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A Unique Type or Variation of Bile Duct Cyst in a 9-Year-Old Girl: A Remarkable Case Study

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Abstract. Patients with bile duct cysts require careful radiological assessment of the hepatobiliary system prior to surgical intervention. This clinical case is uncommon with an atypical clinical presentation and radiological findings. According to the most widely used classification of choledochal cysts, this case presents a combination of Type I and Type IV of choledochal cyst (CC) combining the form of extra, intrahepatic bile ducts and cystic duct dilations.

Keywords: bile duct cyst, Roux-en-Y hepaticojejunostomy, common bile duct, choledochal cyst.

Unikalus tulžies latako cistos atvejis ar jo atmaina. 9 metų mergaitė: išskirtinis tyrimo atvejis

Santrauka. Pacientų, kuriems yra tulžies latako cistų, prieš chirurginę intervenciją būtinas kruopščiai įvertinti hepatobiliarinę sistemą, o tam reikia atlikti jos radiologinį tyrimą. Mūsų klinikinis atvejis yra neįprastas, nes jis pasižymėjo reta klinikine prezentacija ir radiologijos tyrimų rezultatais. Remiantis labiausiai paplitusia choledochinių cistų klasifikacija, nustatyta, kad šiuo atveju buvo choledochinės cistos I tipo ir IV tipo derinys su ekstra- ir intrahepatiniais tulžies latakais bei cistinių latakų išsiplėtimais.

Raktažodžiai: tulžies latako cista, Roux-en-Y hepatikojejunostomija, bendrasis tulžies latakas, choledochinė cista.

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Introduction

A bile duct cyst is a congenital anomaly of bile ducts that often pose diagnostic and therapeutic challenges for paediatric surgeons. Usually, such dilation arises from extrahepatic bile ducts (Type I), but in more complicated cases, intrahepatic bile ducts are affected (Type IV, V) [1]. Biliary cysts are approximately 1:100,000 to 1:150000 in Western countries and 1 in 13,000 in certain parts of Asia 2,3]. In 80% of cases, the anatomical variant of the common bile duct (CBD) cyst by classification of biliary cyst is of Type I. At the same time, Type IV is rare, especially in Baltic countries with a population of only 1.3 million in Estonia, 1.9 million in Latvia, and 2.8 million in Lithuania. In the last two decades, only two patients with Type IV-A choledochal cysts have been diagnosed and treated out of 16 congenital common bile duct cyst cases in Latvia [4]. A report from Finland suggests that the incidence of biliary cysts has increased from 1:128,000 to 1:38,000 over the past 40 years. [5]. We present a clinical case of a type of CC yet to be described in the literature that might be classified under an updated version of the classification of biliary cyst soon.

We aim to describe the characteristics, diagnostics, and postoperative outcome of a child diagnosed with a variant of CC that combines Type I and IV in Latvia.

Case report

A 9-year-old girl presented with three-day long complaints of abdominal pain, vomiting and nausea for the first time. Upon arrival at the Emergency Department (ED), the patient was examined objectively and radiologically. In objective examination, there were no signs of cholestasis, and symptoms of peritoneal irritation; the patient presented some sensitivity to palpation in the periumbilical region. No change in urine color was observed. Laboratory results showed no elevated leukocyte count, inflammatory markers, and blood lipase concentrations; only elevated bilirubin, and alanine transaminase (ALAT) levels were observed.

