- C) 0,5 U/kg
- D) 1,0 U/kg
- E) 2,0 U/kg

11. What dose of insulin will most probably be prescribed to the child who has been suffering from diabetes during the last 5 years?

- A) 0,5-0,6 U/kg
- B) 0,7-0,8 U/kg
- C) 0,25 U/kg
- D) 1,0-2,0 U/kg
- E) 2,-3,0 U/kg

12. The level of albumin in urine in the case of microalbuminuria

- A) 1-10 mg/l
- B) 10-30 mg/l
- C) 30-300mg/l
- D) > 300 mg/l
- E) 0-10 mg/l

13. A child of 5 years old has been suffering from diabetes during 2 years. What is the number of BU that you will recommend?

- A) 12-13 BU
- B) 15-16 BU
- C) 16-17 BU
- D) 17-18 BU
- E) 19-21 BU

14. Which kind of insulin is short-acting?

- A) glulisine
- B) detemir
- C) isophane
- D) glargine
- E) ultralente

15. What is the dose of glucagons in the case of hypoglycemic coma for a ten-year old child ?

- A) 0,5 mg
- B) 1 mg
- C) 2 mg
- D) 5 mg
- E) 15 mg

16. What is the normal level of glycosylated hemoglobin ?

- A) 5-7 %
- B) 10%
- C) 10-15%
- D) 20%

E) 20-25%

17. Refined carbohydrates in dietary management for children with diabetes mellitus should provide less than

- A) 10%
- B) 20%
- C) 30%
- D) 40%
- E) 50%

18. How is it possible to explain the appearance of acetone midair and urine ?

- A) By the increased disintegration of ketogenic amino acids and lipids.
- B) By the disturbance of water - electrolyte balance.
- C) By the disturbance of acid - alkaline balance.
- D) By the disturbance of glucose phosphorylating processes.
- E) By the decreasing of glycolys.

19. The peak activity of Protamine zinc (Ultralente) insulin is...

- A) 1-2 h
- B) 3-4 h
- C) 4-8 h
- D) 10 h
- E) 24 h

20. Treatment of diabetic nephropathy and hypertension includes

- A) angiotensin-converting enzyme inhibitors
- B) blood pressure control.
- C) diabetes control
- D) Angiotensin type 1 blockers
- E) All listed above

Answers: 1-D, 2-C, 3-B, 4-A, 5-A,B, 6-A, 7-A, 8-A, 9-D,10-A,11-B, 12-C, 13-A, 14-A, 15-B, 16-A, 17-A, 18-A, 19-C, 20-E.

Materials of the medical support for the students independent training: a reference chart for organization of students independent work with educational literature.

Tasks	Instructions
To study the pathogenesis of diabetes mellitus in children.	Select the key links of diabetes mellitus pathogenesis type 1
To study the clinical manifestations of diabetes mellitus in children.	To establish the symptoms and gather it to clinical syndromes which enable to make the credible diagnosis of diabetes mellitus.
To study the clinical manifestations of complications of diabetes mellitus in children.	To establish the symptoms and gather it to the clinical syndromes which enable to make the credible diagnosis of

	complications of diabetes mellitus
To study diagnostic criteria of diabetes mellitus	To make a structural plan of disease
To study the additional methods of research (laboratory, instrumental)	To work out a plan of patient's examination.
To study the changes in additional investigational methods which are pathognomonic for diabetes mellitus and its complications.	To enumerate the basic diagnostic criteria of diabetes mellitus according to the data of additional investigational methods.
To conduct differential diagnostics, to establish a final diagnosis	To substantiate the basic components of diagnosis in accordance with modern classification, and to conduct a differential diagnosis.
To prescribe the individual treatment to patient with the diabetes mellitus type 1. To render the first aid in extreme situations: hyperglycemic, hypoglycemic, hyperosmolar, hyperlactacidemic, ketoacidosis coma.	To make the prescribing chart specifying the regimen, diet, medicinal treatment, taking into account the age, severity of patient's state, stage of disease, presence of complications and concomitant diseases.

Basic literature:

1. Nelson Textbook of Pediatrics, ed 16.2000.
2. Bode BW (Ed.): Medical Management of Type 1 Diabetes. 4th ed. Alexandria, VA, American Diabetes Association, 2004
3. Сахарный диабет и его осложнения у детей и подростков : Монография/ Н.В. Филиппова Е.А. и др.-Харьков: Основа 2005-300с.
4. Касаткина Э.Н. Сахарный диабет у детей.-М.: Медицина, 1990-272с.

Additional literature:

1. Klingensmith GJ (Ed.): Intensive Diabetes Management. 3rd ed. Alexandria, VA, American Diabetes Association, 2003
2. Expert Committee on the Diagnosis and Classification of Diabetes Mellitus: Follow-up report on the diagnosis of diabetes mellitus. Diabetes Care 26:3160–3167, 2003

Theme 5. Thyroid gland diseases in children.

Classification of thyroid diseases in children. Etiology, pathogenesis, clinical presentation, diagnostics, differential diagnostics, treatment, prophylaxis of diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children. Degrees of goiter. Emergency in thyroid crisis. Prognosis.

I. Actuality of the theme.

Thyroid functions disturbances is the common state among children. Thyroid diseases is quite various in children age. Thyroid diseases problems are the main relating to Chernobyl disaster because of morbidity increasing among children in autoimmune thyroiditis, hypothyroidism, good-quality and malignant tumors of thyroid. One of major places occupies congenital hypothyroidism that meets in frequency of 1 case to 5000 newborns. Congenital hypothyroidism in 85 – 90% of cases is primary and related to the iodine deficit or thyroid dysgenesis. Thus, the aplasia, hypogenesis or dystopia of thyroid are the more frequent states. Primary hypothyroidism in 5 – 10% of cases unconditioned by dysmorphonose (autosomal – recessive inheritance). Congenital hypothyroidism is the second or tertiary (pathology of hypophysis or hypothalamus) and meets no more than in 3 – 4% of cases. Congenital hypothyroidism leads to the mental and physical development retardation and psychical inability of children, that is why the early diagnosis is very important.

Concrete purposes:

1. To determine the etiologic and pathogenetic factors in diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children

2. To classify and analyse the typical clinical manifestation of diffuse toxic goiter, thyroiditis, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children. To determine the features of congenital hypothyroidism for newborns and children and put a preliminary clinical diagnosis.

3. To make the plan of examination and to analyse the information about laboratory and instrumental data in the classic course of diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, and endemic goiter in children.

4. To demonstrate skills of treatment, rehabilitation and prophylaxis in diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter in children.

5. To diagnose and render an urgent help in thyrotoxic crisis and hypothyroid coma in children.

6. To conduct differential diagnostics of diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children and put a preliminary diagnosis.

7. To determine the prognosis for life in diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis and endemic goiter in children.

8. To demonstrate the skills of medical specialist's moral and deontological principles and principles of professional subordination in pediatrics.

II. Classes (pointing of planned mastering level)

1. A student must have a conception (familiarize): $\alpha 1$

- The place of thyroid diseases in the structure of endocrine system diseases in children, widespread in different age-dependent and ethnic groups;
- Statistical information in relation to morbidity, frequency of complications, lethality, the nearest and remote prognosis in patients with the diseases of thyroid gland;
- The history of scientific studying and the contribution of domestic scientists;

2. A student must know (master): $\alpha 2$

- etiology of diffuse toxic goiter, thyroiditis, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children.
- key links of thyroid diseases pathogenesis ;
- clinical classification of thyroid diseases;
- degrees of goiter;
- classical clinical manifestation of hypothyroidism;
- classical clinical manifestation of diffuse toxic goiter;
- classical clinical manifestation of autoimmune thyroiditis;
- classical clinical manifestation of endemic goiter;
- classical clinical manifestation of diffuse untoxic goiter;
- laboratory diagnosis of hypothyroidism and hyperthyroidism;
- laboratory and instrumental diagnosis of thyroid diseases;
- complications of diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter in children.
- treatment principles of diffuse toxic goiter, congenital hypothyroidism, autoimmune thyroiditis, diffuse untoxic goiter in children.

3. A student must master: $\alpha 3$

Skills:

- Collection of complaints and anamnesis of disease;
- Examination of patient with thyroid diseases and revealing the main symptoms and syndromes.
- To formulate and substantiate the preliminary diagnosis;
- Determination of laboratory and instrumental inspection plan of patient's examination (according to diagnostics' standards);

Abilities:

- To interpret the results of laboratory and instrumental tests.
- To conduct differential diagnosis among acute and subacute thyroiditis, thyroid cancer, to diagnose thyrotoxic crisis and hypothyroid coma are required for emergencies.
- To conduct differential diagnosis with thyroid cancer.
- To give recommendations in relation to the patient's regimen and diet with diseases of thyroid gland, taking into account the stage of the disease, severity of the state and concomitant pathology;
- To complete the treatment plan in thyroid diseases according to the standards taking into account the stage of the disease, complications and concomitant pathology.
- To render the first aid in extreme situations and exigent states.

III. Aims of personality development (educative aims):

- A student must learn to adhere rules of behaviour and principles of medical etiquette and deontology, to develop bedside manner;
- Be able to set a psychological contact with a patient and his family;
- To master the sense of professional responsibility for a timely and adequate medicare.

IV. Interdisciplinary integration:

Subject	To know	To be able
1. Basic		
Human anatomy	Structure of human endocrine system, of thyroid gland , their circulation and innervation.	To determine the location of thyroid projection and palpation, of local lymphatic nodes.
Histology	Structure of thyroid gland vessel system.	
Physiology	Normal physiology of human endocrine system, normative indices of laboratory and instrumental investigational methods and their assessment.	To assess the data of laboratory and instrumental investigational methods and thyroid gland function.
Physiopathology	Key links of pathogenesis of thyroid gland diseases, of hypothyroidism and hyperthyroidism	To estimate the function of thyroid gland and other organs of the endocrine system
Pathologic anatomy	Morphological features of thyroid gland diseases development depending of the stage of the disease .	To analyse and interpret the information about clinical examination and about additional methods of investigation

Pharmacology	Pharmacokinetics and pharmacodynamics, the side effects of preparations (thyroid hormones preparations, thyroidostatics, antibiotics, antiinflammatory drugs etc.)	To prescribe age- dependent treatment of patient, taking into account individual features and period of disease, to establish the individual regimen of preparations taking and dosage. To be able to make a prescription.
Propedeutics of pediatrics.	Basic stages and methods of patient clinical examination	To collect complaints, anamnesis vitae et morbi, to find out the basic risk factors of thyroid diseases ,

		to be able to conduct patient's examination, to reveal the clinical signs of thyroid gland diseases, to interpret the data of additional methods of investigation.
Radiology	Normal parameters of ultrasound and radionuclide diagnostics in thyroid gland diseases.	To interpret the data of ultrasound and radionuclide diagnostics
2. Followings (provided)		
Hospital pediatrics.	Clinical signs of thyroid gland diseases, differential diagnosis and treatment tactics.	To reveal the clinical signs of thyroid gland disease and complications, to conduct differential diagnosis, to be able to prescribe treatment.
3. Interdisciplinary integration		
Acute and subacute thyroiditis	Clinical manifestation of acute and subacute thyroiditis.	To establish specific clinical signs of acute and subacute thyroiditis and to conduct differential diagnosis to other thyroid diseases and acute lymphadenitis.
Thyroid cancer	Clinical manifestation of thyroid cancer	To establish specific clinical signs of thyroid cancer and to conduct differential diagnosis among other manifestations of thyroid diseases

V. Plan and organizational structure of classes.

№ п/п	Basic stages of classes, their function and maintenance	Educational aims are in the levels of mastering	Methods of control and studies	Educational materials	Distributing of time in minutes
1	Preparatory stage Organizational measures Raising of educational aims and motivation	α2	Individual oral	I «Actuality of theme» II «Educational aims»	3 min. 12 min. 20 min.

<p>Control of basic knowledges and skills level:</p> <p>1. Etiology of diffuse toxic goitre, hypothyroidism, autoimmune thyroiditis, endemic goitre, acute and subacute thyroiditis, diffuse untoxic goitre, thyroid cancer in children</p> <p>2.Key links of thyroid diseases pathogenesis; classification of throid diseases;</p> <p>3.Goitre degrees;</p> <p>4.Typical manifestation of diffuse toxic goitre, autoimmune thyroiditis, endemic goitre, acute and subacute thyroiditis, diffuse non toxic goitre in children.</p> <p>5. Laboratory and instrumental diagnosis of hypothyroidism and hypothyroidism:</p> <p>6.Complication of diffuse toxic goitre, autoimmune thyroiditis, endemic goitre, acute and subacute</p>	<p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p> <p>α2</p>	<p>questioning</p> <p>Test control of the second level</p> <p>Individual (oral) questioning</p> <p>Typical situational task of the 2 level</p> <p>Typical situational task of 2 level</p> <p>Typical situational task of 2 level</p> <p>Test control of 2 level</p> <p>Typical situational task of 2 level</p>	<p>The second level tests</p> <p>The table «classification of thyroid gland diseases»</p> <p>Structurally logical chart of thyroid diseases</p> <p>Typical situational task of 2 level</p> <p>Tests of 2 level</p> <p>Typical situational tasks of 2 level</p> <p>Kit of medicines.</p>	
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	thyroiditis, diffuse non toxic goitre in children.; 7.Treatment principles of diffuse toxic goitre, autoimmune thyroiditis, endemic goitre, acute and subacute thyroiditis, diffuse non toxic goitre in children;				
2	Basic stage of professional skills and abilities forming: 1.To conduct the patient's management with thyroid diseases, to take complaints and anamnesis. 2.To conduct the patient examination, to detect main symptoms and syndromes of thyroid disease. 3.To formulate and substantiate the preliminary diagnosis 4.To compose the plan of patient's laboratory and instrumental investigation. 5.Interpret the results of laboratory and	α_3 α_3 α_3 α_3 α_3	Practical professional training Practical professional training Practical professional training Practical professional training Tests and the third level control. The third level test control. The practical professional training in solving of non	Patient Patient Case history A reference chart for forming of professional abilities. Case history. A reference chart for forming of professional abilities. Situational typical tasks of 3 level. The third level tests. Prescribing chart	115 min.

