Choledochal Cysts

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A choledochal cyst (CDC) is an uncommon but correctable cause of biliary obstruction. Patients often present with abdominal pain, pancreatitis, jaundice, and rarely cholangitis. Left untreated, chronic inflammation eventually results in biliary cirrhosis and a risk of malignancy. Cases of cholangiocarcinoma, adenocarcinoma, cystadenoma, and superficial spreading cancer of the bile duct have been reported in patients with a CDC. Operative intervention is warranted in all patients with choledochal cysts.

Choledochal cysts were first described by Vater and Elzer in 1723. The anatomic variations were classified into three types by Alonso-Lej et al. and subsequently modified by Todani et al. into five subtypes (Fig. 91.1). The most common forms are type I and type IV CDC. In the case of a type I choledochal cyst, the dilatation is confined to the common bile duct, whereas the dilatation in patients with a type IV choledochal cyst either extends in continuity with the intrahepatic biliary tree or there is a short portion of normal-caliber common hepatic duct with isolated or multi-focal intrahepatic duct dilatation. In both type I and IV cysts, the dilated duct tapers to a normal diameter, or in the case of anomalous pancreaticobiliary ductal union (APBDU) a stricture of the common bile duct, before it is joined by the pancreatic duct in the head of the pancreas.

Types II, III, and V choledochal cysts are rare variants, comprising less than 2% of the cases. A type II choledochal cyst is a diverticulum arising from the wall of the bile duct, thought to occur due to a localized weakness in a segment of the common bile duct. A type III choledochal cyst, also known as a choledochocele, consists of a dilatation at the distal end of the common bile duct that protrudes into the lumen of the duodenum. Type V choledochal cyst, or Caroli disease, consists of multiple cystic dilatations of the intrahepatic biliary tree, often with strictures between the cysts. The cysts can be confined to one lobe of the liver or found throughout the intrahepatic biliary tree. Recently, a sixth type of choledochal cyst has been described. This type of cyst is defined as a cystic dilation of the cystic duct and has only been described in case reports.

Choledochal cysts are believed to be congenital in origin. The most widely accepted theory as to their pathogenesis is an APBDU, in which the pancreatic duct joins the bile duct proximal to the sphincter complex, forming a long common channel that allows reflux of pancreatic enzymes into the common bile duct. This results in inflammation, ectasia, and stricture just above the pancreatic ductal insertion, and ultimately dilation (Fig. 91.2). The damage to the bile duct is believed to occur in two ways: direct enzymatic injury to the biliary epithelium and damage caused by increased intraluminal pressure, due to a hypertrophic Sphincter of Oddi or the stricture. However, not all CDCs are associated with a long common channel, and not all long common channels are associated with a CDC. Particularly in neonates, CDC can be blind-ending (completely obstructive) or can be present with normal ductal anatomy (non-obstructive). More speculative theories are used to explain these CDC including embryologic proliferation of epithelial cells in the developing bile duct resulting in dilation and sphincter of Oddi spasm resulting in functional bile duct obstruction.

