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## Hemobilia secondary to choledochal cyst

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### ABSTRACT

Non variceal upper gastrointestinal hemorrhage in children is rare. A 5-year-old presented with hematemesis and melena. Pre-referral imaging and exploratory laparotomy did not reveal the source of bleeding. Hemobilia was detected on endoscopy MRI showed a choledochal cyst. The patient underwent successful resection of the cyst and hepaticojejunostomy.

### 1. Introduction

Late presentation choledochal cyst with hemobilia is a rare cause of upper gastrointestinal hemorrhage in children. This case highlights the challenges in diagnosis, definitive care of this rare complication of late presentation choledochal cyst.

### 2. Presentation

A 5-year-old male was transferred with a one-year history of recurrent severe colicky abdominal pain and a six-month history of recurrent hematemesis and melena. His antenatal, birth, growth and developmental history was described as normal. There was no history of umbilical vein catheterization, pancreatitis, worm infestation, abdominal trauma, liver biopsy, use of NSAIDs or herbal medication. The pain was relieved transiently with antacids and did not have any periodicity, aggravating or relieving factors. Upper and lower gastrointestinal endoscopy and CT-scan angiography, done prior to referral, were reported as normal.

Four weeks before transfer the patient developed severe abdominal pain associated with hematemesis and underwent an emergency laparotomy through an upper transverse incision. The surgical exploration was reported as negative and he was managed post-operatively with transfusions and proton-pump-inhibitors.

The patient was transferred due to recurrence of the symptoms. At admission, he was pale. He did not have jaundice. He had a supra-umbilical transverse incision scar. There was no clinical sign of porto-systemic shunting. The liver and spleen were not enlarged.

The key differentials at this stage were oesophageal variceal hemorrhage secondary to portal hypertension, gastric variceal bleed, or an arterio-portal vascular malformation. These did not seem to fit well with the history, previous imaging, and surgical findings.

At this point, stabilization-and-re-investigation in tandem was the best available approach.

#### 2.1. Investigations

He had a very low hemoglobin (5.3g/dl), elevated liver enzymes with normal bilirubin.

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Table 1 shows the patient's hemogram and liver enzymes at admission.

While undergoing blood transfusions, he had a Technetium-99 red cell scan which was negative.

The next day he had an upper endoscopy which found normal oesophageal, gastric, and duodenal mucosa.

On the same evening he had another episode of hematemesis.

A repeat endoscopy during this episode showed blood trickling from the ampulla of Vater (Fig. 1). Subsequently, a hepatobiliary ultrasound (Fig. 2) and magnetic resonance cholangiopancreatography (MRCP) (Fig. 3) showed a type-IA choledochal cyst with intracystic hemorrhage and a thick-walled gallbladder. There was no portal carvenoma, the splenic vein was normal and there was no vascular malformation.

## 2.2. Treatment

The patient continued to receive blood transfusions and prepped for surgery. The abdominal cavity was exposed through an extended right sub-costal incision. The gallbladder was found dilated, thick-walled and filled with altered blood. The common bile duct (CBD) was dilated, thick-walled and contained blood clots (Fig. 4). The hepatic artery and portal vein were normal.

**Table 1**

Blood count, liver enzymes and serum protein. (Low hemoglobin. Elevated GGT, AST & ALT) (WBC-total white blood cell count, HB -Hemoglobin concentration, INR – International normalized ratio, APTT-activated partial thromboplastin time, LDH- lactate dehydrogenase, ALP- alkaline phosphatase, GGT-gamma glutamyl transferase, T.BIL-total bilirubin, D.BIL-direct bilirubin, AST-aspartate transferase, ALT-Alanine transferase).

		Normal values
WBC	10.48	5-15x10 <sup>9</sup> /L
Hb	5.3	11-13g/dl
PLATELETS	292	217-497 x10 <sup>9</sup> /L
INR	0.87	≤ 1.0
APTT	20 secs.	Control = 28sec
TOTAL PROTEIN	59	60-80 g/l
Albumin	36.7	38-54 g/l
LDH	253	120-300 U/l
ALP	232,	93-309 U/l
GGT	320	3-22 U/l
T.bil	3.6	0-21 μmol/l
D.bil	3	0-3.4 μmol/l
AST	62.8	0-36 U/l
ALT	146.1	0-29 U/l
Lipase	23.3	5-31 U/l



