

# Management of choledochal cyst with portal hypertension

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## ABSTRACT

**Portal hypertension (PHT) is a rare complication associated with choledochal cysts. Management issues of PHT patients are inadequately addressed, as its incidence is low and underlying causes variable. We report three cases of choledochal cyst with PHT. All patients had type IVa choledochal cysts, and the causes of PHT were secondary biliary cirrhosis (SBC) (two cases) and alcoholic liver disease (one case). Clinical presentation included jaundice, gastrointestinal bleeding and ascites. One patient with SBC successfully underwent excision with Roux-en-Y hepaticojejunostomy, while the patient with cholangitis was managed with endoscopic retrograde cholangiopancreatography stenting. The last patient with alcoholic liver disease was managed conservatively for seven years and died of liver failure. Management of choledochal cysts depends on the severity of liver disease in cases of cirrhosis of unrelated cause, while those with SBC should be considered for surgical management. Endoscopic stenting may be considered as a temporary measure in high-risk cases.**

**Keywords: choledochal cyst, cirrhosis, hypertension, portal hepaticojejunostomy, secondary biliary**

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## INTRODUCTION

Choledochal cyst is predominantly a disease of the younger age group. Only about 20% of choledochal cysts are diagnosed in adults.<sup>(1)</sup> These cysts are more commonly associated with hepatobiliary complications,<sup>(2-5)</sup> with portal hypertension (PHT) reported as one of its rare complications.<sup>(2-5)</sup> The management issues of such patients are generally inadequately addressed, as its incidence is low and the underlying causes of PHT variable. We describe three patients who had choledochal cysts with associated PHT, and highlight the management of these patients and its related issues. Choledochal cysts were classified according to the Alonso-Lej classification

with Todani modifications, based on radiographic and operative findings.

## CASE REPORT

### Case 1

A 40-year-old woman presented with a single episode of haematemesis and melaena two years ago, which required blood transfusion. For the past year, she had experienced abdominal pain and abdominal distension. Examination showed mild pallor, icterus, no pedal oedema, hepatomegaly (3 cm), splenomegaly (6 cm) and mild ascites. Laboratory parameters revealed haemoglobin 9.8 g/dL, platelet count 135,000 cells/dL, prothrombin time (PT) 18.0 seconds (11.4 seconds for control), serum bilirubin 2.1 mg/dL, serum glutamic oxaloacetic transaminase (SGOT) 153 U/L, serum glutamic pyruvic transaminase (SGPT) 84 U/L, alkaline phosphatase 460 U/L and serum albumin 28 g/L.

Abdominal ultrasonography showed hepatomegaly with a coarse echotexture, dilated intrahepatic biliary radicals (IHBR), common bile duct (CBD) of 2.9 cm, gallstones and cystolithiasis. On Doppler study, the liver was nodular and shrunken, with multiple splenic hilar, lienorenal and periportal collaterals. The patient had Grade II/III oesophageal varices on endoscopy. Magnetic resonance (MR) imaging/magnetic resonance cholangiopancreatography (MRCP) showed a type IVa choledochal cyst with sludge and calculi in the distal CBD and right post sectoral duct, nodular liver, splenomegaly and ascites. Serum-ascites albumin gradient (SAAG) was > 1.1 g/dL, suggestive of PHT. Viral markers for hepatitis B and C were negative.

The working diagnosis of choledochal cyst with cystolithiasis, hepaticolithiasis, PHT due to secondary biliary cirrhosis (SBC), along with gallstones, was kept. The patient underwent two rounds of endoscopic variceal banding. She was started on diuretics, and the PT time was corrected. Preoperative albumin infusion was administered for five days as preoperative preparation. Perioperative findings revealed the presence of a type IVa choledochal cyst (2.5 cm) with multiple pigmented stones in the CBD, a large stone in the posterior sectoral duct, nodular liver with atrophy of the right lobe and hypertrophy of the left lobe, splenomegaly and