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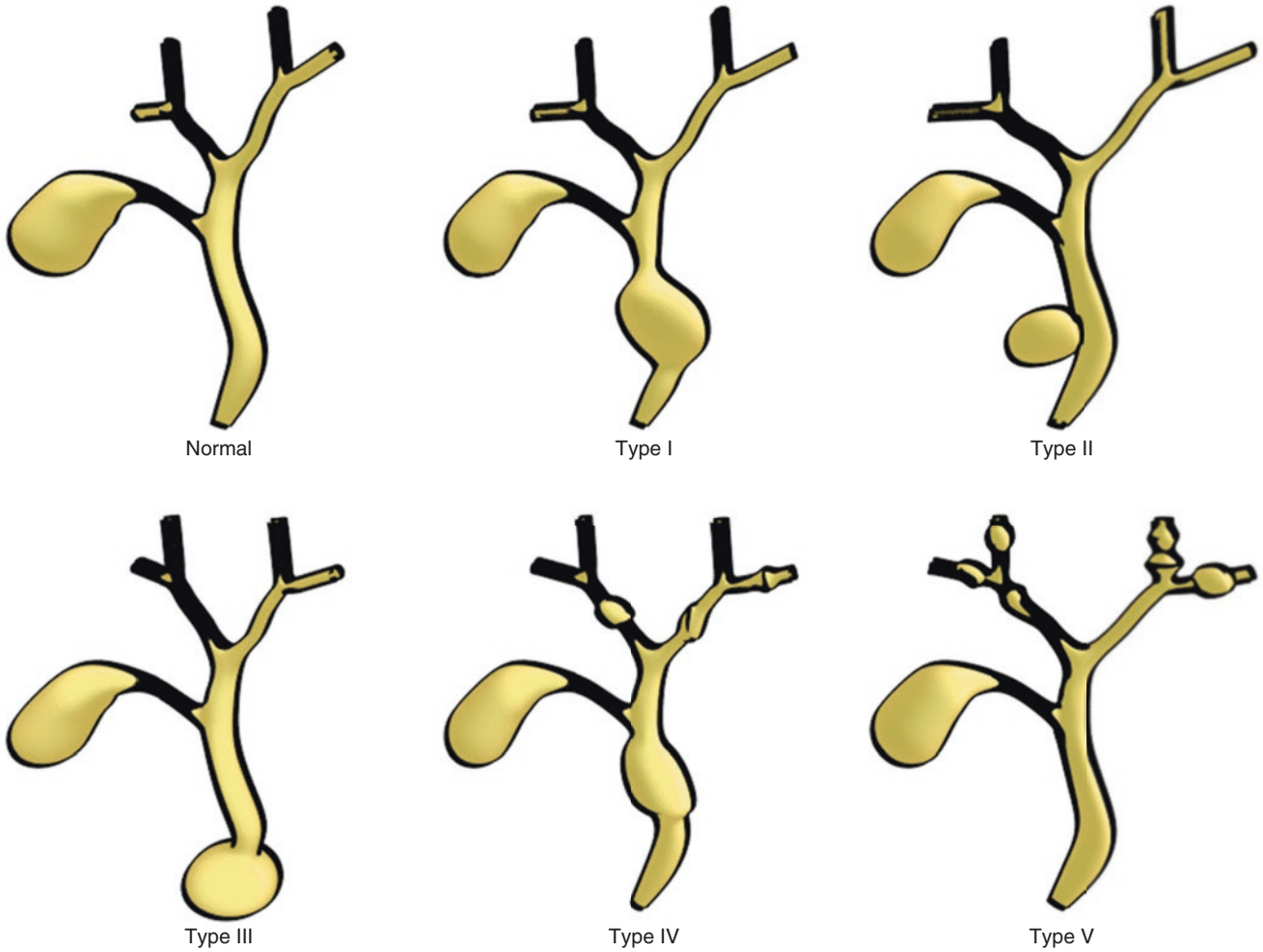

Fig. 91.1 Classification of choledochal cysts, according to Todani et al.: Type I represent approximately 90% of choledochal cysts and is a fusiform dilation of the extrahepatic bile duct. Some have proposed a further classification based on the pattern of disease such that Ia is dilatation of the entire extrahepatic bile duct, Ib is a focal dilatation of extrahepatic bile duct, and Ic is dilatation of the common bile duct itself, though whether these represent different types or simply points along the evolution of a cyst is unclear. Type II is a bile duct diverticulum, an uncommon variant. Type III is a choledochocele, a rare focal intramural dilation of the common bile duct right where it enters the duodenum at the ampulla of Vater. These are often treatable by endoscopic sphincteroplasty. Type IV is characterized by multiple cystic dilations of the intra- and extrahepatic bile duct. Type IVa is fusiform dilation of the entire extrahepatic bile duct and intrahepatic bile ducts, while IVb involves only the extrahepatic ducts. Type V is Caroli disease, in which case the cystic dilation affects a portion or the entirety of the intrahepatic biliary system. This is treated with liver lobectomy or hepatectomy with transplantation, depending on the extent of disease. (Courtesy of Assoc Prof Frank Gaillard, Radiopaedia.org, rID: 8097, used with permission)