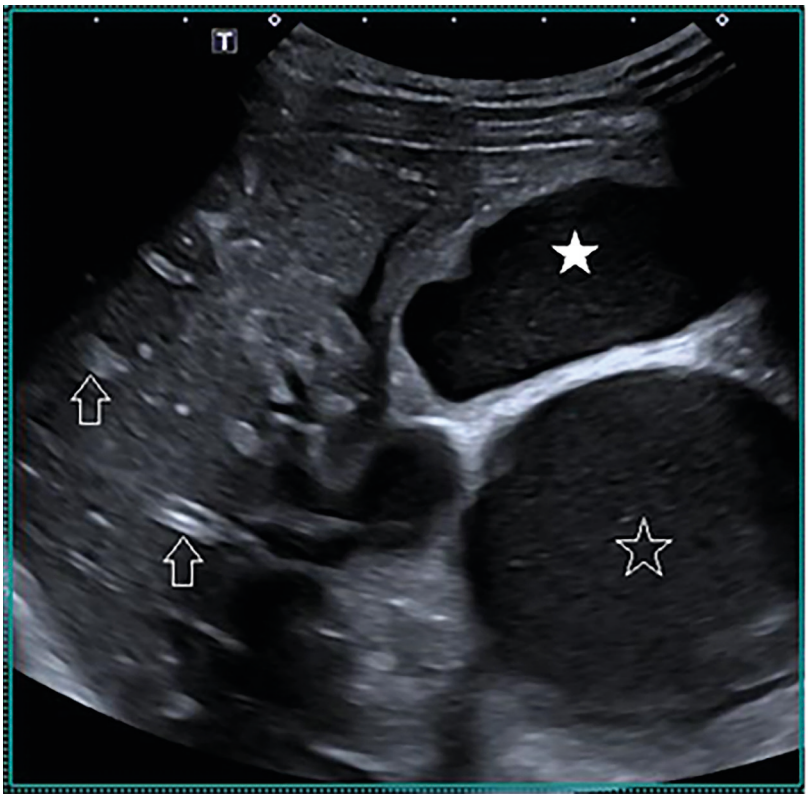

Figure 1. US showed an irregularly shaped large cystic structure at the subhepatic region (open star) with a ventrally located smaller cystic structure (filled star), and moderate intrahepatic bile duct dilatation along with periportal oedema (arrows).

The patient underwent abdominal ultrasonography (US) that showed an irregularly shaped large cystic structure, measuring approximately 6.3 cm × 6.1 cm, with a ventrally located separate smaller cystic structure, possibly, the gallbladder, and moderate intrahepatic bile duct dilatation along with periportal oedema (Fig. 1).

To clarify the US finding, the patient underwent magnetic resonance imaging (MRI) at the emergency department (Fig. 2).

