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Transduodenal resection of a choledochocele (type III choledochal cyst) with sphincteroplasty: A case report

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ABSTRACT

Choledochal cysts are rare congenital anomalies of the biliary tree which may progress to obstruction or malignancy. Of the five Todani variants, choledochocele, or type III choledochal cyst is the rarest. In this case report, we describe a previously healthy 10-year old female who presented with a choledochocele and was treated by near-total excision with transposition of the common channel, resulting in an extended sphincteroplasty.

Choledochal cysts (CDCs) are rare biliary anomalies with an incidence of 1 in 100,000 births [1]. Classified into five subtypes by Todani in 1977 [2], type III choledochal cysts, termed choledochoceles, are the rarest variant [3]. Choledochoceles are a cystic dilation at the distal end of the common bile duct, protruding into the duodenal lumen. Due to the rarity of the disease, a consensus on patient management in the pediatric population does not exist. Since 1975, only thirteen cases of pediatric choledochocele have been reported. The management alternatives for choledochocele have evolved from open transduodenal sphincteroplasty or cystoenterostomy, to include endoscopic treatment. Additionally, this disease process is complicated by the presence of either biliary or intestinal mucosa within the cyst, which may alter treatment. We present a case of choledochocele, in a previously healthy 10-year old child not amenable to endoscopic treatment.

1. Case report

A 10-year old female presented to the emergency department with a three day history of progressively worsening epigastric abdominal pain. The pain was exacerbated by food intake with additional symptoms of anorexia, occasional nausea and non-bloody emesis. The child and her family denied any recent history of fever, weight loss, constipation or diarrhea. Her previous medical history was limited to a tonsillectomy and adenoidectomy, performed two-years prior to presentation, and there was no pertinent family medical history. On physical examination, she was afebrile with unremarkable vital signs. Scleral icterus and jaundice were not evident. Her abdomen however, was tender to palpation of the epigastrium with no palpable masses or hepatosplenomegaly. Biochemical analysis of her blood revealed a leukocytosis of 20.6 (range 5–19.5 K/mL) with elevated values for aspartate aminotransferase (AST) (180, range 16–62 u/L), alkaline phosphatase (372, range 51–332 u/L), amylase (288, range 23–85 u/L) and lipase (2406, range 0–160 u/L). A computed tomography (CT) scan of the abdomen and pelvis identified a dilated second portion of duodenum with an ovoid, 3 cm, low-attenuating structure that was contiguous with the medial wall of the duodenum (Fig. 1).

A magnetic resonance cholangiopancreatogram (MRCP) with intravenous gadoxetate disodium (Eovist®) was subsequently performed to further elucidate the abnormality (Fig. 2).

The lesion was noted to be cystic in structure and located in the medial wall of the second portion of the duodenum, measuring 3.1 × 2.9 × 5.2 cm and appearing to communicate with the biliary

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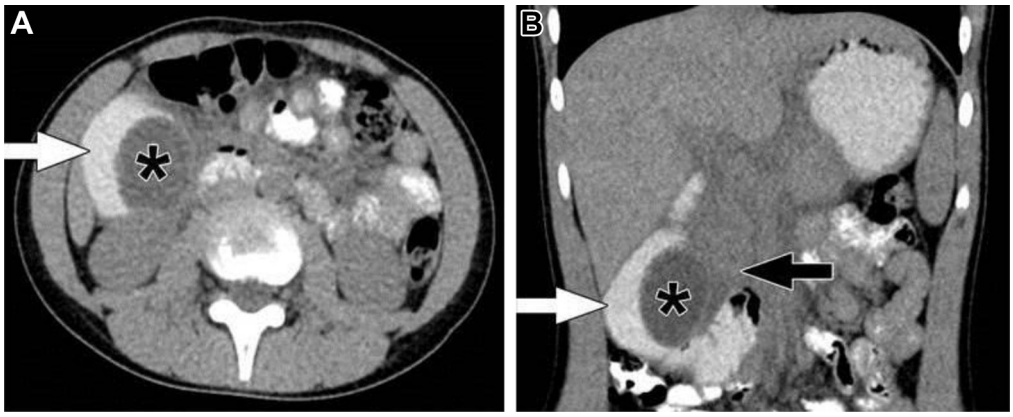