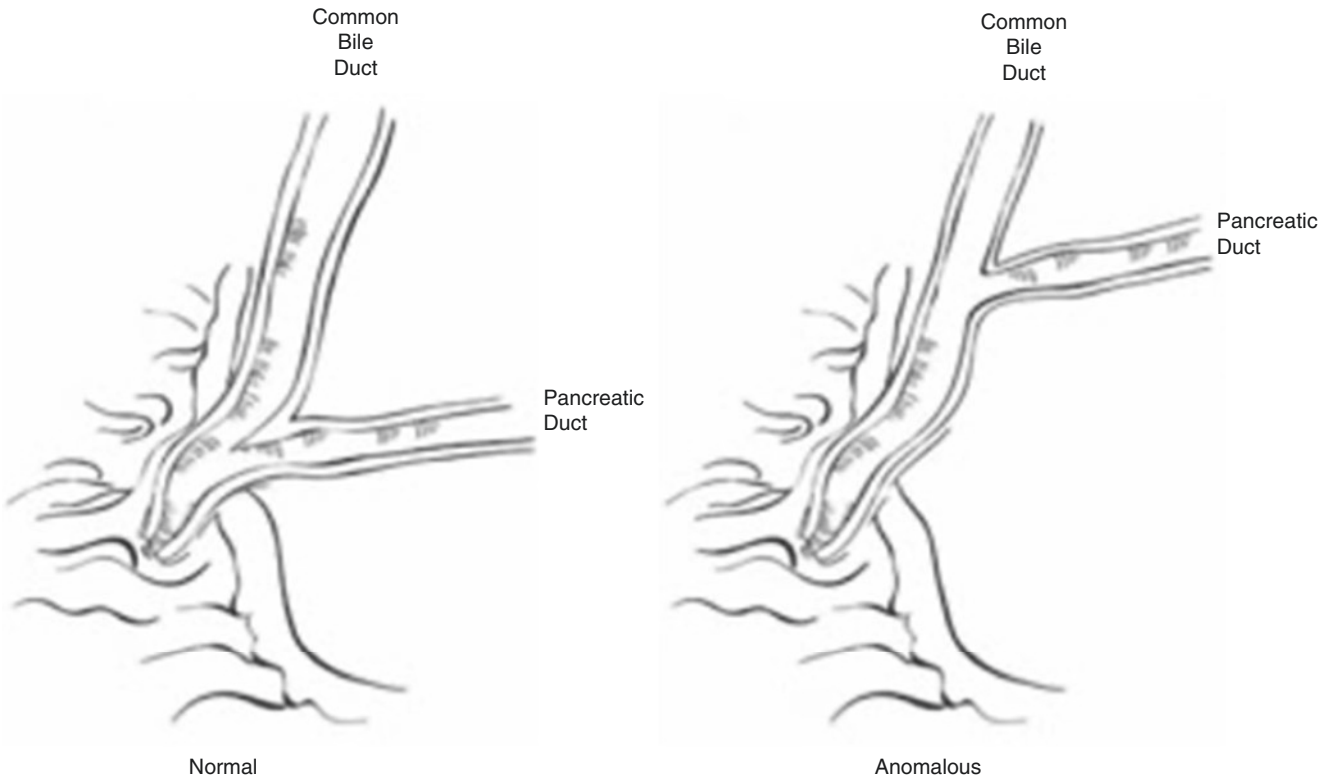

Fig. 91.2 Anomalous pancreaticobiliary ductal union (APBDU), the presumed pathologic basis for the development of a type I and IV choledochal cyst. Note the long common channel, which is thought to allow reflux of pancreatic fluid into the biliary tree where it may damage the lining over time, causing a cyst to form. (Modified with permission from Fischer JE, Bland KI, Callery MP, Clagett GP, Jones DB, editors. Mastery of surgery. 5th ed. Philadelphia, PA: Lippincott, Williams & Wilkins; 2006)

