Hando *et al. Egyptian Liver Journal* (2022) 12:29
https://doi.org/10.1186/s43066-022-00193-0

Egyptian Liver Journal

CASE REPORT Open Access

# Choledochal cyst type I with dilated intrahepatic biliary radicles: a type IVA mimic

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## Abstract

**Background:** A choledochal cyst is a relatively rare congenital anomaly of the biliary tree requiring surgery as the definitive treatment. Amongst the five Todani variants, type I poses a diagnostic and treatment challenge owing to its infrequent, yet clinically significant mimicry for type IVA cysts.

**Case presentation:** We present a case of a 4-year-old female diagnosed to have a giant type IA choledochal cyst that mimicked a type IVA cyst on radiological imaging. The patient was treated by complete cyst excision, cholecystectomy, and restoration of the biliary-enteric communication by a Roux-en-Y hepaticojejunostomy. Regression of the dilated intrahepatic radicles that counterfeited a type IVA cyst was confirmed on follow-up imaging studies.

**Conclusion:** Such an encounter, although rare, can significantly alter the course of management. We recommend extrahepatic cyst excision with biliary reconstruction as the standard treatment when preoperative and intraoperative imaging studies fall short in differentiating the aforementioned variants.

**Keywords:** Choledochal cyst excision, Giant choledochal cyst, Roux-en-Y hepaticojejunostomy, Todani classification

## Background

A choledochal cyst (CC) is a relatively rare congenital biliary tract anomaly characterized by single or multiple cystic dilatations of the extra- and/or intrahepatic biliary tree [1]. The initial description of this condition was done by Vader and Elzer in 1723 and was later classified by Todani et al. in 1977 [2, 3]. The anomaly is more prevalent in Asia, particularly in Japan as compared to other parts of the world [4–6]. Although CC type I presents with dilatation of the extrahepatic bile duct, pressure effect from a giant cyst may result in dilatation of the intrahepatic ducts, thus mimicking a type IVA cyst [7]. We herein report a case of regressed intrahepatic cystic dilatations following excision of a giant type IA CC diagnosed in a 4-year-old female. We also discuss the key aspects of choledochal cyst disease, accentuating the complexity of managing type I and type IV cysts.

## Case presentation

A 4-year-old female child from northern Tanzania presented to our facility with a history of intermittent fever and sclera jaundice for 8 months. She thereafter started to experience intermittent right upper quadrant abdominal pain associated with abdominal distension. A history of passing deep yellow urine, clay-colored stools, and generalized body itching was also reported.

On examination, the child had a tinge of jaundice with no stigmata of chronic liver disease (digital clubbing, palmar erythema, etc.). She was afebrile with normal vital signs for her age. Height for age anthropometry was suggestive of mild stunting; however, other measurements, i.e., mid-upper arm circumference, weight for height, and weight for age, were within the normal limits.

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Abdominal examination revealed asymmetrical distension with non-tender, nodular hepatomegaly measuring approximately 6 cm below the right costal margin in the mid-clavicular line. A mass arising from below the edge of the liver, extending to the right iliac and umbilical region, was noted. It was smooth-surfaced, firm, non-tender, and mobile, measuring approximately 14 cm by its greatest dimension and had a dull note on percussion. The rest of the abdominal examination including a digital rectal exam was essentially normal.

An abdominal computerized tomography (CT) scan revealed multiple dilated intrahepatic biliary radicles and a large common bile duct (CBD) cyst (Fig. 1). The liver, gallbladder (GB), pancreas, spleen, and kidneys appeared normal. Magnetic resonance cholangiopancreatography (MRCP) also showed bilobar dilatations of the central and peripheral intrahepatic biliary radicles and a large cystic dilatation of the extrahepatic biliary tree measuring 11.05 cm by its largest diameter (Fig. 2), suggestive of a giant type IA CC with intrahepatic bile duct dilatations secondary to the compressive effect of the giant cyst, a typical type IVA CC mimic.

