Choledochal Cyst

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82.1 Introduction

Congenital bile duct dilatation is a better term for the spectrum of anomalies known traditionally as choledochal cysts. Choledochal cysts may cause symptoms at any age but typically present with obstructive jaundice and/or abdominal pain in infants and children. Although rare, they are more common in females (female-to-male ratio of about 3–4:1) and in some Asian races.

82.2 Anatomic Classification

Type I, II, and III forms of choledochal cysts were originally described by Alonso-Lej and colleagues. Subsequently, Todani and associates and others have further classified this anomaly into five main types and additional subtypes, based on analyses of cholangiograms. The two relatively common categories of cyst are types I and IV-A. The common varieties are as follows (see Fig. 82.1):

- *Type I* consists of dilatation of the common bile duct, which may be cystic, focal, or fusiform (subtypes A, B, and C, respectively) (90–95% of cases).
- *Type II* is diverticulum of the extrahepatic bile duct.
- *Type III* involves choledochoceles.
- *Types IV:* type IV-A, the second most common type, is defined as both intrahepatic and extrahepatic dilatation of the biliary tree. The rare malformation of multiple extrahepatic cysts is designated as type IV-B.
- *Type V* comprises single or multiple intrahepatic cysts. This type has been referred to as Caroli's disease when associated with hepatic fibrosis.

Pancreaticobiliary malunion may occur without choledochal dilatation, and this has been termed a forme fruste choledochal cyst.