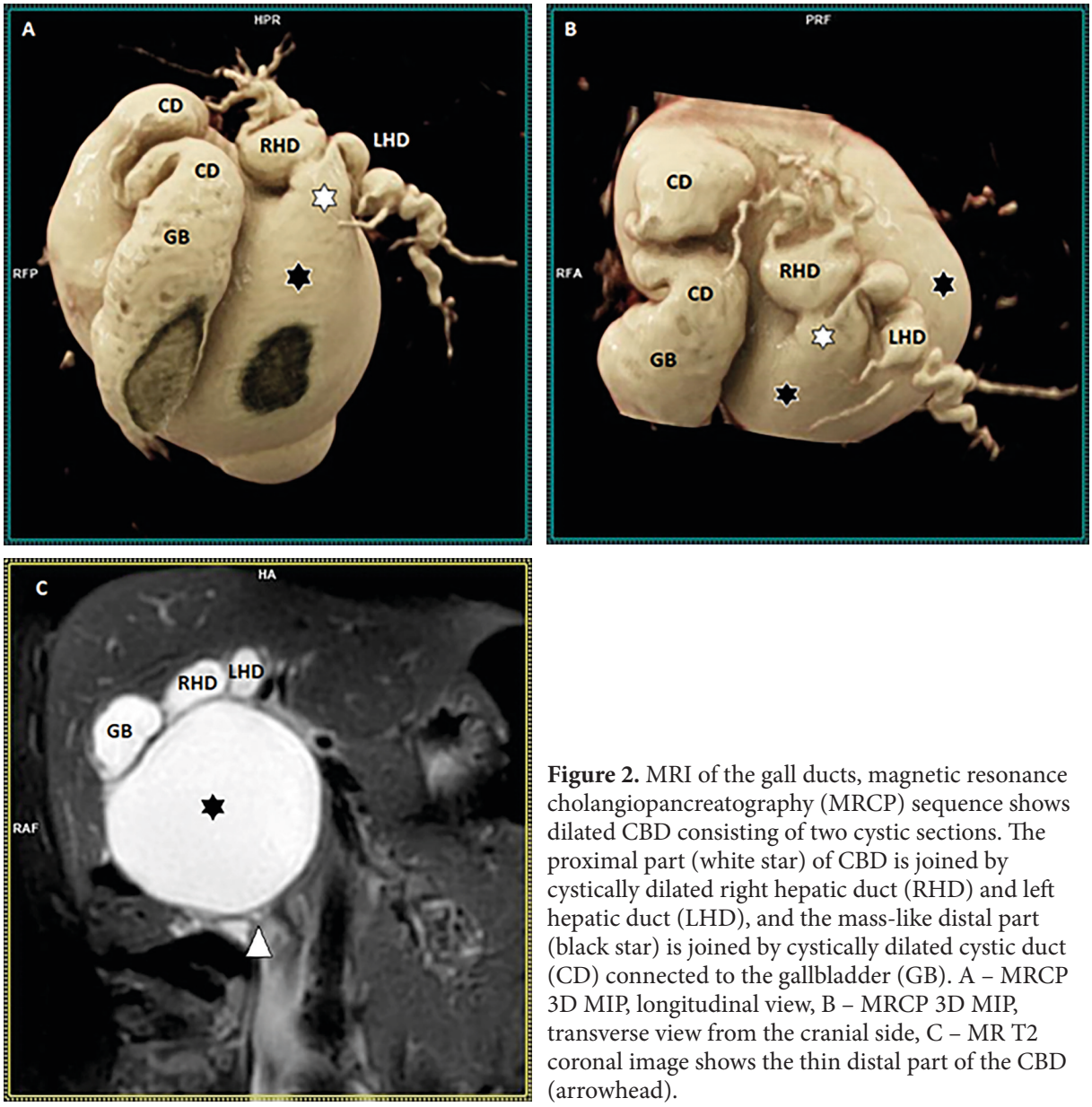

Figure 2. MRI of the gall ducts, magnetic resonance cholangiopancreatography (MRCP) sequence shows dilated CBD consisting of two cystic sections. The proximal part (white star) of CBD is joined by cystically dilated right hepatic duct (RHD) and left hepatic duct (LHD), and the mass-like distal part (black star) is joined by cystically dilated cystic duct (CD) connected to the gallbladder (GB). A – MRCP 3D MIP, longitudinal view, B – MRCP 3D MIP, transverse view from the cranial side, C – MR T2 coronal image shows the thin distal part of the CBD (arrowhead).

MR presented a cystic mass in the location of the CBD of proximally 2 cm in longitudinal section, less than 1.8 cm in cross-section, distally 7 cm in longitudinal section and less than 6.6 cm in cross-section. Gallbladder connects to the widest cystic section through a dilated cystic (2 cm in cross-section). Both main (lobar) bile ducts of the liver are also cystically dilated – the right one less than 0.5 cm, left one 1 cm. The segmental intrahepatic bile ducts are dilated as well.

On MRI, the CBD was dilated and consisted of two cystic sections, of which the subhepatically localized section was 1.8 cm wide; it was joined by cystically dilated right hepatic duct, as well as

up to 1 cm transversely cystically dilated left hepatic duct. The distal part of the CBD was a cystic mass in a cross-section of 6.2 cm, from which an undilated part of the common bile duct was traced distally, its length to the duodenal wall 1.5 cm. The proximal part of the cystic mass was also joined by the cystically dilated cystic duct (Fig 2).

The most dilated part of the CBD dislocated the pancreatic head medially and caudally and compressed the duodenum.

After radiology examinations, the patient was appointed for an open operation: resection extrahepatic bile ducts with open Roux-en-Y hepaticojejunostomy using a circular stapler for entero-entero anastomosis. The surgeon performed a laparotomy incision in the right subcostal region.

