BOHOMOLETS NATIONAL MEDICAL UNIVERSITY

GUIDELINES
for practical classes
for students

Educational discipline: «Pediatric gastroenterology, pulmonology and nephrology»

Field of knowledge: 22 "Health care"

Specialty: 222 "Medicine"

Department of Pediatrics No 2

Approved at the meeting of the Department of Pediatrics No. 2 on August 26, 2024, protocol No. 1

Considered and approved by: Cyclic methodological commission for pediatric disciplines

dated August 29, 2024, protocol No 1

Subject of the lesson:

" The most common congenital diseases of liver in children "

Competencies:

Ability to collect medical information about the child and analyze data (complaints, life history, medical history)
The ability to distinguish and identify leading clinical symptoms and syndromes in hereditary liver diseases in children (Wilson's disease, glycogenoses and others).
The ability to determine the necessary list of laboratory and instrumental studies for hereditary liver diseases in children (Wilson's disease, glycogenosis, and others) and to evaluate their results.
The ability to determine the necessary list of laboratory and instrumental studies for the diagnosis of hereditary liver diseases in children (Wilson's disease, glycogenosis, and others) and to evaluate their results.
Ability to establish a preliminary and clinical diagnosis of hereditary liver diseases in children (Wilson's diseases, glycogenosis and others).
Ability to determine the principles and nature of treatment of hereditary liver diseases in children (Wilson's disease, glycogenosis and others) and prevention of these diseases.
Ability to diagnose emergency conditions.
Ability to determine tactics and provide emergency medical care.
Ability to abstract thinking, analysis.
The ability to master and process modern knowledge.
Understanding the peculiarities of working with children of different ages.
The ability to make decisions when studying the discipline "Fundamentals of pediatric gastroenterology, pulmonology and nephrology"

The purpose of practical class
Formation of students' professional competencies for achieving program learning outcomes by controlling the initial level of knowledge in the process of discussing theoretical issues and testing, performing practical tasks and conducting control of the final level of training in solving situational problems on diagnosis, treatment and prevention of hereditary liver diseases in children (Wilson's disease, glycogenoses and others).

Equipment: PC with appropriate information support, reference materials, methodological recommendations, extracts from medical histories, a set of laboratory test results, manikin.

Lesson plan and organizational structure

Stage name	Description of the stage	Levels of assimilation	Timing

Prepa-ratory	- Organizational issues - Learning motivation: Modern hepatology includes the most problematic issues of etiology, pathogenesis and diagnosis of liver lesions in metabolic diseases in young children. The key role of this organ in the metabolism of proteins, lipids, carbohydrates, trace elements and vitamins determines the significant frequency of liver pathology in hereditary and congenital metabolic disorders, which is manifested by: damage to hepatocytes due to the accumulation of lipids, glycogen or other products, with the development of liver cirrhosis or carcinogenesis; pathological changes in the biliary tract under the influence of increased lithogenicity of bile, associated with the formation of defective bile micelles with an increased level of cholesterol and a reduced content of phospholipids and bile acids, the occurrence of cholelithiasis. *Control of the initial level of knowledge - test control and oral survey.* ***Examples of test tasks:*** **1. The dosage of raw corn starch in infants with GSDIa is:** A. 1.6 g/kg/m/t B. 0.6 g/kg/m/t B. 2.6 g/kg/m/t C. 1.0 g/kg/m/t **2. With which gene mutation is the liver most often affected with alpha-1 antitrypsin deficiency** A.PI*ZZ B. PI*SZ C. PI*SS D.PI*QO **3. Wilson's disease is based on a violation:** A. binding of copper to ceruloplasmin B. accumulation of iron C. insufficient activity of glycogen synthetase D. deficiency of alpha-1 antitrypsin **4. The presence of Kaiser-Fleischner rings corresponds to which score according to the Ferenc scale:**	Introductory Reproductive	15 min

