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## Management of a massive choledochal cyst in a 12 year-old girl: Which imaging modalities should be performed preoperatively?



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### ABSTRACT

A previously well 12 year-old girl presented to the emergency department with a three-day history of fatigue, pruritus, and epigastric discomfort. Abdominal ultrasound revealed a cystic mass in the right upper quadrant measuring 17.0 × 13.2 × 11.7 cm. Magnetic resonance imaging (MRI) confirmed the diagnosis of a choledochal cyst and hepatobiliary iminodiacetic acid (HIDA) scintigraphy demonstrated a communication between the cyst and biliary tree. Percutaneous cholangiography was also performed but did not provide additional diagnostic information. The patient was brought to the operating room a few days later for open resection of the choledochal cyst, cholecystectomy, and Roux-en-Y hepaticojejunostomy. Frozen sections were obtained to ensure complete excision of cyst mucosa. The patient continues to do well more than 18 months post-operatively with no signs of recurrent inflammation or malignancy. Multiple imaging modalities may be used preoperatively to assess the anatomy and subtype of choledochal cysts. The role of invasive options, such as percutaneous cholangiography or endoscopic retrograde cholangiopancreatography (ERCP), remains limited and unclear. Here, we present a potential imaging algorithm to assist with preoperative workup and avoid invasive diagnostic procedures whenever possible.

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Choledochal cysts are dilatations of the biliary tree, which can involve the extrahepatic ducts, intrahepatic ducts, or both. This condition often presents with one or more components of the classic triad: abdominal pain, jaundice, and a palpable mass in right upper quadrant [1]. The Todani classification of choledochal cysts, an expansion of the system originally developed by Alonso-Lej et al., is used to classify the type of cyst based on the location and morphology of bile duct dilatation. Types range from I (true choledochal cyst) to V (Caroli's Disease) [2] (Fig. 1). If left untreated, choledochal cysts can cause significant morbidity and mortality due to recurrent cholangitis and the development of cholangiocarcinoma.

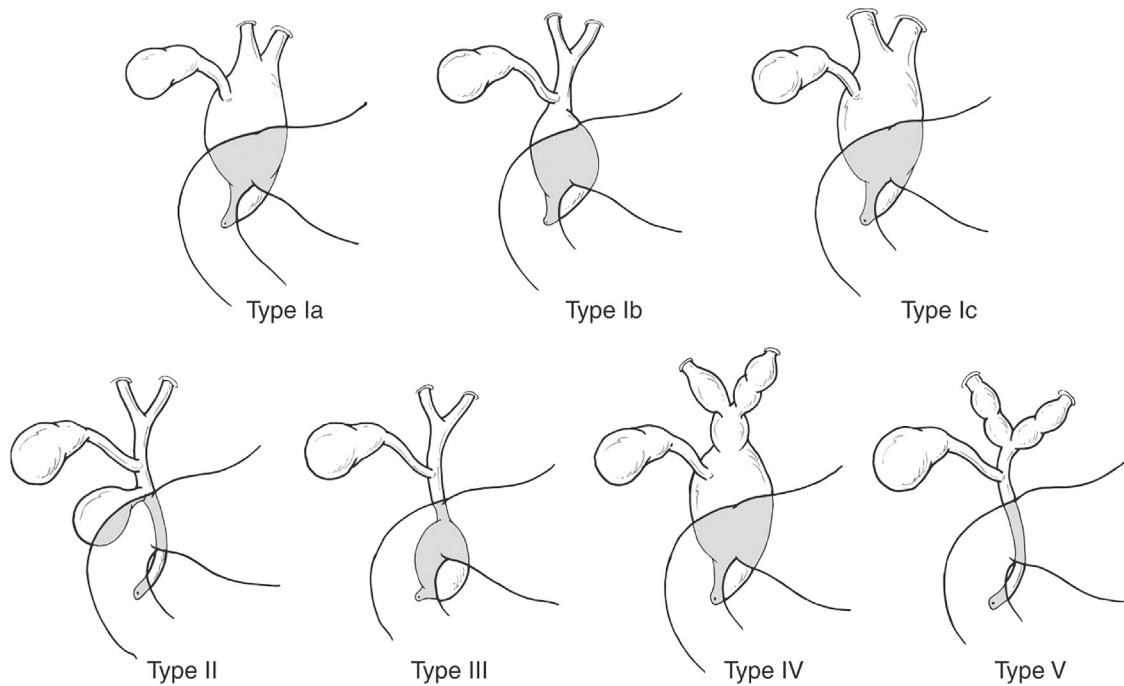
A variety of imaging modalities are available to diagnose the presence of a choledochal cyst and assess its anatomy preoperatively. These include ultrasound, computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP), and biliary scintigraphy (i.e., hepatobiliary iminodiacetic acid (HIDA) scan) [3]. Some clinicians advocate for the use of percutaneous cholangiography and/or endoscopic retrograde cholangiopancreatography (ERCP) as adjuncts, since these procedures provide detailed images of the biliary tree, can rule out the presence of a pancreaticobiliary malformation, and provide thorough evaluation of the cyst itself to aid in classification [4–7]. Unfortunately, these procedures are invasive, require conscious sedation or general anesthesia, and with respect to ERCP, have a non-negligible risk of pancreatitis.