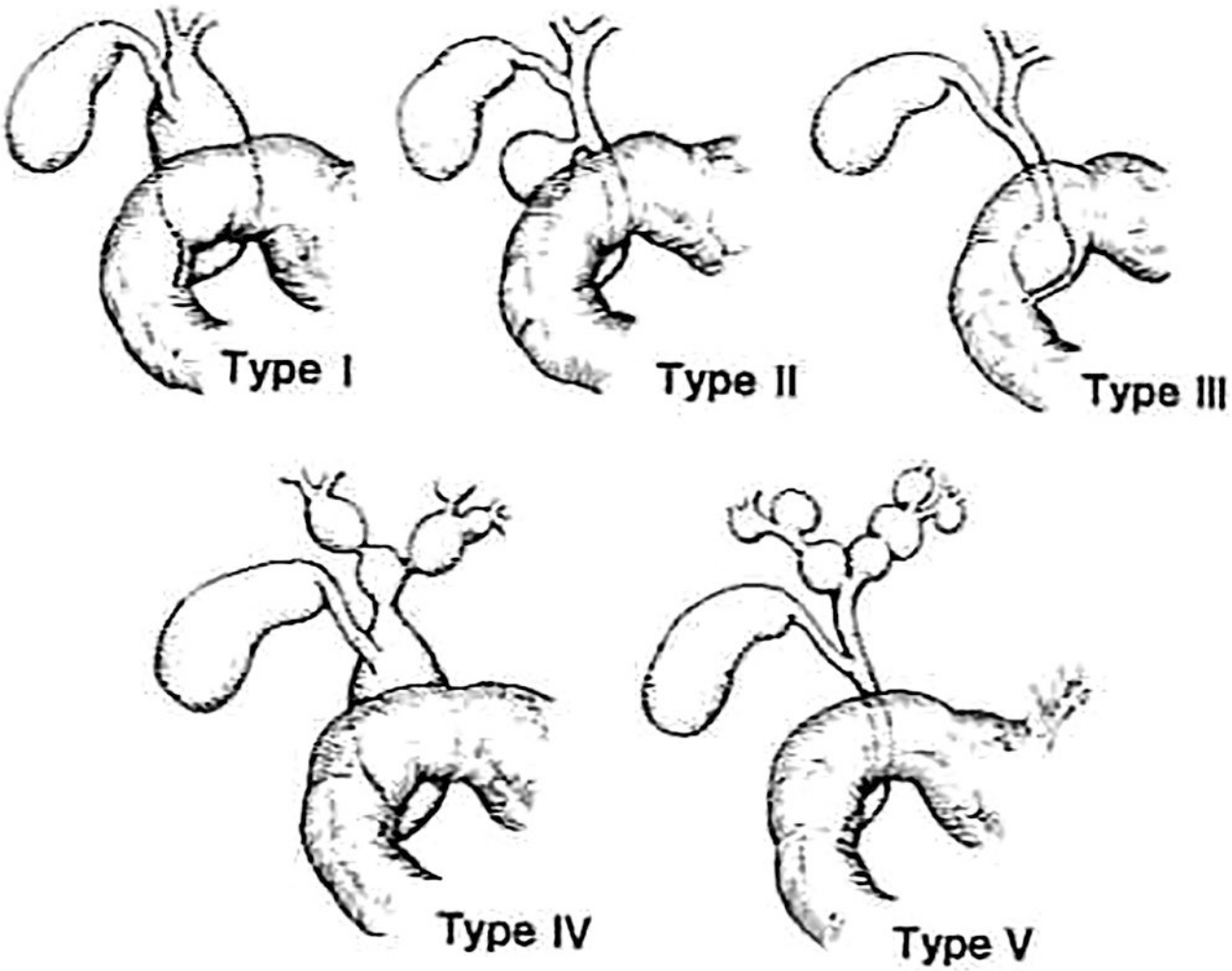

Fig. 82.1 Classification of choledochal cysts

82.3 Pathology

Histologic sections of the wall of extrahepatic choledochal cysts have demonstrated a thick-walled structure of dense connective tissue interlaced with strands of smooth muscle. In most instances, some degree of inflammatory reaction is noted; it is minimal in infants and gradually becomes more marked as patients get older. The histologic appearance of other forms of choledochal cysts is similar, with the exception of choledochocele. In these cysts, the lining is most commonly duodenal mucosa and only occasionally resembles the lining of the bile duct.

The findings on liver biopsy also vary with the age of the patient. In a newborn, the histologic appearance of the liver is usually interpreted as normal or having mild bile duct proliferation consistent with chronic biliary obstruction. Occasionally, in older patients, mild periportal fibrosis is noted. Although the bile duct is usually normal in appearance on histologic section, inflammation may be present, and stones and sludge may be seen in the common bile duct and occasionally in the intrahepatic ducts of older patients. Most patients with choledochal cysts have an anomalous pancreaticobiliary junction.

Carcinomas arising in the wall of choledochal cysts are well recognized and believed to be the result of chronic inflammation. Biliary carcinoma has been noted in patients with an anomalous pancreaticobiliary junction even without a choledochal cyst. Carcinoma in the wall of a choledochal cyst has rarely been reported in a child, being primarily a problem of adults. Although the majority of these malignancies occur in the wall of choledochal cysts, other sites have included the gallbladder and the head of the pancreas in the region of the pancreaticobiliary junction. Because of the long interval over which these lesions seem to develop, it is presumed that they are the result of chronic inflammation from cholangitis. Inflammation is not a prominent feature in patients who have choledochoceles, although mild inflammation may lead to stenosis of the common bile duct and the pancreatic duct.

82.3.1 Etiology

Choledochal cysts are congenital. Two main etiologic theories have been proposed: (1) weakness of the wall of the bile duct due to pancreaticobiliary malunion (PBM) and (2) obstruction of the distal part of the bile duct.

In more than 75% of patients with a choledochal cyst (particularly type I and IV cysts), there is an anomalous junction between the distal common bile duct and the pancreatic duct; the ducts unite outside the duodenal wall some distance proximal to the ampulla of Vater.