A diagnosis of a giant type IA CC was reached. An elective CBD cyst excision and biliary reconstruction were scheduled following patient optimization. Laparotomy was done through a right upper abdominal transverse incision, and a huge CBD cyst was identified together with gross nodulations on the liver surface (Fig. 3A and B). The rest of the viscera was essentially normal. Mobilization of the GB and CBD cyst was done

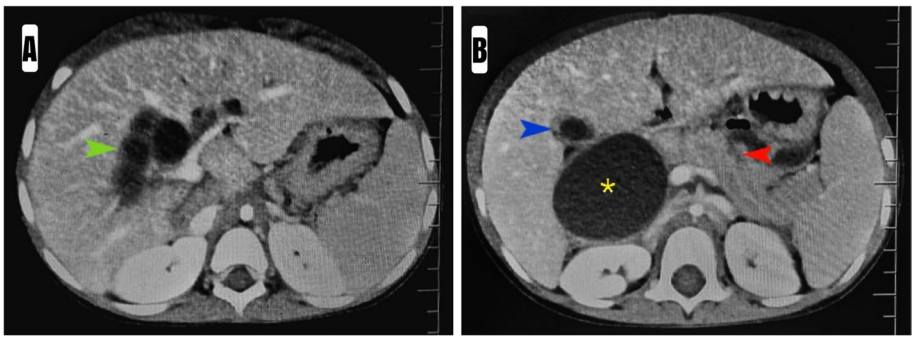


**Fig. 1** **A** and **B**: Preoperative axial CT scan of the abdomen showing (**A**) multiple intrahepatic biliary radicle dilatations (green arrowhead) and (**B**) a giant cystic dilatation of the common bile duct (yellow asterisk). The main pancreatic duct (red arrowhead) and GB (blue arrowhead) are unremarkable

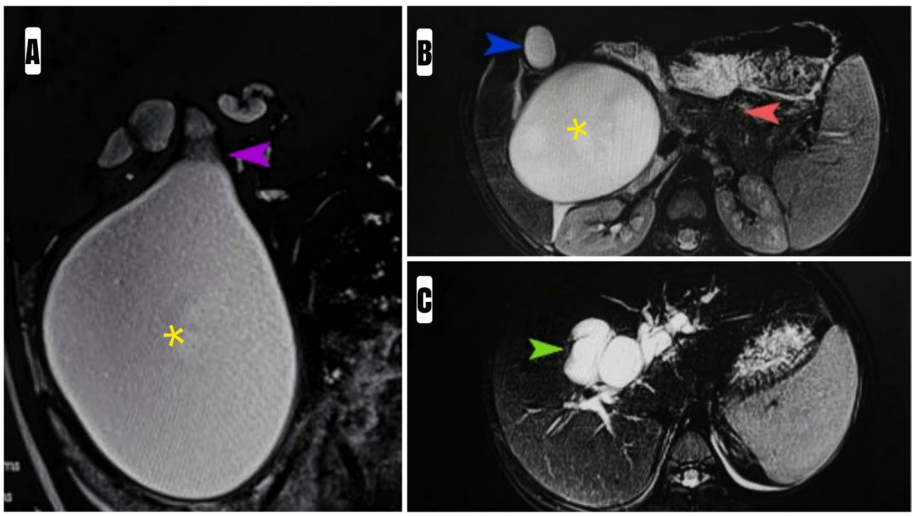


**Fig. 2** **A**–**C**: Preoperative MRCP (**A** and **B**) showing a cysticly dilated common hepatic duct (purple arrowhead) and CBD (yellow asterisk). The main pancreatic duct (red arrowhead) and GB (blue arrowhead) are unremarkable. (**C**) Dilated bilobar central and peripheral intrahepatic biliary radicles (green arrowhead) are present. No T2-weighted hypointense filling defect was seen in the lumen

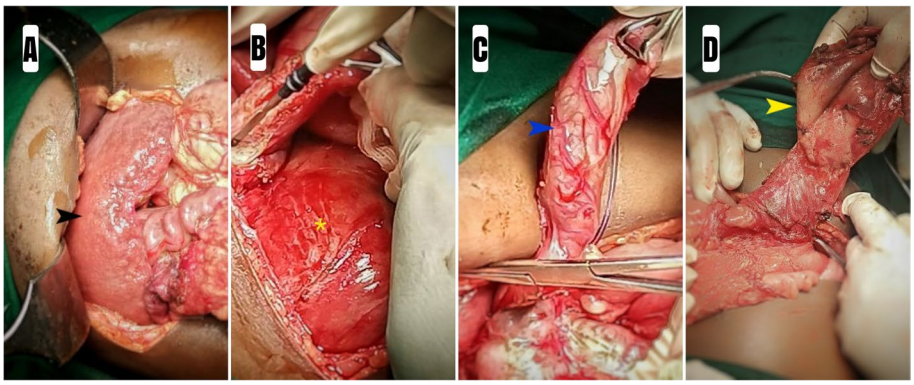


**Fig. 3** **A**–**D**: Intraoperative images. (**A**) Black arrowhead showing a cirrhotic liver. (**B**) Yellow asterisk showing the extrahepatic (CBD) cyst before mobilization. (**C**) Blue arrowhead showing the mobilized GB. (**D**) Yellow arrowhead showing the mobilized choledochal cyst