In this report, we present the case of an adolescent girl diagnosed with an unusually large choledochal cyst. She was assessed with a variety of imaging modalities preoperatively. While she experienced an excellent clinical outcome in hospital and in medium-term follow-up, we wonder whether all of these imaging techniques were helpful or necessary. We hope this case will encourage others to reflect on their treatment decisions with

**Abbreviations:** ERCP, endoscopic retrograde cholangiopancreatography; HIDA, hepatobiliary iminodiacetic acid; MRCP, magnetic resonance cholangiopancreatography; MRI, magnetic resonance imaging.

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**Fig. 1.** Todani classification of choledochal cysts (reprinted with permission from Elsevier [2]): Ia (cystic dilatation), Ib (segmental dilatation), Ic (diffuse dilatation), II (diverticulum), III (choledochoceles), IV (multiple intrahepatic and extrahepatic cysts), V (intrahepatic cysts only).

regards to preoperative imaging of choledochal cysts. We also present a possible algorithm to guide clinicians in selecting imaging modalities that maximize clinical utility, while minimizing risk to patients.

### 1. Case report

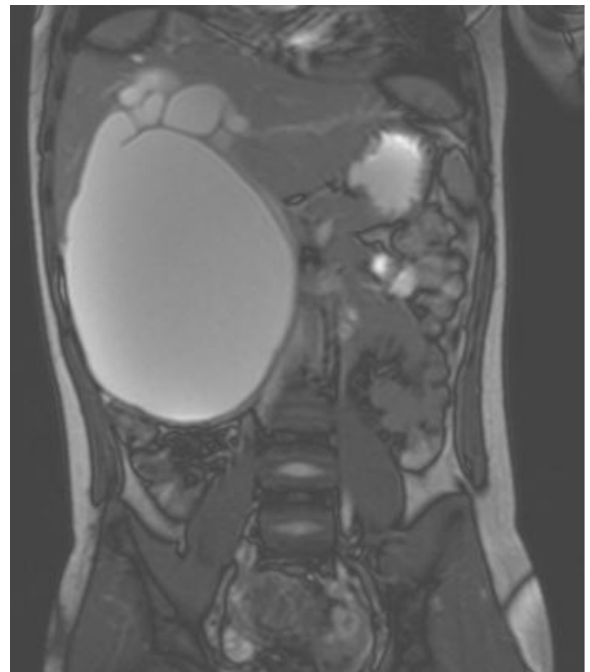
A 12 year-old girl of Asian descent presented to the emergency department in a community hospital with facial cellulitis. She was given a prescription for oral antibiotics and discharged home. Over the next few days, she developed dark urine, pruritus, fatigue, scleral icterus, and mild epigastric pain. There was no fever, nausea, emesis, or weight loss. She returned to the emergency department five days later with ongoing abdominal pain and was found to have elevated liver enzymes, bilirubin, and platelets. An abdominal ultrasound revealed a cystic mass in the right upper quadrant measuring  $17.0 \times 13.2 \times 11.7$  cm.