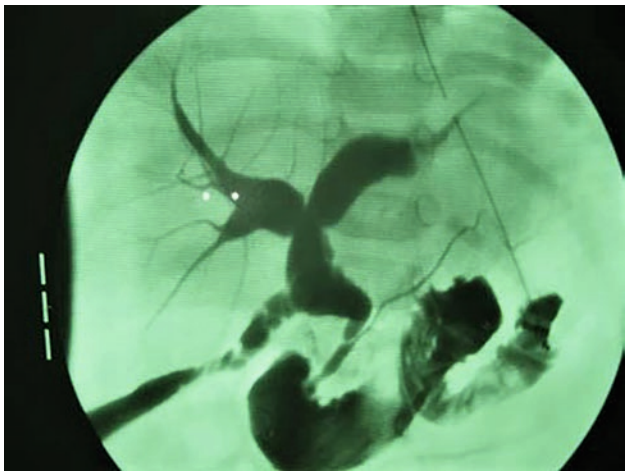

Fig. 82.2 Intraoperative cholangiography showing a long common channel

This common channel often exceeds 5–10 mm in length (Fig. 82.2), and it is not surrounded by the normal sphincter mechanism. Consequently, pancreatic juice refluxes into the biliary tree. Isolated PBM without choledochal dilatation has been implicated in the pathogenesis of gallbladder cancer in adults. A common channel also predisposes to reflux of bile into the pancreatic duct, which may precipitate pancreatitis.

PBM is not found in all patients with congenital choledochal dilatation, and it can occur with a normal caliber bile duct. In addition, choledochal cysts have been detected as early as at 15-week gestation, a time when acinar development of the pancreas is rudimentary, which argues against a significant role for pancreaticobiliary reflux in such cases. An alternative and more plausible explanation is obstruction of the distal common bile duct. A distal obstruction could be functional rather than mechanical and can occur as a result of PBM and an abnormal sphincter of Oddi. Kusunoki et al. have shown that there are abnormally few ganglion cells in the narrow portion of the common bile duct in patients with a choledochal cyst, as compared with controls. Presumably, this would result in functional obstruction and proximal dilatation in the same manner as achalasia of the esophagus or Hirschsprung's disease.

82.4 Presentation

Patients with choledochal cysts usually present in one of two ways, which has led to them being classified as infantile or adult in nature. In the infantile form, patients ranging from 1 to 3 months of age present with obstructive jaundice, acholic stools, and hepatomegaly with a clinical picture indistinguishable from

that of biliary atresia. Occasionally, signs of hepatic fibrosis are present, even in young patients, which is a strong argument for early treatment. Patients with the infantile form of this anomaly tend not to have abdominal pain or a palpable mass. Infants who have been diagnosed with a choledochal cyst prenatally do not ordinarily become jaundiced until 1–3 weeks after birth.

In the so-called adult form of choledochal cyst, clinical manifestations do not generally become evident until after the patient is 2 years of age; and most of these patients have fusiform deformities of the common duct without high-grade or complete obstruction. In this group of patients, the classic triad of abdominal pain, a palpable abdominal mass, and jaundice originally described by Alonso-Lej and colleagues may be noted. At least two features of the triad are found in 85% of children at presentation. Among the more commonly reported presenting features are cholangitis, pancreatitis, and biliary peritonitis from cyst rupture. However, it is important to emphasize that symptoms in older patients tend to be very subtle and intermittent, so the diagnosis frequently goes unrecognized. Because liver damage is progressive, these patients may present initially with cirrhosis and manifestations of portal hypertension.

Rare presentations include gastric outlet obstruction, neonatal bleeding tendency, duodenal intussusception, and portal hypertension.

A choledochal cyst should always be considered in the differential diagnosis of obstructive jaundice or pancreatitis. Differential diagnosis includes duodenal atresia, an ovarian cyst, a duplication cyst, and cystic biliary atresia. Progressive enlargement of the cyst during gestation and the presence of dilated intrahepatic ducts on postnatal scan are indicative of a choledochal cyst rather than biliary atresia, but it may be difficult to distinguish between these two conditions.

