COMPLEX ULTRASONIC DIAGNOSTICS OF BILIARY ATRESIA IN CHILDREN

[1]Yusupalieva G.A., [2]Mursakaeva K.D.
[1,2]Tashkent Pediatrics Medical Institute
https://doi.org/10.5281/zenodo.12609057

Abstract. *Biliary Atresia is the most common cause of persistently direct (conjugated) hyperbilirubinemia in the first three months of life. It is a progressive inflammatory obliteration of the extra- and intrahepatic bile ducts whose pathogenesis remains speculative. This review updates the etio-pathological considerations, clinical manifestations, methods of diagnosis, surgical management, and results of this terrible disease in young infants. We will emphasize that persistent jaundice in the newborn period must be managed urgently, a diagnosis should be established early in life, and Kasai portoenterostomy offered to those infants with Biliary Atresia before their eighth week of life. This will allow more than one-third of children to survive.*

Keywords: *biliary atresia, hepatobiliary system, cholestatic diseases, ultrasound, liver parenchyma, echogenicity, gallbladder, hyperechoic cord, triangular cord sign, acholia, hepatomegaly, liver transplantation, cholestatic diseases.*

Relevance: Biliary atresia (BA), is a rare congenital anomaly of the biliary tract, resulting in impaired patency of the intrahepatic and/or extrahepatic bile ducts in children, leading to subsequent liver cirrhosis (LC). In the development of diseases of the hepatobiliary system in children in the first months of life, BA accounts for 45% and occupies a leading place. The average incidence is 1/10000-1/13000 live births worldwide (WHO, 2018).

Early diagnosis of the disease is extremely important, since timely surgery, and if it is ineffective, liver transplantation can significantly increase the duration and improve the quality of life of these patients.

One of the most important diagnostic methods of modern hepatology is ultrasound examination using Doppler techniques. For newborns and young children, this method is of particular value due to the presence of a number of advantages over other instrumental examination methods (informativeness, non-invasiveness, speed of obtaining results, the ability to use at the patient's bedside) and can significantly complement the differential diagnosis of the causes of neonatal cholestasis.

There is no reliable data on the dynamic changes in intraorgan hemodynamics with the progression of the pathological process. Clear ultrasound criteria have not been developed for a whole group of cholestatic diseases with a similar clinical picture, but requiring different treatment (such as Alagille syndrome, progressive familial intrahepatic cholestasis (PFIC) types 1-5, etc.)

Modern domestic literature does not cover questions about the new possibilities of the ultrasound method in the diagnosis of biliary atresia, about dynamic changes in hepatic hemodynamics in patients at the stage before and after surgery.

If left untreated, BA quickly leads to cirrhosis of the liver, liver failure and death during the first two years of life (Volynets G.V., 2017). This indicates the high relevance of the issue of early detection of this pathology using non-invasive methods.

Objective of the study. Improving the diagnosis of biliary atresia in children through the use of modern methods of complex ultrasound diagnostics.

Materials and methods. For the period 2012–2022 At the TashPMI clinic, 20 children (14 boys and 6 girls) with BA were under observation. All patients were thoroughly examined clinically, life history and illness data were studied, and the results of clinical and laboratory tests were evaluated. The diagnosis was established on the basis of clinical, laboratory and instrumental studies. All children underwent ultrasound examination of the abdominal organs in gray scale mode.

Results. The average age of the children at the time of examination in the clinic was 6.6±2.2 months. In this case, the onset of the disease occurred in the first 3 months of life. Analysis of anamnestic data showed that in 45% of cases (9 children) the mothers' pregnancies were threatened with termination. In 8 (40%) cases, low birth weight of the child was noted. Ultrasound of the abdominal organs revealed hepatomegaly in all children, increased echogenicity of the hepatic parenchyma and its heterogeneity; in 5 children (25% of cases) there was no visualization of the intrahepatic bile ducts.