	<p>instrumental investigation.</p> <p>6.To conduct differential diagnosis among clinical conditions accompanied by thyroid gland changes.</p> <p>7.To give the recommendations for regimen and diet of patient.</p> <p>8.To compose the plan of thyroid disease patient treatment taking into account the stage of the disease and the presence of complications.</p> <p>9.To be able to render the first aid in extreme situations</p>	<p>α3</p> <p>a3</p> <p>α3</p>	<p>typical clinical situations.</p> <p>The third level test control.</p> <p>Practical professional training.</p> <p>The third level test control.</p> <p>The practical professional training on solving of non typical clinical situations.</p>	<p>The third level non typical situational tasks.</p> <p>Treatment algorithm for the thyroid diseases patients.</p> <p>The third level non typical situational tasks.</p> <p>The first aid algorithm in thyroid diseases.</p>	
3	<p>Concluding stage.</p> <p>Control and correction of professional abilities and skills.</p> <p>Working out the totals of class.</p> <p>Home work (basic and additional literature on the topic)</p>		<p>Analysis of clinical work.</p> <p>Solving of non typical tasks and the third level tests.</p> <p>Estimation of the clinical work.</p>	<p>Clinical work.</p> <p>The third level non typical situational tasks.</p> <p>A reference chart for independent work with literature</p>	30 min.

Questions for elementary level of knowledge control

1. The role of thyroid hormones, mechanism of action and regulation of secretion,
- 2.The degrees of thyroid gland enlargement.

3. Definition of struma. Different kinds of struma.
4. Laboratory methods for the diagnosis of thyroid glands disorders.
5. Factors promotes the development of diffuse toxiferous struma.
- 6 What is the classic clinical tirade in diffuse toxiferous struma.
7. The principles of conservative treatment in diffuse toxiferous struma and indications to the surgical treatment.
8. Causes and treatment of thyroid toxic crisis.
9. Laboratory and instrumental criterions of hypothyroidism in children.
10. The causes of development of asquired and congenital hypothyroidism. The principles of treatment. Specialties in early age children.
11. Clinical and laboratory criterions of thyroiditis. Treatment.
12. The criterions of compensation in thyroid gland disorders in children.
13. Methods of early diagnosis of thyroid gland cancer in children.

The primary control tests

1. In child clinical examination there are follow signs revealed - skin humidity, exaltation, irritability, decreasing of body weight, tachycardia, syndromes of Grefe, Stellwag, Moebius, palpitation. For what disease these signs are characteristic?

- A. **Thyreotoxicosis**
- B. Hypothyroidism
- C. Diabetes
- D. Down disease
- E Acromegalia

2. Girl of 14 years old complains of sleeping disturbances, decreasing of body weight, palpitation, cardialgias, and fatigability. A thyroid gland hyperplasia of II degree and exophthalmia is marked.. What changes in hormones level are most typical for this disease?

- A. Decreasing of a thyroxin
- B. **Rising a thyroxin and triiodthyronin**
- C. Increasing of the iodine level connected to protein
- D. Increasing of Thyrotrophic hormone
- E. Increasing of triiodthyronin

3. In the girl of 12 years old after examination the diagnosis of mild diffuse toxic struma established. What dose of thyreostatic Mercazolilum is necessary to administrate for child in this case?

- A. 10-15 mg per day
- B. 5-10 mg per day
- C. **20-30 mg per day**
- D. 1-5 mg per day
- E. 40-50 mg per day

4. Girl, 14 years old, complains of decreasing of body weight in normal

appetite, irritability, sweating, tremor of hands, palpitation,. The thyroid gland enlarged up to II degree, unpainful, elastic. The diagnosis of diffuse toxiferous struma clinically fixed. What from results of examination will confirm your diagnosis?

- A **T3 and T4 is increased**
- B. T3 and T4 is normal
- C. T3 and T4 is reduced
- D. Hypocalcaemia
- E. Hyperphosphatemia

5. In examination of 14 years old girl the nodal struma of III degree is found out. On scenogramm the “hot” unit revealed. Levels of T3 and T4 in a blood are increased. What disease is it possible to think of?

- A. Diffuse toxic struma
- B **Toxic adenoma of a thyroid gland**
- C Cancer of a thyroid gland
- D Autoimmune thyroiditis
- E. Fibrous struma of Riddell

6. In the patient. of 13 years old, relapse of a nephrolithiasis, ostealgia, weakness, fatigability, growing thin are observed. What from the specified diseases can be suspected?

- A. **Hyperparathyroidism**
- B Hypoparathyrosis
- C. Sarcoma of bones
- D. Multiple myeloma
- E. Any of the specified diseases

7. In boy of 15 years old the attacks of seizures in masseters and hands with prevalence of flexors tone are observed. Seizures are painful and symmetric. In examination there are positive signs of Hvostek and Trussot.

What is your diagnosis?

- A. Epilepsy
- B Tetanus
- C. Hyperparathyroidism
- D. **Hypoparathyroidism**
- E. Spasmophilia

8. Examination in the girl of 13 years old in there is I degree thyroid gland enlargement. Does not show any complaints. In palpation the thyroid gland is elastic, painless and of homogeneous consistence, . In investigation the disorders of thyroid gland functions are not revealed, a level of thyroid hormones are normal. What is the diagnosis?

- A. Autoimmune thyroiditis.
- B. **Juvenile struma.**
- C. Cancer of a thyroid gland.

- D. Diffuse toxic struma.
- E. Ridel fibrosal struma.

9. What is typical for the secondary hypothyroidism?

- A. A low level of Adrenocorticotrophin.
- B. A low level of thyrotropin.**
- C. A low level of thyroliberin.
- D.** A high level of thyroliberin.
- E. A high level of thyrotropin.

10. In newborn there are constipations, icterus, flaccidity, sleepiness. In examination: moderate icterus, inflated abdomen, enlargement of liver and lien, puffing in respiration. What is the most probable diagnosis?

- A. Hepatitis
- B. Hypothyroidism.**
- C. Rinitis.
- D** Down syndrom
- E. Rickets.

11. In the patient of 10 years old the delicacy, fatigability, decreasing of progress in school, dry and cold skin, fragility of hair and nails are marked. During the further investigation the hypothyroidism was diagnosed. What therapy is necessary to prescribe for this patient?

- A. Mercazolilum.
- B. Thyroxine.**
- C. Prednisolonum.
- D. DOCSA.
- E. Hidrocortisonum.

12. Child, in 8 months old for the first time the congenital hypothyroidism clinically and according to thyroid hormones tests was confirmed. Now the most expressive manifestation is the serious lag in psychophysical development. What in this case the most rational therapeutic tactics?

- A. L-thyroxine + Pyracetamum**
- B. Thyreoidinum
- C. Ttriodthyroninum
- D. Mercazolilum
- E. L-thyroxine + Retabolilum

Answers to the primary control tests

1-A, 2-B, 3-C, 4-A, 5-B, 6-A, 7-D, 8-B, 9-B, 10-B, 11-B, 12-A

Typical situational tasks of 2 level

Task 1

In 2 week old newborn there are constipations, icterus, flaccidity, sleepiness. You are seeing a 2-week-old boy at a routine visit. The mother complains that he has been constipated, jaundiced, sluggish, and excessively sleepy. The physical examination is normal except for mild jaundice and a distended abdomen in a sleepy infant.

1. What assessment is the most appropriate course to pursue initially?
2. What diagnosis is most likely indicates?
3. What tests are confirmed the suspected diagnosis?
4. Disease treatment.
5. What monitoring of patient condition?

Standard of answer . Task 1.

1. The results of the neonatal metabolic screen
2. Congenital hypothyroidism
3. Levels of thyrotropin-releasing hormone (TRH), thyroid-stimulating hormone (TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4), and thyroglobulin in the infant. The main thrust is directed at the need for speed in the diagnosis and treatment of congenital hypothyroidism because the earlier treatment is started with thyroid hormone, the better the prognosis for intellectual function. Time should not be spent in exhaustive investigation. Regardless of the reason for the hypothyroidism, treatment with replacement thyroid hormone is indicated. If it turns out that the initial diagnosis was erroneous, little harm will be done by treating an infant with a physiologic dose of thyroid hormone for a few days. Waiting for laboratory tests or x-rays to be performed, interpreted, and probably repeated is inappropriate if this will delay treatment. Eventually, they should be done, along with an evaluation of the mother's immune status, her health history, and a complete family history looking for one of the many known, although relatively rare, causes of congenital hypothyroidism. Thyroid dysgenesis is found in 90% of the cases. Neonatal screening for hypothyroidism has allowed for the much earlier diagnosis of hypothyroidism, resulting in improvement of prognosis, so that frank cretinism is now quite rare. Most industrialized countries test for phenylketonuria and hypothyroidism; there is variability in testing for other metabolic and genetic diseases.
4. Begin on the baby oral sodium-L-thyroxine, 10 to 15 mg/(kg_d)
5. Periodic measurement of T3, T4, and TSH is necessary to assess the response to therapy and the need for adjustment of the dose of thyroxine. Careful evaluation of somatic growth by plotting sequential measurements and monitoring bone age is essential

Task 2

In the girl of 7 years old after viral syndrome the temperature up to 39,2 has raised suddenly, the headache, pain in the thyroid gland area is intensified in swallowing and head turning has appeared. The thyroid gland enlarged – II degree, painful in palpation, the hyperemia of the skin above it. In laboratory data the function of thyroid gland is unchanged. In the analysis of a blood the leukocytosis and accelerated BSR.

Questions

1. What is the diagnosis?
2. What examinations are necessary?
3. What changes in hormones level are most typical for this disease?
4. Disease treatment.
5. What monitoring of patient condition?

Standard of answer . Task 2.

- 1 Acute thyroiditis.
2. Levels of thyroid-stimulating hormone (TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4).
3. Decrease of thyroid-stimulating hormone (TSH), increase of tri-iodothyronine (T3), reverse T3, levothyroxine (T4).
4. Glucocorticosteroids.
5. If the nodules are not suppressed by replacement therapy with glucocorticosteroids - Hashimoto's thyroiditis.

Situational Task 3

The girl of 11 years old. Complaints to the general delicacy, fatigability, enlargement of neck. Objectively: thyroid gland in palpation is dense, impure and enlarged up to II degree. In it structure investigation the hyperecho and hypoecho sites were marked. TTH level and of antibodies levels to the thyroid gland are raised.

1. What is the preliminary diagnosis?
2. What of examination will confirm your diagnosis?
3. Conduct differential diagnostics.
4. Disease treatment.
5. What monitoring of patient condition?

Standard of answer . Task 3.

1. Multinodal struma.
2. Puncture biopsy of thyroid gland
3. a. Autoimmune thyroiditis, the hypertrophic type b. Cancer of a thyroid gland c. Fibrose struma of Riedel d. Endemic struma.
4. If the nodules are not suppressed by replacement therapy with T4, surgery is indicated, because malignancy cannot readily be ruled out.

5. Periodic measurement of T3, T4, and TSH is necessary to assess the response to therapy and the need for adjustment of the dose of thyroxine. Careful evaluation of somatic growth by plotting sequential measurements and monitoring bone age is essential

Task 4

The patient of 4 years old retards in mental development. Birth weight is 3900 g, body height is 52 sm. From the first months of life lags behind in development, a head started to hold in one year, to sit in 1, 8 years. Separate words started to speak from 3 years. Objectively: body height is 80 sm, weight is 11kg, face is bloated, amimic and pastose, palpebral fissures are narrow, lips are thick, mouth is slightly opened, tongue is full out and extended from a mouth. Skin acyanotic, dry and shelled, hair dry and infrequent. The big fontanel is still open. There are only 4 teeth. A stomach is normal. Sexual development corresponds to 1 year. Ps is 84 per minute; blood pressure is 85/60 mm Hg. Cardiac tones are weakened. What is the preliminary diagnosis?

1. What is the preliminary diagnosis?
2. What of examination will confirm your diagnosis?
3. What therapy is necessary to prescribe for this patient?
4. What monitoring of patient condition?
5. Prognosis of disease?

Standard of answer . Task 4.

1. Congenital hypothyroidism.
2. Levels of thyrotropin-releasing hormone (TRH), thyroid-stimulating hormone (TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4), and thyroglobulin in the infant.
3. Begin on the baby oral sodium-L-thyroxine, 10 to 15 mg/(kg d)
4. Periodic measurement of T3, T4, and TSH is necessary to assess the response to therapy and the need for adjustment of the dose of thyroxine. Careful evaluation of somatic growth by plotting sequential measurements and monitoring bone age is essential.
5. Prognosis - the mental development retardation.

Situational Task 5

Patient G., 12 years old. Growing weight on 10 kg for 4 months, complains of constant irritability, palpitation, pain in eyes and lacrimation. In examination: skin is warm and wet, mild exophthalmia and hyperemia of conjunctiva, positive signs of Grefe, Koher and Moebius. The thyroid gland is unpainful and diffusively enlarged, that is seen in swallowing,. Pulse 108 per minute, blood pressure is 140 / 66 mm Hg. There is fine tremor in hands fingers.

1. What is the diagnosis?
2. What of examination will confirm your diagnosis?

3. What therapy is necessary to prescribe for this patient?
4. What monitoring of patient condition?
5. Prognoses of disease?

Standard of answer . Task 5.

1. Diffuse toxic struma of II degree with an average thyrotoxicosis
2. Levels of thyrotropin-releasing hormone (TRH), thyroid-stimulating hormone (TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4), antibodies levels to the thyroid gland and thyroglobulin
3. Thyreostatic Mercazolilum is necessary to administrate for child in dose 20-30 mg per day, beta-blokers (anaprilin).
4. Periodic measurement of T3, T4, and TSH is necessary to assess the response to therapy and the need for adjustment of the dose of thyroxine. Careful evaluation of somatic growth by plotting sequential measurements and monitoring bone age is essential.
5. Hashimoto's thyroiditis, hypothyroidism.

VII. Methodical materials to support basic stage class.

Professional algorithm of patient’s management (reference chart) for the practical skills and abilities forming .

