

RADIOLOGICAL MANIFESTATIONS OF CHOLEDOCHAL CYSTS: A PICTORIAL REVIEW

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Introduction

Choledochal cysts are congenital or acquired disproportionate dilatations of any portion of the bile ducts, more commonly found to occur in the extrahepatic bile ducts. The incidence of choledochal cysts is rare, estimated to lie between 1 in 100,000 or 150,000 people in Western countries. However, the incidence is reported to be higher in Asian population. They are found to occur more frequently in females with a 4:1 female to male preponderance.¹ Although most cases are usually diagnosed in infancy and childhood, up to 25% of cases are diagnosed in adulthood.² In this pictorial review, we present cases from our institute that have been diagnosed and classified according to the Todani's classification.

Pathogenesis

Many theories have been postulated about the pathogenesis of choledochal cysts. Congenital cysts are thought to have an association with fetal viral infections or occur due to unequal proliferation of embryologic biliary epithelial cells before bile duct cannulation is complete. Recently, the association between the anomalous union of the pancreaticobiliary duct (AUPBD) and the formation of choledochal cysts has drawn widespread attention. Developmentally, the CBD and pancreatic duct normally unite within the sphincter of Oddi to form a common channel which is usually 0.2-1.0 cm long. When the length of this channel is greater than 1.5 cm, pancreatic juices enter the biliary system and cause cholangitis and bile duct wall destruction, which together with distal stenosis due to scarring, result in formation of a choledochal cyst.^{3,4}

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Clinical Presentation

The triad of pain, jaundice and an abdominal mass are usually noted in most infants. Adults may present with vague abdominal pain and non-specific symptomatology.^{1,3,4} Others may present with complications such as pancreatitis, cholangitis, or biliary lithiasis. Rarely, there may be intra-peritoneal rupture, hemorrhage due to erosion of adjacent vessels, portal hypertension or secondary biliary cirrhosis. In addition, type III cysts can cause gastric outlet obstruction due to the obstruction of the duodenal lumen or intussusception.

Imaging Modalities

Ultrasound is usually the initial modality used for imaging the biliary tree because of its noninvasive and inexpensive nature. There will usually be a cystic lesion in the right upper quadrant at the portahepatis which is separate from the gall bladder. Previous literature reports a sensitivity ranging from 71-97% for ultrasound in making the diagnosis of choledochal cysts.⁵ Furthermore, ultrasound can also be used for follow-up surveillance.

Computed tomography (CT) scans are beneficial in delineating the relationship of the cyst with the biliary system and surrounding structures, evaluation of the intrahepatic ducts (such as in Type V Caroli's disease) and is also useful in ruling out any associated malignancy. Computed tomographic cholangiography has also been used to define the complete anatomy of the biliary system for appropriate surgical management. Another frequently used modality is a technetium-99 HIDA scan, which can be undertaken for demonstrating continuity of the cyst with the bile ducts.⁶

Cholangiography either via endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC) or intraoperative cholangiography remains useful for identifying an abnormal pancreaticobiliary duct junction and presence of lithiasis or malignancy. Given the invasiveness of cholangiography, magnetic resonance cholangiopancreatography (MRCP) is now considered to be the gold standard, with sensitivity for diagnosis being as high as 90% - 100%.⁷

CLASSIFICATION AND RADIOLOGIC FEATURES

Komi et al.⁸ classified AUPBD into three types according to the angle of the ductal union, whether it is at a right angle or forming an acute angle. However, the universally accepted classification system for choledochal cysts is the Todani Classification,^{9,10} (Tab. and Fig.1) based on the morphology, location, and number of intrahepatic and extrahepatic bile duct cysts.