The patient was transferred to a tertiary children's hospital the following day for assessment and further investigations. On arrival, vital signs were within normal limits and the patient was mildly jaundiced without stigmata of liver disease. The abdomen was soft and non-distended, but examination was significant for a tender mass in the right upper quadrant. The facial cellulitis from a few days prior had resolved.

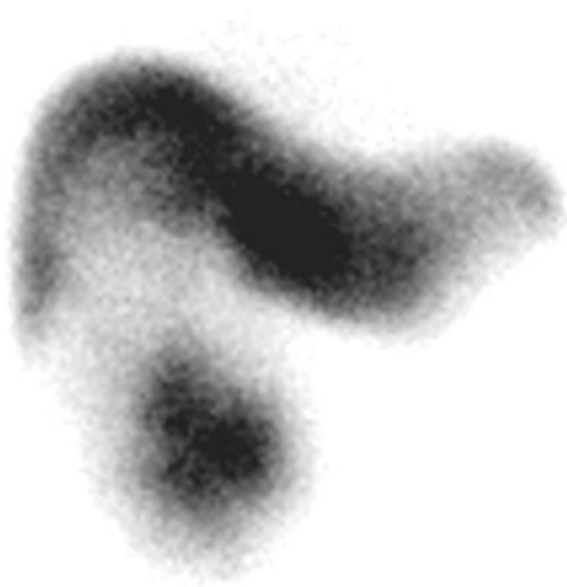
Initial laboratory investigations revealed markedly elevated liver enzymes, including alanine transaminase of 1702 units/L, aspartate transaminase of 695 units/L, alkaline phosphatase of 1087 units/L, and gamma-glutamyl transpeptidase of 657 units/L. Total bilirubin was elevated at 51  $\mu\text{mol/L}$  with a conjugated bilirubin of 39  $\mu\text{mol/L}$ . Lipase was normal at 19 units/L. White blood cell count was also normal at  $4.0 \times 10^9/\text{L}$ . Inflammatory markers were moderately elevated, including a platelet count of  $639 \times 10^9/\text{L}$ , C-reactive protein of 28.9 mg/L, and erythrocyte sedimentation rate of 58 mm/h.

The following day, the patient underwent magnetic resonance imaging (MRI) of the abdomen and MRCP, which confirmed the

diagnosis of a choledochal cyst (Fig. 2). This examination also revealed dilatation of the cystic duct up to 1.7 cm, dilation of common hepatic duct up to 3 cm, and mild dilatation of the intrahepatic ducts. There was no lymphadenopathy, free fluid, or gallstones. The common bile duct could not be adequately visualized. HIDA scan confirmed the presence of a connection between the choledochal cyst and biliary tree (Fig. 3). Delayed



**Fig. 2.** Magnetic resonance images showing a massive choledochal cyst, dilated cystic and common bile ducts, and mildly dilated intrahepatic ducts.



**Fig. 3.** Delayed images from a hepatobiliary iminodiacetic acid (HIDA) scan demonstrating filling of the choledochal cyst and connection with the biliary tree.

images demonstrated drainage of radioactive tracer into the gastrointestinal tract, suggesting that the cyst was either Todani type I or IV.

Percutaneous cholangiography was performed the following day in an attempt to better delineate the anatomy and subtype of the choledochal cyst. The biliary system was accessed through the gall bladder. Injection of contrast once again confirmed the presence of a connection between the choledochal cyst and biliary tree (Fig. 4). A catheter was left in place as a cholecystostomy tube and approximately 1.8 L of bile was drained. Over the next three days, serum liver enzyme levels decreased but remained slightly elevated.

Four days after the percutaneous cholangiogram (and approximately one week after admission to hospital), the patient was brought to the operating room for resection of the choledochal cyst under general anesthesia. Operative steps included laparotomy, open resection of choledochal cyst, cholecystectomy, and Roux-en-Y reconstruction with hepaticojejunostomy. Access to the peritoneal cavity was achieved through a right subcostal incision. The cholecystostomy tube was removed and the transverse colon was mobilized off the inferior edge of the choledochal cyst. The cyst was abutting the hepatic artery, hepatic duct, portal vein, and pancreas.