	Task	Sequence of implementation	Remarks and warnings related to self-control
	To conduct examination of the patient with thyroid disease.	<ol style="list-style-type: none"> 1.To conduct the complaints and disease anamnesis. 2.To take thoroughly the patient’s life anamnesis. 3.To conduct examination of the patient. 4.To investigate cardiovascular system of the patient (palpation, percussion). 	<p>To pay attention to the features of disease course , underlying factors, concomitant diseases etc.</p> <p>To establish the risk factors which can cause the development of disease.</p> <p>To assess patient’s general condition, position in bed, color and humidity of skin and mucose, presence of neck veins and extremities’ swelling.</p> <p>To pay regard to rhythm of pulse, it tension and size on both hands, apex shove, it properties, margins of absolute and relative cardiac dullness, it changes, HR(tachi- or bradycardia, extrasystole), BP.</p>

		<p>5.To conduct auscultation of the heart and of the main vessels .</p> <p>6.To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the system of digestion.</p> <p>9. To conduct examination and palpation of thyroid gland and local lymphatic nodes.</p>	<p>To pay regard to heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To pay attention to features of percussion and auscultation in children of different age.</p> <p>To pay attention to changes in hyper- and hypothyroidism.</p> <p>To determine the degree of thyroid gland enlargement.</p>
	To formulate the preliminary diagnosis.	<p>1.To formulate the preliminary diagnosis</p> <p>2.To substantiate all components of preliminary diagnosis taking as a basis complaints,anamnesis, and examinations.</p>	To formulate the based on modern classification preliminary diagnosis of thyroid disease and to substantiate each component of it.
	To evaluate the parameters of additional laboratory tests.	<p>1.To evaluate the blood count data.</p> <p>2. To evaluate the biochemistry data.</p> <p>3.To evaluate the blood hormonal profile.</p>	<p>To pay attention to signs of anemia, leucocytosis, changing of formula, elevation of sedimentation rate.</p> <p>To pay attention to cholesterol, lipids and glucose levels.</p> <p>To pay attention to TSH and thyroid hormones changing.</p>
	To evaluate the data of additional examination.	To understand the data of thyroid ultrasound.	To pay special attention to the thyroid volume depending of age, tissue characteristics, presence of nodes.
	To conduct differential diagnosis.	1.Consistently to find the common signs in complaints, life and disease anamnesis, the data of examination, the	Special attention must be paid to differential diagnosis among the acute and subacute thyroiditis, thyroid cancer, tuberculosis of lymphatic nodes,

		<p>data of laboratory and instrumental examination in patients with similar status.</p> <p>2. To find the differences among complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumental methods in similar nosology.</p> <p>3. To find out the differences for excluding similar diseases from the list of probable diagnoses, being based on this algorithm.</p> <p>4. To conduct differential diagnostics according to the algorithm among all of nosologies are having the similar signs, among other diseases of thyroid gland.</p> <p>5. Taking into account the impossibility to exclude the diagnosis of thyroid disease from the list of probable diagnoses to draw a conclusion about most probability of such diagnosis</p>	<p>systemic diseases of connective tissue, systemic blood diseases, in congenital hypothyroidism – among physical and mental retardation of child.</p>
	<p>To formulate the concluding clinical diagnosis.</p>	<p>1. To formulate the concluding clinical diagnosis.</p> <p>2. Basing on preliminary diagnosis, additional investigations data, conducted differential diagnosis to substantiate all elements of concluding clinical</p>	<p>Being based on modern classification of thyroid diseases to formulate the diagnosis, complications of disease and presence of concomitant diseases.</p>

		diagnosis.	
	To prescribe treatment for patients.	<p>1.To prescribe non medicinal treatment</p> <p>2.To prescribe the medicinal treatment.</p>	<p>To specify the regimen and detailed diet according to the disease.</p> <p>Taking into account age, severity of patient state, the stage of disease, the presence of complications and concomitant pathology, to prescribe modern medicinal treatment in accordance with the standards of thyroid diseases therapy.</p>

Materials of control for conclusive classes stage:

The secondary control tests

1. In the child of 4 years old the basic exchange is 28 %, a level of a cholesterol in a blood is 8,6 mmol/l, inclusion of a radioactive iodine in a thyroid gland after 6 hours is 2,1 %, after 24 hours is 3,0 %, after 48 hours is 3,5 %.

For what disease such laboratory parameters are characteristic?

- A. Diabetes
- B. Hypothyroidism
- C. Diseases of metabolism
- D. Hyperthyroidism
- E. Pituitary nanism

2. 3. In the child of 1, 5 years old the activity is reduced, does not walk, does not talk. Objectively: skin acyanotic, dry and hydropic, the tongue is big, saddle-like nose, a voice is low and rasping and hair is thick and rasping. The large fontanel is 3, 0x3, 0 sm. Teeth are not present. What diagnosis is it possible to think of?

- A. Down disease
- B. Hypothyroidism
- C. Rachitis
- D.Pituitary nanism
- E. Diabetes

3. In examination of 14 years old girl the nodal struma of III degree is found out. On scenogramm the “hot” unit revealed. Levels of T3 and T4 in a blood are increased.

What disease is it possible to think of?

- A. Diffuse toxic struma
- B Cancer of a thyroid gland

- C Toxic adenoma of a thyroid gland
- D Autoimmune thyroiditis
- E. Fibrous struma of Riddell

4. Patient G., complains of irritability, sweating, a tremor of hands, palpitation, body weight reduction in normal appetite. The thyroid gland is enlarged up to I – II degree, unpainful, elastic. The specified symptomatology most of all corresponds to:

- A. to a diffuse toxiferous struma
- B. to nervosisms
- C. to a hypothyroidism
- D. to a nodal toxic struma
- E. to a hypoparathyrosis

5. In patient G. of 15 years old in examination the enlargement of thyroid gland seen in a swallowing and infringement of eyes convergence are revealed.

What from eye signs is found out in the patient?

- A. Moebius
- B Schtelwag
- C Koher
- D Krause
- E Grefe

6. How long antithyroid therapy of a diffuse toxic struma in children in condition of achievement and preservation of euthyroidism can be conducted?

- A. during 3 months.
- B. during 6 months.
- C. during 1-1,5 years.
- D. during 1 month
- E. during 2 months

7. A 11-year-old girl has a mass in her neck. Physical examination reveals a thyroid nodule, but the rest of the gland is not palpable. A technetium scan reveals a “cold” nodule. The child appears to be euthyroid. Which of the following diagnoses is the *least* likely?

- a. Simple adenoma
- b. Follicular carcinoma
- c. Papillary carcinoma
- d. Cyst
- e. Dysgenetic thyroid gland

8. You are seeing a 4-week-old boy at a routine visit. The mother complains that he has been constipated, jaundiced, sluggish, and excessively sleepy. The physical examination is normal except for mild jaundice and a distended abdomen in a sleepy infant. The most appropriate next step is to

- a. Repeat all the baby's abnormal laboratory results (if any)
- b. Obtain x-rays of the baby's skull, wrists, and knees
- c. Begin on the baby oral sodium-L-thyroxine, 10 to 15 mg/(kg_d)
- d. Evaluate the neonate in 2 weeks for the results of symptomatic treatment
- e. Obtain a pediatric endocrinology consultation within 2 weeks

9. Boy, 13 years old, complains of irritability, sweating, tremor of hands, palpitation, decreasing of body weight in normal appetite. The thyroid gland enlarged up to II degree, unpainful, elastic. The diagnosis of diffuse toxic struma clinically fixed. What from results of examination will confirm your diagnosis?

- A. Hyperphosphatemia
- B. T3 and T4 is normal
- C. T3 and T4 is reduced
- D. Hypocalcaemia
- E. T3 and T4 is increased

10. The diagnosis of mild diffuse toxic struma established in the girl of 12 years old. What dose of Mercazolilum is necessary to administrate for child in this case?

- A. 10-15 mg per day
- B. 5-10 mg per day
- C. 20-30 mg per day
- D. 1-5 mg per day
- E. 40-50 mg per day

11. A 14-year-old girl with enlarged thyroid gland up to 3 degree from non endemic region. She states that the findings demonstrated began more than a year ago, asymptomatic. The most likely diagnosis is

- a. Iodine deficiency
- b. Congenital hypothyroidism
- c. Graves disease
- d. Exogenous ingestion of synthroid
- e. Lymphocytic (Hashimoto) thyroiditis

12. Girl of 10 months old with complaints on retardation in physical and psychomotor development. In examination the rasping face, dry skin, get hoarsen voice, enlarged tongue with the impresses of gingives, bradycardia, enlarged stomach, umbilical hernia are marked. For what disease these signs are characteristic?

- A. Rachitis.
- B. Down disease.
- C. Congenital hypothyroidism.
- D. Endemic struma.
- E. Sporadic struma.

13. The mother of 2-week-old boy complains that he has been constipated,

jaundiced, sluggish, and excessively sleepy. The physical examination is normal except for mild jaundice and a distended abdomen in a sleepy infant. The most appropriate next step is to

- a. Repeat all the baby's abnormal laboratory results (if any)
- b. Obtain x-rays of the baby's skull, wrists, and knees
- c. Begin on the baby oral sodium-L-thyroxine, 10 to 15 mg/(kg_d)
- d. Evaluate the neonate in 2 weeks for the results of symptomatic treatment
- e. Obtain a pediatric endocrinology consultation within 2 weeks

14. In the girl of 15 years old the attacks of seizures in masseters and hands with prevalence of flexors tone are observed. Seizures are painful and symmetric. In examination there are positive signs of Hvosstek and Trussot.

What is your diagnosis?

- A. Epilepsy
- B Hypoparathyroidism
- C. Hyperparathyroidism
- D. Tetanus
- E. Spasmophilia

15. Patient, 11 y.o., the strumectomy one year ago has been made, was taken with complaints on delicacy and flaccidity. Decreasing in studying progress , memory impairment were admitted. In examination the dryness of skin, fragile and dim hair, bradycardia, predilection to constipations are marked. For what disease these signs are characteristic?

- A. Hypothyroidism.
- B. Adenoma of thyroid gland.
- C. Diffuse toxic struma.
- D. Subacute thyroiditis.
- E. Fibrose struma of Riedel

Answers to the secondary control tests

1-B, 2-E, 3-C, 4-A, 5-A, 6-C, 7-E, 8-C, 9-E, 10-C, 11-E, 12-C, 13-C, 14-B, 15-A.

**Materials of the medical support for the students' self training:
a reference chart for organization of students' independent work with
educational literature.**

Tasks	Instructions
To study the etiology and pathogenesis of diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse nontoxic goiter, thyroid cancer in children. Be able to detect the degrees of goiter.	To enumerate basic etiologic factors, to select the key links of thyroid gland disease pathogenesis.
To study clinical manifestations of	To establish the symptoms and to gather

diffuse toxic goiter, hypothyroidism, autoimmune thyroiditis, endemic goiter, acute and subacute thyroiditis, diffuse untoxic goiter, thyroid cancer in children.	it in the clinical syndromes to put the probable diagnosis of thyroid gland disease.
To study diagnostic criteria of thyroid gland diseases	To make the flow diagram of disease
To study the additional methods of examination (laboratory, instrumental)	To work out a plan of patient examination.
To study the changes in additional investigational methods which are pathognomonic for thyroid diseases.	To enumerate the basic diagnostic criteria of thyroid gland diseases according to the data of additional investigational methods.
To conduct differential diagnostics, to establish concluding diagnosis	To substantiate the basic components of diagnosis in accordance with the modern classification, and to conduct a differential diagnosis.
To prescribe the individual treatment to patient with the thyroid gland disease. To be able to render the first aid in thyroidotoxic crisis for children.	To make the prescribing chart specifying the regimen, diet, medicinal treatment, taking into account the age, severity of patient's state, the stage of disease, the presence of complications and concomitant diseases.

THE RECOMMENDED LITERATURE

Basic:

1. Nelson Essentials of Pediatrics, fifth edition, Copyright © 2007
/ edited by Richard E. Behrman, Robert M. Kliegman, Ann M. Arvin;
senior editor, Waldo E.
2. Rudolph's Pediatrics, 21st Edition.

Additional:

1. Майданник В.Г. Педиатрия. Учебник (2-е издание, испр. и доп.)- Харьков: Фолио, 2002. - 1125 с.
2. Волосовец А.П., Кривопустов С.П., Криворук І.М., Черній О.Ф. Навчальний посібник з дитячої ендокринології. - Тернопіль: Укрмедкнига, 2004. -495 с
3. Наказ МОЗ України від 27.04.2006 № 254 Про затвердження протоколів надання медичної допомоги дітям за спеціальністю "Дитяча ендокринологія"

THE DISEASES OF HYPOTHALAMIC PITUITARY SYSTEM AND SEXUAL GLANDS IN CHILDREN.

Etiology, pathogenesis, classification, clinic, diagnostics, differential diagnostics, treatment, prophylaxis of different clinical forms of growth disorders (exogenous constitutional, pituitary, somatogenic); obesity (exogenous constitutional, subthalamic), pubertal dispituitarism, in children, different forms of sexual glands, disorders, pathology of sexual glands for children (disturbances of boys and girls sexual development). Prognosis.

I. Actuality of the theme.

Hypothalamic – pituitary system is one of the major links in adjusting and control of endocrine glands. The disorders hypothalamic – pituitary glands are the cause of pathological processes origin in organism and the development of many endocrine diseases. It predetermines the necessity of studying and improvement of knowledge of these problems for precise diagnostics and institution of adequate therapy.

II. Classes (studies pointing with mastering level planned)

1. 1. An of student must know (to familiarize with): α 1
 - About the diseases of hypothalamic – pituitary system in the structure of endocrine diseases in children, prevalence in different age groups;
 - About statistical information in relation to morbidity, frequency of complications origin, nearest and remote prognosis of patients.
 - About history of problem scientific studying and contribution of domestic scientists;
2. A student must know (master): α 2
 - Anatomic physiological features endocrine system of healthy children endocrine system; the features of metabolism;
 - Structure and functions to the hypothalamus and hypophysis; hormones, mechanism of their action; regulation of hemadens functions.
 - To familiarize with the modern state of problems for diagnostics and treatment of the hypothalamic – pituitary system and obesity.
 - Etiology, pathogenesis and clinical displays of hypothalamic – pituitary system diseases and different forms of obesity in children.
 - Methods of treatment of different clinical forms of obesity, growth disorders, diabetes insipidus, pubertal dispituitarism, disorders of sexual development.
 - Urgent condition in pathology of hypothalamic – pituitary system and different forms of obesity in children. Pathogenesis clinic and treatment methods of prophylaxis in hypothalamic – pituitary system and obesity; organization of outpatient clinical observation..

- Differential diagnosis.