Diagnosis

Eighty percent of patients with choledochal cysts present before age 10. In clinical practice, the classic triad of abdominal pain, jaundice, and right upper quadrant mass occurs infrequently. Presentation in infancy is more commonly from an abdominal mass or jaundice, whereas after 2–3 years of age, most patients present with abdominal pain related to pancreatitis or ductal inflammation. A few patients are diagnosed during an episode of cholangitis when they present with jaundice, fever, and right upper quadrant pain (Charcot's triad). Older children, teenagers, and adults commonly present with symptomatic cholelithiasis or acute cholecystitis as a result of biliary stasis. In these cases, the common bile duct may be only minimally dilated, the so called *forme fruste* or *fusiform* CDC, and the underlying cause related to APBDU can be missed, leading to a misguided cholecystectomy and delayed diagnosis of the underlying pathophysiology. When APBDU is documented, all patients should undergo surgical treatment to prevent the consequences of APBDU pathophysiology, regardless of the size of their common bile duct. Prolonged pancreatico-biliary reflux with secondary ductal inflammation results in biliary epithelial damage, eventually leading to metaplasia or frank malignancy. Although this rarely occurs prior to adulthood, it should be addressed in childhood, as malignancies stemming from choledochal cysts carry a poor prognosis. In severe cases, the epithelium is replaced by granulation tissue and fibrosis, resulting in abnormalities of liver function, pancreatitis, and portal hypertension. A giant choledochal cyst can spontaneously perforate such that a patient presents with biliary ascites and sepsis. These patients have chemical peritonitis and require urgent care. These cases have become increasingly rare due to the widespread availability of US.

The diagnosis of a cystic anomaly of the biliary tract can often be made early in life. For types I, II, and IV, this is visualized as a cyst in the right upper quadrant. The diagnosis may be further delineated by the presence of a communication between the cystic duct and biliary tree. Further delineation of the anatomy—and particularly the diagnosis of APBDU—requires the use of MRCP. If detailed delineation of the long common channel or pancreatic duct is indicated, ERCP might be necessary. The gold standard diagnostic modality for a type III CDC is ERCP, which can also be therapeutic (endoscopic sphincterotomy).

Prenatal diagnosis of CDC is increasing in frequency. The differential diagnosis for a cystic lesion arising in the hilum of the liver detected in utero includes choledochal cyst and

the cystic variant of biliary atresia, which are difficult to definitively differentiate. It is therefore very important to counsel parents for both and to emphasize their different clinical course and prognosis. These patients require assessment of the patency of the extra-hepatic biliary tract soon after birth. If the biliary tract is obstructed, biliary atresia is likely. If the biliary tract is patent, the more likely diagnosis is choledochal cyst. Total and direct bilirubin levels need to be assessed in the first few weeks of life—an elevated total bilirubin with a greater than 20% direct fraction should lead to an early operative cholangiogram and either a Kasai portoenterostomy if biliary atresia is confirmed or resection of the CDC with reconstruction. If the bile ducts are patent, the patient can have normal bilirubin and enzyme levels after birth, in which case the patient can be followed by serial liver function tests (LFTs) and US. I typically recommend resection of the CDC before 3 years of age or prior to that if the LFTs or US (sludge or stones) show evolution of cholestatic pathophysiology. These patients are ideal, as operative repair can be performed prior to the onset of the inflammatory changes that can make operative dissection more difficult and dangerous.

Laboratory studies performed in patients with a choledochal cyst are highly variable. They may be normal early on or during quiescent periods, or serum conjugated and unconjugated bilirubin and hepatocellular transaminase levels may be elevated in the presence of significant cholestasis, cholangitis or cirrhosis. In patients who present with pancreatitis, serum amylase and lipase levels will usually be elevated.

Surgical Therapy

The first choledochal cyst excision with hepaticojejunostomy was performed by McWhorter in 1924. Thereafter, various procedures including external drainage or internal drainage through cyst-enterostomy were attempted, but with significant morbidity and mortality. Today, the treatment of a patient with a choledochal cyst depends on the subtype. The majority of patients with choledochal cysts undergo excision of the cyst and re-establishment of bile flow through a biliary-enteric anastomosis. For most, so as to limit the likelihood of progressive inflammatory changes within the CDC, we perform definitive correction soon after diagnosis. Exceptions to this include infants with asymptomatic and non-cholestatic CDCs where a period of growth is desirable, patients with active cholangitis who should be treated with an appropriate course of intravenous antibiotics before definitive repair is undertaken, and those who present with acute pancreatitis and who should be treated to allow for resolution of pancreatic inflammation prior to definitive resection. We administer prophylactic antibiotics before incision and continue them for 24 h.