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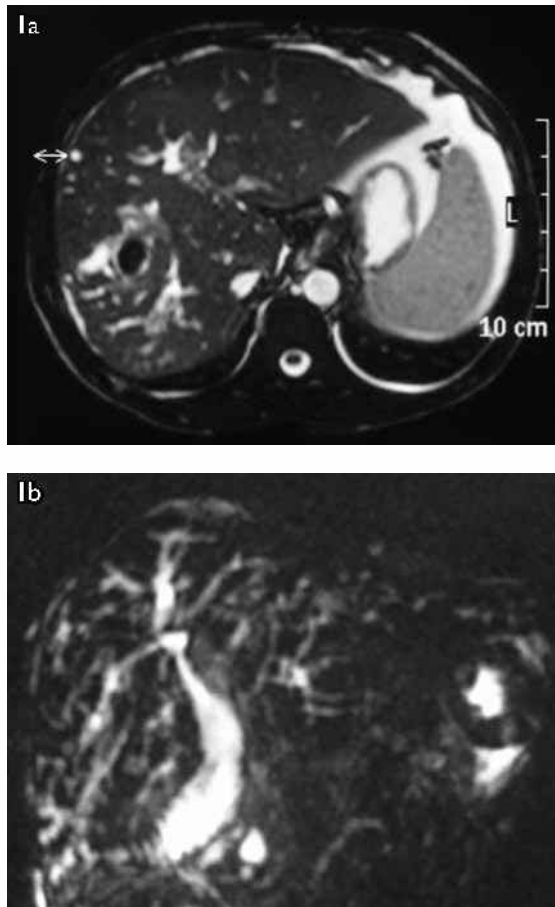
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**Fig. 1** Case 1. (a) Postoperative T2-W MR image shows right and left lobe hypertrophy, retained calculi in the right sectoral duct, splenomegaly and mild ascites. (b) Postoperative MRCP image shows hepaticojejunostomy, with mild intrahepatic biliary dilatation on the right side.

multiple collaterals in the hepatoduodenal ligament and omentum.

The patient underwent excision of the choledochal cyst with Roux-en-Y hepaticojejunostomy. While retrieving the large intrahepatic stone, bleeding occurred and was managed conservatively, requiring two units of blood. Postoperatively, her ascites increased despite being given diuretics, and she developed wound dehiscence on postoperative Day 9. The patient subsequently underwent secondary suturing. Operative biopsy showed fibrous tissue septa with periductular fibrosis, ductular proliferation and intraductular cholestasis, suggestive of obstructive pathology as the cause of cirrhosis. At the one-year follow-up, the patient was well and the ascites were controlled with diuretics. MR imaging/MRCP showed a dilated right post sectoral duct with stone, a normal left system and hepaticojejunostomy (Fig. 1). Endoscopy showed Grade II varices.

#### Case 2

A 66-year-old man presented with complaints of recurrent episodes of jaundice without pruritus



**Fig. 2** Case 2. (a) CT image shows an uncomplicated choledochal cyst with multiple collaterals, patent portal vein and mild ascites. (b) MRCP image shows a type IVa choledochal cyst without any stones and a normal pancreatic duct with mild ascites.

or clay-coloured stool, which had been associated with abdominal distention since 2002. The patient was diagnosed with a choledochal cyst in 2003 and offered surgical management for which he refused. He again developed jaundice for three months, which was accompanied with abdominal distension and an episode of melaena. He had been a chronic alcoholic for the past 50 years, consuming about 80–120 g of alcoholic beverages per day. However, he had been abstaining from alcohol since the previous year. On examination, he had moderate icterus, gynaecomastia, hepatomegaly (6 cm) and moderate ascites. Laboratory

parameters revealed total/direct bilirubin 7.6/4.1 mg/dL, SGOT 145 U/L, SGPT 38 U/L, alkaline phosphatase 207 U/L, total protein/albumin 74/28 g/L and PT 19 seconds (14 seconds for control).

Abdominal ultrasonography demonstrated hepatomegaly (16 cm) with a coarse echotexture and nodular surface, dilated IHBR with fusiform dilatation of the CBD (4 cm), portal cavernoma with ascites. SAAG was > 1.1 g/dL, suggestive of PHT. Endoscopy revealed Grade III/IV esophageal varices and normal papilla. MR imaging/MRCP confirmed the ultrasonographic findings of choledochal cyst type IVa with PHT (Fig. 2). Viral markers for hepatitis B and C as well as autoimmune markers were negative.

The patient was managed conservatively for seven years, during which he had two episodes of jaundice with ascites. Based on the working diagnosis of alcoholic liver disease with Child Class C cirrhosis and uncomplicated choledochal cyst, he was scheduled for primary prophylaxis of varices and endoscopic retrograde cholangiopancreatography (ERCP) stenting to assess and relieve the jaundice due to any obstructive component. Counselling regarding liver transplant was also scheduled. Unfortunately, the patient developed hepatic encephalopathy with hepatorenal syndrome before he could undergo ERCP, and subsequently died of liver failure.