followed by cholecystectomy, complete extrahepatic cyst excision, and Roux-en-Y hepaticojejunostomy reconstruction (Fig. 3C and D). Abdominal drains were kept in situ alongside the anastomoses sites. The resected cyst and GB together with a liver parenchymal tissue were submitted for histopathological analysis. Results revealed features suggestive of liver cirrhosis and excluded malignancy of the biliary tree (Fig. 4).

Following surgery, the patient was admitted to the pediatric intensive care unit and was later transferred to the surgical pediatric ward on postoperative day 3. Oral feeds were initiated on the third postoperative day after the resolution of postoperative paralytic ileus. Abdominal drain and wound stitches were removed on postoperative days 4 and 7, respectively, and the patient was discharged thereafter. Preoperative laboratory investigations that revealed deranged liver function coupled with elevated markers of acute pancreatitis normalized postoperatively (Table 1).

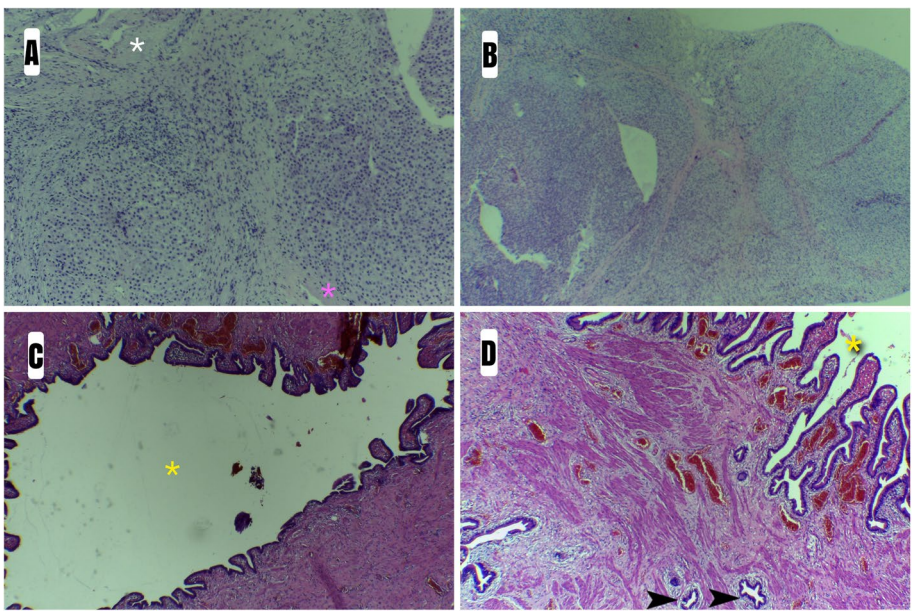


**Fig. 4** **A**–**D**: Hematoxylin and eosin (H&E) stain of hepatic and bile duct tissue. (**A**) 20× showing nodularity with portocaval fibrosis (white and purple asterisk showing the portal triad and central vein, respectively). (**B**) 4× showing fibrous bands separating hepatic tissue ingue nodules. (**C**) 4× showing a cysticly dilated duct (yellow asterisk) with subepithelial chronic inflammatory cell infiltrates and fibrosis. (**D**) 20× showing a section of the bile duct with a portion of the main cysticly dilated lumen (yellow asterisk) and smaller ducts (black arrowheads) lined by columnar epithelium. The duct walls are composed of dense fibrous tissue as well as scattered smooth muscle cells