3. A student must master: α3

Skills:

- Collection of complaints and anamnesis of disease;
- Examination of patients with the diseases of hypothalamic – pituitary systems and revealing of basic symptoms and syndromes;
- Formulate and substantiate the preliminary diagnosis.
- Determination of laboratory and instrumental plan of patient examination (according to diagnostics standards);

Abilities:

- to interpret the result of laboratory and instrumental tests.
- To conduct differential diagnosis
- Using the sygmal and centile tables to detect the indexes of physical development, the degree of delaying and acceleration, weight excess and deficiency in children.
- To detect the bone age of children.
- To give recommendations in relation to the patient regimen and diet in diseases of hypothalamic – pituitary systems
- taking into account the stage of disease to specify the severity of the state and concomitant pathology;
- to complete the treatment plan in leukemias and lymphadenomas according to standards taking into account the stage of disease, complications and concomitant pathology.
- To render first aid in extreme situations and exigent states.

III. Aims of personality development (educative aims):

- A student must learn to adhere to the rules of behavior and principles of medical etiquette and deontology to develop bedside manner in patients with leukemias and lymphadenomas ;
- to lay hands on ability to set a psychological contact with a patient and his family;
- to master the sense of professional responsibility for a timely and adequate medicare.

IV. Interdisciplinary integration:

Subject	To know	To be able
1. Previous (providing)		
anatomy	Anatomic-physiologic features of hypothalamic – pituitary system of healthy children.	
Histology	Structure and functions of hypothalamus and	

	hypophysis.	
Physiology	Hormonal regulations of of hemadens functions	To asses the data of laboratory and instrumental investigational methods
Pathologic physiology	Pathogenesis of of hypothalamic – pituitary system diseases.	
Pathologic anatomy	Morphological features of development of diseases of the hypothalamic – pituitary system are depending on the stage of process	To analyse and interpret the information about clinical examination and about additional methods of investigation
Pharmacology	Pharmacocinetics and pharmacodynamics, the side effects of preparations (hormonal drugs, metabolic preparations, etc.), are using in treatment of patients with the hypothalamic – pituitary system diseases.	To prescribe age dependent and patient individual features treatment, period of disease, to establish the individual regimen of preparations taking and dosage. To prescribe recipes.
Internal diseases propedeutics.	Basic stages and methods of patient clinical examination	To collect complaints, anamnesis vitae et morbi, to find out the basic risk factors, to coduct patient examination, to reveal the clinical signs of pituitary hypothalamic diseases, to interpret the data about additional methods of investigation.
To detect the hormones of hypothalamic –pituitary-suprarenal axes.	Normal ranges of hormones of hypothalamic – pituitary –suprarenal axes.	To interpret the data of hormonal investigations.
3. Interdiscipline integration		
Primary obesity (exogenous – constitutional).	Clinical manifestation of exogenous – constitutional obesity.	To establish the specific signs of exogenous – constitutional obesity, to conduct differential diagnosis among hypothalamic syndrome of pubertal age.
Terner syndrome.	Clinical signs of Terner syndrome.	To establish the clinical signs of Terner syndrome and conduct the differential

		diagnosis with dwarfism.
Hypohonadism,	The clinical signs of hyponadism.	To determine the clinical signs of hypohonadism and conduct the differential diagnostics among different forms of sexual development delaying.

Methodical materials to support basic stage

professional algorithm of patients management implementation (reference chart) for the practical skills and abilities forming.

№	Task	Sequence of implementation	Remarks and warnings related to self-control
1	To conduct of patient examination with obesity, sexual development disorders, diabetes insipidus, growth disorders.	<p>1.To conduct the complaints and disease's anamnesis gathering.</p> <p>2.Carefully gathering the life anamnesis of patient.</p> <p>3.To conduct examination of the patient.</p> <p>.</p> <p>.</p> <p>4.To investigate the state of hypodermic fatty layer, estimate anthropometric information, sexual development of child.</p>	<p>To pay attention to features of disease course , underlying factors, concomitant diseases etc.</p> <p>Pay attention for features of disease course , underlying factors, concomitant diseases etc.</p> <p>to establish the availability of risk factors which facilitate the disease occurrence.</p> <p>To assess patient general condition, position in the bed, color and wetness of skin and mucous, presence of neck veins and extremities swelling.</p> <p>To pay a regard to rhythm of pulse, it tension and size on both hands, apex shove, it properties, margins of absolute and relative cardiac dullness, it changes, HR(tachi-or bradycardia, extrasystoly),BP.</p>
		<p>5.To conduct of heart and of main vessels auscultation.</p> <p>6..To investigate the pulmonary system (percussion, bronchophony).</p> <p>7.To conduct lungs auscultation.</p> <p>8.To investigate the system of digestion.</p>	<p>To pay regard to heart tones weakening or amplifying, appearance of murmurs and additional III, IV tones.</p> <p>To pay attention to features of percussion and auscultation in children of different age.</p>

2	To formulate the preliminary diagnosis.	1.To formulate the preliminary diagnosis. 2.To substantiate all components of preliminary diagnosis based on complaints, anamnesis, and examinations.	Based on modern classification to formulate the preliminary diagnosis of hypothalamic-pulmonary system disease and to substantiate each component of it.
3	To evaluate the parameters of additional laboratory tests..	1.To evaluate the blood count data, to determ the bony age, the body mass index, indexes of masculinisation and feminization. 2.To interpret the additional investigations data. .	To pay attention to the presence of changes of anthropometric information, information of harmoniousness of physical and sexual development. To pay a regard to presence of delayed in sexual development from the age from age-old ranges, information about lipidogrammes, to maintenance of cholesterol, biochemical indexes.
4	To understand the data of additional and laboratory investigation.	To understand the chest X-Ray data, the data of ECG, and of ultrasound.	To turn the special attention on the signs of changes in cella turcica configuration, information of X-ray of the skull and the long bones, the presence of additional formations in the area of cella turcica, information about specific gravity of urine, and others like that, changes of ECG
5.	To conduct differential diagnosis.	1.Consistently to find the common signs in complaints,life and disease anamnesis, data of examination, data of laboratory and instrumental tests in patient and in similar states. 2.To find differences among complaints, information of life and disease anamnesis, examination data, information about the laboratory and instrumental methods of research and in similar nosology. 3.On the basis of found out differences to exclude similar diseases from the list of probable diagnoses. 4.To conduct differential diagnostics according to the above mentioned algorithm with all of nosologies which have an alike clinical picture with a patient, including with the signs of hypothalamus-pituitary diseases. 5.Taking into account the impossibility to exclude the	The special attention need to be spared to differential diagnostics with exogenous-constitutional obesity, diabetes, inherited syndromes are accompanied by the growth delaying.

		diagnosis of leukemia from the list of probable diagnoses to draw a conclusion about most probability of such diagnosis.	
6	To formulate the final clinical diagnosis.	1.To formulate the final clinical diagnosis. 2.Based on initial diagnosis, additional investigations data, conducted differential diagnosis to substantiate all elements of concluding clinical diagnosis.	Being based on modern classification of obesity, diabetes, growth disorders, disorders of sexual development, to formulate a previous diagnosis, complications of basic disease and presence of concomitant diseases.
7.	To prescribe treatment for patients.	1.To prescribe not medicinal treatment 2.To prescribe the medicinal treatment.	Expressly to specify the regimen and detailed diet according to a disease. Taking into account age, severity of patient state, the stage of disease, the presence of complications and concomitant pathology, to prescribe modern medicinal treatment in accordance with standards of obesity, diabetes, growth disorders, disorders of sexual development treatment.

THE DISEASES OF HYPOTHALAMIC PITUITARY SYSTEM AND SEXUAL GLANDS IN CHILDREN.

Tests – 1 V 1

- 1.The clinical signs of diabetes insipidus could be all listed below except for
 - A) disuria;
 - B) thirst;
 - C) poliuria
 - D) dryness of skin and mucoses;
 - E) diminishing of appetite.
- 2.The most reliable sign of diabetes insipidus will be:
 - A) hypotonic poliuria ;
 - B) decreasing of blood plasma osmolarity.
 - C) decreasing of cortisol level in a blood;
 - D) increasing of aldosteron level in a blood;
 - E) increasing of glucose level in the urine
3. The main signs of central diabetes or diabetes insipidus are all, except for:
 - A) increasing of antidiuretic hormone level in a blood;
 - B) decreasing of urine density;
 - C) absence of kidneys disease;
 - D) increasing of plasma osmolarity ;
 - E) thirst, poliuria.
- 4.Glucosuria inherent by all diseases listed below, except for:
 - A) Diabetes mellitus;
 - B) Fanconi nephrophtysis;

- C) to the hepatic illness;
 - D) idiopathic family renal glucosuria;
 - E) protracted starvation.
5. The most reliable sign of diabetes insipidus will be all, except for:
- A) hypotonic polyuria ;
 - B) decreasing of blood plasma osmolarity.
 - C) decreasing of cortisol level in a blood;
 - D) decreasing of aldosterone level in a blood;
 - E) increasing of urea level in the blood.
6. The main signs of central diabetes or diabetes insipidus are all, except for:
- A) increasing of antidiuretic hormone level in a blood;
 - B) decreasing of urine density;
 - C) absence of kidney disease;
 - D) increasing of plasma osmolarity ;
 - E) thirst, polyuria.
7. Glucosuria inherent by all diseases listed below, except for:
- A) Diabetes insipidus;
 - B) Fanconi nephropathy;
 - C) to the hepatic illness;
 - D) idiopathic family renal glucosuria;
 - E) protracted starvation.

THE DISEASES OF HYPOTHALAMIC PITUITARY SYSTEM AND SEXUAL GLANDS IN CHILDREN.

Tests – 1 V 2

8. In the regulation of ADH secretion the main factor will be:
- A) osmolarity of plasma;
 - B) level of glucose in a blood;
 - C) level of electrolytes in a blood;
 - D) level pH in a blood;
 - E) level of urea in a blood.
9. The inherited diabetes insipidus is more frequently diagnosed at children of:
- A) first year;
 - B) pubertal age;
 - C) in a period of babyhood;
 - D) in prepubertal period;
 - E) in a period of senior age.
10. The causes of the acquired diabetes insipidus must be all, except for:
- A) mutations of ADH receptor gene;
 - B) primary pyelonephritis;
 - C) secondary pyelonephritis;
 - D) amyloidosis;
 - E) hypercalcemia, hypokalemia.
11. For treatment of nephrogenic diabetes insipidus are used:
- A) thiazide preparations;
 - B) osmotic diuretics;
 - C) preparations of ADH;
 - D) preparations of potassium;

E) antiinflammatory preparations.

12. Nephrogenic diabetes insipidus is characterized by the all, except for:

- A) hyperosmolarity of urine;
- B) elevated or normal level of ADH in a blood;
- C) hypostenury;
- D) not effective treatment with adiurecrin
- E) normal parameters of glomular filtration and tubular reabsorbtion

13. The causes of acquired central diabetes insipidus are the all, except for:

- A) genetic defect of ADH transport synthesis;
- B) craniocerebral trauma;
- C) neuroinfection (encephalitis, meningitis);
- D) operations in the area of hypophysis;
- E) tumors (craniopharigeomas, meningeomas).

14. The basic factors of the poluria are:

- A) all reasons;
- B) central diabetes incipidus;
- C) nephrogenic diabetes insipidus;
- D) psychogenic polidesum;
- E) dipsogenic diabetes incipidus.

Answers: 1-a,2-a,3-a,4-a,5-a,6-a, 7-a, 8-a,9-a,,10-a, 11-a, 12-a, 13-a,14-a.

V. The questions for the control of secondary knowledge level of abilities and skills:

VII.1 Materials of control for the preparatory stage of class.

A questions for control of initial level of knowledge of skills and abilities:

1. Hormones of hypothalamus and hypophysis, mechanism of action.
2. Syndromes of growth disorders (after the method of sygmal deviations).
3. Diagnostics of growth acceleration signs.
4. The definition of bony age and it detecting.
5. The causes of origin and clinical signs of hypophysial dwarfism.
6. Differential diagnosis of pituitary nanism.
7. What functional tests uses for the detecting of pituitary somathotropin reserves?
8. The principles of pituitary nanism treatment ?
9. The causes of origin, clinical and laboratory criteria for the diagnosis of gigantism and acromegaly.
10. Name the symptoms are characteristic for progress of hypophysis adenoma growth.
11. Partial gigantism in children. The principles of treatment.
12. The causes of origin and clinical signs of pubertal dispithuitarism.
13. What clinical signs reveals in adiposogenital dystrophy?
14. Classification of obesity. Degrees of obesity.
15. Clinical signs of exogenous - constirtutional and subthalamic obesity.
16. The principles of different clinical forms therapy in obesity in children.
17. The causes and clinical signs of diabetes insipidus in children.

18. Laboratory and instrumental criteria for diagnostics of diabetes insipidus. Functional tests in children.
19. The treatment of diabetes insipidus in children.
20. Hormones of sexual glands and the hypothalamic-pituitary system, mechanism of action, regulation of secretion, principles of feedback links.
21. The definition of genetic and gonad sex. Passport sex.
22. The definition of intersexualism.
23. Physiology of sexual maturation. (formation of gonads, of internal and external genitalia (formation of gonads, internal and external genitalia, secondary sexual signs.)
24. The criteria for girls sex maturation.
25. The criteria for boys sex maturation.
26. Factors are impaired the process of sex maturation.
27. The pathogenesis of sexual apparatus congenital anomalies
28. The classification of sexual anomalies congenital anomalies.
29. The diagnosis of sexual maturation disorders.
30. True agenesis and dysgenesis of gonads. Diagnosis and treatment.
31. Turner syndrome. Causes of origin, manifestation, diagnosis, treatment.
32. Dysgenesis of testicles and ovaries. Causes of origin. Diagnosis, treatment.
33. The syndrome of incomplete masculinisation. Etiology. Clinical forms. Treatment.
34. Syndrome of testicular feminization. Etiology. Clinical signs, diagnosis, treatment.
35. Klinefelter syndrome. Etiology. Manifestation. Diagnosis. Treatment.
36. The main principles of intersexualism treatment.
37. Organization of outpatient observation in children with sexual maturation disorders.

THE DISEASES OF HYPOTHALAMIC PITUITARY SYSTEM AND SEXUAL GLANDS IN CHILDREN.