Fig. 1. Axial (A) and coronal (B) images from the CT of the abdomen and pelvis with intravenous and oral contrast media performed at presentation. The axial reformatted image (A) shows dilation of the contrast filled second portion of the duodenum (white arrow) with a low attenuation, cystic mass (asterisk) apparently within the lumen along the medial wall. The coronal reformatted image (B) again shows the dilated, contrast filled duodenum (white arrow) with the intraluminal cystic mass along the medial wall (asterisk). This image better shows the relationship of the cystic mass to the head of the pancreas with a claw of pancreatic tissue along the left wall of the cystic mass (black arrow).

tree. The common bile duct (CBD) and intrahepatic biliary tree proximal to the lesion were of normal caliber. The gallbladder also appeared without abnormalities. Primary differential diagnoses for this finding included intestinal duplication cyst and a type III CDC.

The child's symptomatology resolved with hydration, and she began tolerating oral intake ad lib without symptom recurrence. She was discharged home on a prophylactic antibiotic (Trimethoprim-Sulfamethoxazole) and ursodiol with a scheduled endoscopic

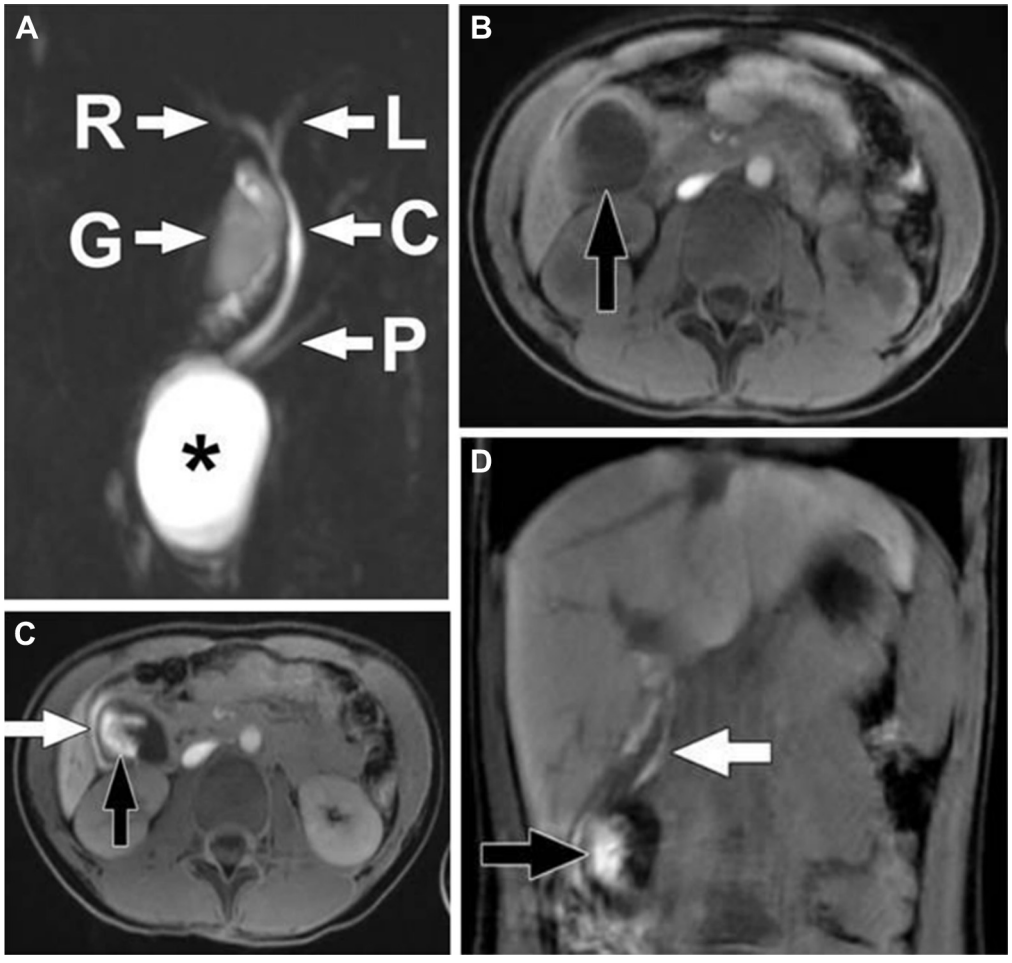

Fig. 2. MRCP performed to better characterize the cystic lesion. (A) Coronal maximum intensity (MIP) 3D MRCP image shows the cystic lesion (asterisk) near the junction of the normal caliber common bile (c) and pancreatic (p) ducts. The intrahepatic ducts (r, l) are also normal in caliber and the gallbladder (g) is normal. Axial pre-contrast (B) and axial (C) and coronal (D) 20 min post-contrast LAVA images show a cystic mass along the medial wall of the second portion of the duodenum (black arrow in B) with accumulation of biliary excreted contrast media at after a 20 min delay (black arrows in C and D). Excreted contrast media is also visible in the second portion of the duodenum (white arrow in C) and in the non-dilated common bile duct (white arrow in D). Note how the cystic lesion changes in size over the course of the exam appearing smaller in (C) than in (B).

retrograde cholangiopancreatography (ERCP) to better define her pancreaticobiliary anatomy in relation to the cystic lesion and help guide surgical intervention.

During the interval between hospital discharge and ERCP, the child developed brief, self-limiting occurrences of epigastric pain. At the time of the ERCP, endoscopic visualization within the duodenum identified a large submucosal cystic structure in the expected region of the major papilla obscuring the normal anatomic definition of the ampulla. A small orifice into the cyst was cannulated followed by contrast injection, leading to opacification of the cystic cavity and secondary filling of normal appearing biliary and pancreatic ductal structures (Fig. 3). It was felt at this time that the cyst was too large to be treated by sphincteroplasty alone.