The goal of surgical intervention should always be complete excision by whatever technique is utilized, whether open, laparoscopic, or robot-assisted. For patients with a type I or IV cyst, complete excision with reconstruction by either Roux-en-Y hepaticojejunostomy (RYHJ), or hepaticoduodenostomy (HD)can be performed. Hepaticoduodenostomy is gaining popularity due to its more physiologic anatomy, avoidance of an additional anastomosis, and the complications inherent in Roux limbs, shorter operative times, endoscopic access to the anastomosis, and the relative simplicity of the laparoscopic procedure. Prior concerns related to bile reflux and intrahepatic malignancy have not been substantiated. The laparoscopic technique is becoming more prevalent and has been used exclusively in my practice for over a decade. The laparoscopic approach is demanding and requires technical proficiency, but there are numerous reports documenting the safety and efficacy of the procedure in skilled hands with equivalent or better results than the open procedure. Typically, four ports are required. Excellent exposure of the hilum is obtained by a central transabdominal traction suture through the umbilical vein remnant at the liver, and a transabdominal suture through the retracted (cephalad) gall bladder. The cystic duct is divided and the distal end used for traction to safely get around the cyst at the level of the common hepatic duct. Once the duct has been encircled, it can be divided and the proximal dissection to the hepatic confluence performed. The hepatic duct is divided at the confluence. The distal duct can then be dissected to the point of narrowing above the pancreatic duct and then ligated and divided. Superiorly applied traction to the cyst and close adherence to the wall of the cyst allow for safer dissection into the head of the pancreas with visualization of the pancreatic duct in the rare circumstance that it is encountered within the cyst. An infant cystoscope can be utilized at this point through one of the epigastric ports to visualize and flush the proximal and distal ducts if stones are present. The choice of RYHJ or HD depends on the ease of approximation of the second part of the duodenum to the hepatic confluence after an extensive Kocher maneuver has been performed. It is almost always possible to do a HD without tension in children less than 12 years old. If tension is present, a RYHJ should be performed. In either case, transabdominal corner sutures should be placed to align the anastomosis. It is important to perform the hepaticoduodenostomy well distal to the pylorus to avoid bile reflux—I place mine just beyond the curvature of the duodenum into the second part. Fine monofilament sutures should be used to achieve mucosal to mucosal apposition of the anastomosis with either interrupted or running technique depending on the size of the duct. The traction sutures are then released and the cholecystectomy completed.

Complete resection of the cyst is important. If a portion remains, the risk of cancer may be as high as 50%. It is there-

fore important to dissect proximally to the hepatic ductal confluence and distally to the point where the duct is normal in caliber or strictured (above the entrance of the pancreatic duct). Historically, in patients with a long-standing choledochal cyst, recurrent bouts of cholangitis could result in significant inflammation throughout the hilum, fibrosis of the ducts, and adhesions, making dissection around the choledochal cyst quite difficult and creating the potential for injury to the hepatic artery or portal vein. The recommendation in this circumstance was to dissect the mucosa off of the adherent cyst, leaving the fibrous back wall in place adherent to the vasculature. In the current age, this appears to be rarely necessary. Dissection high on the cyst at the level of the common hepatic duct allows significant room behind the cyst to avoid the vessels and allow a careful dissection distally after division of the cyst. Using this technique, I have never needed to leave the wall of the cyst behind even in severely adherent cases. For a type IV cyst, the intrahepatic ductal dilatation usually resolves after CDC resection; but if the confluence is narrow, the lateral walls of the hepatic ducts can be spatulated to ensure a wide anastomosis. It is important to realize that even with complete CDC excision, there is a slightly increased (the magnitude of which is undefined) lifetime risk of intrahepatic cholangiocarcinoma over that in the general population, presumably due to intrahepatic ductal metaplasia from pancreatic fluid reflux prior to resection, and parents and patients must be counselled appropriately. Patients who have had previous internal drainage procedures for choledochal cysts should undergo reoperation with cyst excision, even if they remain asymptomatic. This is due to the continued risk of malignancy and symptom recurrence.

