

The Role of Radiation Research Methods in the Diagnosis of the Biliary Tract Cystic Transformations in Children

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Abstract The aim of the study is to improve diagnostic and treatment results of various forms of choledochal cyst transformations in children through the use of highly informative diagnostic methods and the introduction of new treatment technologies. **Introduction.** Choledochal cysts transformation is a relatively rare abnormality of the biliary tract. In 80% of cases choledochal cysts transformation is detected in childhood and adolescence. In most cases, this pathology is diagnosed in infants and children (in 25% of cases - in children of the first year of life and in 60% - at the age of 10 years). **Material and methods.** The research is based on the analysis, diagnostics and treatment of 78 patients aged from 2 months to 18 years with choledochal cysts transformation, who were observed in the Department of Pediatric Surgery of the Republican Research Centre of Emergency Medicine for the period from 2005 to 2021. **Results and discussion.** 78 children were examined and treated: 56 (71.8%) girls and 22 (28.1%) boys. Ultrasound investigation of the liver and abdominal organs was performed in all patients of the control and main groups. Great importance was attached to retrograde endoscopic examinations in the diagnostics of various clinical forms and types of choledochal cysts. All patients were performed the following surgical procedures: in 39 (50.0%) cases - resection of the choledochal cyst with the imposition of a hepaticenteroanastomosis with a skeleton ring on the Roux loop of the small bowel, in 32 (41.0%) patients - resection of the choledochal cyst with the imposition of hepaticoenteroanastomosis on the Roux loop of the small bowel, in 6 (7.6%) cases, supraduodenal choledohoduodenoanastomosis was applied, in 1 (1.4%) patient - Kerr's drainage (due to perforation of the choledochal cyst). **Conclusion.** Timely recognition of congenital dilation of the common bile duct presents significant difficulties. Choledochal cysts in children belong to the group of rare diseases which stipulates its difficulty in diagnosing them. A thorough assessment of clinical data and anamnesis study allow to suspect the disease, and endoscopic retrograde cholangiopancreatography, percutaneous and transhepatic cholangiography, magnetic resonance cholangiopancreatography helps to clarify the diagnosis, to determine the nature and type of lesion.

Keywords Choledochal cysts, Anastomosis, Children

1. Introduction

Choledochal cysts transformation (CCT) is a relatively rare abnormality of the biliary tract. In 80% of cases choledochal cysts transformation is detected in childhood and adolescence [1-6]. In most observations this pathology is diagnosed in infants and children (in 25% of cases - in children of the first year of life and in 60% - at the age of 10 years). In the adult population, the disease is more common in young women, the ratio of women/men is approximately 4/1 [1-2,5-7]. The morbidity rate is 1 per 100,000 population in Western countries [1-2,4-6,8] and 1 per 13,000 population in Japan [2,5-6].

The pathogenetic basis of the disease is considered to be congenital hypoplasia of the muscular layer of the duct, neuromuscular dysfunction of the sphincter of Oddi [2].

The clinical picture of CCT has a characteristic triad of symptoms: abdominal pain (90-92%), intermittent jaundice (43-70%), palpable tumor-like formation in the right hypochondrium (25-60%). In 20% of patients, the clinical picture may be absent for a long time [9,2-3].

The difficulty of diagnosing CCC is due to the fact that it can be asymptomatic in the early stages. The lack of clear clinical manifestations significantly complicates the early diagnostics of congenital diseases of the bile ducts [10].

Full-fledged diagnostics of bile duct cysts is possible only with the help of special research methods. It is important to have the most complete understanding of the cyst size in various parts of the hepatobiliary tract, its contents, the relationship of the cyst with the surrounding organs and structures. This is important not only for assessing the degree of existing pathological changes, but also for choosing the optimal method of surgical correction of pathology [11].

The introduction of ultrasound investigation (USI), multi-spiral computed tomography (MSCT), magnetic resonance imaging (MRI), including magnetic resonance

cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography (ERCP), as well as percutaneous and transhepatic cholangiography and cholangiostomy (PTHC and PTHCS) into clinical practice has allowed not only to early diagnose of cystic transformation of the common bile duct, dynamic monitoring of patients in order to control the size of dilatation, but also to choose the optimal period of surgical intervention [12-13]. They allow us to get data not only about the existence of the cyst, its size and position, but also gives an idea of the cyst walls, its contents, relationships with the surrounding organs, the nature of their changes. Their efficiency is 75.8—79.2%. The most reliable data is provided by ERCP, but recently it is rarely used due to its high invasiveness [3].