The child subsequently underwent exploratory laparotomy for planned transduodenal cyst excision and sphincteroplasty. Examination of the duodenum revealed no external mass, although upon palpation of the second portion of the duodenum, the cystic structure was readily apparent. An intraoperative cholangiogram was then performed via cannulation of the gallbladder (Fig. 4), revealing a cystic outpouching filling with contrast distal to the confluence of the CBD and pancreatic duct (PD). The ampulla was intact, with contrast draining from the cyst into the duodenum through a short common channel.

The anti-mesenteric wall of the duodenum was opened and the cystic mass was palpated in the head of the pancreas (HOP) protruding into the medial aspect of the duodenal wall. The ampulla of Vater was visualized within the splayed duodenal mucosa. The antegrade tube placed via the gallbladder (for cholangiogram) was removed and a feeding tube was placed in retrograde fashion via the ampulla into the cyst and CBD (Fig. 5A). The mucosa along the mesenteric side of the duodenum was opened, and the cystic mass was dissected from the mucosal wall and the HOP, beginning in the third portion of the duodenum and proceeding toward the feeding tube-containing ampulla (Fig. 5B). In order to aid in the identification of the CBD and PD during dissection, the feeding tube entering the ampulla of Vater was exchanged for two stents placed through the ampulla into the orifices of the CBD and PD. The stents could be palpated in the superior most aspect of the cyst. The cyst was opened and found to contain a mucosal lining with the stents visualized in a common channel comprised of the distal component of the CBD, the PD and the choledochocele as the stents traversed out of their respective ducts into the duodenum via the intact ampulla (Fig. 5C). It was determined that complete cyst excision would compromise the orifice of both the CBD and PD, potentially necessitating a pancreaticoduodenectomy for safe reconstruction. The majority of the cyst was excised and a frozen section of the lining revealed histology consistent with intestinal mucosa. This reduced our concern for possibility of malignant transformation in the future. The ampulla was opened, resulting in a full thickness mucosa-lined oval of intestine with the CBD and PD entering at the most medial aspect of the oval. The mucosa of the medial duodenal wall was anastomosed to the conjoined cyst wall and ampulla, resulting in a large sphincteroplasty, transposing the ampulla to where the biliary and pancreatic stents maintained the patency of the CBD and PD openings at the medial most portion of the mucosa-to-mucosa anastomosis (Fig. 5D). The anti-mesenteric duodenotomy was closed in two layers. A cholecystectomy completed the operative procedure.

The patient's postoperative course was uncomplicated. The patient was discharged on postoperative day ten, tolerating a regular diet with complete resolution of abdominal symptoms. An esophagogastroduodenoscopy (EGD) was performed six weeks after her operative procedure, and the pancreatic and biliary stents were removed uneventfully. One year surveillance EGD has been performed with a normal appearing "ampulla of Vater." No further surveillance studies are planned.