**Fig. 1.** Gastro-duodenoscopy image. Blood trickling into the duodenum through the ampulla (arrow).

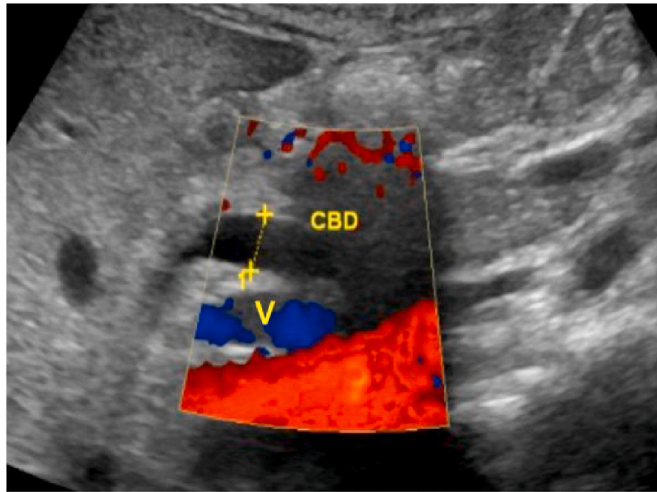


Fig. 2. Hepatobiliary ultrasound. Dilatation of the common bile duct (CBD). Normal Portal vein (V).

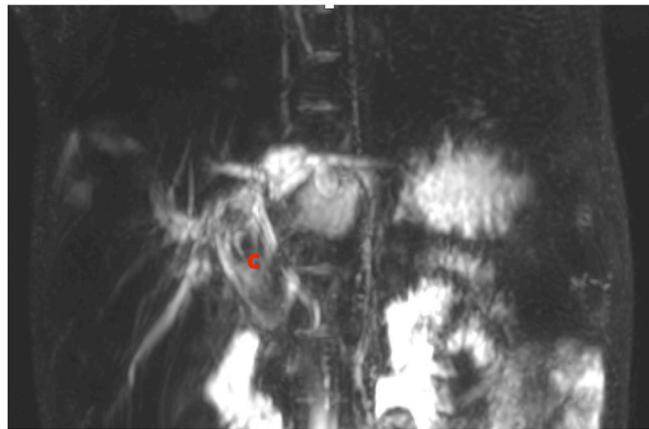


Fig. 3. Coronal MRI-scan image. Dilated CBD (C).

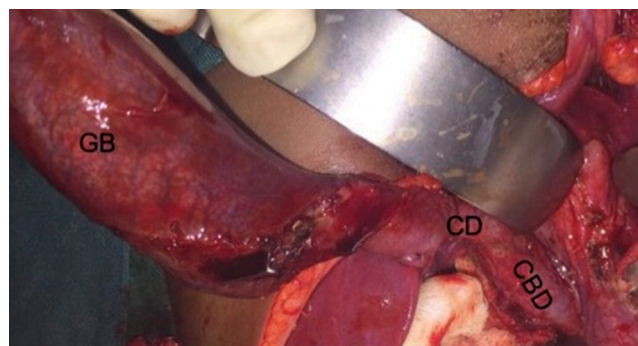


Fig. 4. Subcostal laparotomy findings. Dilated gallbladder with thickened wall and blood content (GB). Attenuated, short and thickened cystic duct (CD), Fusiform dilated common bile duct with thick wall and blood content (CBD).

The gallbladder was mobilized off the liver bed, the dilated CBD mobilized gently off the portal structures and vascular loops slinged around the portal vein, hepatic arteries and choledochal cyst.

The cyst-CBD complex was excised in continuity. The points of transection were cranial to a small section of cyst-CBD interface at the hepatic duct confluence proximally and within the pancreas caudal to the point of transition from cystic wall to common bile duct distally with stay sutures to the proximal and distal stumps. The right and left hepatic ducts and distal duct stumps were gently lavaged with warm saline via a 6F feeding tube to clear any clots, bile sludge and crystals. The distal stump was oversewn with poly-

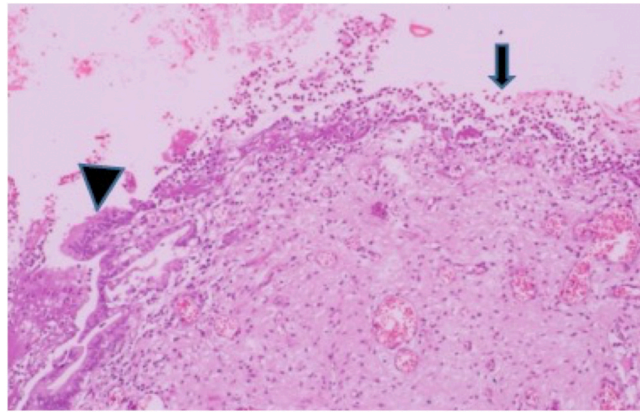


Fig. 5. Histology of the resected cyst: Image shows part of a columnar mucosal lining (arrowhead) with ulceration and a chronic inflammatory cell infiltrate (arrow) (x20 magnification).

dioxanone suture (PDS) 6/0. Bilio-enteric continuity was re-established via a Roux-N-Y retro-colic end-to-end interrupted suture hepaticojejunostomy.