Type I	Involvement of the extrahepatic bile ducts
Type IA	Entire extrahepatic duct involved
Type IB	Focal segment of extrahepatic duct involved
Type II	True diverticulum of extrahepatic duct
Type III	Choledochocele; cystic dilatation of distal portion of extrahepatic duct
Type IVA	Involvement of both extra and intrahepatic ducts
Type IVB	Multiple segmental dilatations of the extrahepatic ducts only
Type V	Caroli's disease; cystic dilatation of intrahepatic ducts only

Table 1: Todani's classification of choledochal cysts

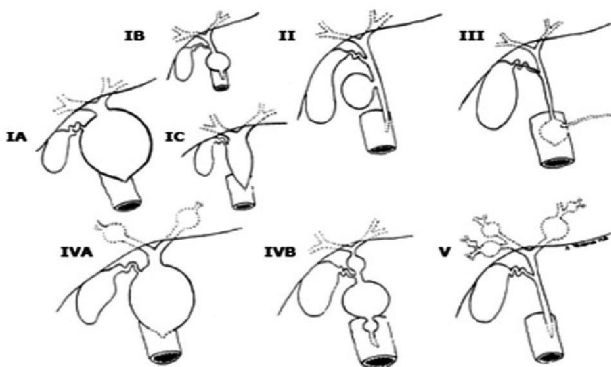


Figure 1: Schematic diagram of Todani classification of choledochal cysts^{9,10}

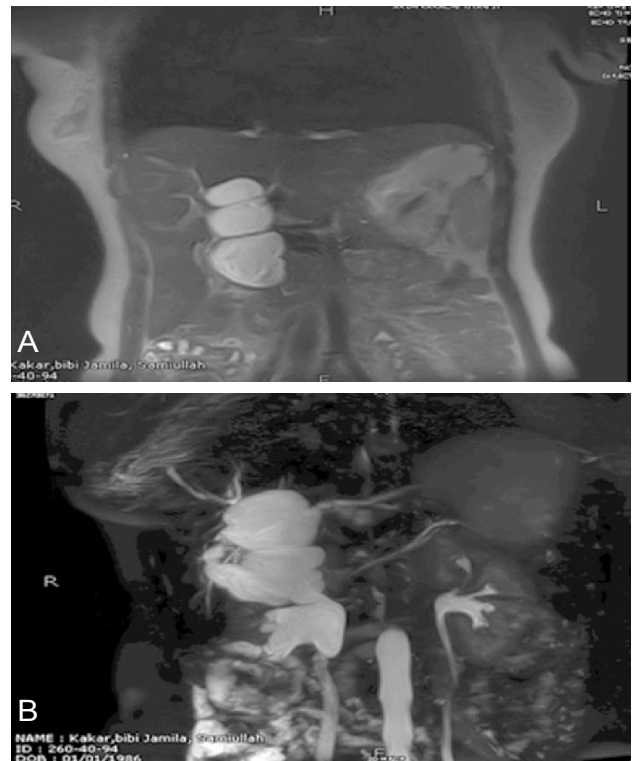


Figure 2(A and B): Choledochal cyst type IA. MRCP examination was performed in a 27 years old female with right upper quadrant pain. Fusiform and tortuous dilatation of extrahepatic biliary tree in keeping with type I choledochal cyst. No evidence of intrahepatic biliary dilatation.

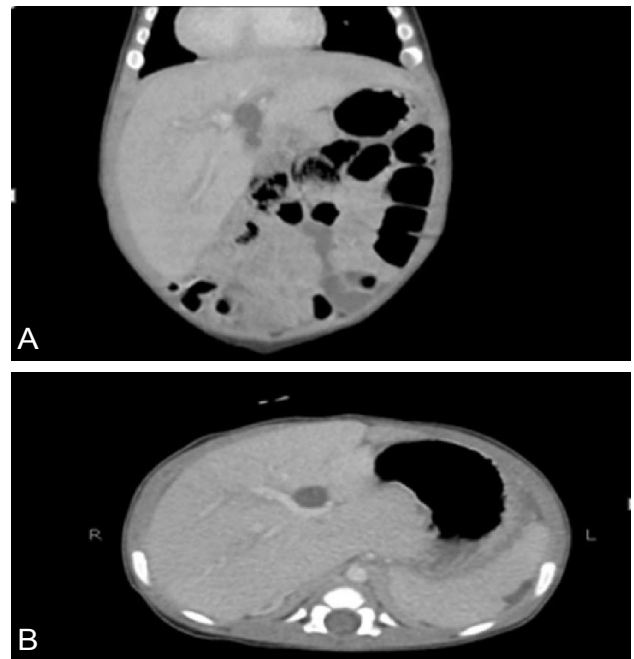


Figure 3(A and B): Choledochal cyst type IC. Axial and coronal images from enhanced CT abdomen of a 4 month old boy with jaundice. There is cystic dilatation of the common bile duct at the portahepatis without evidence of intrahepatic biliary dilatation.