The liver in the children studied had a typical location, clear, even contours, and sharp edges. The liver parenchyma was homogeneous and of medium echogenicity. When conducting a correlation analysis, a direct strong relationship was found between the linear dimensions of the liver and the growth of infants ($r=+0.86$; $p<0.05$), a less pronounced relationship ($r=+0.81$; $p<0.05$) between the linear dimensions liver and body surface area of the child. Indicators of the anteroposterior size of the right and left lobes depending on height are presented in the table

Dynamics of increase in the linear dimensions of the liver depending on the growth of the child

height	53-55 cm	56-58 cm	59-61 cm	62-64 cm	65-68 cm
RL (mm) M±o	61,2±3,9	61,7±5,4	63,1±3,8	64,2±3,6	65,9±3,6
LL (mm) M±o	43,7±2,9	44,7±2,9	44,7±3,8	43,5±2,2	47,0±5,0

Designations: RD - right lobe (antero-posterior size); LD - left lobe (antero-posterior size).

The gallbladder was identified in all 20 cases. It had the correct shape, thin (no more than 1 mm) walls, and anechoic contents. The average length of the gallbladder in children of the control group was 24.3±3.6 mm, width - 5.9±1.8 mm. The study of the common bile duct in this age group is associated with a number of technical problems (the impossibility of examining the patient while holding his breath, restless behavior of a hungry child, an increased amount of gases in the abdominal cavity, as well as the small size of the object being studied). The common bile duct in the study group was reliably determined in 4 (20%) children. The best visualization of the CBD was carried out when scanning in the position of the sensor perpendicular to the right costal arch (in the projection of the hepatoduodenal ligament). In this case, the CBD is defined as a tubular echo-negative structure with echogenic walls, located in close proximity to the trunk of the portal vein, more neutral than it. When using the color flow mode and ED mode, the color signal was not detected in it. The CBD diameter in children of this age group was 1.0±0.1 mm.

Coagulopathy, characterized by bleeding from injection sites, the appearance of bruises on the body and a petechial rash, was observed in 12 (60%) children at the age of the first 3 months of life, as well as at the onset of the disease. In the remaining 8 (40%) children, no episodes of coagulopathy, clinically and laboratory confirmed, were recorded before surgical treatment.

Biochemical analysis revealed a moderate increase in cytolytic enzyme activity: ALT 150.8±22.6 U/l, AST 240.2±36.6 U/l. The level of total bilirubin increased to 153.7±17.6 mcmol/l, with a predominance of the direct fraction of bilirubin (84.0±10.1 mcmol/l). The total protein content was 61.7±1.9 g/l, the albumin level was 35.1±1.2 g/l.

In order to determine the diagnostic value of individual ultrasound signs, their specificity, sensitivity and predictive value were determined. According to the results of our study, the most specific signs are: absence of the gallbladder (90%), hyperechoic cord at the site of the gallbladder (90%), triangular cord sign-TCS (90%). The sensitivity of the listed signs does not exceed 78.5% (absence of the gallbladder). The maximum positive predictive value was noted for the sign "gallbladder in the form of a hyperechoic cord" (90.9%), negative predictive value for STR (78.3%).

Diagnostic value of individual ultrasound signs and their combinations in the diagnosis of asthma (%).

Ultrasound signs	Sensitivity	Specificity	Positive, predictive value	Negative, predictive value
Absence of gallbladder	78,5	90	89,5	36
Gall Bladder Length < 19 мм	63,2	74	66,7	63,7
GB in the form of a hyperechoic cord	66,7	90	90,9	72
Hepatomegaly	75,7	60	84	63,2
Splenomegaly	69,8	70	88	73,7
TCS	66,7	90	84,3	78,3
TCS + Absence of gallbladder	80	95,6	80	90,1
TCS + GB in the form of a hyperechoic cord	74.1	93,2	78,7	82,4