82.5 Investigations

82.5.1 Ultrasound Scanning

Ultrasound scanning (US) can diagnose choledochal cysts with a specificity of 97% in children. Ultrasound is therefore an excellent first-line investigation of neonatal jaundice persisting more than 2 weeks after birth and may help to differentiate choledochal cysts from biliary atresia. Antenatal diagnoses can be made on ultrasound, although diagnostic accuracy from this technique has been reported to be as low as 15%; also, it is not possible to differentiate between biliary atresia and choledochal cysts with antenatal ultrasound.

82.5.2 Radionuclide Scintigraphy

Scintigraphy is safe and atraumatic and has been used for a long time in the diagnosis of choledochal cysts. Scintigraphy follows the progression of an isotope from the biliary tract into the small intestine and is reported to distinguish with 100% accuracy between choledochal cysts and biliary atresia. Type I cysts may be diagnosed with a sensitivity of 100%, but only two-thirds of type IV disease is detected, and the extent of the intrahepatic disease may be underestimated on scintigraphy.

82.5.3 Computed Tomographic Scan

Although there are reports of computed tomographic (CT) scans diagnosing choledochal cysts, others have found that cysts are missed on CT scans (especially in small-size cysts or choledochocele) but picked up on magnetic resonance cholangiopancreatography (MRCP). A comparative study of 14 patients with choledochal cysts was performed, in which each patient had both CT cholangiography and MRCP performed. The MRCP investigation was superior at detecting and defining lesions. A better role for CT scanning may be in the postoperative period, where it has been shown to be superior to MRCP in locating the biliary-enteric anastomosis and in defining any stenosis thereof.

82.5.4 Magnetic Resonance Cholangiopancreatography

For the reasons outlined above, MRCP represents the current gold standard in the imaging of choledochal cysts. However, there are a few caveats to this. Although the technique is excellent for diagnosing and characterizing the cysts themselves, it is not so good at detecting anomalous pancreaticobiliary union, but this is probably not that important in determining patient management. Also, MRCP may not be as sensitive a tool in pediatric cases as it is in adults, where ultrasound has a preeminent role.

82.5.5 Endoscopic Retrograde Cholangiopancreatography

Investigation with endoscopic retrograde cholangiopancreatography (ERCP) is excellent for defining biliary anatomy and as such has been used to diagnose choledochal cysts. MRCP has been shown to be just as good if not better than ERCP, however, without the potential complications of the ERCP invasive technique.

82.6 Preoperative Preparation

Preoperative correction of any bleeding tendency with either parenteral vitamin K administration or even fresh frozen plasma is a must. Intravenous (IV) antibiotics are given to treat any attack of cholangitis.

82.7 Surgical Management

Historically, a cyst-enterostomy internal drainage procedure, either cyst-duodenostomy or cyst-jejunostomy, was considered the surgical management option for choledochal cysts. These approaches for the most part have been abandoned in high-income countries due to complications, including malignancy in the remaining cyst, pancreatitis, and cholangitis. In many low-middle-income countries, patients may present at a significant delay, with malnutrition and some degree of liver dysfunction. Even with the best medical optimization, a cyst-jejunostomy may be the most feasible option either as a palliative procedure or to stage definitive surgical treatment. Unfortunately, many patients present with cirrhosis and may not be operative candidates for what would otherwise be a readily treatable disease. Liver transplant is also generally unavailable in such settings.

The ideal procedure is cyst excision followed by Roux-en-Y hepaticojejunostomy or choledochojejunostomy, with the former thought to reduce the incidence of stricture formation postoperatively. Alternatively, hepaticoduodenostomy has also been recommended so that the anastomosis is accessible to ERCP in the event of postoperative complications. Hepaticoduodenostomy has not been widely adopted due to the potential for biliary reflux and cholangitis. The technique of appendix or free jejunal interposition hepaticoenterostomy similarly failed to gain widespread acclaim, as these grafts underwent stenosis with resultant hepatic fibrosis.