**Table 1** Summary of the pre- and postoperative laboratory investigations

| Laboratory tests and parameters (SI and CU) | Preoperative values | Day 7 Postoperative values | 6 months Postoperative values | Reference ranges |
|---|---|---|---|---|
| **Full blood count** | | | | |
| White blood cell count (cells 10^9/L) | 16.0 | 9.8 | 5.5 | 5.0–15.5 |
| Hemoglobin (gm/dL) | 10.7 | 11.5 | 11.3 | 10.4–13.6 |
| **Liver function test** | | | | |
| Total bilirubin (μmol/L) | 60.5 | 21.4 | 6.1 | 3.4–20.5 |
| Direct bilirubun (μmol/L) | 44.6 | 13.5 | 3.1 | 0.0–8.6 |
| Alkaline phosphatase (U/L) | 1000.0 | 512.0 | 242.0 | 0.0–500.0 |
| Gamma-glutamyl transferase (U/L) | 866.0 | 219.3 | 33.2 | 9.0–36.0 |
| Aspartate transaminase (U/L) | 228.0 | 68.0 | 52.3 | 5.0–34.0 |
| Alanine transaminase (U/L) | 127.0 | 35.0 | 71.9 | 0.0–55.0 |
| Serum albumin (gm/L) | 30.4 | 33.0 | 37.0 | 35.0–55.0 |
| Prothrombin time (s) | 10.6 | 10.9 | 11.3 | 9.4–12.5 |
| International normalized ratio | 0.9 | 0.9 | 1.0 | 0.8–1.2 |
| Partial thromboplastin time (s) | 40.2 | 25.3 | 33.1 | 25.4–36.9 |
| **Markers of acute pancreatitis**[a] | | | | |
| Serum lipase (U/L) | 229.0 | 38.0 | 37.1 | 13.0–60.0 |
| Serum amylase (U/L) | 236.0 | 90.0 | 89.0 | 25.0–125.0 |
| **Renal function test** | | | | |
| Blood urea nitrogen (mmol/L) | 1.8 | 1.7 | 2.2 | 2.5–6.7 |
| Serum creatinine (μmol/L) | 38.3 | 39.2 | 39.4 | 27.0–88.0 |

*μmol/L*, micromole/liter; *CU*, conventional units; *gm/dL*, gram/deciliter; *gm/L*, gram/liter; *L*, liter; *mmol/L*, millimoles/liter; *s*, seconds; *SI*, International System of Units; *U/L*, units/liter. [a]Nonspecific

No complaint was reported throughout the 6-month post-discharge follow-up period. Physical examination was essentially normal except for nodular hepatomegaly measuring approximately 2 cm (preoperatively, 6 cm) below the right costal margin in the mid-clavicular line and a healed surgical scar. An MRCP performed 6 months postoperatively (Fig. 5) revealed regression of the dilated intrahepatic biliary radicles that were initially seen on preoperative imaging studies (Figs. 1 and 2).

## Discussion

The pathogenesis of CC is controversial, with several proposed theories postulating the possible etiologies. Congenital weakness of the bile duct wall, defective

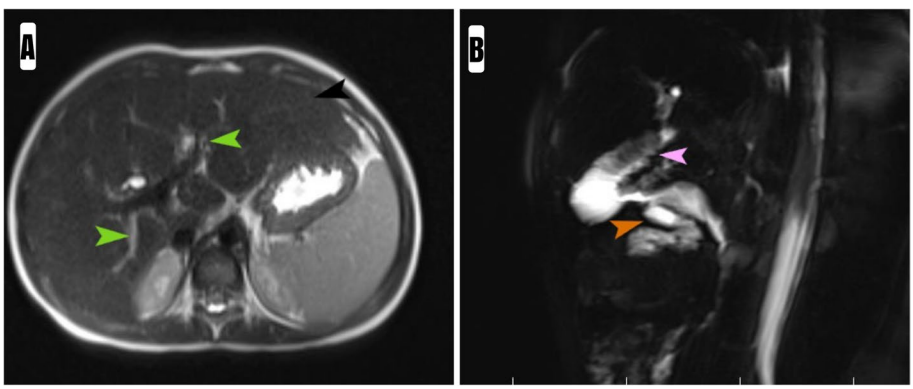


**Fig. 5** **A** and **B**: Postoperative MRCP showing (**A**) enlarged liver parenchyma with cirrhotic changes seen as irregular borders (black arrowhead). Bilobar central and peripheral intrahepatic biliary radicles appear normal (green arrowheads). An empty GB fossa seen. No T2W hypointense filling defect is seen in the lumen. (**B**) Roux-en-Y hepaticojejunostomy reconstruction in situ with a patent Roux (pink arrowhead) and Y (brown arrowhead) limb