Test -2 V1

1. What is not typical for exogenous - constitutional type of primary obesity?
 - A) Changes of a skin with a pigmentation, folliculitis, striae on hips and breeches;
 - B) Early terms of superfluous body mass occurrence;
 - C) The subcutaneously - fatty layer is distributed in regular intervals;
 - D) Slow, gradual progress of obesity;
 - E) Family predilection to obesity.
2. Attributes of an Itsenko-Cushing syndrome are all signs, except for:
 - A) Premature sexual development;
 - B) Lunar face;
 - C) Osteoporosis;
 - D) Premature sexual pilosity;
 - E) Distribution of subcutaneously - fatty layer non-uniformly, in the top part of a body.

3. What percent of superfluous body mass is characteristic for III degree of obesity?
 - A) 50-100 %;
 - B) 5-10 %;
 - C) 10-15 %;
 - D) 25-50 %;
 - E) 15-30 %

4. Typical attributes of Lawrence -Moon-Barde-Bidl syndromt are everything, except for:
 - A) Cataract;
 - B) Pigmentary retinopathy;
 - C) Uniform obesity;
 - D) Oligophrenia;
 - E) Polysyndactylia, congenital anomalies of a skeleton.

5. The laboratory data in Itsenko – Cushing syndrome will have the following signs, except for:
 - A) Decreasing of hydrocortisone derivates in urine
 - B) Increasing of cortisone concentration in a blood
 - C) Rising in a blood of a cholesterol level;
 - D) Rising in a blood of sodium and chlorines levels
 - E) Rising in a blood of a glycemia.

6. What is not typical for a clinical signs of Itsenko - Cushing syndrome?
 - A) Premature ossification of bones;
 - B) Obesity;
 - C) Lunar face;
 - D) Osteoporosis;
 - E) Arterial hypertension.

7. In treatment of an initial obesity all is used, except for
 - A) Increased exercise stresses;
 - B) Balneotherapy;
 - C) Dieteticses;
 - D) Hydrotherapy;
 - E).Fangothrapy

8. What disease is characterized of obesity, retardation in mental and physical development, hypotonia since early years and cryptorchism?
 - A) Prader – Willy syndrome
 - B) Down.syndrome
 - C) Itsenko – Cushing syndrome.
 - D) Lawrence - Moon sendrome.
 - E) Pubertal subthalamic.syndrome

9. What disease is characterized of obesity, retardation in mental and sexual development, polydactyilia and pigmentary retinitis?
- A) Lawrence - Moon syndrome.
 - B) Down syndrome
 - C)) Itsenko – Cushing syndrome
 - D)) Prader – Willy syndrome
 - E) Pubertal subthalamic.syndrome
10. What are the relative contraindications for treatment of obesity?
- A) Tuberculosis;
 - B) Chronic renal failure;
 - C) Psychoneuroses;
 - D) HIV-infection;
 - E) All listed above

THE DISEASES OF HYPOTHALAMIC PITUITARY SYSTEM AND SEXUAL GLANDS IN CHILDREN.

Test -2 V2

11. What wide-spread disturbances of a metabolism could find in patients with obesity?
- A) Hyperinsulinemia, hypercholesterolemia.
 - B) Hypoinsulinemia , hypocholesterolemia.
 - C) Hyperinsulinemia, hypocholesterolemia
 - D) A normal level of blood glucose and hypercholesterolemia.
 - E) A normal level of cholesterol, β – lipoproteids and disproteinemia.
12. Name the preparations promotes of appetite diminishing
- A) Phepranonum;.
 - B) Thyreoidinum;
 - C) Furosemidum;
 - D) ATP;
 - E) Aloe.
13. What diseases are more wide-spread among the patients suffering of obesity?
- A) Hypertension.
 - B) Diabetes.
 - C) Osteoarthrosis.
 - D) Coronary failure.
 - E) All listed above
14. What malignant neoplasms more wide-spread among the patients suffering of obesity?
- A) Cancer of gold bladder and cholic ducts.
 - B) Lungs cancer

- C) A cancer of prostate, direct and colonic intestines in men.
- D) A cancer of endometrium and mamma in women.
- E) All listed above

15. For 2 degree of obesity excess of weight makes

- A) 30-50 %;
- B) 50 %;
- C) 10-15 %;
- D) 15-20 %;
- E) 20-25 %.

16. Risk factors of paratrophy development are everything, except for:

- A) Nutritional;
- B) Constitutional;
- C) A hypokinesia;
- D) Endocrine diseases of mother;
- E) A long antibiotic therapy.

17. Calculation of nutrition in paratrophy is conducted on:

- A) Approximately - appropriate weight;
- B) Appropriate weight;
- C) Actual weight;
- D) Weight-height index.

18. For what endocrine disease the increasing of body weight is not typical ?

- A) Itsenko - Cushing syndrome
- B) Hypothyroidism.
- C) Hypogonadism.
- D) Hyperdysinsulinism.
- E) Typically for all listed,

19. For 1 degree of obesity excess of body weight makes:

- A) 14-25 %;
- B) 5-10 %;
- C) 15-30 %;
- D) 10-15 %;
- E) 10-23 %.

20. An otherwise healthy 7-year-old girl is brought to your office by her father because she has some acne, breast development, and fine pubic hair. The most likely etiology for her condition is

- a. A feminizing ovarian tumor
- b. A gonadotropin-producing tumor
- c. A lesion of the central nervous system
- d. Exogenous estrogens
- e. Early onset of "normal" puberty (constitutional)

Answers: 1-a,2-a,3-a,4-a,5-a,6-a,7-e,8-a, 9-a,10-b,11-a,12-a,13-e,14-a,15-a,16-e, 17 – a, 18-e, 19-c, 20-c.

Tasks:

1. The parents of a 14-year-old boy are concerned about his short stature and lack of sexual development. By history, you learn that his birth weight and length were 3 kg and 50 cm, respectively, and that he had a normal growth pattern, although he was always shorter than children his age. The physical examination is normal. His upper-to-lower segment ratio is 0.98. A small amount of fine axillary and pubic hair is present. There is no scrotal pigmentation; his testes measure 4.0 cm³ and his penis is 6 cm in length. What is the probable diagnosis. In this situation you should.

Answers: 1. Delaying in sexual development, constitutional type.
2. Measure pituitary gonadotropin

2. The parents of a 14-year-old boy are concerned about his short stature and lack of sexual development. By history, you learn that his birth weight and length were 3 kg and 50 cm, respectively, and that he had a normal growth pattern, although he was always shorter than children his age. The physical examination is normal. His upper-to-lower segment ratio is 0.98. A small amount of fine axillary and pubic hair is present. There is no scrotal pigmentation; his testes measure 4.0 cm³ and his penis is 6 cm in length. Compose the plan of laboratory investigations. Compose the plan additional investigation.

1. Hormonal profile (FSH, GSH, testosterone), blood count, urine tests, biochemical tests.

2. ECG, cranial X-ray, ECHO-EG.

3. A 10-year-old obese boy has central fat distribution, arrested growth, hypertension, plethora, purple striae, and osteoporosis. Which of the following disorders is most likely to be responsible for the clinical picture that this boy presents? What tests are confirmed the diagnosis?

1. Craniopharyngioma

2. Cella turcica tomography, CT, ECHO-EG, cortisol, ACTH, testosterone levels, MRI of adrenals,

4. A 10-year-old obese boy has central fat distribution, arrested growth, hypertension, plethora, purple striae, and osteoporosis. What conditions must be included in differential diagnosis of disease? What therapeutic approach must be applied in this case.

1. Adrenal glands hyperplasia, adrenal adenoma, adrenal carcinoma.

2. Neurosurgery.

5. An otherwise healthy 7-year-old child is brought to you to be evaluated because he is the shortest child in his class. Careful measurements of his upper and lower body segments demonstrate normal body proportions for his age. Which of the following disorders of growth is likely? What investigations must be prescribed

for diagnosis confirming?

1. Pituitary nanism
2. Cranial X-ray, pituitary hormones tests.

6.A 4-year-old child has mental retardation, shortness of stature, brachydactyly (especially of the fourth and fifth digits), and obesity with round facies and short neck. The child is followed by an ophthalmologist for subcapsular cataracts, and has previously been noted to have cutaneous, subcutaneous, and perivascular calcifications of the basal ganglia. What diagnosis is more likely? This patient is likely to have which of the following features?

1. Lawrence-Moon syndrome.
2. Elevated concentrations of parathyroid hormone

7. Child, 14 years old, complaints of fast body weight increasing during the last 3 years accompanied with appetite increasing, thirst, fatigue. The diagnosis of II degree obesity with alimentary genesis established.

What are the main features of dietary treatment? What is the plan of investigation?

1. Restriction of culinary salt entering.
2. Blood count, cholesterol level, ECG, lipidogramme, cranial X-ray, ECHO-EG, fast glucose, glucose tolerance test.

8. In 13 year old child with III degree of obesity in the glucose tolerance test obtained follows data: glucose on empty stomach is 5,4 mmol/l, after 1 hours of carbohydrates loading is 10 mmol/l, after 2 hours of carbohydrates loading is 7,8 mmol/l. What condition being established? What measures are necessary for carbohydrates metabolism normalization?

1. Impaired tolerance to carbohydrates.
2. To administrate a diet, to encourage active movements aimed to normalize body weight

9.Patient C., 12 years old. The obesity, fatigue, sleepiness, headache disturbs.

Objectively: body height of 171 cm, weight of 106 kg, and the adiposity is mainly on arms and trunk.

A skin dry with a crimson - mottled shade. On arms, breast and hips there are crimson - cyanotic strips of a stretching. Pulse is 76 per min., blood. pressure is 160 / 102 mm Hg.

What is the more likely diagnosis?

What kind of excess is the main cause of hypertension in the patient?

1. Cushing disease
2. ACTH, epinephrine, hydrocortisone, aldosteronum, androstendion

10. In the girl of 15 years old the obesity, mainly on brachiums and trunk, and hirsutism, disturbances of a menses is observed. On brachiums, breast and on both sides of a stomach and on her hips there are crimson - cyanochroic strips of skin stretching

What diagnosis is the most authentic?

What investigations could confirm the diagnosis?

Cushing disease.

ACTH, cortisol, cranial X-ray.

VII. Materials of the medical support for the students independent preparation: a reference chart for organization of students independent work with educational literature.

Tasks	Instructions
To study the etiology and pathogenesis of hypothalamus- pituitary diseases.Be able to detect the risk group for the obesity, diabetes insipidus, growth disorders, sexual maturation disorders..	To enumerate basic etiologic factors of hypothalamus- pituitary diseases, select the key links of hypothalamus- pituitary diseases pathogenesis.
To study clinical manifestations of hypothalamus- pituitary diseases pathogenesis in children.	To establish the symptoms and gather it to clinical syndromes are enable to put the credible diagnosis of diabetes insipidus, obesity, growth disorders and sexual maturation disorders.
To study diagnostic criteria of hypothalamus- pituitary diseases.	To make the flow diagram of disease
To study the additional methods of research (laboratory, instrumental)	To work out a plan of patient investigation.
To study the changes in additional investigational methods are pathognomonic for hypothalamus- pituitary diseases.	To enumerate the basic diagnostic criteria of l hypothalamus- pituitary diseases. according to the data of additional investigational methods.
To conduct differential diagnostics, s to establish a concluding diagnosis	To substantiate the basic components of diagnosis in accordance to modern classification, and to conduct a differential diagnosis.
To prescribe the individual holiatry to patient with the diabetes insipidus, obesity, growth disorders and sexual maturation disorders. Able to render the first aid in hypothalamus- pituitary diseases.	To make the prescribing chart specifying the regimen, diet, medicinal treatment, taking into account the age, severity of patient state, stage of disease, presence of complications and concomitant diseases.
To study the etiology and pathogenesis of diabetes insipidus, obesity, growth disorders and sexual maturation disorders.in children.	To enumerate basic etiologic factors, select the key links of diabetes insipidus, obesity, growth disorders and sexual maturation disorders pathogenesis.

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DISEASES OF BLOOD AND ENDOCRINE SYSTEM IN CHILDREN

Tests to prepare for the module control

1. The acromegalia is diagnosed for the patient of 13 years old. What other signs are characteristic for this disease?

- A. Giantism.
- B. Obesity.
- C. Infantilism.
- D. Premature sexual development.
- E. All answers are true.

2. The girl of 8 y.o. is in the clinic because of growth inhibition. From the anamnesis reported that child is from socially unsuccessful family. In examination the STH level of 22 IU per ml revealed. After conducting the loading test level of hormone has raised up to 30 IU per ml.. What is your diagnosis?

- A Pituitary nanism.
- B Psychological nanism
- C Laron syndrome
- D Somatogenic nanism
- E Thyroid nanism

3. In conducting of ACTH test (with Synacthenum) in the patient with primary chronic adrenal failure the level of 17-KS in urine must be follows:

- A. Unchanged.
- B.Increased in 50 %.
- C.Increased in 100 %.
- D.Reduced in 50 %.
- E.Decresed in 100 %.

4. The boy of 3 months old was taken to the hospital because of delayed icterus and persistent constipations. He is sick from birthday. Mother's pregnancy has been complicated with a hestosis. In examination is poorly active, the hydroptic face, macroglossy, icteric skin. Narrow palpebral fissures.Muscle tone is reduced. Bradycardia. What is the most probable diagnosis?

- A. Rachitis
- B. Down diseaese
- C. Congenital hypothyroidism
- D. Intestinal form of mucoviscidosis
- E. Hirshprung disease

5. The boy of 3 months old was taken to the hospital because of delayed icterus and persistent constipations. He is sick from birthday. Mother's pregnancy has been complicated with a hestosis. In examination is poorly active, the hydroptic face, macroglossy, icteric skin. Narrow palpebral fissures.Muscle tone is reduced. Bradycardia. What is the most probable diagnosis?

- A. Rachitis
- B. Down disease
- C. Congenital hypothyroidism
- D. Intestinal form of mucoviscidosis
- E. Hirshprung disease

6. Girl, aged 14, about one year ago the irritability and tearfulness has appeared. Then, the diffusively enlarged thyroid gland of II degree was detected. This state was considered as pubertal age manifestation.. Treatment was not conducted. Irritability was gradually changed by complete apathy. Bradycardia, constipations, bloated face, pastose skin, have appeared, .The paleness of a skin with waxy shade, inspissation of thyroid gland were increased. What disease must be assumed?

- A. Cancer of a thyroid gland
- B. Diffuse toxic struma
- C. Autoimmune thyroiditis
- D. Subacute thyroiditis
- E. Hypothyroidism .