Figure 4(A, B, C and D): Choledochal cyst type III. Axial and coronal images from CT KUB (A and B) and MRCP examination (C and D) of a 29 year old female who was being evaluated for recurrent UTI. A small cystic lesion is seen in the region of the head of pancreas which is inseparable from the distal portion of the CBD. No filling defect is identified within the diverticulum or the CBD. No intrahepatic biliary dilatation is seen.

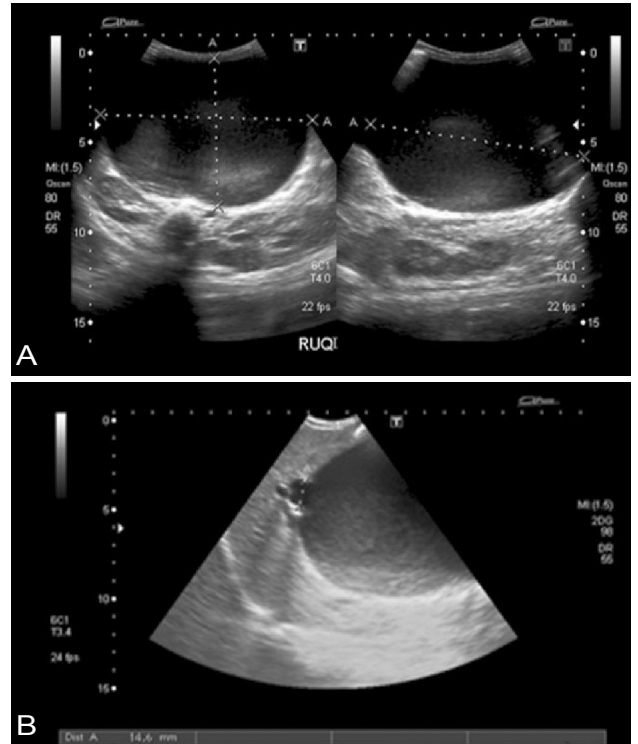


Figure 5(A and B): Choledochal cyst type IVA. A 3 month old female patient presented with an abdominal mass. Ultrasound showed a large cystic structure arising from the porta hepatis along with intrahepatic and extrahepatic biliary dilatation.

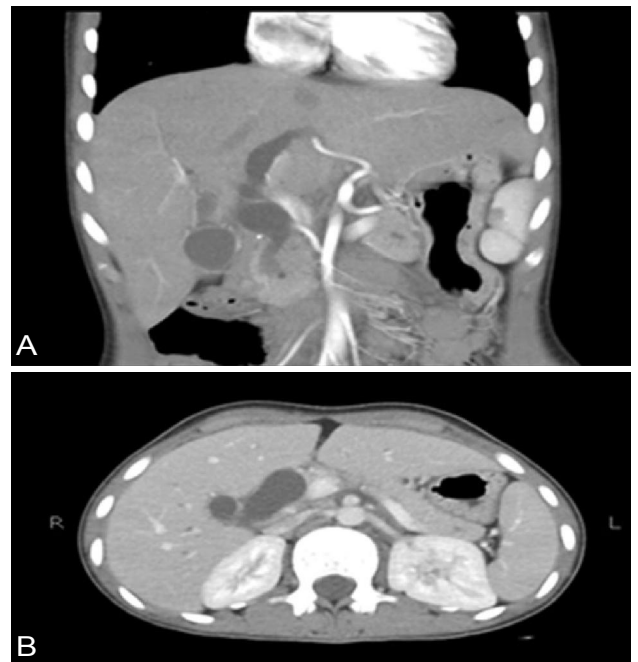


Figure 6 (A and B): Choledochal cyst type IVA. A 12 years old boy with jaundice and right upper abdominal pain underwent contrast enhanced CT examination. The coronal and axial images show a Type IV choledochal cyst with intrahepatic and extrahepatic biliary ductal dilatation. High density noted in the distal CBD, which may represent inspissated secretions vs. calculus.