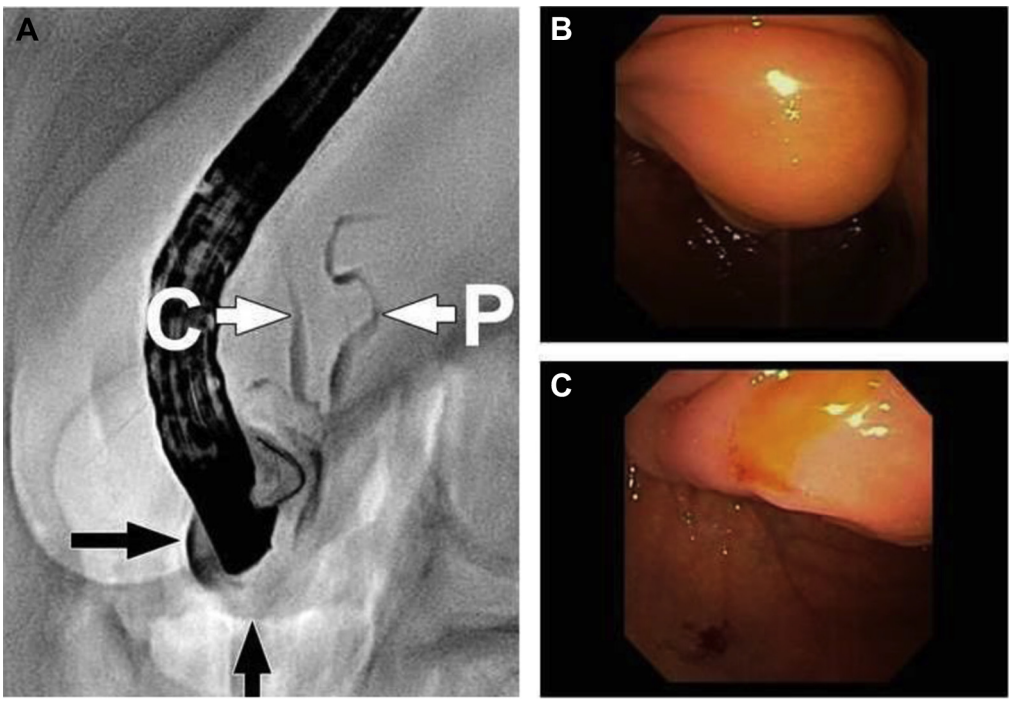

Fig. 3. (A) Fluoroscopic spot image from ERCP shows and endoscope at the level of the ampulla with partial contrast opacification of the cystic lesion (black arrows) and contrast opacification of normal caliber common bile (c) and pancreatic (p) ducts. (B, C) Endoscopic imaging from ERCP showing a large submucosal structure, involving the Ampulla of Vater, at the second portion of the duodenum.