7. Patient, 13 y.o., the strumectomy one year ago has been made, was taken with complaints on delicacy and flaccidity. Decreasing in studying progress, memory impairment were admitted. In examination the dryness of skin, fragile and dim hair, bradycardia, predilection to constipations are marked. For what disease these signs are characteristic?

- A. Hypothyroidism.
- B. Adenoma of thyroid gland.
- C. Diffuse toxic struma.
- D. Subacute thyroiditis.
- E. Fibrose goiter of Riedel.

8. Child of 7 months old with complaints on retardation in physical and psychomotor development. In examination the rasping face, dry skin, get hoarsed voice, enlarged tongue with the impresses of gingives, bradycardia, enlarged stomach, umbilical hernia are marked. For what disease these signs are characteristic?

- F. Rachitis.
- G. Down disease.
- H. Congenital hypothyroidism.
- I. Endemic struma.
- J. Sporadic struma.

9. In the patient of 13 years old the relapse of a nephrolithiasis, ostealgia, delicacy, loss of body weight, increased fatiguability are observed. What from the listed diseases can be suspected?

- A. Sarcoma of bone.
- B. Hyperparathyroidism.
- S. Hypoparathyroidism z.
- D. Pedge disease I

E. Multiple myeloma.

10. Child, in 9 months old for the first time the congenital hypothyroidism clinically and according to thyroid hormones tests was confirmed, . Now the most expressive manifestation is the serious lag in psychophysical development. What in this case the most rational therapeutic tactics?

- A. Mercazolilum
- B. Thyreoidinum
- C. Ttriodthyroninum
- D. L-thyroxine + Pyracetamum
- E. L-thyroxine + Retabolilum

11. In the patient of 10 years old the delicacy, fatigability, decreasing of progress in school, dry and cold skin, fragility of hair and nails are marked. During the further investigation the hypothyroidism was diagnosed. What therapy is necessary to prescribe for this patient?

- F. Mercazolilum.
- G. Prednisolonum.
- H. Thyroxine.
- I. DOCSA.
- J. Hidrocortisonum.

12. Girl of 13 years old complains on increased body weight during last year. Father is predilected to obesity and suffers of diabetes. Girl's body weight exceeds norm for the age by 50 %. Allocation of fat is irregular. Skin dry. Set of striae. Arterial blood pressure is 125/75 mm Hg. In the blood rising level of ACTH and corticosteroids are revealed. Ultrasonic of paranephroses has revealed their uniform enlargement. On skull bones X-ray the signs of osteoporosis. What is your diagnosis?

- A. Cushing disease
- B. Cushing syndrome
- C. The exogenous obesity
- D. Subthalamic obesity
- E. Pubertal basophilism of young age

13. In the child of 10 months old, is feeding with the cow milk constantly, the serious systolic apex murmur, moderate cardiomegaly, pastose skin, Crocq's disease are marked. In ultrasound the heart disease is not revealed. Laboratory data: Hb 38 g/l, C.I.- 0,7, total albumin - 50 g/l, serumal iron 2 mcM /l. What therapy first of all must be prescribed?

- A. Fractional transfusion of packed red cells.
- B. Urgent blood transfusion .
- C. Enteral introduction of iron preparations.
- D. Cardiac glycosides by the fast saturation regimen.
- E. Transfusion of colloids (Albuminum).

14. Girl, aged 12, sick of a diabetes, after sportive lesson in school the acute

feeling of exaltation, famine, nausea, headache, tremor of extremities, doubling in eyes have appeared. In examination: respiration is superficial, profuse sweat, tachycardia, arterial pressure is 90/60 mm.Hg, positive Babinski symptom. The girl has suddenly lost her consciousness, there was an attack of tonic and clonic cramps.

What is your diagnosis?

- A. Vagoinsular crisis.
- B. Hyperglycemic coma.
- C. Vascular collapse.
- D. Asthenic syndrome.
- E. Hypoglycemic coma

15. Girl aged 4, during the month complaints on decreased appetite, thirst, polyuria. Within last day a soporous state appears. What investigation must be conducted first of all?

- A. Detecting of glucose.
- B. Detecting of creatinine.
- C. Lumbar puncture.
- D. X-ray of skulls.
- E. Ultrasonic examination of kidneys.

16. Boy aged 4, has arrived to hospital in a serious state without consciousness. Fatigue and vomiting gradually accrued. Toxic respiration, moderate tachycardia, skin is dry, turgor is diminished, tongue is dry and rough, smell of acetone from a mouth. Blood's glucose level is 26,4 mmol/l, of sodium is 120 mmol/l, pH - 7,1, glucosuria and ketonuria. What is the diagnosis?

- A. Acetonemic vomiting.
- B. Hypovolemic shock.
- C. Hypoglycemic coma.
- D. Ketoacidotic coma.
- E. Meningocephalitis.

17. Girl, aged 6, was hospitalized in clinic without consciousness. Sick of diabetes during one year. In examination: the skin is dry and pale; mucous of lips is in bright red color; pupils narrowed, sunken eyes; respiration is noisy, tachypnea; the smell of acetone; cardiac sounds are muffled, tachycardia; arterial pressure is reduced, stomach inflated. A level of glucose in the blood is 22,4 mmol/l. What dose of insulin should be entered for deducing the child from coma?

- A. 0,1 U/kg
- B. 0,05 U/kg
- C. 0,2 U/kg
- D. 0,25 U/kg
- E. 0,15 U/kg

18. Boy aged 14, was hospitalized in children's hospital with complaints on thirst, raised appetite and accelerated emiction. On the eve the vomiting and abdominal pain were marked. It is known, that 10 days ago has transferred a flu. In examination the consciousness is confused; pupils narrowed, weak reaction to light;

skin is pale, dry and cold by touch, hyperemia on the cheeks; smell of acetone in the air; respiration noisy, cardiac tones are muffled, tachycardia, tachypnea; stomach inflated. What test must be conducted first of all?

- A. Detection of glucose and ketonic particles in the urine
- B Conducting the tests of glycemic and glucosuric profiles
- C Detecting of blood osmolarity
- D Detecting of basis excess
- E Detection of glucose and ketonic particles in the blood

19. Boy, aged 14, takes 10 U of insulin before breakfast and 6 U before dinner. During the lunchtime has eaten a little, in 30 minutes after dinner has lost consciousness, cramps, paleness, serious humidity of skin and jaws masticatory spasm have appeared. Cardiac tones are muffled, tachycardia up to 105 per minute, arterial hypotonia. What it is necessary to enter first of all ?

- A intravenously by jet to enter 40 % glucose solution
- B Subcutaneously 0,1 % epinephrine solution of an epinephrine
- C Glucocorticoids intravenously
- D 10 % Sodium chloridum intravenously
- E it is intravenous by drops the 5% glucose solution

20. Girl of 1 month age is sick just after birthday. Complaints on weight loss, flaccidity, repeated regurgitation and vomiting are irrespective of feeding. Was born with weight 3700,0, now weight is 3000,0. Child is pale, hypodermic fat is absent, dryness of mucosas, retraction of fontanel. Penis-like clitoris. Tachycardia, cardiac sounds are muffled. Sodium of blood is 86 mmol/l, potassium is 9,1 mmol/l. What is the forecast to this child?

- A. Favorable in case of diet administration
- B Favorable in case of enzymes administration
- C Favorable in case of oral rehydratation
- D Favorable in case of glucocorticoids administration
- E Favorable in case of a diet and enzymes administration.

21. . Child, aged 8, is hospitalized with complaints on thirst and polyuria. During 5 years is sick of a diabetes. In the anamnesis the diabetic coma developed three times before . Level of Glucose is 15,6 mmol/l. In the urine is 5 %. By ophthalmologist the retinopathy is revealed. What dose of insulin will be prescribed to the patient?

- A. 2,0 U/kg .
- B. 0,25 U/kg.
- C. 0,5 U/kg .
- D. 1,5 U/kg.
- E. 5,0 U/kg.

22. Boy, aged 12, sick of diabetes during 8 years. Hospitalized in intensive care department in a coma of 2 degrees. The consciousness is absent. Skin and mucous are pale and dry. Respiration noisy Smell of acetone from the mouth. Anuria. is fixed: glucose of blood is 35 mmol/l, Serumal potassium is 2,5 mmol/l,

sodium is 120 mmol/l, urea is 9,5 mmol/l, ketonic particles in the serum is 7,5 mmol/l. Acetone test in urine is +++++, glucose is 120 mmol/l. For deducing the patient from a coma it is necessary to conduct the following actions:

- A. To enter hydrocortisone
- B. Hemotransfusion.
- C. To conduct of neuroleptanalges
- D. To conduct a dehydration.
- E. To conduct the correction of acid basic equilibrium.

23. To the girl who shows complaints on undue fatiguability, polydipsia, polyuria and relapsing pustular enanthesises the glucose tolerance test has been conducted. The following results are obtained: glycemia on empty stomach is 6,8 mmol/l, after 2 hours is 17,5 mmol/l. What is your conclusion?

- A. The data are doubtful.
- B. Normal data.
- C. Diabetes.
- D. Disordrered tolerance to glucose.
- E. The data are uncertain.

24. Girl, aged 16, sick of diabetes during 8 years. Hospitalized in intensive care department in a coma of 2 degrees. The consciousness is absent. Skin and mucous are pale and dry. Respiration noisy Smell of acetone from the mouth. Anuria. is fixed: glucose of blood is 35 mmol/l, Serumal potassium is 2,5 mmol/l, sodium is 120 mmol/l, urea is 9,5 mmol/l, ketonic particles in the serum is 7,5 mmol/l. Acetone test in urine is +++++, glucose is 120 mmol/l. All listed below can result to coma except for:

- A. Smoking.
- B. Insulin overdosage .
- C. Infectious diseases.
- D. Serious physical work.
- E. Use of fat nutrition.

25. Boy, 8 years old, takes to the clinic concerning to growth inhibition. Was born asphyxiated with weight 2800 g. In school studied well. Parents have average body height. Objectively: body height - 107 cm, weight - 23 kg, face looks like a doll. Hair are thin, skin dry and has icteric shade. Subcutaneously - fatty tissue is well developed on the neck, thorax, and stomach. Muscles developed insufficiently. Specify the most probable diagnosis:

- A. Hondrodystrophia
- B. Craniopharingioma
- C. Pituitary nanism
- D. Down syndrome
- E. Fanconi syndrome

26. In tall patients with hypergonadotropic hypogonadism and with combined contents in the buccal epithelium of X and Y chromatin for final diagnostics of disease it is necessary to investigate parameters as follows :

- A. Somatotropin production
- B. Morphology of pituitary body
- C. Karyotype
- D. Production of insulinoid growth factors
- E. Genealogic investigation

27. To the child sick with acute lymphoblast leukemia, for correction of anemia the hemotransfusion with a packed red cells conducted. What laboratory investigations should be done necessarily after a hemotransfusion?

- A. Determine of a hemoglobin, RBC, urine tests.
- B. Coombs test, functional liver tests..
- C. Proteinogramme, coagulogramme.
- D. Electrolytes in a blood and urine.
- E. Urinary acid of blood, acid and alkaline condition of blood.

28. The basic pathogenetic factor of diabetic angiopathies development are:

- A. Duration of a diabetes.
- B. A chronic hyperglycemia.
- C. Relative insulinic insufficiency
- D. Absolute insulinic insufficiency.
- E. Occurrence of a diabetes at children's age

29. Patient L., without consciousness. Mother reports he suffers of diabetes during 12 years. 2 weeks ago was ill of lacunar angina. Skin is dry. Respiration is frequent. The smell of an acetone in exhaled air is absent. Blood pressure is 80/40 mm Hg. What coma most likely takes place in the patient?

- A. Hyperosmolaric
- B. Hyperlactatacidemic
- C. Hypoglycemic
- D. Ketoacidic.

30. Patient K., 8 years old, was taken to endocrinologic department in connection to nonperishable blood pressure rising (150/90 mm Hg), thirst, polyuria, a polydipsia, periodic attacks of cramps in various muscular groups. Hypokalemia, hypernatremia, decreasing of urine concentrational ability and the resistance to antidiuretics are found out. What is the most probable diagnosis?

- A. Adenoma in the suprarenal cortex.
- B. Idiopathic hyperaldosteronism.
- C. Diabetes.
- D. Diabetes insipidus.
- E. Pheochromocytoma.

31. In the girl of 10 years old, complaints to irritability, sweating, pains in the area of heart, headache. Enlargement of a thyroid gland. In examination the III degrees nodal struma is found out. Skin is wet, hot by touch, tachycardia 104 b. per minute. On a scanning image the hot node reveals. Level of thyroid hormones is high.

- A. Diffuse toxic goiter.
- B. Autoimmune thyroiditis.
- C. A cancer of a thyroid gland.
- D. Toxic adenoma.
- E. Ridel fibrosal struma.

32. In the boy of 5 years old after viral syndrome the temperature up to 39,2 has raised suddenly, the headache, pain in the thyroid gland area is intensified in swallowing and head turning has appeared . .The thyroid gland enlarged, painful in palpation, the hyperemia of skin above it. In laboratory data the function of thyroid gland is unchanged. In the analysis of a blood the leukocytosis and accelerated BSR. What is the diagnosis?

- A. Autoimmune thyroiditis.
- B. Acute thyroiditis.
- C. Toxic adenoma of a thyroid gland.
- D. Ridel fibrosal struma
- E. Sporadic struma.

33. The girl of 11 years old. Complaints to the general delicacy, fatigability, enlargement of neck. Objectively: thyroid gland in palpation is dense, impure and enlarged up to I degree. In it structure investigation the hyperecho and hypoecho sites were marked. TTH level and of antibodies levels to the thyroid gland are raised. What is the preliminary diagnosis?

- A. Autoimmune thyroiditis, the atrophic type.
- B. Autoimmune thyroiditis, the hypertrophic type.
- C. A diffuse nontoxic struma of I degree.
- D. Subclinical hypothyroidism.
- E. Multinodal goiter.

34. In what parts of endocrine system in Cushing disease the function is primary changed ?

- A. Sexual glands.
- B. Hypothalamus
- C. Suprarenal glands.
- D. Pituitary body.
- E. Thyroid gland.

