

DIAGNOSTIC FEATURES OF CHOLEDOCHAL CYSTS IN CHILDREN

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Abstract: The paper presents an analysis of the data of ultrasound, CT, MRCP and intraoperative cholangiography in patients with bile duct cyst. Echographic data are presented in 86 patients, CT in 61, MRCP in 8 and intraoperative cholangiography in 73 patients with this pathology. Ultrasound is a highly sensitive and informative screening method for the diagnosis of choledochal cysts in children. To increase the specificity of ultrasound with cysts of the common bile duct, a dynamic study of the contractility of the biliary system after taking a "choleretic breakfast" is proposed. Currently, in the diagnosis of pathology of the biliary tract, CT of the abdominal cavity and MRCP are effective non-invasive methods, alternative to invasive ERCP. It has been established that the final verification of the form of bile duct cysts, abnormal fusion of the pancreatic duct into the cyst cavity is determined using MRCP and intraoperative cholangiography, which allows you to choose the optimal method of surgical treatment.

Keywords: choledochal cysts, diagnosis, anatomical forms, children.

Congenital choledochal cyst (CC) is a relatively rare abnormality in the development of bile ducts (DBD). In recent years, the number of published messages has been increasing among children and adults [1,7]. This is due to the introduction of modern highly informative diagnostic methods for malformations of gastrointestinal tract in children [4,5,8,9,11]. At present, with the help of ultrasound, it is possible to establish a diagnosis of CC by antenatal [6]. However, ultrasound also has disadvantages. These include the difficulties in visualizing the distal parts of the gastrointestinal tract, the relationship of the cyst with the pancreatic duct. Endoscopic retrograde cholangiopancreatography (ERCP) is widely used for the diagnosis of chronic obstructive heart disease [8]. However, some authors are skeptical of conducting ERCP in children, given the complexity of the method, as well as the possible risk of infection of the bile duct and pancreatic ducts [2,10,11]. In recent years, computed tomography (CT) or magnetic resonance cholangiopancreatography (MRCP) has been used to diagnose choledochal cysts. According to some authors, the informative value of CT in choledochal cysts is 80 - 90% [1]. However, the CT scan is not without negative aspects, since it requires a large exposure, which leads to blurring of the image of the object and, in addition, gives significant exposure. MRCP is an informative and non-invasive diagnostic method [9]. It has been reported that MRCP has a sensitivity of 90% - 100% for detecting CC [7]. The advantages of this method are the absence of radiation exposure, the possibility of visualization in a three-dimensional image, and a contrast agent is not used, which excludes the possibility of allergic reactions, ascending infection and reactive pancreatitis. However, according to some authors, MRCP is much lower sensitivity for determining the fusion of the pancreatic duct and the common pancreatobiliary canal, also has an age restriction [7].

Preoperative diagnostic methods for CC begins with ultrasound and is supplemented by modern research methods (CT, ERCP, MRCP), and intraoperative cholangiography (IOCH) during surgery [3]. However, the diagnostic value of MRCP and ERCP in childhood causes certain contradictions and to date in pediatric practice has not been widely used.

However, to date, in the diagnosis of this peculiar and relatively rare defect of the biliary system, there are many unsolved problems that require further improvement. In some publications, difficulties in diagnosing this pathology, various methods of completing operations, even tactical errors made during the operation are noted. CC is often not recognized not only during clinical examination, but even during surgery. From the foregoing, it can be noted that the problem of diagnosing patients with CC is currently not sufficiently developed.

The purpose of the study was to determine the role of various instrumental studies in the diagnosis and determination of anatomical forms of choledochal cysts in children according to the clinic.

Materials and research methods. 86 patients with choledochal cyst, as well as 4 children with diseases that simulated this anomaly (solitary cyst of the liver - 2, doubling of the gallbladder - 1, dropsy of the gallbladder - 1) were treated in the clinical bases of the Department of Hospital Pediatric Surgery of Tashkent Pediatric Medical Institute in 1979 - 2020. The age of patients ranged from 15 days to 16 years. Among the patients, girls predominated - 76.7%, the ratio of boys and girls was 1: 3, which corresponds to literature data.