After excision of the cyst, the intrahepatic ducts should be probed and lavaged with saline to rid the ductal system of sludge and possible stones. Additionally, on occasion, obstruction may be found in the proximal biliary system, which can be dilatated. Therefore, intraoperative cholangiography before cyst excision is a must.

It is preferable to excise the choledochal cyst in its entirety. However, for many patients who have had recurrent bouts of cholangitis and pericystic inflammation, the cyst may be fibrosed to adjacent vascular structures. If the cyst is adherent to the portal vein or hepatic artery and removal would risk damaging these structures, then the cyst can be opened and the adherent part of the cyst left behind. The mucosa of the remaining cyst wall should be cauterized to prevent future carcinoma development. Alternatively, a mucosectomy may be performed.

Patients with choledochoceles are not usually diagnosed until they are at least 5 years of age because the characteristic clinical symptom is abdominal pain of an intermittent nature that is not specific. A longitudinal duodenotomy permits complete exposure of an intraduodenal choledochocele. Once the choledochocele is exposed, it should be unroofed, and then the mucosa is reapproximated with multiple interrupted absorbable sutures. It is important to identify and calibrate the entry points of the common bile duct and the pancreatic duct to determine whether a sphincteroplasty of these ducts will be needed.

Patients with intrahepatic cysts or Caroli's disease are difficult to manage because they tend to develop severe recurrent bouts of cholangitis, subsequent biliary cirrhosis, and progressive segmental ductal ectasia. For this reason, these patients require frequent follow-up with US over a period of many years. A variety of techniques may be needed for management of intrahepatic cysts. Partial hepatic lobectomy may be done when the disease is localized and amenable to resection, but unroofing with drainage into a Roux limb of jejunum may be needed when proximal ductal obstruction is encountered. Otherwise, multiple cysts and Caroli's disease that are not amenable to localized resection or drainage may be indications for liver transplantation.

82.8 Timing of Surgery

The timing of surgery should be early after diagnosis, even in asymptomatic prenatally diagnosed neonates (within the first 1–3 months of age), to reduce the incidence of complications and particularly to prevent liver fibrosis in neonates.

82.9 Postoperative Complications

82.9.1 Intrahepatic Cholelithiasis

Intrahepatic stones are a particular problem in cases of type IV disease with residual intrahepatic cysts. Choledochoscopy is used at the time of surgery to detect and remove intrahepatic stones at operation. Intraoperative cholangiography with ductal probing and washout of debris reduces postoperative complications of stones retained in the biliary radicals.

82.9.2 Malignant Change After Cyst Excision

Even after cyst excision, there are reports of malignancy, usually from incomplete cyst excision. However, in a comprehensive review, the incidence of post-excision

malignancy has been estimated at only 0.7%. Since malignancy may occur in the residual intrapancreatic portion of the choledochal cyst, close long-term follow-up for those patients with intrapancreatic extension is recommended, with resection if needed.

The extent of the resection in type IV-A cysts is controversial. Several authors advocate management by excision of the extrahepatic component only, with hepaticoenterostomy. However, malignancy has been reported to arise in the intrahepatic cysts, as described above, and it has also been reported to occur after resection of the extrahepatic cyst with hepaticojejunostomy. Clearly, when the intrahepatic cysts are widespread, they cannot be excised; however, when the intrahepatic disease is localized, it is reasonable to perform a partial hepatectomy. Similarly, partial hepatectomy has been practiced for Caroli's disease.

82.9.3 Cholangitis

Patients who have had cyst excision with internal drainage have a lower incidence of postoperative cholangitis than patients with biliary atresia who have had a similar type of drainage procedure. Hepaticoduodenostomy after cyst excision has long been claimed to be associated with a higher incidence of cholangitis and biliary reflux; however, our group (Ain Shams University in Cairo) has been using this type of biliary reconstruction after cyst excision for more than 50 cases with a follow-up period now approaching 6 years, and preliminary data refute the claim of higher cholangitis and biliary reflux rates with hepaticoduodenostomy.