### Case 3

A 49-year-old known diabetic man presented with jaundice that was associated with cholestatic symptoms and weight loss for three months. Examination revealed icterus, hepatomegaly (4 cm) and a palpable gallbladder. Laboratory parameters showed haemoglobin 12.9 g/dL, total bilirubin 12.5 mg/dL, SGOT 81 U/L, SGPT 56 U/L, alkaline phosphatase 528 U/L and albumin 32 g/L.

Abdominal ultrasonography showed hepatomegaly with a coarse echotexture and nodular surface as well as a type IVa choledochal cyst with multiple calculi. Doppler study revealed portal vein (13.5 mm) with multiple collaterals at the splenic hilum. Endoscopy revealed Grade II varices and normal papilla. MRCP confirmed the ultrasonographic findings.

The working diagnosis of choledochal cyst with cystolithiasis, jaundice and PHT was kept. The patient was originally scheduled for stone removal with ERCP but underwent stenting instead, as a large stone could not be removed. Two months later, he underwent repeat ERCP, and multiple stones were removed after lithotripsy. Three months later, the patient was re-admitted due to cholangitis with renal failure. He was

started on intravenous fluids and antibiotics, and stent change was planned. Despite the stent change and endoscopic nasobiliary drain placement, the patient died from sepsis nine months after his diagnosis.

### DISCUSSION

Choledochal cyst is a surgical problem typically related to infancy and childhood.<sup>(1)</sup> The incidence of diagnosis in adulthood is increasing with advances in imaging techniques. In contrast to children, choledochal cysts in adults are associated with a higher incidence of cyst-related complications. These include cystolithiasis, hepaticolithiasis, calculous cholecystitis pancreatitis, malignancy and cirrhosis with PHT.<sup>(2-5)</sup> PHT is a rare condition that complicates the management of choledochal cysts, especially when diagnosed in the fifth or sixth decade of life. It manifests clinically in the form of hepatosplenomegaly, jaundice, haematemesis, melaena or ascites.

The various causes of PHT include extrahepatic biliary obstruction leading to SBC, recurrent inflammation leading to portal vein thrombosis, direct compression of the portal vein,<sup>(6)</sup> and other unrelated aetiologies of cirrhosis. The mechanism responsible for development of PHT determines whether surgical intervention may be beneficial. Direct compression of the portal vein is reported as the main mechanism in children. Surgical decompression of the cyst by internal drainage has been shown to reverse PHT in these patients.<sup>(6)</sup>

Portal vein thrombosis may be suspected in patients who have a history of recurrent cholangitis. In these cases, the cyst may be densely adherent to the portal vein. It can be diagnosed on preoperative Doppler ultrasonography. Modified cyst resection, leaving behind the posterior wall of the cyst that is adherent to the portal vein, with cauterisation of the mucosa, can be employed in these patients.

SBC and unrelated cirrhosis are the main mechanisms responsible for the development of PHT in adults. In our series, two had SBC and one had alcoholic liver disease as the causes of PHT. In adults, PHT due to SBC should be differentiated from that with unrelated causes, as the management differs in the two situations. Since surgical treatment in the presence of PHT is associated with increased morbidity and mortality,<sup>(7)</sup> patients with no evidence of obstruction and definitive evidence of cirrhosis due to other causes can be managed conservatively. One reason for surgery in uncomplicated cyst is the increased risk of hepatobiliary malignancy; this may not be significant, especially when an uncomplicated