The aim of the study is to improve diagnostic and treatment results of various forms of choledochal cyst transformations in children through the use of highly informative diagnostic methods and the introduction of new treatment technologies.

2. Material and Methods

The research is based on the analysis, diagnostics and treatment of 78 patients aged from 2 months to 18 years with choledochal cysts transformation, who were observed in the Department of Pediatric Surgery of the Republican Research Centre of Emergency Medicine for the period from 2005 to 2021.

In the research material, along with uncomplicated forms of CCT, we included complicated forms of this pathology with clinical manifestations of mechanical jaundice and purulent cholangitis. Complicated forms of cystic changes in choledochus had the peculiarity of progressing according to their own laws, acquired the features of an independent disease and, accordingly, required a separate treatment program.

Depending on the tactics and method of therapeutic and diagnostic procedures, all patients with various forms of CCT were divided into 2 groups: control and main. The control group included 39 (50%) patients who were treated with traditional therapeutic and diagnostic tactics. In 39 (50%) patients of the main group, the treatment and diagnostic process was supplemented by modern methods of radiation and endobiliary diagnostics, as well as a new method of surgical treatment of choledochal cysts transformations in children using an external metal frame.

All patients with various forms of CCT were examined urgently. The diagnostic process at the availability of contact began with finding out the complaints of patients. In parallel, a thorough but rapid physical examination was carried out using the methods of questioning, examination, palpation, auscultation, percussion.

Further, the examined patients were performed laboratory examination and instrumental diagnostics, the purpose of which was: to clarify or confirm the presence of CCT with determination of its clinical form and type, to assess the

general condition of patients and to choose treatment tactics with the determination of an adequate method of surgical intervention.

3. Results

78 children were examined and treated: 56 (71.8%) girls and 22 (28.1%) boys. In total, X-ray examination of the chest and abdominal organs was performed in 52 (66.6%) patients.

Ultrasound investigation (USI) of the liver and abdominal organs was performed in all patients of the control and main groups. USI was performed in 100% of all sick children, which allowed to diagnose a choledochal cyst in 79.6% of cases (n=60). The most characteristic signs were: the diameter of the extrahepatic and intrahepatic bile ducts, the nature of their contents, the presence of acoustic shadows, as well as the state of the liver, pancreas and neighboring organs (Tab. 1).

Table 1. Frequency of ultrasound signs occurrence in patients with cystic bile duct extensions, n=72

Ultrasound sign	Detection frequency	
	absolute	%
Congenital CCT with dilation of the main intrahepatic ducts	33	42.3
Dilation of the extrahepatic bile ducts diameter	60	76.9
The presence of echoes in the bile ducts lumen	24	30.7
With acoustic shadow	32	41.0
Without acoustic shadow (putty-like bile))	48	61.5
Liver cyst	10	12.8
Cyst of the pancreas head	3	3.8

ERCP for the purpose of diagnosing CCT was performed in only 18 (23.0%) patients including 8 patients who were conducted repeated investigations. We used special investigations in the main group of patients in the form of modern methods of radiation and endovisual diagnostics to improve the diagnostics and treatment results of CCT various clinical forms and types. The special methods of radiation and endovisual diagnostics and treatment used by us in the main group of patients included MSCT-cholangiography, MRCP, as well as PTHCG and PTHCS.

MSCT was performed in 27 (34.6%) patients according to the following protocol: scanning of the abdominal organs with thin sections (0.625 mm) after contrast of the biliary tract (BT). Then 3D reconstruction was performed in the frontal plane using a minimum intensity projection with a layer thickness of 3-5 mm (Fig. 1).

In 17 (21.8%) patients of the main group, we used the MRCP technique, which is based on the use, in contrast to ionizing radiation in radiography, of a strong magnetic field that is harmless to humans.

It should be noted that in 6 (7.7%) patients, cannulation of

the major duodenal papilla (MDP) for ERCP was not possible due to technical reasons. In these patients, the PTHCG technique was used as an alternative method to diagnose and determine the nature of CCT.



