



Review Article

CHOLEDOCHAL CYST AND NEW ERA IN ITS MANAGEMENT

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Introduction:

Choledochal cyst (bile duct cyst) is an important cause of surgically treated jaundice in infants and children. It is a congenital condition involving cystic dilatation of bile ducts.¹ They are uncommon in western countries,² incidence ranges from 1 in 100,000 to 150,000 live births with the incidence in the United States as high as 1 in 13,500 but not as rare in East Asian nations like Japan and China with rates as high as 1 in 1000 in Japan. There is a well-documented female dominance (3 to 4:1) that contributes to the belief that choledochal cyst (CC) is sex-linked.¹⁻³ Familial cases have been recognized, but no genetic basis has been evident. The majority (60%) of CCs are diagnosed in the first decade of life, 20% remain undiagnosed until later in childhood or adulthood, and the remaining 20% to 25% of cases are diagnosed prenatally. Prenatal diagnosis of choledochal cysts is increasing in frequency,² in the United States and Europe, perhaps due to improved imaging techniques.

Etiology

The etiology of choledochal cyst remains unknown. Various hypotheses of the etiologic basis exist since 1852 by Douglas and Yotsuyanagi which are now regarded as unlikely. Distal obstruction, weakness of the duct wall, or a combination of the two are the predominant hypotheses.⁴ In 1916 Kozumi and Kodama recognized an anomalous junction between the bile and pancreatic ducts during an autopsy case

with choledochal cyst.⁵ In 1969 Babbitt described the "long common channel" theory, also known as pancreaticobiliary maljunction (PBM)^{6,7} which is a rare congenital anomaly described as a proximal insertion of the pancreatic duct (PD) into the common bile duct (CBD). In 1984 Todani and colleagues were able to show PBM through analysis of endoscopic retrograde cholangiopancreatography (ERCP).⁸ In fetal development, PBM creates a nidus for reflux of pancreatic enzymes into the CBD that causes damage to the ductal wall and leads to cyst formation. Distal obstruction at the level of the duodenum is an additional factor leading to damage of the ductal wall and causing formation of a saccular dilation.⁸ The majority of choledochal cysts are associated with PBM; however, PBM can be seen without an associated choledochal cyst in 20% to 30% of cases.

Embryogenesis:

In normal development, a hepatic diverticulum forms from the ventral aspect of the foregut at the fourth week of gestation which progresses to cranial and caudal buds. The liver and extrahepatic biliary tree form from the cranial bud and the caudal bud divides into the superior and inferior buds. The gall bladder and cystic duct are derived from the superior bud, and the inferior bud gives rise to the right and left ventral pancreas. At the sixth week, the ventral pancreatic bud and CBD rotate around the duodenum clockwise by 180 degrees. The CBD at this point enters the duodenum at the left posterior surface. In the seventh week the main PD (Wirsung duct) and CBD junction ends in the developing duodenum as closed cavities through elongation to form the ampulla of Vater. The junction retracts in the eighth week of gestation to reside in the submucosa of the duodenal

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wall. A concentric ring of mesenchyme forms around the junction of the pancreatic and biliary ducts, beginning the formation of the sphincter of Oddi. In the twelfth week of gestation, the main PD and CBD are obliquely arranged in the duodenum. PBM is believed to arise from a misarrangement of the PD and CBD.⁹

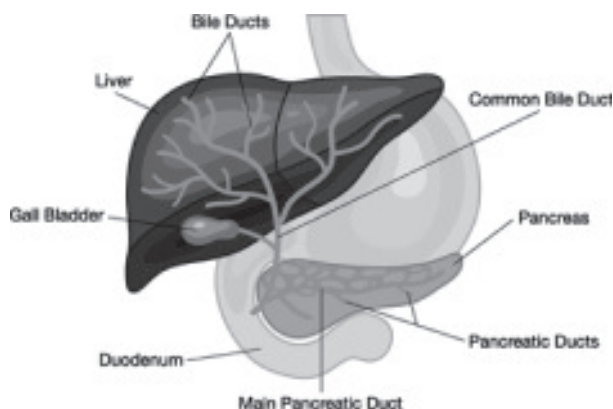


Fig.-1: Biliary system

Classification:

The original anatomic classification of choledochal cysts included types I, II, and III.¹⁰ After review of

cholangiograms, Todani and colleagues broadened the classification into five types with some subtypes (Table 1, Fig. 2).^{10,11} Type I is the most common, accounting for 70% to 90% of cases¹⁰ and is divided into three subtypes (types I A, I B, and I C).