### 2.3. Outcome and follow-up

The post-operative recovery was un-eventful, and he was discharged 6 days after surgery.

Histology of the cyst showed chronic lymphoplasmacytic infiltrate with focally denuded mucosa and non-metaplastic columnar epithelium (Fig. 5).

The chief complaint was occasional abdominal discomfort in the first 6 months follow-up. There were no episodes of hematemesis or melena. An MRCP at 1-year showed normal intrahepatic bile ducts with normal drainage into the Roux limb. The hemoglobin and liver function have remained normal at 5 years post-surgery. The plan is to continue annual reviews and counselling for life-long follow-up for signs of cholangitis and ultrasonography for long-term surveillance.

### 3. Discussion

Cases of haemobilia have been reported in patients who have had blunt liver trauma, liver biopsy, liver transplant and vascular malformations[1–7]. Other rare causes of haemobilia in children include worm infestations involving the biliary tree[8–10], choledochal cyst with pseudoaneurysm formation, cyst wall erosion into an adjacent vessel or hematemesis due variceal bleed secondary to undiagnosed choledochal cyst associated portal hypertensions[11–13]. Cases of liver abscess with erosion into the bile ducts and gastric duplication with vascular fistula presenting with haemobilia have also been described[14,15].

In developing settings, the infective/infestation causes and late presentation complicated choledochal cysts are an important consideration in the diagnostic work-up in a child with non-variceal upper gastrointestinal hemorrhage. In these scenarios, the approach to non-traumatic haemobilia as described by Bairagi and Aronson is valuable[10] The emergent nature of the presentation requires one to undertake stabilization and diagnostic measures in tandem.

Early diagnosis of choledochal cyst is also problematic due to the non-specific presentation with recurrent abdominal pain. Late presentation is common and more likely to target diagnostic maneuvers in the setting of biliary obstruction or pancreatitis [16–18].

In this case the patient was five years old, had longstanding episodes of abdominal pain and subsequently presented with significant hematemesis. Biliary obstruction in choledochal cyst is possible due to biliary stasis from poor clearance by a mechanically less efficient dilated duct which in turn results in bile sludge and crystals. Acute cyst distension and obstruction from the blood clots may have worsened the abdominal pain. The cause of bleeding is less clear. Histology confirmed cyst mucosal erosion. Therefore, the working theories to explain the distension and bleeding due to mucosal erosion are as follows. First, choledochal cysts have been associated with an abnormal pancreatobiliary union (pancreaticobiliary malunion – PBMU). PBMU is characterized by a proximal confluence of the common bile duct and the pancreatic duct resulting in a long common channel between the sphincter of Oddi and the point of confluence[19]. The long channel leads to higher intraluminal pressures resulting in reflux into the distal bile duct and cyst (pancreaticobiliary reflux – PBR) [20]. Second, the distension of the cyst may lead to diminished mucosal blood flow due to obstruction of the vessels traversing the serosa to the mucosa get stretched by increased cyst wall tension. In this case the pancreatic duct, point of confluence and common channel were not visible on the MRCP which is more sensitive in older patients. Endoscopic cholangiopancreatography (ERCP) or intra-operative cholangiography are more sensitive in delineating these anatomical variations. Both were not viable options in this clinical scenario.

While haemobilia is a rare cause of upper gastrointestinal bleeding in most children, the common causes need to be ruled out while actively looking for associated hepatobiliary pathology[10,21,22]. An upper GI endoscopy during an active bleed in a hemodynamically stable patient is probably the only definitively confirmatory diagnostic measure where previous imaging has failed. The other tests such as CT-scan angiography and nuclear scan have low sensitivity and specificity respectively because they require a high flow rate for angiography and active bleeding in both. Once the diagnosis of haemobilia is confirmed, ultrasound with doppler and

Cross-sectional imaging with intravenous contrast are necessary to check for arterio-venous malformations and to define the biliary and vascular anatomy. The ultrasound is reliable in detecting biliary pathology and vascular malformations. The main challenge with ultrasound is its user-dependence. MRCP is the best non-invasive investigation for hepatobiliary and pancreatic anomalies but can miss the pathology due to technical (poor image acquisition) or interpreter limitations.

Exploratory laparotomy for these patients, as a last resort, can be quite challenging and one might miss the source of bleeding if the porta-hepatis is not adequately assessed.

Definitive management, once the diagnosis is confirmed involves complete excision of the cyst and establishment of bilio-enteric drainage through a Roux-N-Y hepaticojejunostomy. It is a difficult operation especially in the setting of recurrent inflammation and complete excision of the mucosa as described by Lily is an alternative [23]. Long-term follow-up is necessary in these patients to detect and treat cholangitis which may not be associated with a stricture at the hepatico-jejunostomy and also lifelong surveillance for cholangiocarcinoma of the remnant duct [24].

### Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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