Figure 1. 3D-reconstruction of cystic dilation of the common bile duct of type I

4. Discussion

A mandatory set of laboratory diagnostic methods for accurate and timely diagnosis of all patients with various clinical forms of CCT was carried out, including the determination of clinical and biochemical blood parameters, characteristics of the acid-base state (ABS). It was mandatory to assess the level of hemoglobin, red blood cells, hematocrit, to determine the leukocytosis with the leukocyte formula and the rate of erythrocyte sedimentation (ESR).

The most important advantage of ultrasound investigation was its non-invasiveness, speed of execution (3-5 minutes), high sensitivity in detecting direct and indirect signs of various clinical forms and types of CCT, safety and ease of execution, allowing for a repeat study with an assessment of the process dynamics. At the same time, the resolution capabilities of echography in patients with CCT are significantly reduced in some complex clinical situations. In this regard, USI in the diagnostics of CCT was considered as a primary, screening investigation.

In the study of patients with CCT, the presence of a cavity formation in the the liver gate area was determined in all cases (Fig. 2), but it was not always possible to clearly differentiate where it came from (Fig. 3). At the large size of the cyst it was difficult to determine whether it comes from the liver itself (solitary or parasitic cyst) (Fig. 4), the head of the pancreas, or it is directly a hepaticocholedochal cyst.

When performing the ERCP, strict compliance with the technique was carefully monitored. In order to prevent "artificial" pneumobilia and obtain artifacts on cholangiograms, the catheter was filled with saline solution

before MDP cannulation to prevent air entering to the lumen of the choledochus. Retrograde catheterization was performed taking into account the angle of confluence of the bile and pancreatic ducts at the mouth of the MDP and without excessive effort. The catheter position was controlled by injecting 2-3 ml of contrast medium without pressure. ERCP for the purpose of diagnosing CCT was performed in only 18 (23.0%) patients, including 8 patients who were conducted repeated investigations (ERCP). In 6 patients of the control and main groups, ERCP revealed the presence of various clinical forms and types of CCT, which was an indication for performing decompression of the BD by endoscopic papillosphincterotomy (EPST) (Fig. 5, 6).