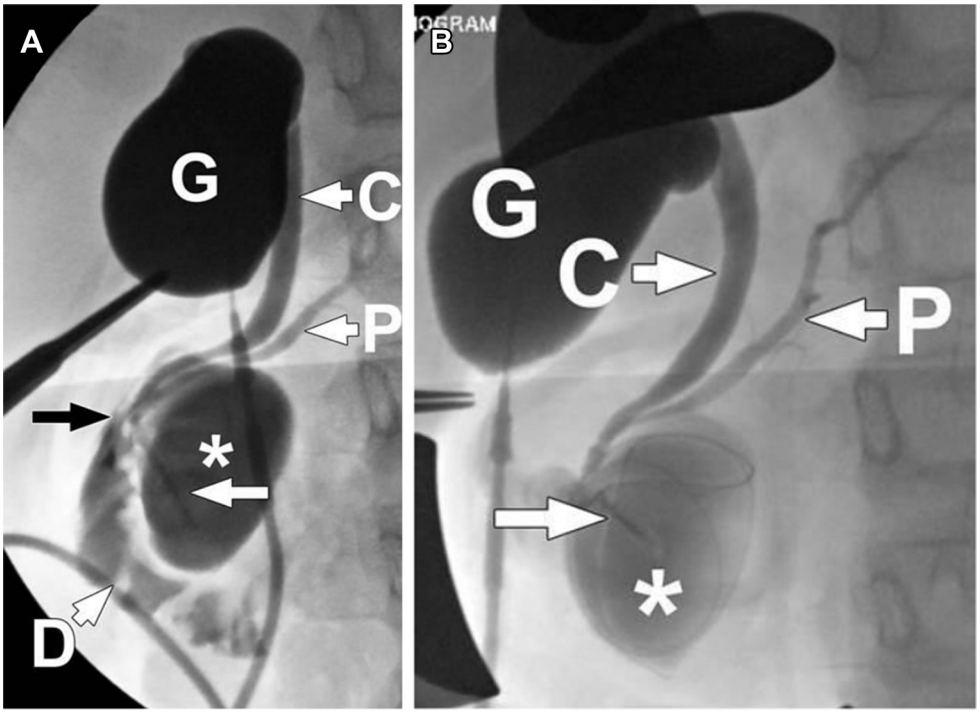

Fig. 4. (A) Selected fluoroscopic spot images from the intraoperative cholangiogram show contrast opacification of the gallbladder (G) and normal caliber common bile (C) and pancreatic (P) ducts. The common bile duct and pancreatic duct join just above the level of the ampulla (black arrow in A). A jet of contrast originating from the normal common channel of the common bile and pancreatic ducts (white arrows) fills the cystic lesion (asterisk). Contrast subsequently drains into the duodenum (D in (A)). (B) A wire has been introduced via the ampulla and is looped in the cystic lesion. More of the pancreatic duct is opacified by contrast. Asterisk displays cystic lesion. G = gallbladder, C = common bile duct P = pancreatic duct.