A markedly distended gallbladder was found in the abdominal cavity, with a distended, cystic mass medially to the hepatoduodenal ligament. A snuff-box suture was placed in the gallbladder, and a catheter inserted for intraoperative cholangiography. During surgery, a water soluble contrast medium (CM) was injected into the gallbladder showing connection with a cystically dilated CBD; at this stage no drainage of the CM to the intrahepatic bile ducts and duodenum was visible. Due to the large amount of the CM (~80 ml) and the overfilled choledochal duct, it was decided to mobilize the cyst, to prevent a further rupture. The gallbladder was gradually skeletonized from the liver bed, the anterolateral wall of the cyst was cleared of adhesions and decompressed. Moving along the caudal part of the cyst, which was closely with the head of the pancreas, the distal part of the CBD being 3 mm wide. It was contrasted to check its patency through the major duodenal papilla. A small catheter was inserted into this narrow distal part of the CBD indicating permeability and connection with the major duodenal papilla. The thin part of the CBD was ligated. Further, another separate catheter for administration of CM was inserted into the proximal part of the biliary cyst ductus hepaticus communis, where it was contrastingly differentiated and was approximately 1.5 cm wide, and the bifurcation of the bile duct showed a stricture of the left hepatic bile duct, followed by prestenotic dilatation, corresponding to an intrahepatic cystic dilatation (Type IVA). The right hepatic bile duct did not show marked cystic enlargement. After administration of the CM in the the proximal part of the biliary tree, the proximal part of the cyst continued to be skeletonized up to the common hepatic duct – approximately 1 cm from the bifurcation. Upon dissecting the common hepatic duct 1 cm from the bifurcation, no stricture was visualized in the right hepatic bile duct maintaining bile drainage, but a stricture with a cystic dilatation was found in the left hepatic bile duct, retaining the bile drainage. No calculi or other thickenings were found in the bile ducts. Roux-en-Y hepaticojejunostomy was performed. An entero-entero anastomosis was created with a circular stapler. A drain was placed in the abdominal cavity after the laparotomy was completed. There were no early or late complications after the surgery. The patient was discharged from the hospital after five days. At the last follow-up six months postoperatively, no dilated intrahepatic biliary ducts or elevated serum bilirubin, pancreatic or hepatic enzimes, nor anemia were found.

The surgical sample was a fragment from the proximal and distal parts of the bile duct cyst wall and the resected gallbladder. Microscopically, the cyst wall consisted of oedematous connective tissue and a single layer of cylindrical epithelial lining. The connective tissue of the wall included small granular lymphocytes as well as neutrophilic and eosinophilic leucocytes with a moderate inflammatory infiltration. Gallbladder without significant structural pathology. All layers of the gallbladder wall marked oedema, vascular congestion, and multiple haemorrhagic foci. The histological finding was consistent with clinical diagnosis of a CBD cyst.

Discussion

Biliary cysts could be congenital or an acquired anomaly affecting the biliary tree involving both extrahepatic and/or intrahepatic segments [6]. The original clinical classification was revised in 1977

to include intrahepatic cysts and further refined in 2003 to incorporate the presence of an abnormal pancreaticobiliary junction (APBJ). Biliary cysts are associated with significant complications such as ductal strictures, stone formation, cholangitis, pancreatitis, rupture, and secondary biliary cirrhosis. Congenital biliary cysts arise not only from the choledochal duct but also everywhere in the biliary tree, and are frequently accompanied by abnormal pancreatobiliary junction or pancreatobiliary malunion [7]