Resection was performed using a combination of blunt dissection and electrocautery. Suture ligation and surgical clips were used to ligate larger vessels. A small rim from the inferior aspect of the cyst was left adherent to the posterior aspect of the pancreas due to persistent bleeding from smaller vessels. The cyst was opened and we proceeded with submucosal resection in this area (Fig. 5). Complete excision of cyst mucosa from the pancreas was confirmed with frozen sections. The gall bladder was then mobilized off the liver and left attached to the cyst as a single specimen.

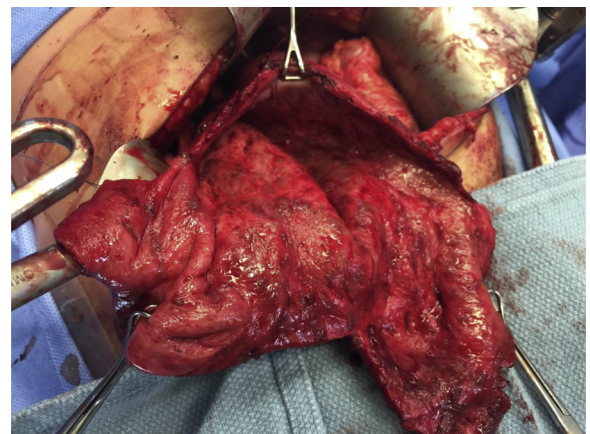
The gall bladder and choledochal cyst were removed en bloc by dividing the common hepatic duct just below the confluence of four segmental hepatic bile ducts. Frozen sections from the proximal margin of the common hepatic duct revealed residual choledochal cyst mucosa. As a result, an additional 4 mm of common hepatic duct was excised. Additional frozen sections from this area were clear of cyst mucosa. Reconstruction was performed by creating a Roux-en-Y retrocolic jejunal limb. A Jackson Pratt (JP) drain was left



**Fig. 4.** Percutaneous cholangiogram confirming the presence of a connection between the choledochal cyst and biliary tree.

in the peritoneal cavity near the hepaticojejunostomy. The abdomen was closed and the patient was taken to the pediatric intensive care unit in stable condition.

On postoperative day 2, the patient developed tachycardia and hypotension. Serum hemoglobin was 58 g/L. The patient received two units of packed red blood cells but did not require any further interventions. Following this, she was transferred to the surgical ward, the JP drain was removed, and diet was advanced. The patient was discharged home on post-operative day number 6. Final pathology revealed complete resection of a type 1 choledochal cyst. There was evidence of chronic inflammation but no signs of malignancy. The patient continues to do well more than 18 months



**Fig. 5.** Giant choledochal cyst opened to facilitate excision of cyst mucosa near the pancreas.

post-operatively with no signs of recurrent inflammation or malignancy.

## 2. Discussion

Choledochal cysts occur with an incidence of 1 in 100,000–150,000 live births in Caucasians and as high as 1 in 1000 among those of Asian descent [8]. Nearly 25% of patients present within the first year of life, and 60% within the first decade [9]. The size of cysts can vary, but rarely exceed 9 cm [10].

In cases of suspected choledochal cyst, CT, MR imaging sonography, ERCP, cholescintigraphy and percutaneous cholangiogram have all been reported to be useful for classification and surgical planning [3]. In this case, determining the subtype of the cyst preoperatively using the Todani system proved challenging. It was not possible to definitively classify the lesion as either type I or IV. Some groups, such as Visser et al., have argued that types I and IV are the same disease and that making a distinction between the two is artificial and a source of confusion [11].

In many cases, choledochal cysts are first detected on imaging performed for abdominal pain or jaundice, such as abdominal ultrasound or computed tomography. When a choledochal cyst is

suspected, MRI with MRCP is a reasonable next step. This technique can confirm the diagnosis of a choledochal cyst, demonstrate the presence of a communication with the biliary tree, and has no ionizing radiation. If the association between the cyst and biliary tree remains unclear, then a HIDA scan may be a helpful adjunct [12,13].