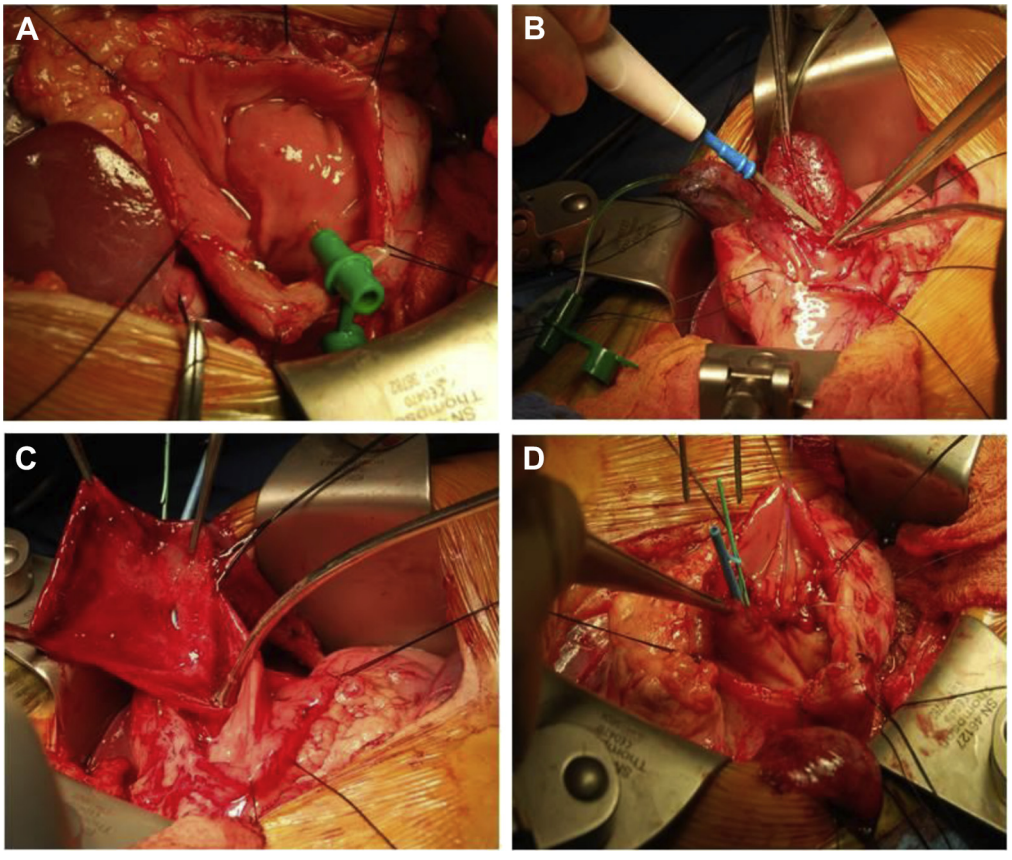

Fig. 5. Intraoperative gross imaging of choledochocele excision and transduodenal sphincterotomy. (A) A feeding tube was placed in a retrograde fashion through the ampulla of Vater and common bile duct, following the tract of the antegrade tube from the gallbladder. It is seen here entering through the Ampulla of Vater. (B) Dissection of the cystic mass from the wall of the duodenum was performed, proceeding toward the stent-containing ampulla. (C) The cyst was opened and stents from common bile and pancreatic ducts visualized at the superior aspect of the mucosa into the duodenum. (D) A large sphincteroplasty and ampulla transposition was performed, taking care to maintain the stents, thus patency of the common bile and pancreatic duct openings.

2. Discussion

A choledochocele is a rare variant of choledochal cyst. In a review by Yamaguchi of 1433 adult and pediatric patients with choledochal cysts, choledochocele comprised only 1% of all cysts [4]. Children with choledochocele, as in the case of our patient, often present with abdominal pain [5–8], though they may also present with jaundice [7,9] and emesis [6,10]. These biliary anomalies are diagnosed through a combination of ultrasonography and ERCP [7,9,11], though no gold-standard exists for choledochocele diagnostic modality.

Due to the rarity of disease, a treatment algorithm for pediatric choledochocele has not yet been established. In some cases, the choledochocele may be amenable to endoscopic sphincterotomy, thus sparing the patient an open surgical procedure [10,12]. In our patient, the 5 cm choledochocele was determined to be too large for endoscopic management, and thus, exploratory surgery was performed. The primary objective of the operative procedure was to relieve the obstructive component of the choledochocele which was causing pancreatic and biliary symptomatology with the ideal outcome encompassing excision of the entire cyst. There were two important secondary considerations – to maintain normal egress of biliary and pancreatic secretions and minimize the risk of long term malignant transformation.

During the operative exploration, it became clear that there was a common channel composed of the wall of the distal CBD, PD and the choledochocele. The cyst fluid had collected inferiorly to the common channel creating a cystic mass in the HOP. The common channel outflow was directed through an intact Ampulla of Vater. Excision of the entire cyst including the common channel would require re-implantation of the CBD and PD, each of which was less than 3 mm in diameter. Given the technical challenges of the re-implantation, an alternative could have been a pancreaticoduodenectomy. Additionally, we were concerned that incomplete excision of the cyst would leave the patient at risk for malignant transformation in the future.

Although malignant transformation of choledochocele is a rare occurrence, several cases have been reported in the adult population [13,14]. Choledochoceles may be lined by either biliary or intestinal mucosa [11], thus an intraoperative frozen section histological analysis of cyst lining was important in order to dictate optimal management. In adult series, a 14.3–27% risk for malignancy has been reported, including adenocarcinoma, anaplastic carcinoma, undifferentiated cancer and squamous cell cancer [13,15–17]. Carcinoma arising in adult choledochoceles has been attributed to pancreaticobiliary reflux allowing for activated pancreatic secretions to irritate biliary mucosa, leading to chronic mucosal inflammation which could progress to malignancy [14,18]. Fortunately, our patient's choledochocele was lined with duodenal mucosa making the potential risk of malignant transformation less likely. As a result, we chose to leave the superior most aspect of the cyst wall *in situ*. By opening the Ampulla of Vater at its most inferior edge, we created an oval of full thickness, mucosa-lined intestine made up by the wall of the Ampulla of Vater and the common channel with the CBD and PD orifices at the medial most aspect of the oval. The surrounding duodenal mucosa was then anastomosed to this full thickness oval of intestine, resulting in a giant transposing sphincteroplasty. If the mucosa of the choledochocele had been biliary in nature we would have considered a pancreaticoduodenectomy, in order to achieve complete cyst resection and minimize the remote risk of malignancy. Alternatively, if we had performed a subtotal cystectomy with biliary epithelium present, the patient would have required indefinite long term surveillance of the oval of intestine to ensure that malignant transformation did not occur.

Eighteen months after surgical intervention, our patient remains well, with no further abdominal pathology or complications. The rarity of a choledochocele and the variability of mucosal findings make the treatment of this type of choledochal cyst complex.

Conflicts of interest

The authors have no disclosures of funding or conflicts of interest.

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