Patients underwent comprehensive clinical, laboratory and radiation diagnostic methods: ultrasound and CT of the abdominal organs, MRCP, IOCH, histomorphological examination of the removed biomaterial.

Ultrasound of the liver and gastrointestinal tract is a screening diagnostic method for CC. Ultrasound was performed according to the standard method using Aloka SSD 1400 devices with multi-frequency sensors: vector - 1.4 MHz, convex - 3-8 MHz and linear 5-8 MHz, as well as the Voluson R 730 Pro. CT was performed in 61 (67.8%) of 90 patients. The main indications for CT were abnormalities of the gastrointestinal tract diagnosed by ultrasound. The study was carried out in the patient's position on the back using a Philips "Brillians-64" apparatus (matrix 1024x1024, slice thickness - voxel size - x, y, z - 0.33x0.33x0.34 mm). The radiation load per patient, on average, was 1.2 mSv. MRCP was performed in 8 (8.9%) patients. The study was carried out on a Samsung Magnetom Espree magnetic resonance imager with a superconducting magnet of a magnetic field of 1.5 T using a coil for body weight (Whole body). IOCH was performed in 73 (84.9%) of 86 operated patients with CC, in 13 (15.1%) cases, for various technical reasons, it was not possible to conduct it.

Results and its discussion. The rarity of the pathology and the polymorphism of clinical signs of CC leads to significant difficulties in the diagnosis of the disease. Most hospitalized patients were referred to the clinic with erroneous diagnoses. Mistakes in referral diagnoses occurred in 38 (44.1%) cases. The reason for such diagnostic errors at the prehospital stage becomes understandable, given that, due to its rarity, this pathology is little known to pediatricians and general practitioners, even surgeons.

An ultrasound scan of the abdominal organs examined the liver, extra- and intrahepatic bile ducts, pancreas, pancreas. Moreover, in patients with CC, an enlarged choledoch located along the portal vein was revealed, exceeding normal age parameters with fluctuations from 1.0 cm to 18.0 cm, with various forms of expansion throughout the choledoch, communicating with an increased or decreased volume of the gallbladder, often with calculi characteristic acoustic track (Fig. 1).



Fig. 1. Echoscopic picture of spherical expansion of the common bile duct with a reduced gallbladder in volume (a); with an increase in the volume of the gallbladder (b); calculous cyst of the common bile duct (c).

An ultrasound scan was performed on 15 (17.4%) of 86 patients who underwent gallbladder size, and 12 of them had signs of chronic inflammation of its wall with deformation in the cervical ductal segment. 5 (6%) patients who had previously undergone inadequate primary operations (cyst drainage) in other hospitals, upon admission to our clinic, did not allow us to differentiate the gallbladder due to the pronounced adhesive process and its intimate fusion with the dilated duct. In 14 (16.7%) of 84 patients with CC in the gall bladder, dense inclusions and calculi with a characteristic acoustic pathway were revealed. In 11 (78.6%) of 14 patients with bile duct cyst, stones were established before surgery as a complication of bile duct cyst, and in 3 (21.4%) cases it was found during surgery. Echography in 8 out of 14 patients with no inclusions in the gallbladder revealed different densities in the lumen of the enlarged bile duct with a pronounced acoustic pathway, indicating the presence of calculus - choledocholithiasis, confirmed during surgery. Stones in 3 patients were localized in the gall bladder, in 8 - in the enlarged common bile duct, in 3 - in the gall bladder and in the cavity of the cyst.