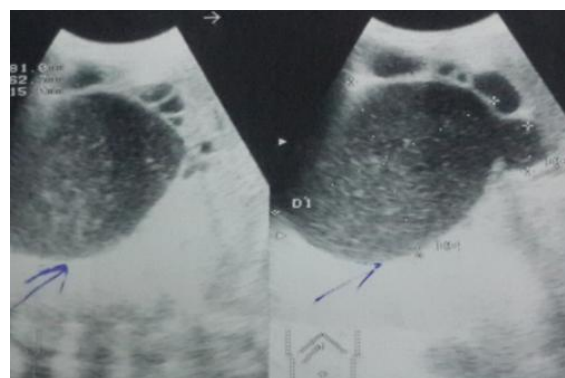


Figure 2. Cystic formation of hepaticocholedochus



Figure 3. Cystic formation of the intra-and extrahepatic bile ducts

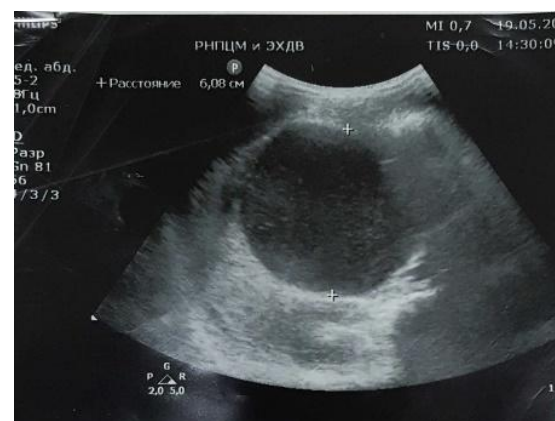


Figure 4. Volume formation in the IV-V-VI segments of the liver

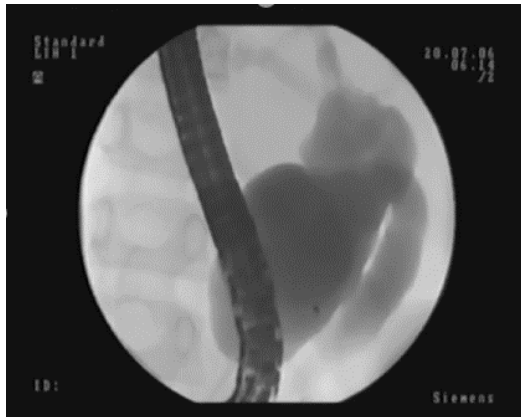


Figure 5. ERCP. Cystic dilation of the intra-extrahepatic bile duct

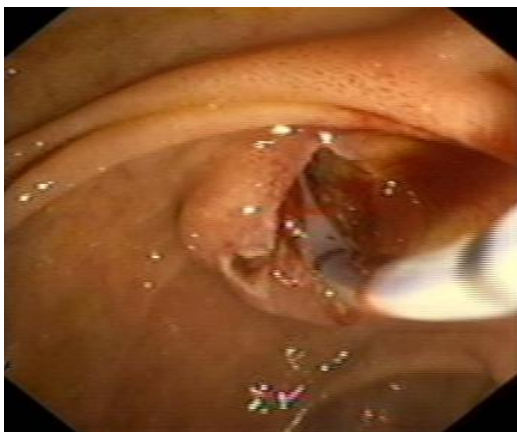


Figure 6. EPST by L. Demlihg and K. Kawai method (1980)

EPST was performed according to the method described by L. Demlihg, K. Kawai (1980). The intervention was performed with the help of a string papillotome as follows: a sphincterotomy was inserted into an ampoule of MDP and oriented in such a way that the direction of the incision corresponded to 11 o'clock. When the whitish tissues of the choledochus or the hole from which the bile flows appeared in the field of view, a second attempt of cannulating was made (Fig. 7).

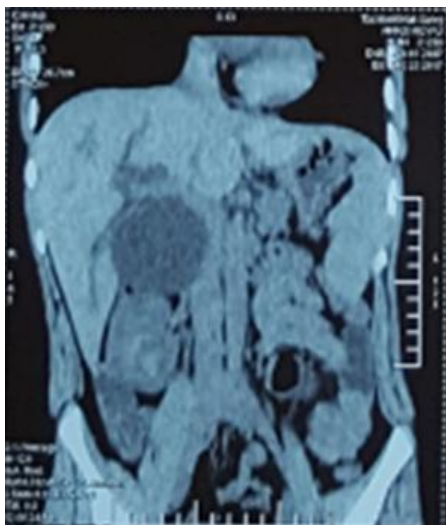


Figure 7. MSCT. The common bile duct cystic dilation of type I

Cystic dilations of the extrahepatic bile ducts on MSCT were volumetric cavities with a diameter of 5.0 to 12.0 cm, with 2-3 mm of wall thickness, with smooth, clear contours. We performed a contrast of the bile ducts at a complex interpretation of the detected cysts origin (Fig. 8).

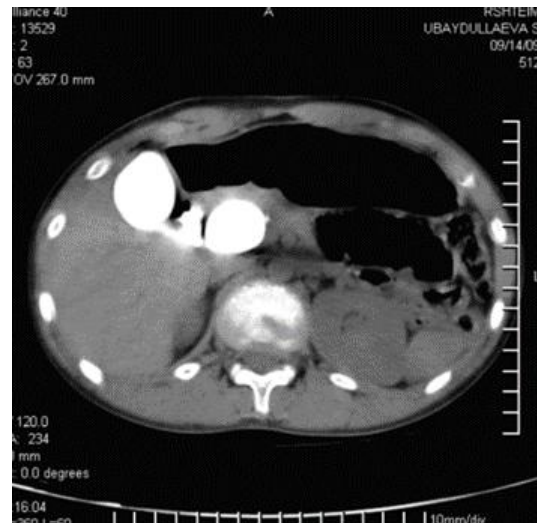


Figure 8. After ERCP

In our observations, the cyst was located almost from the liver gate to the confluence of the hepaticocholedochus in the duodenum.

MRCP was performed in the horizontal position of the patient in the following pulse sequences: T-2 coronar (T-2 fatsat) and dual-echo (T-1 infaseoutfase) in the axial projection, followed by 3D-MRCP- reconstruction) (Fig. 9, 10, 11).



Figure 9. MR-signs of a huge cyst (echinococcal?) in the mesenteric zone with compression of the choledochus and the common bile duct

The analysis of the results obtained in 23 (29.5%) patients clearly shows that the diagnostic value of MRCP in all evaluation criteria is almost equal to the methods of direct X-ray cholangiography. MRCP clearly visualized the gallbladder, intra- and extrahepatic bile ducts, cholelithiasis, regardless of the concretions location. In this regard,

we were able to determine the presence of CCT in all the examined patients, and in its complications with mechanical jaundice, it allows us to choose the optimal method of decompression.