embryologic ductal proliferation, congenital distal obstruction, and sphincter Oddi dysfunction are some of the suppositions [1, 8]. However, anomalous pancreaticobiliary duct union (APDU) is the widely accepted hypothesis. The theory postulates that APDU results in pancreaticobiliary reflux which causes activation of pancreatic enzymes within the duct with subsequent inflammatory response resulting in defective bile duct wall integrity with or without downstream stenosis [1, 2, 9, 10].

The generally accepted Todani classification of bile duct cysts is based on the anatomic locations and the extent of biliary tree involvement (Fig. 6) [2, 6, 9]. Type I is solitary extrahepatic cysts which can either be cystic (IA), saccular (IB), or fusiform (IC) and is the most prevalent type encountered in 60–80% of the cases [6, 9]. Type II cysts are diverticulums of the supraduodenal portion of the CBD, accounting for 1–2%, whereas type III (choledochocele) are intraduodenal cysts that account for 0.5–4% and lack the female predilection as compared to the rest of the variants [5, 9, 10]. Both malignancy and APDU are seldom in type III cysts, thus dictating their management as highlighted in the subsequent passages [8]. Type IV cysts are characterized by multiple intrahepatic and extrahepatic cysts (IVA; 15–30%) or multiple extrahepatic duct cysts (IVB; 1–2%) [2, 5, 9]. Type V disease (Caroli's disease) is rare; it presents with single or multiple intrahepatic duct cysts [2, 5, 9, 10]. CCs barely exceed 6 cm in diameter; giant cysts measuring 10 cm or more are thus extremely rare [11, 12]. The presented case was diagnosed to have the commonest variant, a giant type IA CC.

CCs commonly present before 10 years of age with a female predilection (3–4:1 female to male); nonetheless, the condition has also been reported in adults to a comparatively lesser extent, 25% being initially identified in adulthood [2, 9, 13–15]. The reported case represents the 20% of patients who present with the classic triad of CC most commonly seen in children; symptoms include abdominal pain, jaundice, and a palpable right upper quadrant abdominal mass [2, 16]. Of these symptoms, jaundice is a common presentation in Todani's types I and IV [2, 5]. Furthermore, about 85% of children and adults will present with two or one of the three classic triad symptoms, respectively [17]. Clinical presentation, laboratory workups, and intraoperative findings in this case also pointed towards CC-related complications which are sometimes the only presenting feature especially in the adult population. These included features of ascending cholangitis, acute pancreatitis, defective liver excretory and synthetic function, cholestasis, and liver cirrhosis. Other complications that can result from