The data presented are characteristic, but non-specific for CC, since these changes are also observed in other pathologies of the liver, biliary tract and diseases of the abdominal organs, accompanied by the presence of a tumor-like formation. In our observations, diagnostic difficulties were observed in patients with liver echinococcosis (3), a solitary cyst of the liver (2), doubling (1) and dropsy (1) of the gallbladder, pancreatic cysts (1). For differential diagnosis with the above diseases, to increase the reliability of ultrasound of CC, we suggest conducting dynamic functional echocholecystography after a "choleretic breakfast." In 66 (73.3%) cases, the contractile and evacuation functions of the biliary system (size, shape of the gallbladder and cystic dilated bile ducts) with choleretic breakfast were additionally investigated. The study was carried out in dynamics: on an empty stomach and after 30 minutes, after giving an egg yolk or a solution of magnesium sulfate, which have a cholekinetic effect. At the same time, the gallbladder and the cyst wall contract, which manifests itself in a change in their size (or one of them) and in a shape different from the original ones, which increases the specificity characteristic of cystic transformation. Similar changes are not observed with solitary or parasitic cysts of the liver and pancreas.

Ultrasound is a sufficient study for CC surgery. However, the expansion can be localized in any part of the biliary tract in the form of total expansion or its individual sections. In addition, it is not possible to judge the abnormal flow of a separate segmental duct into the cystic dilated choledoch or the presence of an abnormal biliary-pancreatic fusion, often with cystic expansion of the bile ducts (11 - 12.7% of our 86 observations). An indirect echoscopic sign of this condition can be considered an expansion of the pancreatic duct. Biliary-pancreatic cysts have features in the clinical course, there is a possibility of iatrogenicity (the probability of damage when the cyst is excised, full or partial clamping

during ligation) during surgery. The data presented indicate the advisability in such cases to supplement the diagnostic tests of CT and MRCP before surgery, and during the operation - IOCH. Currently, in the diagnosis of pathology of the biliary tract, CT of the abdominal cavity and MRCP are effective non-invasive methods, alternative to invasive ERCP. For the diagnosis of CC in 61 (67.8%) patients, MSCT was used, which adds additional information to the ultrasound data. With MSCT, it is possible to assess the condition of the extra- and intrahepatic bile ducts, the Wirsung duct, the relationship of the cystic formation with adjacent organs, and determine the form and type of cystic expansion (Fig. 2).

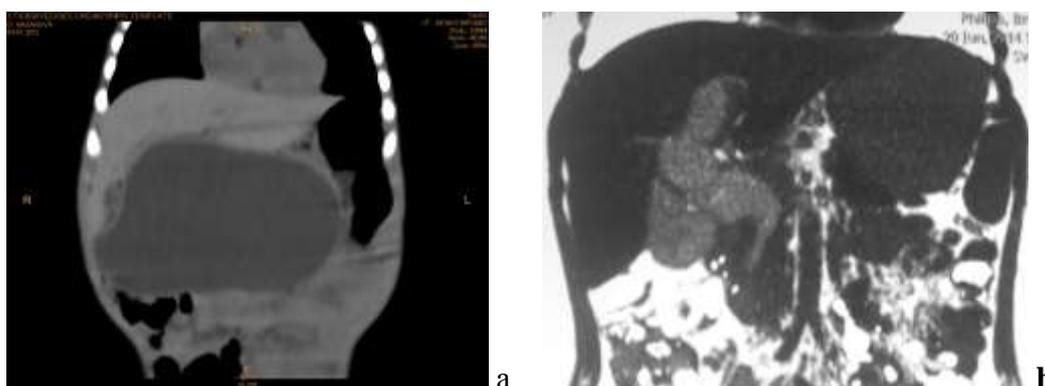


Fig. 2. a) MSCT picture of a giant saccular expansion of the common bile duct with a reduced size of the gallbladder. Compression of nearby organs of the pancreatobiliary zone is noted; b) MSCT cholangiography. Intrahepatic bile ducts are asymmetrically dilated. The common bile duct is cystically dilated, type IV. Wirsung duct not visualized.

The value of the CT diagnostic information increases with the image in 3D reconstruction.