Type	Description	Frequency
I	Choledochal cyst: Cylindrical or segmental dilatation	70% - 90%
I-A	Cystic dilatation of entire CBD	
I-B	Cystic dilatation of a segment of CBD	
I-C	Fusiform dilatation of CBD	
II	Choledochal diverticulum: supraduodenal segment	2% - 5%
III	Choledochocele: intraduodenal diverticulum	4%
IV-A	Multiple cysts at intra and extrahepatic ducts	10% - 20%
IV-B	Multiple cysts at extrahepatic ducts only	
V	Multiple cysts at intrahepatic ducts only (Caroli's Disease: segmental or diffuse)	1%

Table 1 – Todani's Classification of Choledochal Cysts

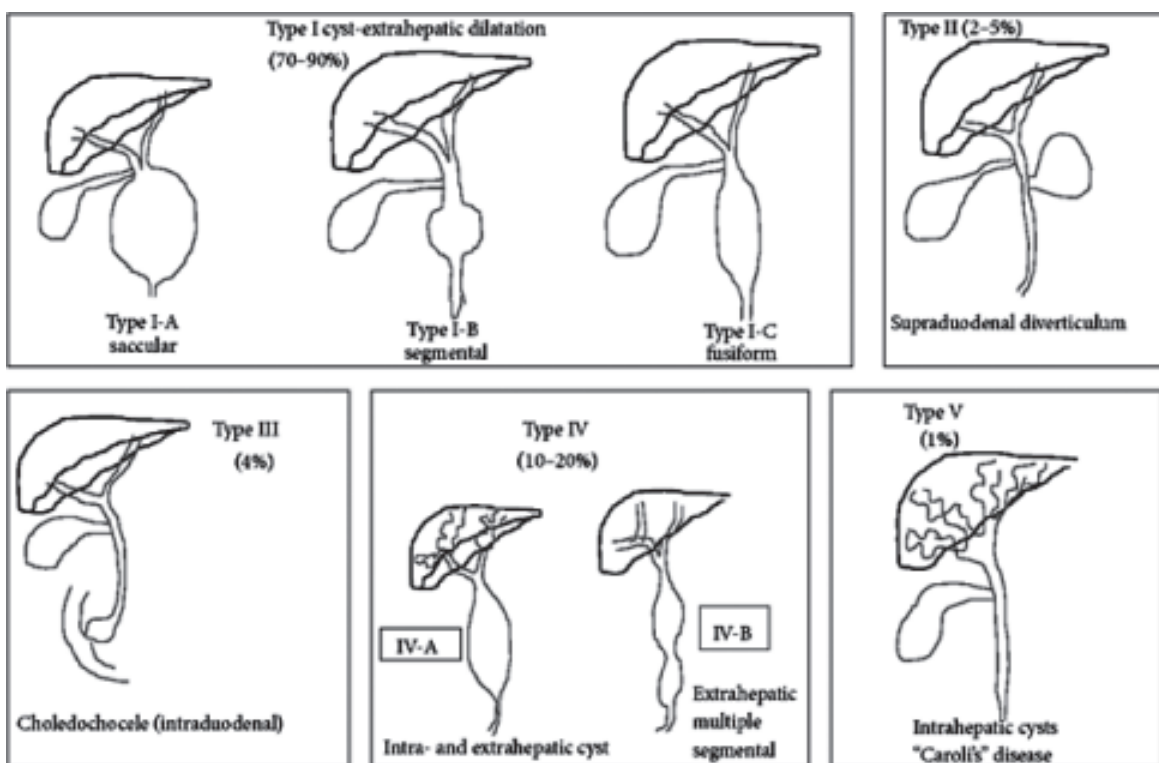


Fig.-2: Todani's classification of Choledochal Cysts

Clinical Presentation:

1. Patients with choledochal cysts can manifest clinical symptoms at any time during their life, with 80% of patients being symptomatic before the age of 10 years.¹²
2. Abdominal pain, jaundice, and a palpable right upper quadrant abdominal mass is the classic triad for patients with choledochal cyst, reported only in about 20% of cases.
3. Two of the three symptoms are seen in two thirds of patients at the time of diagnosis.¹²
4. There are two forms according to age at presentation.
5. The **infantile form** occurs before 12 months of age, and presents with obstructive jaundice, acholic stools, and hepatomegaly similar to biliary atresia.¹³
6. The **adult form** usually presents anytime after 12 months usually after 2 years of age and has a greater number of symptoms including fever, nausea, vomiting, and jaundice.¹⁴
7. Signs of hepatic fibrosis may be present in the infantile form, and these patients benefit the most from early treatment.
8. Perforation of choledochal cyst is rare (1% to 12%) and thought to be due to a fragile cystic wall usually at the junction of the cystic and common bile ducts from inflammation, increased ductal pressure, or increased intraabdominal pressure. These patients present with abdominal pain, sepsis, and peritonitis.¹⁵
9. Patients with adult form tend to have a fusiform dilation of CBD without complete obstruction of the distal CBD and are more likely to present with classic triad.
10. Symptoms arise from biliary obstruction from a mucous plug or biliary sludge leading to ascending cholangitis and complications of pancreatitis.
11. Bile stasis, sludge, stone formation, inflammation, and recurrent super infection from persistent bacterial colonization have all been identified as complications. Secondary biliary cirrhosis is noted in 40% to 50% of patients.¹²

12. In cases of CC not diagnosed until adulthood, patients may present with cholelithiasis and symptoms mimicking biliary colic or cholecystitis.

Diagnosis:**Laboratory Studies**

Laboratory data are usually reserved primarily for evaluating the clinical condition not diagnosis of the patients with choledochal cysts.

Serum markers:

1. S. bilirubin – total, direct, indirect. Conjugated hyperbilirubinemia is typical in the infantile form but may or may not be present in the adult form due to intermittent or incomplete biliary obstruction.
2. S. alkaline phosphatase
3. BT, PT, APTT - in chronic cases an abnormal coagulation profile may be evident due to hepatic injury.

Imaging Studies

Diagnosis of choledochal cysts is made by imaging studies.

1. Ultrasound of abdomen is the first imaging modality used. It is an excellent screening tool and is used by some as the only imaging study in infants. It carries a 71% to 97% sensitivity.¹⁶ The best imaging techniques for Caroli disease (type V cysts) are ultrasound, CT, and MRI.¹²
2. Magnetic resonance cholangiopancreatography (MRCP) is now considered the gold standard for imaging choledochal cyst, especially given the complications and concerns with invasive cholangiography (Fig.3a). The diagnostic sensitivity of MRCP is 90% to 100%.¹⁷ It also avoids radiation associated with CT scan. Administration of secretin increases pancreatic secretion and dilates the pancreatic duct. Thus some centers administer secretin before MRCP to increase diagnostic yield.¹⁸ The type and extent of the choledochal cyst are well visualized, and images can be reconstructed in three dimensions (Fig.3b).



Fig.-3a

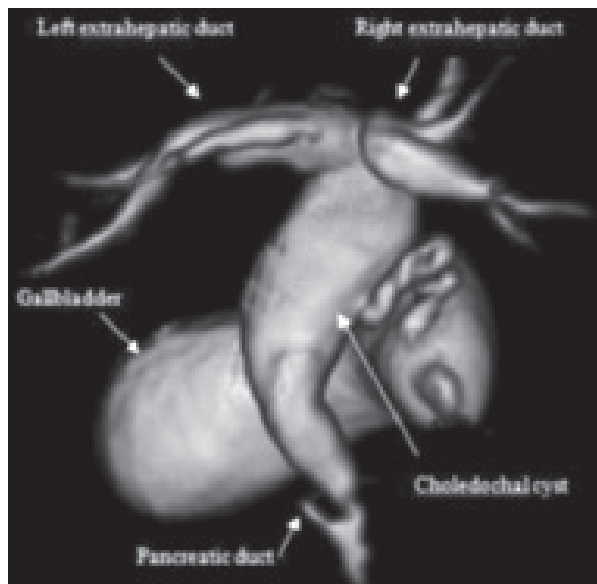


Fig.-3b : Three-dimensional reconstruction of a MRCP image of a choledochal cyst with mild dilation (6 mm) of the right and left extrahepatic ducts. Pancreatic duct is seen entering the normal caliber distal common bile duct.