	A. -1 B. 1 C. 0 D.2 **5. What kind of monitoring is required when taking the drug Trientin:** A. ALT B. AST C. urinary copper excretion and zinc levels D. total bilirubin, creatinine		
Main	Formation of professional competences: - demonstration of a thematic patient or review of extracts from medical histories of patients with the most common diseases of hereditary liver diseases in children (Wilson's disease, glycogenoses and others); - evaluation of the results of laboratory studies; - on the basis of anamnesis, data of a clinical examination and the results of laboratory studies, the establishment of a preliminary clinical diagnosis - determining of factors and pathogenetic mechanisms of disease development; - appointment of treatment and management of the disease;	Introductive Reproductive Creative Reproductive Creative Reproductive Creative	100 min
Final	Control of the final level of preparation Clinical cases A 14-year-old boy was diagnosed with Wilson's disease. Define P. Ferentz's score scale according to the features of the clinical course. Child from 1B., 1P, GV 40 years old. According to the parents, the boy grew and developed without peculiarities according to his age. At the age of 12, periodic abdominal pain accompanied by decreased appetite began to bother him. Signs of changes in neurological symptoms increased: alertness and concentration decreased, learning difficulties arose, tremors, dystonia appeared. Laboratory tests revealed (SAC without significant changes, BAC - total protein -70; ALT -64, AST - 59, total bilirubin – 18, ceruloplasmin level in the blood – 0.18, creatinine and urea	Creative	20 min

	within age norms). An ophthalmological examination revealed a Kaiser-Fleischer ring. Liver elastography revealed an increase in liver density. ***Answer standard*** 5 points – ceruloplasmin level 0.18, Kaiser-Fleschner ring, severe neurological symptoms. At the medical commission at the Military Commissariat, a 15-year-old boy was found to have the following changes in the laboratory blood test (total bilirubin - 21, ALT -300, AST - 280, creatinine, urea without abnormalities). Ophthalmological examination revealed yellow-green rings encircling the cornea. 1. What are the doctor's tactics and justify them. 2. Make a preliminary diagnosis. ***Answer standard*** Further referral to the hepatocenter or the department of gastroenterology is necessary to conduct the following studies for verification in the direction of HBV: determining the level of ceruloplasmin in the blood, the daily level of copper in the urine, copper in the liver biopsy, determining the genetic mutations of ATP7B in the biopsy, the presence of a Kaiser ring -Fleishner's during ophthalmological examination, neurological symptoms, anemia with a negative Coombs test. Wilson's disease. A 13-year-old girl complains of slight facial puffiness, swelling of the lower extremities, mainly in the morning. Wilson's disease was diagnosed a year ago. The patient received D-penicillamine at the rate of 150 mg/day in 3 doses. 1. Justify the occurrence of these complaints. 2. Further tactics. ***Answer standard***		

	the complaint data – side effects of taking D-penicillamine, which require a change in treatment – use of trientine in initial doses of 225 mg/day in 2-4 doses, control of ALT, AST, INR indicators; daily excretion of copper. General assessment of educational activity		

Recommended Books

1. Nelson Textbook of Pediatrics, 2-Volume set, 21-th edition. By Robert M. Kliegman, Bonita M.D. Stanton, Joseph St. Geme and Nina F Schor. – Philadelphia, PA : Elsevier Inc., 2020 - 4264 p. (pp. 2101-2105)
ISBN-10 : 032352950X ISBN-13 : 978-0323529501
2. S.Guandini, A. Dhawan Texbook of Pediatric Gastroenterology, Hepatology and Nutrition (Second edition), Chicago,IL, USA, London UK 2022; 880-894.

Questions for student self-preparation for practical classes

1. Definition, etiology, pathogenesis of WD.
2. Peculiarities of the course of WD.
3. Diagnosis of WD (P. Ferentz scoring scale).
4. Treatment of WD, modern tactics of drug selection.
5. Classification of glycogenoses, characteristics of types.
6. Features of the clinical course of GSDIa, Ib.
7. Features of the clinical picture of subtypes and laboratory diagnostics of GSD IV.
8. Features of the course of the hepatic form of glycogenosis type IX (CSD IX)
9. Deficiency of alpha-1 antitrypsin - features of the clinical picture, treatment.

Methodical guidelines have been created as.prof. Horobets N.I., as prof. Iemets O.V.