Currently, we often used MRCP for the diagnosis of CC in children of older age groups. In our observations, 8 (8.9%) patients underwent an MRCP study. This method is the most promising and informative way to diagnose CC with abnormal pancreatobiliary fusion (Fig. 3).

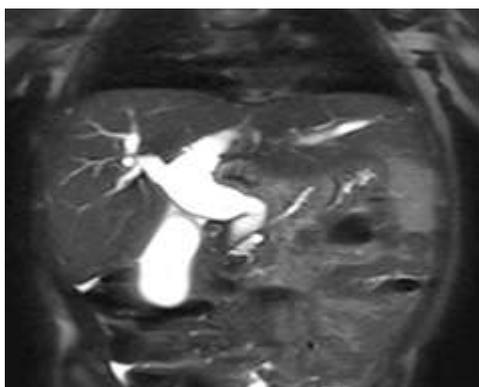


Fig. 3. MRCP. There is a biliary and pancreatic cyst of the common bile duct. Wirsung duct opened into the cyst cavity.

The absence of radiation exposure, low invasiveness, and the need for special training are the main advantages of the method. However, the disadvantages of this method is the limitation when used in children of younger age groups. Therefore, the final diagnosis and method of completing the operation was established using IOCH. The study was performed after the application of microdrainage cholecystostomy in the bottom of the gallbladder. For contrast, 76% water-soluble preparation (urographin, verographin) was diluted with saline in

a ratio of 1: 1. In order to obtain an image of the intrahepatic bile ducts, to determine the contrast in the duct of the pancreas and duodenum, the contrast medium was injected to moderate tension of the gallbladder and cyst without clamping the hepatoduodenal ligament. X-ray images were taken using a portable X-ray unit at the time of artificial apnea and in the regimes selected according to the age and constitution of the patient. After performing an x-ray, the contrast agent was evacuated by suction. This study was conducted in 73 (84.9%) of 86 operated patients with CC, in 13 (15.1%) cases for various technical reasons it was not possible to conduct it. With IOCH, it becomes possible to detail the results of preoperative ultrasound and CT examinations to more accurately determine the shape, size of the cystic formation, the nature of the expansion of the extra- and intrahepatic bile ducts, the symmetry of the expansion of the lobar ducts, and the possibility of additional abnormal bile ducts opening into the lumen of the cystic expansion.

The data of complex preoperative (ultrasound, MSCT) and IOCH studies showed the heterogeneity of the forms, sizes and length of the cystic expansion within the extra- and intrahepatic ducts and finally verify its anatomical version. Cystic enlargements are most often localized in the extrahepatic segment of the bile ducts of various lengths and diameters in the form of types I and IV and correlates with literature data on the rarity of types II, III, V, and VI of cystic lesions of the biliary tract. With IOCH in 54 (74%) children, the shape and size of the cystic expansion usually coincided; in 19 (26%) cases, there was a slight increase in the size of the cystic expansion, probably due to stretching of its wall during tight filling with contrast medium during cholangiography (Fig. 4).



Fig. 4. IOCH. Cystic expansion of extrahepatic bile ducts with symmetric expansion of intrahepatic bile ducts.

At IOCH performed in 73 patients, type I cystic enlargement was observed in 54 (74%) as: expansion in a limited area or along one of the ducts - the common bile or hepatic in 16 (29.6%) patients. We characterize it by extrahepatic localized segmental expansion. In 38 (70.4%) patients, the expansion covered the common bile and hepatic ducts - the general extrahepatic cystic expansion. In 17 (23.3%) of 73 cases, expansion of the extra- and intrahepatic ducts (type IV) was simultaneously observed - diffuse expansion of the bile ducts, and in 6 symmetrical: the same expansion of the right and left hepatic ducts; 11 asymmetric: the predominance of expansion of the right or left duct. In 2 (2.7%) patients, a slight expansion of extrahepatic ducts and intrahepatic cystic lesion was observed throughout - atypical form - FF type.