Conclusion. Based on an analysis of the first clinical symptoms of BA in young children, it is shown that one of the most important in the diagnosis of atresia is the combination of stool acholia with prolonged jaundice and hepatomegaly. The presence of these clinical symptoms can be considered the first step in the algorithm for diagnosing BA in children in the first 3 months of life, which necessitates the need for a biochemical blood test, which is the second step in diagnosing BA. If coagulopathy, hyperbilirubinemia is detected mainly due to the direct fraction, increased levels of GGTP and alkaline phosphatase, it is necessary to carry out the third step in the diagnosis of biliary atresia - ultrasound of the abdominal organs, which reveals increased echogenicity of the hepatic parenchyma and its heterogeneity, impaired visualization of the intrahepatic bile ducts and gallbladder.

REFERENCES

1. Акилов Х.А., Матякубов Х.Н .УСОВЕРШЕНСТВОВАНИЕ ДИАГНОСТИКИ И ХИРУРГИЧЕСКОЙ ТАКТИКИ ЛЕЧЕНИЯ МЕХАНИЧЕСКОЙ ЖЕЛТУХИ У ДЕТЕЙ Вестник экстренной медицины, 2017, X (4) ст.44.

2. Акилов Х.А., Урманов Н.Т., Акилов Б.Б., Примов Ф.Ш., Асадав Ш.Я., Ходжаяров Н.Р. БИЛИАРНАЯ АТРЕЗИЯ КАК ПРИЧИНА МЕХАНИЧЕСКОЙ ЖЕЛТУХИ Вестник экстренной медицины, 2017, X (4), ст.81-82.
3. Ортиқбоева Н.Т., Абдухалик-заде Г.А. ҲОМИЛА ИЧИ ИНФЕКЦИЯСИНИНГ ҲОМИЛА ИММУНОЛОГИК ҲИМОЯСИГА ТАЪСИРИ Журнал репродуктивного здоровья и уро-нефрологических исследований 2021 СТ. 311.
4. Акилов Х.А., Матякубов Х.Н , Диагностика и хирургическая коррекция кистозной трансформации внепеченночных желчных протоков у детей, Актуальные вопросы детской хирургии, анестезиологии и реаниматологии 2017 ст. 7-8.
5. Ramachandran P, Safwan M, Srinivas S, Shanmugam N, Vij M, Rela M. The extended Kasai portoenterostomy for biliary atresia: A preliminary report. J Indian Assoc Pediatr Surg. 2016 Apr-Jun;21(2):66-71. [PubMed]
6. Bezerra JA, Wells RG, Mack CL, Karpen SJ, Hoofnagle JH, Doo E, Sokol RJ. Biliary Atresia: Clinical and Research Challenges for the Twenty-First Century. Hepatology. 2018 Sep;68(3):1163-1173. [PMC free article] [PubMed]
7. Schooler GR, Mavis A. Cystic biliary atresia: A distinct clinical entity that may mimic choledochal cyst. Radiol Case Rep. 2018 Apr;13(2):415-418. [PMC free article] [PubMed]
8. Wang L, Yang Y, Chen Y, Zhan J. Early differential diagnosis methods of biliary atresia: a meta-analysis. Pediatr Surg Int. 2018 Apr;34(4):363-380. [PubMed]
9. Chung PHY , Zheng S ,Tam PKH.Biliary atresia: East versus west. *Seminars in Pediatric Surgery.* 2020; 29150950
10. Bezerra JA , Wells RG ,Mack CL ,Karpen SJ ,Hoofnagle JH ,Doo E et al. Biliary Atresia: Clinical and Research Challenges for the Twenty-First Century.*Hepatology.* 2018; 68: 1163-1173
11. Tam PKH ,Chung PHY ,St Peter SD ,Gayer CP ,Ford HR ,Tam GCH et al.Advances in paediatric gastroenterology.*The Lancet.* 2017; 390: 1072-1082