Figure 7 (A and B): Type IVB choledochal cyst. 7 year old female with abdominal pain and vomiting. Coronal sections of CT abdomen shows multiple segmental dilatation of the extrahepatic ducts.



Figure 8 (A and B): Type V choledochal cysts: Caroli's disease. Cystic dilatation of the right lobe intrahepatic ducts with central dot sign. Normal extrahepatic channels.

Complications

Complications of choledochal cysts in adults include cholecystitis, recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis (Fig. 9), and malignant transformation into cholangiocarcinoma with incidence increasing with age.^{1,4} Moreover, the diagnosis of adult choledochal cysts is often delayed due to nonspecific clinical symptomatology or symptoms masked by other secondary hepatobiliary conditions. This results in an increased risk of malignant degeneration which occurs most often in Todani type I and IV cysts.¹ While most of the reported cases of cancer arising in choledochal cysts are cholangiocarcinomas within dilated extra-hepatic bile ducts, 10% are gall bladder carcinoma.¹¹ Gall bladder carcinoma in patients with AUPBD and without CBD dilatation is shown to be more common than patients with gall bladder carcinoma with choledochal cysts.¹²

Therefore, if a suspicious abnormality is detected in the wall of a choledochal cyst on imaging studies, further evaluation should be performed to exclude the presence of AUPBD.¹²

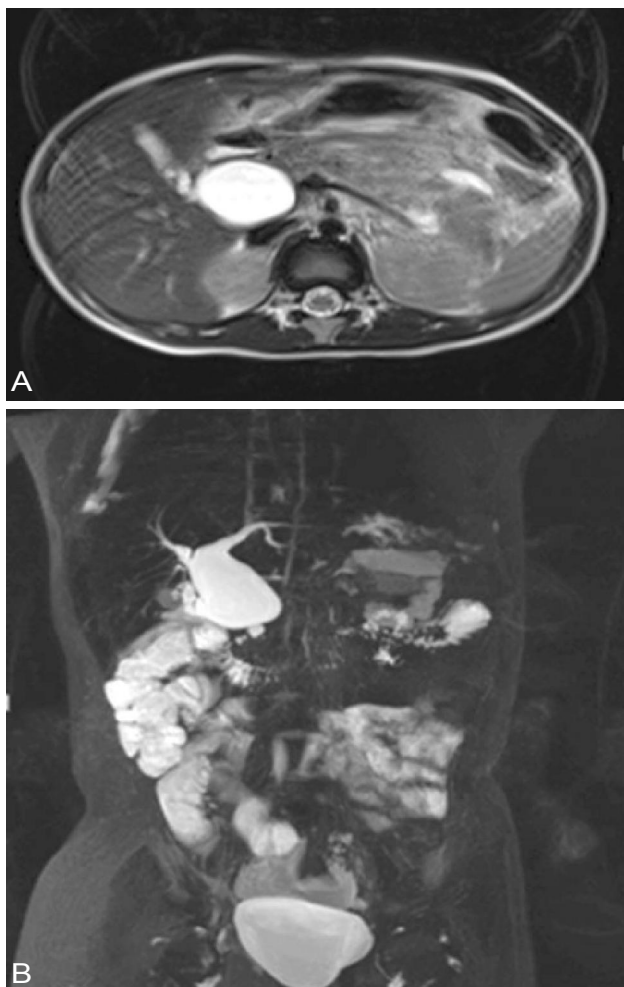


Figure 9(A and B): Axial and coronal images from MRCP examination of a 7 years old girl with recurrent pancreatitis. There is fusiform/saccular dilatation of the proximal and mid CBD with dilatation of the right and left hepatic ducts. The findings representing type IVA choledochal cyst. Swollen pancreas with peripancreatic free fluid seen representing acute pancreatitis.


Treatment

Optimal treatment option is usually surgical intervention. Excision of the extrahepatic biliary tree, including cholecystectomy, with a Roux-en-Y hepaticojejunostomy is ideal options for Type I, II and IV cysts. Type III cysts can often be managed with endoscopic sphincterotomy or endoscopic resection. Additional segmental resection of the liver may be necessary

in patients with Type IVA cysts. Moreover, patients with Type IVA cysts or Type V cysts will eventually require liver transplantation.¹³

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