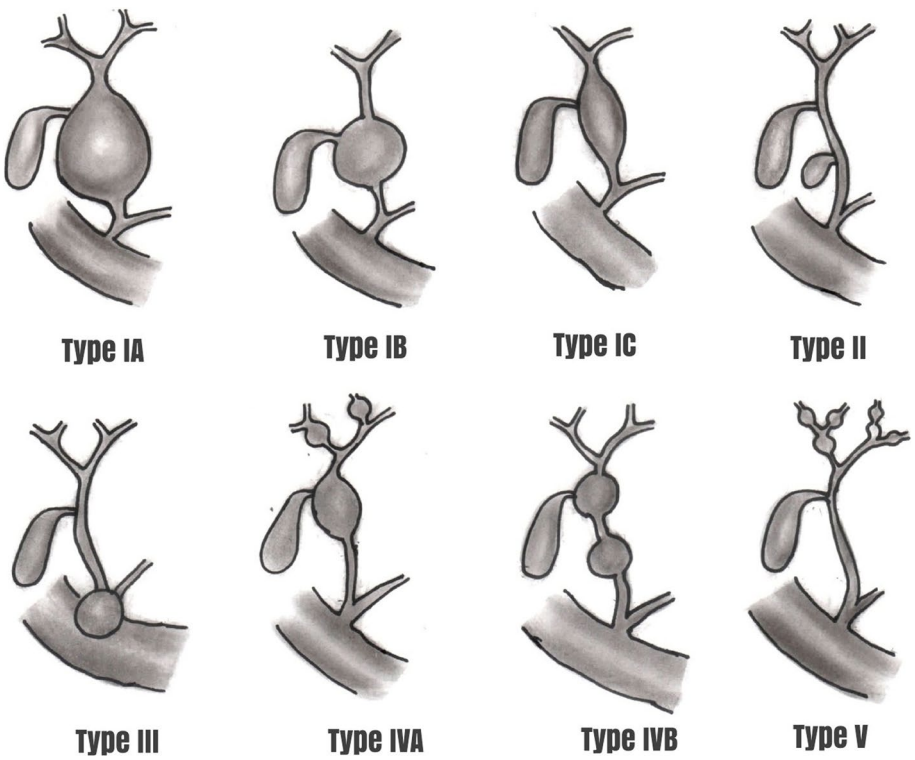


**Fig. 6** Illustration of the Todani classification of bile duct cysts

delayed diagnosis include hepatocellular injury without cirrhosis, cholelithiasis, choledocholithiasis, cystolithiasis, bile duct stricture, bile peritonitis secondary to cyst rupture, portal hypertension secondary to liver cirrhosis, and hepatobiliary malignancy [2, 18]. The most alarming complication confirmed in the current case was liver cirrhosis; however, macroscopic and histological regression of biliary cirrhosis following choledochal cyst drainage has been reported in the literature [19, 20], a possibility that might happen on long-term follow-up.

Abdominal ultrasonography (US) is the most common initial diagnostic investigation given its wide availability and cost-effectiveness [2]. The sensitivity of US for diagnosing CCs ranges from 71 to 97% [8]. MRCP is the diagnostic modality of choice due to its high sensitivity and specificity that ranges from 73 to 100% and 90–100%, respectively; nonetheless, limited capacity to detect minor ductal anomalies or small choledochoceles has also been reported [21]. MRCP is excellent in characterizing the cyst anatomy and its relation to the biliary tree, thus aiding in surgical planning [2, 9, 22]. In comparison to MRCP, abdominal CT scan is relatively disadvantaged in delineating the biliary tree and pancreatic duct anatomy and is reserved for situations where an associated tumor is suspected or in absence of other sophisticated imaging modalities [8, 23]. However, parallel testing that involves augmenting diagnostic tools with an alternative as applied to the presented case has the advantage of increasing the test sensitivity [24]. Endoscopic retrograde cholangiopancreatography (ERCP) was formerly one of the diagnostic modality, but its role has been replaced by MRCP due to its invasive nature and procedure-associated complications such as pancreatitis and cholangitis which may further complicate an anticipated surgery. In a case when the anatomic details of the biliary tree cannot be demonstrated by MRCP, intraoperative cholangiography is indicated [23]. The technetium-99 hepatobiliary iminodiacetic acid (HIDA) scan has a limited role in the diagnosis of CC and is particularly useful in delineating continuity of the cyst with bile ducts and in diagnosing cyst rupture [8]. Invasive procedures such as ERCP and percutaneous transhepatic cholangiography are reserved for the management of choledochal cyst-associated complications such as cholangitis [21, 25]. The role of diagnostic laparoscopy for types I–III CCs which are seldom missed on MRCP is yet to be explored [21].

The surgical treatment of CC has evolved from simple aspiration and marsupialization to more complex cyst excisions and biliary-enteric reconstruction over the past decades. This treatment evolution curtails the incidence of complications encountered as a result of simple drainage procedures and choledochocystoenterostomies, i.e., cystojejunostomy and cystoduodenostomy. Such complications included cholangitis due to bacterial contamination in repeated cyst aspiration, duodenal/jejunal reflux or anastomotic strictures, stones formation, and development of intracystic malignancy that commonly (≈95%) occurs in the extrahepatic component [8, 26–28]. Moreover, since the advent of minimally invasive surgery for hepatobiliary diseases, laparoscopic surgeries have been performed with the benefit of minimal blood loss and shorter hospital stay at an expense of longer operative time and overall higher costs [2, 5, 29]. However, there is no significant difference in the incidence of postoperative complications such as bile leakage or wound infection rate when compared to open procedures [29].