82.9.4 Stricture Formation

Performing a higher anastomosis at the level of the confluence, as proposed by Todani and coworkers, may reduce the incidence of anastomotic stricture formation. Other investigators disagree and believe that conventional drainage at the level of the hepatic hilum is sufficient. We agree that an anastomosis to the common hepatic duct is sufficient, provided that a stricture of the right or left hepatic duct is not left in place.

82.9.5 Pancreatitis

Rarely, patients may develop pancreatitis after cyst excision and internal drainage. Acute pancreatitis due to protein plug formation is observed in more than 20% of patients who were followed up in the long term. The morphology of the pancreatic duct and ductal dilatation, possibly caused by long-standing stagnation of pancreatic juice, may be associated with postoperative pancreatitis in choledochal cyst patients. Overall, however, pancreatitis is an uncommon event after excision and internal drainage of a choledochal cyst.

82.9.6 Laparoscopic Excision

Recently, laparoscopic cyst excision and hepaticojejunostomy have been described. It is too early to assess the long-term results of this approach in terms of anastomotic strictures and malignancy arising in residual cyst tissue. The principle of laparoscopic surgery for choledochal cysts is similar to that of open surgery, although it is much more technically demanding, especially in small children in whom the peritoneal space is very limited. The magnification of the laparoscope allows excellent visualization of the anatomy and, in turn, facilitates meticulous mobilization of the cyst from surrounding structures. Fashioning of the jejunal Roux loop can be performed through the enlarged umbilical wound. This enables meticulous bowel anastomosis, just like open surgery, and avoids intra-abdominal contamination.

82.10 Postoperative Follow-Up

Our routine is to keep the patients on low doses of ampicillin or trimethoprim-sulfamethoxazole for approximately 6 weeks postoperatively to protect against cholangitis, after which the potential for this complication appears to diminish. Postoperative follow-up should be every 3 months for the first year and annually thereafter in asymptomatic patients. At each visit, liver function studies and serum amylase levels should be determined. Ultrasound of the liver and pancreas is done annually or when necessary if patients become symptomatic. US of the liver is particularly important in patients who are found to have intrahepatic ductal dilatation preoperatively. US is also helpful for long-term evaluation of the Roux-en-Y ductal anastomosis because occasional patients will develop late anastomotic strictures or stones. Of note, patients who had hepaticoduodenostomy were followed up with a hepatobiliary iminodiacetic acid (HIDA) scan and barium meal and follow-through in the Trendelenburg position to assess biliary alkaline reflux; findings were nonsignificant.

82.11 Evidence-Based Research

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In this study, 68 fetuses with ultrasound-detected choledochal cysts were randomized into two groups: early operation group (<1 month) and late operation group (>1 month). During the first month of life, 32 infants became symptomatic (jaundiced) and were excluded from the trial. Of the 36 infants who were asymptomatic, 16 had an early operation and 20 had a late operation. Hepatic fibrosis was more commonly seen in those infants operated in the late group. There was no mortality, bile leak, or anastomotic stricture seen in either group. Liver function normalization was delayed in the late operative group. Therefore, the study concluded that surgical intervention is warranted in the neonatal period for asymptomatic choledochal cysts.

Key Summary Points

1. Choledochal cysts are uncommon, but when encountered, they may appear nonspecifically rather than classically. A high index of suspicion will avoid a delay in diagnosis.
2. The imaging modality of choice for diagnosing and characterizing choledochal cysts is magnetic resonance cholangiopancreatography (MRCP).
3. Delayed diagnosis may have a variety of undesirable sequelae, including biliary cirrhosis, cholangiocarcinoma, pancreatitis, and cholangitis.
4. To avoid these complications, choledochal cysts should be treated by complete excision, whenever possible, with reconstruction using internal drainage.
5. Follow-up is essential to detect development of any complications, such as cholangitis, anastomotic stricture, or intrahepatic cholelithiasis.

Suggested Reading

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