3. A technetium-99 HIDA scan may provide more definitive data. It has varying sensitivities, with 100% for type I cysts and 67% for type IV.¹⁶ A HIDA scan may also be helpful for distinguishing between choledochal cyst and biliary atresia. Biliary atresia on a HIDA scan is characterized by lack of contrast emptying into the duodenum, whereas a choledochal cyst will have contrast entering the duodenum (Fig.4). A HIDA scan may also be useful in the diagnosis of cystic rupture, in which case contrast would empty into the peritoneal cavity.¹² On HIDA scan a beaded appearance due to intraductal bridging may be indicative of Caroli disease.
4. An abdominal computed tomography (CT) scan may be another useful modality. A CT can show

the intrahepatic ducts, distal CBD, and the pancreatic duct, features that are not reliably identified on ultrasound, making CT scan highly useful in identifying type IV and type V cysts.

5. Endoscopic retrograde cholangiopancreatography (ERCP) plays a role in diagnosis and evaluation of the biliary anatomy. Choledochoceles (type III cysts) can be diagnosed with various imaging modalities such as upper gastrointestinal series (UGIS), endoscopy, ERCP, MRCP, and CT cholangiography. The advantage of ERCP over MRCP and CT cholangiography for choledochoceles is the ability to perform a therapeutic procedure such as a sphincterotomy.

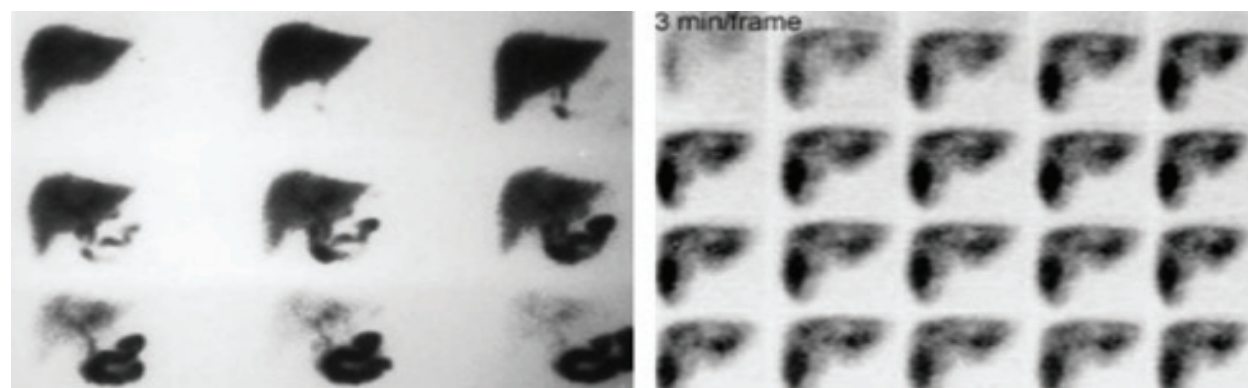


Fig.-4: HIDA scan

Complications:

1. Pancreatitis
2. Cholangitis
3. Biliary cirrhosis
4. Malignancy e.g. cholangiocarcinoma and gall bladder cancer

Timing of Surgery:

The timing of surgery should be early after diagnosis to reduce the incidence of complications described,¹⁹ and particularly to prevent liver damage in neonates.¹³

Management:

Primary cyst excision combined with biliary reconstruction is the standard treatment of choledochal cysts. In earlier decades, an operation known as cystenterostomy was performed that only drained the cyst and the biliary reconstruction left the cyst behind. That surgery proved ineffective, leaving many patients with recurrent cholangitis, bowel obstruction, and chronic inflammation in the remaining cyst leading to the substantial risk of portal hypertension and malignant transformation into cholangiocarcinoma and gall bladder cancer.¹⁹ Biliary reconstruction may be achieved by one of several techniques based on the surgeon's personal preference and the current circumstances of each

case. Today, complete excision of the extrahepatic cysts in type I and IV choledochal cysts followed by Roux-en-Y hepatico-jejunostomy (RYHJ) (Fig. 5a, 5b) is the most commonly used techniques for biliary reconstruction. It involves two anastomoses, namely jejunojejunostomy and hepatico-porto-jejunostomy with a long segment (40 cm) of defunctioning jejunum as the biliary conduit. Open as well as laparoscopic procedure is performed. Several authors reported that complications related to RYHJ are not uncommon. Stone formation in elongated pouch, intrahepatic bile duct stone formation, bowel obstruction caused by adhesions between the elongated blind pouch and small bowel, duodenal obstruction caused by compression by a high Roux-en-Y limb vascular arch and bile stasis caused by an abnormally elongated Roux-en-Y jejunal limb causing stone formation in the Roux-en-Y jejunal limb.^{20, 21, 22}

Some surgeons prefers hepaticoduodenostomy (HD) (Fig. 6a, 6b) because it is simple to perform, had fewer complications such as anastomotic leakage, postoperative adhesive bowel obstruction, and peptic ulcer.²³ Delivery of bile into the duodenum, rather than into a Roux-en Y limb of the jejunum is considered to be more physiological and is another advantage of HD.²³ One of the noticed advantages of HD in the different studies was the statistically significant shorter operative time compared to the RYHJ group.²³

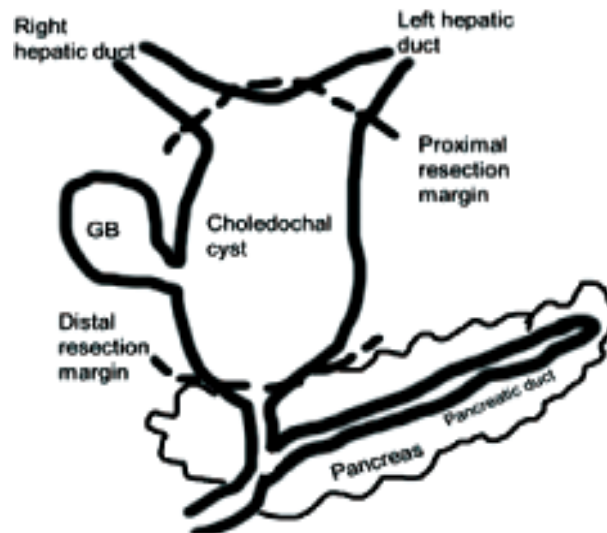


Fig.-5a: Choledochal Cyst



Fig.-5b: Roux-en-Y hepaticojejunostomy



Fig.-6a: Choledochal Cyst



Fig.- 6b: Hepaticoduodenostomy

Another new surgical technique of biliary reconstruction using native gall bladder as biliary conduit is preferred by some surgeons.²⁴ Here, choledochal cyst is excised as standard technique but gall bladder with its neck is kept in situ (Fig. 7a, 7b). The gall bladder neck is anastomosed with the common hepatic duct stump and gall bladder fundus is anastomosed with the antero-inferior wall of the first part of distal duodenum (Fig.7c) (Choledocho-Cholecysto-Duodenostomy).²⁴ These two operative approaches eliminate the potentially premalignant epithelial cyst lining and also separates the pancreatic drainage from the biliary drainage. The new technique also maintains a normal anatomy more than the Roux-en-Y Hepatico-jejunostomy. Gall bladder has

been used in place of an intestinal conduit and hence more physiological. Dissection is done only in sub-hepatic region thus reducing the area of adhesion. Small incision, less reflux due to small cholecysto-duodenostomy (< 10mm), less operative time, less possibility of gall stone formation as gall bladder works as a conduit with continuous flow of bile rather than a normal reservoir, less intestinal complications than RYHJ, the newer technique may be a good alternative of the standard procedure.²⁴ Some other procedures such as jejunal interposition hepaticoduodenostomy (ChicagoBeijing procedure) and appendix interposition have also been reported as operative procedures.²³ But many authors have reported that jejunal interposition is a complex