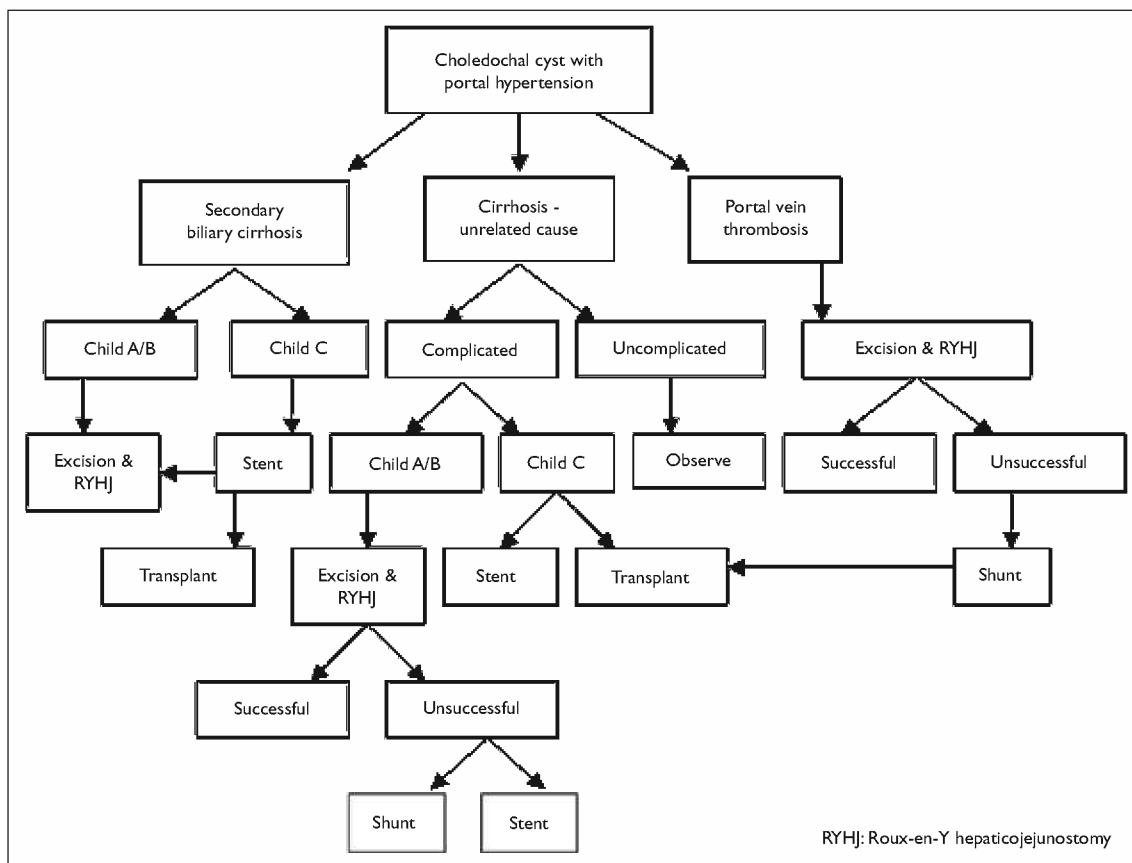


Fig. 3 Algorithm for management of choledochal cyst with portal hypertension.

cyst is detected in the fifth or sixth decade of life. Case 2 was managed conservatively for seven years after the diagnosis of uncomplicated cyst with alcoholic liver disease was made, but he eventually succumbed to liver failure.

Management of choledochal cyst with SBC would depend on the Child Class, the presence of portal vein thrombosis and amount of collaterals around the cyst. Decompression of the cyst prevents further deterioration of liver function, and may even improve or completely reverse cirrhotic changes.<sup>(8)</sup> Surgical decompression appears to be more effective than endoscopic stenting, as observed in our patients. Endoscopic drainage may serve as a temporary measure in patients who are unfit for surgery.<sup>(9)</sup> Prolonged endoscopic drainage may be associated with complications like stent blockage, and may require stent change, as seen in Case 3. Therefore, patients may be considered for definitive surgical treatment once endoscopic stenting has improved the liver function and reduced the surgical risk in these patients.<sup>(9)</sup>

Although decompression by Roux-en-Y drainage can result in adequate long-term functional improvement

in 60%–70% of cases, excision and hepaticojejunostomy is the treatment of choice.<sup>(10)</sup> Surgical treatment requires preoperative preparation, including correction of the coagulation status, improvement of albumin level, and control of ascites and varices. Portal decompression may be required before biliary reconstruction,<sup>(2)</sup> especially in the presence of portal vein thrombosis and collaterals along the hepatoduodenal ligament. On the other hand, a shunt may cause further deterioration of liver function by diverting the portal flow, especially in patients with Child Class C cirrhosis. Liver transplantation should be offered to patients with Child Class C cirrhosis. In the absence of portal vein thrombosis, careful ligation of the collaterals may enable the surgical procedure to be completed.<sup>(11)</sup>

In the presence of partial thrombosis with Child Class A/B cirrhosis, excision of the choledochal cyst may be attempted first, with the shunt kept as reserve option in the event that access to the hepatoduodenal ligament is not possible as planned, as was the case in our first patient. Choledochal cyst with PHT is rarely encountered. Although it is not possible to draw conclusions from a small series of choledochal cyst with PHT, we had followed up our cases for long periods

and reviewed other reports in the literature in order to suggest an algorithm for the management of such patients (Fig. 3).

In adults, the cause of PHT in choledochal cyst guides the management of the disease. Surgical management with adequate preoperative preparation should be considered for patients with choledochal cyst and SBC. ERCP and stenting may be considered as temporary measures in high-risk cases. In cases where there is an unrelated cause of cirrhosis with an uncomplicated cyst, management should be based on the extent of liver disease.

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