In patients who present with a perforated choledochal cyst, the historical recommendation was a two-stage procedure of external drainage, followed by resection of the cyst. A recent large series from China compared the results of single-stage resection with immediate reconstruction and the two-stage approach and found that the single-stage approach was safe and effective for most patients and dramatically reduced hospital stay. While this condition is rare in North America, primary cyst resection and reconstruction should be the strategy in most cases.

For patients with a type II choledochal cyst, my procedure of choice is to excise the diverticulum, unless it is also associated with an APBDU or if excision and closure risks narrowing the common bile duct. In those circumstances resection of the extrahepatic ducts with reconstruction should be performed.

The recommendations for treatment of the patient with a type III choledochal cyst vary according to the type of epithelium found within the cyst. An ERCP should be performed prior to surgical intervention of the mucosal lining of the cyst biopsied. If the biopsy reveals mucosa of duodenal origin, the lesion can be treated by sphincterotomy or meatotomy to enlarge the opening and relieve the obstruction. This is most commonly performed endoscopically though it can also be done open using a transduodenal approach. In the rare cases that the epithelium lining the lesion appears to be of biliary tract origin, or the cyst is large and not amenable for sphincterotomy, excision of the cyst with re-implantation of both the bile duct and the pancreatic duct has been recommended. Here again, it is essential to excise all of the biliary epithelium so as to minimize the likelihood of malignancy.

In patients with Caroli disease (type V choledochal cyst), the extent of disease dictates the type of surgical procedure performed. In patients with disease confined to one lobe of the liver, formal lobectomy is recommended. If a patient has bi-lobar or diffuse disease with recurrent cholangitis, liver failure, fibrosis, or malignancy, liver transplantation is the treatment of choice. The timing for transplantation has not been established but it must take place prior to the development of a cholangiocarcinoma. Therefore, these patients require close long-term surveillance even if asymptomatic.

Postoperative Care

Postoperative care in the acute phase is as for any major abdominal procedure with a low threshold to obtain imaging to rule out an anastomotic leak, such as for persistent fever or prolonged ileus. In the longer term, complications following repair of a choledochal cyst include stricture at the hepatic anastomosis, intrahepatic lithiasis or proteinaceous debris in the long common channel, and cholangiocarcinoma, any of which can occur many years later although most strictures occur in the first 2–3 years. Cholangitis should be very rare and usually is indicative of stricture or other obstruction to normal bile flow. The vast majority of patients have an uneventful postoperative course and do extremely well on long-term follow-up. My routine follow-up consists of LFTs and an US every year for 3 years to detect a stricture, and a yearly US of the liver every year thereafter for the ongoing risk of intrahepatic cholangiocarcinoma. As the absolute risk of cholangiocarcinoma in these patients is unknown, this follow-up is admittedly empiric.

Editor's Comment

Excision of the most common varieties of choledochal cyst can safely be performed laparoscopically or robotically—at many centers this is now routine. We have also shown that hepaticoduodenostomy is a safe and effective alternative to Roux-en-Y hepaticojejunostomy. Surgeons who worry about bacterial contamination and the risk of cholangitis still espouse routine use of a Roux-en-Y. However, bacterial contamination alone is usually well-tolerated in the absence of

obstruction or stasis. This is borne out by the fact that cholangitis after hepaticoduodenostomy remains quite rare.

In the absence of chronic inflammation, the operation to excise a choledochal cyst is generally straightforward but, as always, scrupulous attention to the technical details is critical. It is imperative that the mucosal lining of the cyst is completely excised, even if the fibrous wall of a chronically inflamed and adherent cyst must be left behind. It is also important during creation of the anastomosis that the mucosa of the biliary tree is meticulously approximated to that of the duodenum (or Roux limb). Finally, the entire distal common bile duct must be dissected, into the pancreas if necessary, while avoiding injury to the pancreatic duct. Stents and drains are not usually necessary, but patients should be closely monitored for signs of an anastomotic leak, which usually resolves with percutaneous drainage and 1–2 weeks of bowel rest and parenteral nutrition. As with most anastomoses involving the gastrointestinal (GI) tract, leaks are often followed by strictures, which sometimes respond to percutaneous balloon dilation or stent placement but more often require surgical revision.

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