35. The child of 3 years old arrived in a hospital with complaints on paleness, total delicacy, sharp decreasing of appetite. Parents consider the child to be ill just after birth. In examination child is very pale, subnutritional, hypertelorism is marked. Child is blonde . Peripheric lymph nodes are small-sized, liver and lien are not enlarged. In the routine blood analysis: erythrocytes- $1,7 \times 10^{12}/l$, Hb- 48 g/l, the color index - 0,9, reticulocytes- $0,0001 \times 10^9$, thrombocytes- 200×10^9 , leucocytes- $7,8 \times 10^9/l$, blood sedimentation rate -18 mm / hour. In bone marrow puncture there is a sharp depression of erythroid locus. Bilirubin - 17,1 $\mu\text{mol}/l$ due to indirect fraction. What preliminary diagnosis in this case?

- A. Iron deficiency anemia

- B. Acquired hypoplastic anemia
- C. Congenital Fanconi aplastic anemia
- D. Congenital hypoplastic anemia of Diamond -Blackfan
- E. Hemolytic anemia

36. The child in age of 6 months was hospitalized. Mother of child shows complaints on paleness and deterioration of him appetite. The child was born with weight of 2100 g in term and in gestational age of 35 weeks. Is on the mixed nutrition. Objectively: skin is pale, a subnutrition take place. Cardiac tones are muffled and systolic murmur auscultated on an apex. A liver + 2 cm. In the analysis of a blood: erythrocytes- $2,8 \times 10^9/l$, Hb-86 g/l, color index - 0,9, reticulocytes- $0,000810^9$, blood sedimentation rate - 9 mm / hour. Serumal iron - 4,36mcmol/l. A bilirubin of a blood is 4,6 mcmol/l due to indirect fraction. What is the optimal method of therapy in this case?

- A. Preparations of iron
- B. Hemotransfusion
- C. Vitamines B_6, B_{12} , Acidum folicum
- D. Packed red cells transfusion
- E. Corticosteroids

37. The child in age of 8 months. During last month the insufficiency of weight increasing and deterioration of appetite are marked, delicacy has appeared, the fragility and transversal striation of nails, a xeroderma, the phenomena of angular stomatitis, atrophy of tongue papillas has appeared. Routine blood test: erythrocytes- $3,0 \times 10^{12}/l$, Hb-68 g/l. What is the most probable diagnosis?

- A. Hemolytic anemia
- B. Posthemorrhagic anemia
- C. Iron deficiency anemia
- D. Infecton - toxic anemia
- E. Hypoplastic anemia

38. The boy of 3 years old has arrived in a hospital. Parents of the child show complaints on paleness of him skin and decreasing of appetite. In a ration dairy products prevail. In routine blood test: erythrocytes- $1,8 \times 10^{12}/l$, haemoglobin-60 g/l, leucocytes- $4,6 \times 10^{12}/l$, segmented -44 %, eosinocytes - 6 %, lymphocytes - 46 %, monocytes - 4 %, blood sedimentation rate 10 mm / hour. A level of serumal iron - 6 mcmol/l. What preparation is the most rational for starting treatment?

- A. Ferri lactas
- B. Vitamin B_{12}
- C. Packed red cells
- D. Folic acid
- E. Vitamin B_6

39. Patient C., 12 years old. The obesity, fatigue, sleepiness, headache disturbs.

Objectively: body height of 171 sm, weight of 106 kg, and the adiposity is mainly on arms and trunk. A skin dry with a crimson - mottled shade. On arms, breast and hips there are crimson - cyanotic strips of a stretching. Pulse is 76 per min., blood pressure is 160 / 102 mm Hg.

What kind of excess is the main cause of hypertension in the patient?

- A. Epinephrine
- B. Hydrocortisone
- C. Aldosteronum
- D. Androstendion
- E. All hormones are listed above

40. What is not typical for exogenous - constitutional type of primary obesity?

- A. Changes of a skin with a pigmentation, folliculitis, striae on hips and breeches;
- B. Early terms of superfluous body mass occurrence;
- C. The subcutaneously - fatty layer is distributed in regular intervals;
- D. Slow, gradual progress of obesity;
- E. Family predilection to obesity.

41. . Attributes of an Cushing syndrome are all signs, except for:

- A. Premature sexual development;
- B. Lunar face;
- C. Osteoporosis;
- D. Premature sexual pilosity;
- E. Distribution of subcutaneously - fatty layer non-uniformly, in the top part of a body.

42. Typical attributes of Lawrence -Moon-Bardet-Bidl syndrome are everything, except for:

- A. Cataract;
- B. Pigmentary retinopathy;
- C. Uniform obesity;
- D. Oligophrenia;
- E. Polydactyly, congenital anomalies of a skeleton.

43. In child clinical examination there are following signs revealed - skin humidity, exaltation, irritability, decreasing of body weight, tachycardia, syndromes of Grefe, Stellwag, Moebius, palpitation. For what disease these signs are characteristic?

- A. Acromegalia
- B. Hypothyroidism
- C. Diabetes
- D. Down disease
- E. Thyroid storm.

44. In the child of 1, 5 years old the activity is reduced, does not walk, does not talk. Objectively: skin acyanotic, dry and hydropic, the tongue is big, saddle-like nose, a voice is low and rasping and hair is thick and rasping. The large fontanel is 3, 0x3, 0 sm. Teeth are not present. What diagnosis is it possible to think of?

- A. Down disease
- B. Hypothyroidism
- C. Rachitis
- D. Pituitary nanism
- E. Diabetes

45. In examination of 14 years old girl the nodal struma of III degree is found out. On scenogram the "hot" unit revealed. Levels of T3 and T4 in a blood are increased.

What disease is it possible to think of?

- A. Diffuse toxic goiter.
- B. Cancer of a thyroid gland
- C. Toxic adenoma of a thyroid gland
- D. Autoimmune thyroiditis
- E. Fibrous struma of Riddell

46. In the patient of 13 years old, relapse of a nephrolithiasis, ostealgia, weakness, fatigability, growing thin are observed.

What from the specified diseases can be suspected?

- A. Sarcoma of bones
- B. Hypoparathyrosis
- C. Hyperparathyroidism
- D. Multiple myeloma
- E. Any of the specified diseases

47. In the girl of 7 years old the fatty tissue on the face has disappeared. On the lower half of body adeps is postponed well.

How such condition refers to?

- A. Oligotrophy
- B. Lipoma.
- C. Lipoatrophia.
- .D. Dystrophia of nutritional genesis.
- E. Illness of Symonds.

48. What wide-spread disturbances of a metabolism could find in patients with obesity?

- A. Hyperinsulinemia, hypercholesterolemia.
- B. Hypoinsulinemia, hypocholesterolemia.
- C. Hyperinsulinemia, hypocholesterolemia
- D. A normal level of blood glucose and hypercholesterolemia.
- E. A normal level of cholesterol, β – lipoproteids and disproteinemia.

49. Name the preparations promotes of appetite diminishing

- A. Furosemidum;
- B. Thyreoidinum;
- C. Phepranonum;.
- D. ATP;

E, Aloe.

50. What diseases are more wide-spread among the patients suffering of obesity?

- A. Hypertension.
- B. Diabetes.
- C. Osteoarthrosis.
- D. Coronary failure.
- E. All listed above

51. What malignant neoplasms more wide-spread among the patients suffering of obesity?

- A. Cancer of gall bladder and cholic ducts.
- B. Lungs cancer
- C. A cancer of prostate, direct and colonic intestines in men.
- D. A cancer of endometrium and mamma in women.
- E. All listed above

52. Patient G., 12 years old. Growing weight on 10 kg for 4 months, complains of constant irritability, palpitation, pain in eyes and lacrimation. In examination: skin is warm and wet, mild exophthalmia and hyperemia of conjunctiva, positive signs of Grefe, Koher and Moebius. The thyroid gland is unpainful and diffusively enlarged, that is seen in swallowing,. Pulse 108 per minute, blood pressure is 140 / 66 mm Hg. There is fine tremor in hands fingers.

What is the diagnosis?

- A. A nodal toxic struma of IV degree with a mild thyrotoxicosis
- B. Diffuse toxic struma of I degree with a serious thyrotoxicosis
- C. A nervosism
- D. A subacute thyroiditis
- E. Diffuse toxic goiter of II degree with the mild thyroid storm.

53. The girl of 13 years old complains of a xeroderma and decreasing of memory. In examination: tongue enlarged and reflexes are time-lapsed.

For what disease these signs are characteristic?

- A. A diffuse toxic struma
- B. A hypothyroidism
- C. A subacute thyroiditis
- D. An adenoma of a thyroid gland
- E. Endemial Struma with euthyroidism

54. In patient G. of 15 years old in examination the enlargement of thyroid gland seen in a swallowing and infringement of eyes convergence are revealed.

What from eye signs is found out in the patient?

- A. Moebius
- B. Schtelwag
- C. Koher
- D. Krause
- E. Grefe

55. How long antithyroid therapy of a diffuse toxic struma in children in condition of achievement and preservation of euthyroidism can be conducted?

- A. during 3 months.
- B. during 6 months.
- C. during 1-1,5 years.
- D. during 1 month
- E. during 2 months.

56. The parents of a 14-year-old boy are concerned about his short stature and lack of sexual development. By history, you learn that his birth weight and length were 3 kg and 50 cm, respectively, and that he had a normal growth pattern, although he was always shorter than children his age. The physical examination is normal. His upper-to-lower segment ratio is 0.98. A small amount of fine axillary and pubic hair is present. There is no scrotal pigmentation; his testes measure 4.0 cm³ and his penis is 6 cm in length. In this situation you should

- a. Measure pituitary gonadotropin
- b. Obtain a CT scan of the pituitary area
- c. Biopsy his testes
- d. Measure serum testosterone levels
- e. Reassure the parents that the boy is normal

57. A 13-year-old asymptomatic girl with enlarged thyroid gland up to 3 degree from non endemic region. She states that the findings demonstrated began more than a year ago. Treatment for the patient in the previous question includes

- a. Iodine
- b. Synthroid (L-Thyroxin)
- c. PTU (propylthiouracil)
- d. Psychiatry consult
- e. Surgical removal of thyroid

58. The 2-week-old female is noted to have a thin membrane adhering together the upper portion of the labia minora. The most appropriate course of action for this condition is to

- a. Apply estrogen cream daily
- b. Refer for surgical repair
- c. Apply traction to the opposing labia until the membrane breaks
- d. Evaluate the patient for congenital adrenal hyperplasia
- e. Do nothing, as the lesions are of no consequence.

59. A 12-year-old girl has a mass in her neck. Physical examination reveals a thyroid nodule, but the rest of the gland is not palpable. A technetium scan reveals a “cold” nodule. The child appears to be euthyroid. Which of the following diagnoses is the *least* likely?

- a. Simple adenoma
- b. Follicular carcinoma
- c. Papillary carcinoma
- d. Cyst

e. Dysgenetic thyroid gland

60. A 15-year-old boy has been immobilized in a double hip spica for 6 weeks after having fractured his femur in a skiing accident. He has become depressed and listless during the past few days and has complained of nausea and constipation. He is found to have microscopic hematuria and a blood pressure of 150/100 mmHg. You should
- Request a psychiatric evaluation
 - Check blood pressure every 2 h for 2 days
 - Collect urine for measurement of the calcium-creatinine ratio
 - Order a renal sonogram and intravenous pyelogram (IVP)
 - Measure 24-h urinary protein
61. A 7-day-old boy is admitted to a hospital for evaluation of vomiting and dehydration. Physical examination is otherwise normal except for minimal hyperpigmentation of the nipples. Serum sodium and potassium concentrations are 120 meq/ L and 9 meq/ L, respectively; serum glucose is 120 mg/dL. The diagnosis can be confirmed in this patient by
- Obtaining a barium swallow
 - Measurement of 17-hydroxyprogesterone
 - Measurement of T3, T4, and TSH
 - Somatomedin C measurement
 - Measurement of serum renin levels
62. What is more likely in electrolytes disorders (mmol/l) for 2-year-old girl with nephrogenic diabetes insipidus
- Na⁺ 118, K⁺ 7.5
 - Na⁺ 125, K⁺ 3.0
 - Na⁺ 134, K⁺ 6.0
 - Na⁺ 144, K⁺ 2.9
 - Na⁺ 155, K⁺ 5.5
63. The first sign of pubertal development in a 13-year-old boy
- Enlargement of the testes
 - Development of pubic hair
 - Acne
 - h. Growth spurt
 - Penile enlargement
64. You are seeing a 2-week-old boy at a routine visit. The mother complains that he has been constipated, jaundiced, sluggish, and excessively sleepy. The physical examination is normal except for mild jaundice and a distended abdomen in a sleepy infant. The most appropriate course to pursue initially is assessment of
- The mother's serum for autoantibodies to thyroid gland
 - Levels of thyrotropin-releasing hormone (TRH), thyroid-stimulating hormone (TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4), and thyroglobulin in the mother
 - The results of the neonatal metabolic screen
 - Levels of thyrotropin-releasing hormone (TRH), thyroid-stimulating hormone

(TSH), tri-iodothyronine (T3), reverse T3, levothyroxine (T4), and thyroglobulin in the infant

e. The effects on growth and symptoms with increasing feeds by 20% per day

65. You are seeing a 2-week-old boy at a routine visit. The mother complains that he has been constipated, jaundiced, sluggish, and excessively sleepy. The physical examination is normal except for mild jaundice and a distended abdomen in a sleepy infant. The most appropriate next step is to

- a. Repeat all the baby's abnormal laboratory results (if any)
- b. Obtain x-rays of the baby's skull, wrists, and knees
- c. Begin on the baby oral sodium-L-thyroxine, 10 to 15 mg/(kg_d)
- d. Evaluate the neonate in 2 weeks for the results of symptomatic treatment
- e. Obtain a pediatric endocrinology consultation within 2 weeks

66. For patients with Klinefelter syndrome the following clinical signs are characteristic, except for:

- A. High body height.
- B. Presence of a gynecomastia.
- C. Azoospermism.
- D. Body height is lower than average.
- E. All listed above.

67. Mother of the 5 years old girl complains of small weight and low body height of the child. Anamnesis: the child from II pregnancy with threat of abortion in 5-7 weeks. Labors in 38 weeks. Child's body weight at birth is 1400 g, body height is 30 cm. Objectively in examination: body height of child is 80 cm and weight is 11 kg. There are dismorphic features and congenital heart disease. What diagnosis is most probable in this case?

- A. Pituitary nanism.
- B. Cerebral nanism.
- C. Premordial nanism.
- D. Pseudopremordial nanism.
- E. Chondrodystrophy.