Currently, surgical options are guided by the type of cyst and associated hepatobiliary pathology [26, 30]. As performed in the presented case, complete cyst excision with cholecystectomy and Roux-en-Y hepaticojejunostomy reconstruction is the standard therapy in type I and the extrahepatic component of types IVA and IVB cysts [18, 26, 31, 32]. The extent of resection in type IVA cysts is still controversial with concerns over the long-term complications (i.e., recurrent cholangitis, hepatolithiasis, secondary biliary cirrhosis, and malignant transformation) related to the residual intrahepatic disease following the standard aforementioned approach [33, 34]. With such apprehension, some authors advocate segmentectomy, sectionectomy, or hemihepatectomy with Roux-en-Y biliary-enteric reconstruction for an intrahepatic disease localized to a resectable portion of the liver [5, 33, 35, 36]. Conversely, in patients with diffuse involvement of the liver, biliary drainage procedures have been deemed ineffective, and liver transplant is considered an ideal option [5, 27, 35]. This also applies in the face of associated liver cirrhosis as for the presented case and in Caroli's disease, provided that the patient satisfies the criteria for liver transplantation.

In technically challenging circumstances that preclude cyst excision such as associated inflammatory adhesions, the preferred alternative is a Roux-en-Y choledochocystojejunostomy. Repeated cholangitis with marked pericystic inflammation may also prevent a safe complete cyst excision. This can be managed by resection of the anterolateral part of the cyst followed by mucosectomy of the inner epithelial lining prior to reconstruction as described by Lilly 1978 [37]. Lilly's technique avoids damage to the portal vein in difficult posterior dissection. External drainage is indicated for a perforated cyst in patients whose condition is too unstable for cyst excision and a bilioenteric anastomosis [23]. Because of the age-related risk of malignancy in adults and the likelihood of late anastomotic strictures in individuals treated without cyst resection, long-term follow-up is required.

Treatment options for types II, III, and V cysts have not been extensively explored due to the rarity of these variants. Options for type II cysts include diverticulectomy with or without T-tube decompression of the common bile duct or cystoenterostomy for cysts arising from the intrapancreatic portion of the common bile duct [26]. Endoscopic sphincterotomy and cyst unroofing have become the treatment of choice for type III cysts [38, 39]. This procedure overrides the theoretical malignant transformation risk-elimination benefit of transduodenal cyst excision due to the subtle risk of cancer in choledochoceles [8].

## Conclusion

The startling finding picked on follow-up MRCP that showed regression of the dilated intrahepatic biliary radicles following surgery acquaints clinicians on the possibility of misclassifying a type I cyst for a type IVA cyst. Unless an alternative indication is present, we recommend refraining from aggressive surgeries that involve hepatic resections when preoperative imaging studies are inconclusive and when intraoperative cholangiography is unavailable. Nonetheless, further studies are needed to eliminate the uncertainties on the extent of resection of type IVA cysts.

#### Abbreviations

APDU: Anomalous pancreaticobiliary duct union; CBD: Common bile duct; CC: Choledochal cyst; CT: Computerized tomography; ERCP: Endoscopic retrograde cholangiopancreatography; GB: Gallbladder; MRCP: Magnetic resonance cholangiopancreatography; US: Ultrasonography.

#### Acknowledgements

We acknowledge the effort of Victorin Karia Amani, a medical student from the Muhimbili University of Health and Allied Sciences, who sketched the illustrations in Fig. 6.

#### Authors' contributions

DJH gathered the clinical information, conceptualized and drafted the initial manuscript, did an extensive literature review, and was a major contributor in writing the final manuscript. DWK did an extensive literature review and critically revised, prepared, and copyedited the final manuscript. MDB reviewed, analyzed, and interpreted the radiological information and also contributed in revising the final manuscript. JLM contributed in gathering the clinical information and preparing the initial draft. MG and HMC reviewed, analyzed, and interpreted the pathological information. AHM, ZMB, and PJN selected the case, supervised, and reviewed the final manuscript. All authors approved the final manuscript as submitted.

#### Funding

None

#### Availability of data and materials

Not applicable

## Declarations

#### Ethics approval and consent to participate

Ethical clearance was obtained from the Institutional Review Board of Muhimbili University of Health and Allied Sciences (Ref. No. DA.25/111/01B/168).

#### Consent for publication

Written informed consent was obtained from the child's parent for publication of this case report and accompanying images.

#### Competing interests

The authors declare that they have no competing interests.

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Received: 15 January 2022 Accepted: 15 April 2022
Published online: 26 April 2022

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