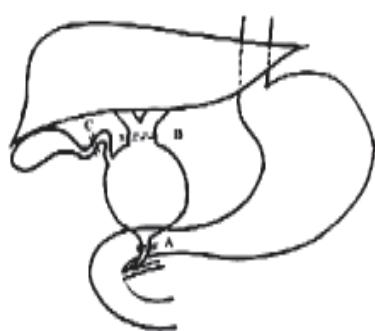


Fig.-7a. Choledochal cyst and lines of excision. A, division of lowest end of terminal common bile duct or choledochal cyst. B, Division at the site of common hepatic duct just above the choledochal cyst. C, Division of gall bladder neck.



Fig.-7b. After the excision of the choledochal cyst

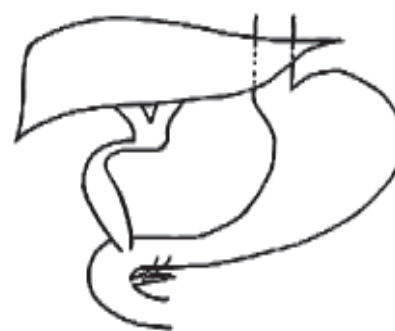


Fig.-7c. Anastomosis of gall bladder neck with common hepatic duct and anastomosis of fundus of the gall bladder with antero-inferior wall of distal first part of duodenum.

procedure that may carry the possibility of stricture at the anastomosis beside it often leads to bile reflux into the stomach.²³

Type II choledochal cysts are rare but appear to have a low malignant potential. This type requires simple cyst resection, complicated dissection and reconstruction are not required.

Type III choledochal cysts, or choledochoceles, are intraduodenal or intrapancreatic dilations of the distal common bile duct. Management has traditionally been operative marsupialization of the cyst, usually through a transduodenal approach.²⁵ Choledochoceles can be treated by sphincterotomy or cyst marsupialization during an ERCP.²⁶⁻²⁸

For patients with type IV disease, most surgeons recommend cyst resection to the hepatic ducts, leaving in place the dilated intrahepatic ducts because they may decrease in size without distal obstruction.

Surgical treatment of type V (Caroli disease) is challenging. Segmental resections can be performed if the disease is localized to a portion of the liver. Liver transplantation has also been performed.^{29,30}

Postoperative Care:

Advancement of surgical treatment results minimal morbidity and mortality and reduces the number of late complications. The most common late complication continues to be anastomotic stricture. Early diagnosis and cyst excision results in low complication rates in most experienced centers. The technique of Roux-en-Y hepaticojejunostomy is favored by most, although comparable results can be achieved by hepaticoduodenostomy and Choledochcho-Cholecysto-Duodenostomy.

Follow-up:

Studies suggest that long-term follow-up is indicated due to the potential for problems such as anastomotic stricture, cholangitis, intrahepatic stone formation, and malignancy.³¹ This is particularly important for patients with type IV disease because malignancy can occur in incompletely resected cystic hepatic ducts or recurrent cysts.³¹ The incidence of malignancy of the gallbladder and bile ducts remains high in patients with PBM compared with the general population and occurs at a younger age.

Conclusion:

The management of choledochal cysts is performed predominantly by pediatric surgeons. Because of the high risk of serious sequelae including malignancy, early surgical excision is warranted even in

asymptomatic patients. Total excision of choledochal cyst is usually feasible. Both RYHJ and HD are effective techniques for biliary reconstruction with satisfactory and comparable results on both early and long-term follow up. Hepatico-duodenostomy may be preferred due to shorter operative time and avoidance of intestinal anastomosis; however more patients with HD are required before reaching a final conclusion. The other surgical technique (Choledochcho-Cholecysto-Duodenostomy) is more anatomical and physiological than standard RYHJ.²⁴ There is no significant disadvantage or complication observed in this short period study.²⁴ A long term follow-up of patients is needed to look for future probable complications of stricture, recurrent cholangitis or any other untoward complications. Even if such complications occur there is scope for a redo-surgery or a Roux-en-Y Hepaticojejunostomy.²⁴ Because of the rare nature of this disease, extensive knowledge of the different variants and experience with advanced biliary tract surgery is critical in attaining good outcomes.

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