68. What includes the genetic sex concept of?

- A. Presence secondary sexual characters.
- B. A corresponding structure of external genitals.
- C. A corresponding structure of internal genitals.
- D. Presence of corresponding set of sexual chromosomes (XX or XY).
- E. All listed above

69. The child of 14 years old, arrived in endocrinology department due to superfluous adjournment of subcutaneously fatty layer mainly in the area of face and upper half of trunk, progressing muscular delicacy, resistant rising of arterial blood pressure, hyperpigmentation of skin. In laboratory tests revealed hypokalemia and raised plasma ACTH level. The excretion in urine of 17 - KS and 17 - OKS are increased and diminished after taking of Dexamethasone. What is the most probable diagnosis?

- A Cushing disease .
- B Dermatomyositis.
- C Corticosteroma.
- D Subthalamic obesity..
- E Addison diseaseI.

70. Child of 12 years old, sick aof autoimmune thyroiditis, there are changes in the blood count: RDC.: $2,1 \times 10^{12}/l$; Hb: 82 g/l; the CI: 0,9; thrombocytes: $310 \times 10^9/l$; reticulocytes.: 30 ‰; WBC.: $4,2 \times 10^9/L$; eosinophiles.: relating to stab neutrophile 2 %; segmented neutrofiles.: 58 %; lymphocytes.: 28 %; monocytes.: 6 %; ESR: 28 mm / h. total bilirubin: 115 mmol/l, direct.: 12,5 mmol/l, AST: 0,2 mmol/l, ALT: 0,3 mmol/l. Coombs test is positive. What is the preliminary diagnosis?

- A Autoimmune hemolytic anemia.
- B Hypoplastic Fanconi anemia.
- C Minkovski - Schoffer hemolytic anemia.
- D B_{12} - folic acid scarce anemia.
- E Hypoplastic anemia.

71. At the child of 9 years old the following changes in a blood count are revealed: RBC.: $2,5 \times 10^{12}/l$; Hb: 85 g/l; the CI: 0,85. Determine a degree of the anemia?

- A IV.
- B II.
- C I.
- D III.
- E O.

72. . The child of 10 years old, has bitten by the dace steppe viper .Blood count: RBC.: $2,1 \times 10^{12}/l$; Hb: 92 g/l; the CI: 0,9; reticulocytes.: 20 ‰; thrombocytes: $210 \times 10^9/l$; leucocytes.: $6,2 \times 10^9/l$; eosinophiles.: 2 %; relating to stab neutrophile : 2 %; segmented neutrophiiiles.: 78 %; lymphocytes.: 22 %; monocytes.: 6 %; ESR: 15 mm / h. What is the preliminary diagnosis?

- A Acute hypoplastic anemia
- B Acute hemolytic anemia
- C Iron deficiency anemia.
- D B_{12} - folic acid scarce anemia
- E Sickle cells anemia

73. The girl of 10 years old was taken to the hospital in an unconsciousness. Suffers of type 1 diabetes. Takes the combined insulin therapy. In the morning, going to school, after introduction of insulin has had a meal less usual. After 2 hours the anxiety, tremor has appeared. Soon she has lost consciousness. In hospital have assumed the hypoglycemic coma. What is the medical tactics in relation to this child?

- A. To adjust i.v. drop introduction of 10 % glucose solution.
- B. To indicate the glucose test and to wait the results
- C. To take a blood for glucose test and at once to enter i.v., trickling 20,0 ml of 40 % glucose solution.

- D. I.v. trickling introducing of 20,0 ml 40 % glucose solution.
- E. To enter i.m. of 1 % epinephrine solution

74. Nine years old girl was hospitalized with complaints on skin paleness, yellowness and dark color of urine. In examination hepatosplenomegaly detected. In the routine blood test: Hb-54 g/l, erythrocytes- $1,1 \times 10^{12}/l$, color index- 1,0, reticulocytes -20%, thrombocytes- 200×10^9 , leucocytes- $12,0 \times 10^9/l$, eosinocytes -3 %, relating to stab neutrophiles - 7 %, segmented- 70 %, lymphocytes - 16 %, monocytes - 4 %, blood sedimentation rate 22 mm / hours, direct bilirubin -10 $\mu\text{mol}/l$, indirect bilirubin 62 $\mu\text{mol}/l$, thymol turbidity test - 3 units. Direct and indirect Coombs tests are positive. What therapy is necessary to administrate in this case?

- A. Immunodepressants
- B. Splenectomy
- C. Ferrum lek
- D. Phenobarbitalum
- E. Karsil

75. The girl of 3 years old. In examination there are retardation in physical development, paleness of mucosas, disembriogenetical stigmata, gingival bleedings, « coffee- like stains » in the top part of a trunk, polydactyilia, systolic apex murmur. In the routine blood analysis there is a pancytopenia. What is preliminary diagnosis?:

- A. Fanconi anemia
- B. Diamond-Blackfan anemia
- C. Klinefelter syndrom
- D. Down syndrom
- E. Acquired aplastic anemia

76. The child of 3 years old arrived in a hospital with complaints on paleness, total delicacy, sharp decreasing of appetite. Parents consider the child to be ill just after birth. In examination child is very pale, subnutritional, hypertelorism is marked. Child is blonde. Peripheric lymph nodes are small-sized, liver and lien are not enlarged. In the routine blood analysis: erythrocytes- $1,7 \times 10^{12}/l$, Hb- 48 g/l, the color index - 0,9, reticulocytes- $0,0001 \times 10^9$, thrombocytes- 200×10^9 , leucocytes- $7,8 \times 10^9/l$, blood sedimentation rate -18 mm / hour. In bone marrow puncture there is a sharp depression of erythroid locus. Bilirubin - 17,1 $\mu\text{mol}/l$ due to indirect fraction. What preliminary diagnosis in this case?

- A. Iron deficiency anemia
- B. Acquired hypoplastic anemia
- C. Congenital Fanconi aplastic anemia
- D. Congenital hypoplastic anemia of Diamond -Blackfan
- E. Hemolytic anemia

77. In child of 8 years there are the increasing paleness, delicacy, hemorrhages on a skin has appeared. In sternal puncture the depression of all locuses of hemopoiesis. What basic method of therapy is indicated in aplastic anemia during the subacute period?

- A. Corticosteroids + bone marrow transplantation
- B. Splenectomy
- C. Hemotransfusion + cytostatics
- D. Cytostatics + bone marrow transplantation
- E. Antibiotics + hemotransfusion

78. Child with the diagnosis of Minkovsky - Schoffer hemolytic anemia admitted to the hospital because of hemolytic crisis. What parameter of the minimal osmotic resistance of erythrocytes will confirm the diagnosis?:

- A. 0,7
- B. 0,42
- C. 0,39
- D. 0,4
- E. 0,53

79. Girl of 13 years old complains of a long-term and abundant menses and general delicacy. In examination her general state is serious, lengthways the body there are hemorrhagic rashes are varies from spots up to ecchymomas and petechias were detcted and on mucous there are hemorrhages. Two weeks ago has transferred respiratory infection, has taken Sulfanilamides. What is more probable result to this state?

- A . Hemorrhagic vasculitis
- B Werlhof's disease
- C Disseminated intravascular coagulation syndrome
- D Meningococemy
- E Cristmas disease

80. An otherwise healthy 7-year-old girl is brought to your office by her father because she has some acne, breast development, and fine pubic hair. The most likely etiology for her condition is

- a. A feminizing ovarian tumor
- b. A gonadotropin-producing tumor
- c. A lesion of the central nervous system
- d. Exogenous estrogens
- e. Early onset of "normal" puberty (constitutional)

Answers:

1	A	11	B	21	D	31	B	41	A	51	A	61	B	71	D
2	B	12	A	22	E	32	D	42	A	52	E	62	E	72	B
3	A	13	A	23	C	33	E	43	E	53	B	63	A	73	C
4	C	14	E	24	A	34	B	44	B	54	A	64	C	74	B
5	C	15	A	25	C	35	D	45	C	55	C	65	C	75	A
6	C	16	D	26	C	36	A	46	C	56	E	66	D	76	D
7	A	17	A	27	A	37	C	47	C	57	B	67	C	77	B
8	C	18	E	28	D	38	C	48	A	58	A	68	D	78	B
9	D	19	A	29	A	39	A	49	A	59	E	69	A	79	B

10	C	20	D	30	D	40	C	50	C	60	B	70	B	80	E
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Situational Task 1. A girl of 10 years old firstly hospitalized in the department with complaints of pagophagia. On examination tachycardia and cardiac dilatation occur, and systolic murmurs are present. The spleen is enlarged. Laboratory findings: microcytosis with increasing deficiency the RBCs, hypochromia, poikilocytosis, and increased red cell distribution width (RDW). Reticulocytes are moderately elevated. White blood cell counts are normal. The bone marrow is hypercellular, with erythroid hyperplasia. Leukocytes and megakaryocytes are normal.

1. What is the preliminary diagnosis?
2. Prescribe treatment.
3. Is there necessary to prescribe vitamins in this case?
4. How to count the therapeutic dose?
5. How absorption of ethiotropic drugs will affect the treatment efficiency?

Standard of answer

1. Iron-deficiency anemia
2. Oral administration of simple ferrous salts (sulfate, gluconate, fumarate) provides inexpensive and satisfactory therapy.
3. There is no evidence that addition of any trace metal, vitamin, or other hematinic substance significantly increases the response to simple ferrous salts.
4. For routine clinical use the physician should be familiar with an inexpensive preparation of one of the simple ferrous compounds. The therapeutic dose should be calculated in terms of elemental iron; ferrous sulfate is 20% elemental iron by weight. A daily total of 6 mg/kg of elemental iron in three divided doses provides an optimal amount of iron for the stimulated bone marrow to use.
5. Better absorption may result when medicinal iron is given between meals. Intolerance to oral iron is uncommon. A parenteral iron preparation (iron dextran) is an effective form of iron and is usually safe when given in a properly calculated dose, but the response to parenteral iron is no more rapid or complete than that obtained with proper oral administration of iron, unless malabsorption is present.

Situational Task 2. A boy of 16 years old. Since 2-years-old age has been suffering from diabetes, got alcohol, hospitalized with complaints of weakness, consciousness,

On examination : the skin is pale and cyanotic , consciousness, sweating.

Breathing is superficial , the smell of alcohol, tachycardia, cramps.

1. What is the preliminary diagnosis?
2. To render the first aid
3. What are the main effects of prescribed preparation?
4. What is alternative therapy?
5. All treatments for hypoglycemia provide recovery in approximately 10 minutes

Standard of answer

1. Hypoglycemic coma.
2. Manage mild hypoglycemia by giving rapidly absorbed PO carbohydrate or glucose; for a comatose patient, administer an intramuscular injection of the hormone glucagon,

3. Glucagon stimulates the release of liver glycogen and releases glucose into the circulation.
4. Appropriate and alternative therapy is intravenous glucose (preferably not more than a 10% glucose solution).
5. What time is need for recovery in case of adequate treatment?

Situational Task 3 A 3-year-old child presents with a petechial rash but is otherwise well and without physical findings. Platelet count is 20,000/L; hemoglobin and WBC count are normal.

1. What is the most likely diagnosis?
2. Conduct differential diagnosis.
3. What therapy is necessary to prescribe for this patient?
4. What is the mean age of presentation in this condition?
5. What the difference among petechia and ecchymoses?

Answers:

1. Idiopathic thrombocytopenic purpura.
2. Patients with acute lymphoblastic leukemia frequently have symptoms of pallor and fever in addition to bleeding. Nearly 50% of them have hepatomegaly and splenomegaly. CBC reveals anemia, leukocytosis or leukopenia, and thrombocytopenia. Disseminated intravascular coagulopathy (DIC) is secondary to a severe underlying disease, such as fulminant bacterial sepsis with hypotension or profound hypoxia. Patients with Henoch-Schonlein purpura have symptoms of skin rash and abdominal or joint pain, lupus erythematosus (SLE) is very rare in a 3-year-old child. Findings include fever, joint pain, and skin rash. CBC can reveal anemia, leukopenia, and thrombocytopenia.
3. Heparin, glucocorticoids, desagregants.
4. The mean age of presentation of ITP is 6 years.
5. Ecchymoses is more than 0,3 cm in diameter compared to petechiae.

Situational Task 4. Having performed a complete history and physical examination of the patient, you proceed with a diagnostic workup. Initial laboratory results are as follows: hemoglobin 8 g/dL; hematocrit 24%; leukocyte count 11,000/L with 38% neutrophils, 7% bands, 55% lymphocytes; hypochromia on smear; free erythrocyte protoporphyrin (FEP) 110 g/dL; lead level 7 g/dL whole blood; platelet count adequate; reticulocyte count 0.5%; sickle cell preparation negative; stool guaiac negative; and mean corpuscular volume (MCV) 65fl.

1. What is the most likely diagnosis?
2. Prescribe the treatment.
3. Which dietary modifications are indicated in this case?
4. What indication for hemotransfusion in this case?
5. What the diagnostic value of free protoporphyrins?

Answers are:

1. Iron deficiency anemia
2. Response to a therapeutic trial of iron is an appropriate and cost-effective method of diagnosing iron deficiency anemia. A prompt reticulocytosis and rise in hemoglobin and hematocrit follow the administration of an oral

preparation of ferrous sulfate. Intramuscular iron dextran should be reserved for situations in which compliance cannot be achieved. This is because this treatment is expensive, painful, and less effective than oral iron.

3. Dietary modifications, such as limiting the intake of cow's milk and including iron-fortified cereals along with a mixed diet, are appropriate as long-term measures, but they will not make enough iron available to replenish iron stores.
4. The gradual onset of iron-deficiency anemia enables a child to adapt to surprisingly low hemoglobin concentrations. Transfusion is rarely indicated unless a child becomes symptomatic or is further compromised by a superimposed infection.
5. When the iron available for production of hemoglobin is limited, free protoporphyrins accumulate in the blood. Levels of erythrocyte protoporphyrin (EP) are also elevated in lead poisoning. Iron-deficiency anemia can be differentiated from lead intoxication by measuring blood lead, which should be less than 10 g/dL.

НАВЧАЛЬНО-МЕТОДИЧНЕ ВИДАННЯ

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ПАШКОВА ОЛЕНА ЄГОРІВНА
ГИРЯ ОЛЕНА МАКСИМІВНА
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ВРУБЛЕВСЬКА СВІТЛАНА ВОЛОДИМИРІВНА
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