In the past, some centers advocated for the use of more invasive techniques, such as percutaneous cholangiography or ERCP. Improvements in MRI technology suggest that these investigations may no longer be necessary. A 30-year review of choledochal cysts in 19 children and 73 adults found that percutaneous drainage was performed routinely in the past but less frequently since 2000 [14]. ERCP can also be avoided in most cases. This technique typically does not provide additional diagnostic information compared to MRCP alone, requires general anesthesia in a child, and carries a non-negligible risk of pancreatitis [12,13].

We wonder whether percutaneous cholangiography may have a role in draining especially large choledochal cysts, such as in this case. Allowing the cyst to drain for a few days may facilitate complete surgical excision from a purely technical standpoint [15] or alleviate symptoms in the preoperative period [16]. We have incorporated our thinking and approach into an algorithm (Fig. 6).

## 3. Conclusion

There are multiple imaging modalities available to determine the anatomy and subtype of choledochal cysts. The assessment of larger cysts can be complicated by distortion of surrounding structures and obscured anatomy. Even in these difficult and unusual cases, using a combination of ultrasound, MRI, and HIDA is a reasonable strategy that minimizes radiation, adequately delineates the anatomy, and can confirm the presence of a communication between the cyst and biliary tree. Invasive procedures such as percutaneous cholangiography and ERCP should be avoided whenever possible, and only used in cases where the anatomy remains unclear and when the results will change clinical management.

## Conflict of interest

The authors declare that they have no conflict of interest.

## Consent

Written consent was obtained from the patient's parents to publish this report.

## Authors' contributions

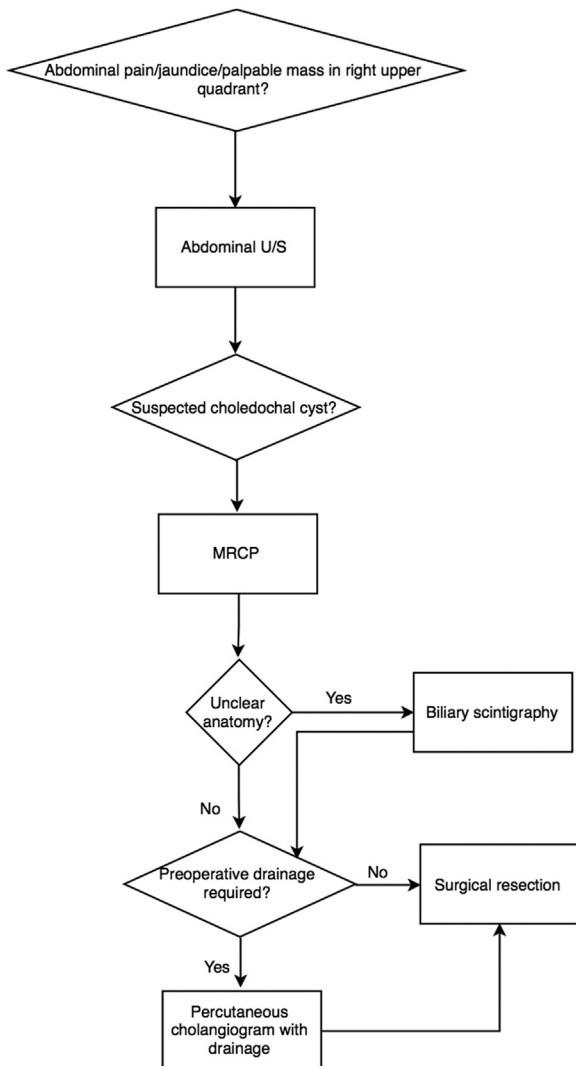
All authors contributed to the study concept, data collection and/or data analysis, and writing and/or revising the manuscript. All authors have reviewed and approved the final manuscript as submitted.

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**Fig. 6.** Possible algorithm to guide preoperative imaging and management of choledochal cysts.

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