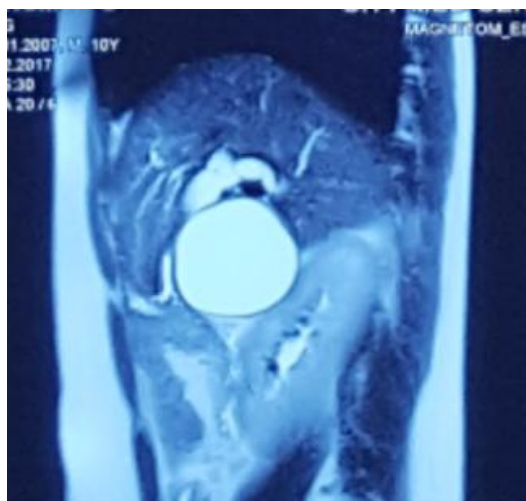


Figure 10. MR-signs of hepaticocholedochus cystic transformation



Figure 11. MRI-cholangiography. CCT and choledocholithiasis

The analysis of the above data testifies that the use of MRCP contributes to the early and reliable recognition of CCT presence and the development of its complicated forms (mechanical jaundice (MJ), purulent cholangitis (PC)). MRCP was performed in only 6 (7.7%) patients of the control and main groups using the Lunderquist Guide Wire. The study was conducted under general intravenous anesthesia under X-ray and television control using lateral access from the 7-8 intercostal space along the right axillary line. The liver and bile ducts were punctured with thin-walled Chiba needles (22G).

In PTHC, the main information was obtained by contrasting the ductal system by determining the degree of violation of the bile passage in the form of a partial or complete block of the common bile duct (CBD) with the expansion of the bile ducts proximal to the block (Fig. 12, 13).



Figure 12. PTHC. CCT at the level of porta hepatis

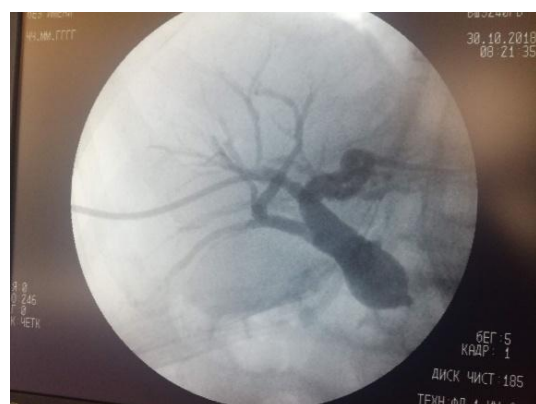


Figure 13. PTHC

It should be noted that in addition to diagnostics, percutaneous-transhepatic endobiliary interventions such as PTHCS were used in 8 (10.2%) patients of the control and main groups at complications of CCT with MJ and GC for therapeutic purposes as the first stage of surgical correction of this pathology. PTHCS in these patients proved to be an effective and quite adequate method of external bile excretion for the correction of the MJ and PC phenomena in these patients. All patients were performed the following surgical procedures: in 39 (50.0%) cases - resection of the choledochal cyst with the imposition of a hepaticenteroanastomosis with a skeleton ring on the Roux loop of the small bowel, in 32 (41.0%) patients - resection of the choledochal cyst with the imposition of hepaticenteroanastomosis on the Roux loop of the small bowel, in 6 (7.6%) cases, supraduodenal choledohoduodenoanastomosis was applied, in 1 (1.4%) patient - Kerr's drainage (due to perforation of the choledochal cyst).

5. Conclusions

Timely recognition of congenital dilation of the common bile duct presents significant difficulties. Choledochal cysts in children belong to the group of rare diseases which stipulates its difficulty in diagnosing them. A thorough assessment of clinical data and anamnesis study allow to suspect the disease, and endoscopic retrograde

cholangiopancreatography, percutaneous and transhepatic cholangiography, magnetic resonance cholangiopancreatography helps to clarify the diagnosis, to determine the nature and type of lesion.

A radical operation for cysts of the extrahepatic bile ducts is considered to be the maximum excision of the cystically altered walls of the hepaticocholedochus and the imposition of a hepaticenteroanastomosis with a skeleton ring on the Roux loop of the small bowel.

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