The classification defines six types of biliary cysts (Type I-V), but, according to our clinical case, it is important to understand the difference between Type I and IV. 50–85% of all cysts belong to Type I, characterized by cystic dilation of the CBD. This type does not involve the intrahepatic bile ducts. There are also subtypes: IC – the dilation which extends from the pancreaticobiliary junction to the extrahepatic portions of the left and right hepatic ducts, associated with an APBJ, and Type ID – cystic dilation of the common duct and cystic duct. Only 2% of cysts belong to Type II being true diverticula of the extrahepatic bile ducts; they are not associated with APBJ. Up to 5% of cysts belong to Type III, being cystic dilations limited to the intraduodenal portion of the distal CBD. Also, there are subtypes A and B. 15–35% of cysts belong to Type IV, being multiple intrahepatic and extrahepatic dilations. Type IVA has both intrahepatic and extrahepatic cystic dilations. Type IVA is the second most common type of biliary cysts and is often associated with a distinct change in duct calibre and/or a stricture at the hilum which are features that help to differentiate it from Type IC cyst. 20% of cysts belong to Type V and are characterized by one or more cystic dilations of the intrahepatic ducts without involvement of extrahepatic ducts. Radiology imaging shows a dilation of the hepatic duct and a cystic duct. When tailoring this clinical case for the current classification of the bile duct cysts, a combination of Type IC and ID and Type IVA was found [7,8].

Their development is associated with both congenital and acquired causes. Congenital cysts may result from an unequal proliferation of embryologic biliary epithelial cells before bile duct cannulation is complete [9], but acquired cysts occur due to reflux of pancreatic enzymes into biliary tracts. Some biliary cysts may be the result of an APBJ. While APBJ is a rare congenital anomaly, it is present in 50 to 80% of patients with biliary cysts [10]. The APBJ may allow reflux of pancreatic juice into the biliary tree resulting in damage of the biliary epithelium and cyst formation [11].

The approach to managing patients with biliary cysts depends on the cyst type. Patients with Type I or IV cysts usually undergo surgical resection of the cyst due to the significant risk of malignancy and are good surgical candidates. Complete surgical excision is recommended for Type I and IV biliary cysts to remove all tissue of the cyst if possible. The approach is advocated because of the risk of malignancy associated with these cysts. In addition to decreasing the risk of malignancy, cyst removal can reduce complications, such as recurrent cholangitis, cystolithiasis, choledocholithiasis, and pancreatitis. Surgical treatment by cyst-enterostomy rather than resection is associated with poorer short- and long-term outcomes, with a higher risk of subsequent malignancy [12,13,14].

Pancreaticobiliary maljunction (PBM), in combination with choledochal cysts, is a rare and unique disorder. Several definitions have been proposed for the PBM, but the most widely accepted is an excessive length of the common pancreaticobiliary duct due to the abnormal convergence of the pancreatic and biliary ducts out of the duodenal wall. Acute pancreatitis is more common in children with PBM (30% of patients) than in adults with PBM (9%), irrespective of the presence of biliary dilatation [15].

MRI was performed more frequently during the two decades than endoscopic retrograde cholangiopancreatography and intraoperative cholangiography. MRI was the most helpful modality to detect PBM by showing an extra duodenal wall junction between the choledochus and the central pancreatic duct, even in patients with the standard common duct. Magnetic resonance cholangiopancreatography, or MRCP, uses a powerful magnetic field and radio waves to evaluate the liver,

gallbladder, bile ducts, pancreas, and pancreatic duct for disease. This radiology modality has several benefits, being a non-invasive imaging technique that does not involve radiation exposure and can provide detailed images of the soft-tissue structures of the body, such as the liver and pancreas [16,17].

Conclusions

CC is a congenital anomaly of bile ducts that poses a diagnostic and therapeutic challenge to paediatric surgeons. MRI is a non-invasive radiology imaging method capable of discovering and demonstrating the subcategorized types and configurations of CC before the surgical treatment and to prevent its complications. Since MRI is not related to exposure to ionizing radiation, it is an excellent modality for children.

All the authors declare no conflict of the interest.

The patients and the families have signed the permission to publish this case report. The Committee of the Ethics of University of Latvia has approved the case to be published. The authors of this article have confirmed that there is no conflict of interest to be reported.

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