The form of cystic expansion in each patient is peculiar: bag-shaped (19), spindle-shaped (17), spherical (13), cylindrical (24). It is practically difficult to find the same cysts in two different patients.

Local expansion of individual segments of the extrahepatic bile ducts often has a rounded or cylindrical shape. Expansion within the common bile duct - spindle-shaped, diffuse expansion outside and inside the hepatic ducts - has a spherical or saccular shape.

IOCH allows also to determine non-extended local narrowing or expansion. It is necessary to distinguish between cystic expansion of the bile ducts without stenosis of the terminal section of the common bile duct (52 - 71.2%), when the violation of the outflow of bile is short-term and is due to functional discoordination of the sphincter of Oddi, and cysts with stenosis of the terminal section of the common bile duct (21 - 28, 8%). Some authors, apparently, exaggerate the importance of stenosis of the distal part of the common bile duct in the formation of suprastenotic expansion, since in the presence of severe stenosis there should be a violation of the outflow of bile and, accordingly, obstructive jaundice. However, we observed 3 cases of cystic expansion with gigantic cyst sizes, but without the effects of obstructive jaundice. With true stenosis of the terminal section of the common bile duct in 21 (28.8%) cases, despite the tight filling of the cyst, the passage of contrast into the duodenum did not occur. With this option, a secondary suprastenotic expansion of the external and intrahepatic ducts is possible. The uneven expansion of the bile ducts at various levels primarily indicates an anomaly in its structure. Depending on the degree of impaired patency in the terminal section of the common bile duct, a cystic anomaly can manifest itself only as a pain syndrome, or, with a significant degree of stenosis, the pain syndrome is combined with obstructive jaundice and signs of biliary hypertension. The presence in some cases of pain without signs of obstructive jaundice and the periodic development of biliary hypertension also confirms impaired patency of various origins and degrees in the terminal section of the common bile duct.

More valuable information during cholangiography was obtained about the relationship between the common bile duct and pancreatic ducts. If there is a message of cystic expansion of the bile ducts with the pancreatic duct, a biliary-pancreatic cyst is formed, which occurred in 11 (12.8%) of 86 operated children with CH. This confirms the need to distinguish between biliary cysts, the content of which is bile, and biliary-pancreatic cysts, in which pancreatic enzymes enter the cystic dilated common bile duct due to the fusion of the Wirsung duct. With a biliary-pancreatic cyst, the flow of pancreatic juice into the cavity of the cyst promotes the development of a pathological vicious circle - enzymatic cholecystocholangitis; and stagnation and difficulty of the outflow from the cavity of the cyst causes the development of reactive pancreatitis with the peculiarities of the clinical course. In 11 patients, a frequently recurring pain syndrome is caused by reactive pancreatitis due to biliary pancreatic fusion. At the same time, the flow of pancreatic juice into the cavity of the cyst under conditions of bile stasis causes circulatory disorders and necrosis of the walls of the cyst of the bile ducts and contributes to spontaneous rupture of the common bile duct cyst. In our observations, out of 8 patients with perforation of the bile duct cyst, 2 were diagnosed with biliary pancreatic cyst during cholangiography.

Thus, the diagnosis of cystic anomalies of the bile ducts, despite the successes of modern medicine, is often still a difficult task. This is due to the lack of clear symptoms, a peculiar dynamism of clinical symptoms, which leads to a multidimensional focus of research and differential diagnosis.

Conclusions:

1. Ultrasound is a highly sensitive and informative screening method to diagnose choledocha cysts in children. In differential diagnosis of choledocha cysts with cystic formations of the hepatobiliary zone, functional dynamic echolecystography with "choleretic breakfast" increases the specificity of diagnosis of choledocha cysts.
2. To establish a biliary pancreatic cyst and abnormally inflowing segmental ducts into cystic expansion, ultrasound data are not informative enough, which requires additional CT, MRCP and IOCH.

3. The complex approach to diagnostics allows to orientate with greater accuracy in various anatomical forms of choledocha cysts, to choose tactics and optimal term of surgery.

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