# Imaging of the Choledochal Cyst[1]

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Choledochal cysts are rare and usually manifest in infancy and childhood. They are considered congenital because they occur in fetuses and neonates. Their origin may be related to an abnormal connection between the pancreatic duct and common bile duct and chronic reflux of pancreatic juice into the bile duct, resulting in irritation of the duct and subsequent dilatation. Choledochal cysts appear as cystic or fusiform dilatation of the common bile duct at radiography. Ultrasonography is the best initial method of evaluating dilatation of the intra- and extrahepatic bile ducts. Computed tomography is considered to be more accurate in delineating the intrahepatic biliary tree. Hepatobiliary scintigraphy with technetium-99m disofenin provides physiologic information on hepatic uptake and accumulation of radionuclide in the dilated biliary tree. Endoscopic retrograde cholangiopancreatography, percutaneous transhepatic cholangiography, and intraoperative cholangiography are definitive studies that demonstrate anatomic details of the biliary tree and the pancreaticobiliary ductal junction.

## ■ INTRODUCTION

Cystic dilatation of the common bile duct, also known as a choledochal cyst, is an uncommon but serious condition that requires surgical treatment. After publication of the first comprehensive review by Alonso-Lej et al (cited in references 1 and 2), this entity became widely known and has been reported throughout the world. Although choledochal cysts are generally considered a disorder of childhood and infancy, the ages in reported cases have ranged from newly born to 80 years old; however, 60% of such cysts are diagnosed in patients less than 10 years old (1–3). The origin of choledochal cysts has been a matter of considerable investigation and debate. The common channel theory proposed by Babbitt et al (4) is the most widely accepted theory, and an anomalous pancreaticobiliary ductal union has aroused interest as a cause of dilatation of the common bile duct.

**Abbreviations:** ERCP = endoscopic retrograde cholangiopancreatography, PTC = percutaneous transhepatic cholangiography

**Index term:** Bile ducts, cysts, 766.1492

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In this article, the possible origin of choledochal cysts from embryonic malformation of the pancreaticobiliary ductal union is discussed. An overview of the generally accepted classification of choledochal cysts is presented, and characteristic imaging features are shown with a combination of modalities, including upper gastrointestinal series radiography, ultrasonography (US), computed tomography (CT), and hepatobiliary scintigraphy. Endoscopic retrograde cholangiopancreatography (ERCP), percutaneous transhepatic cholangiography (PTC), and intraoperative cholangiography are reliable imaging methods that show the extent of bile duct dilatation and the anatomy of the pancreaticobiliary ductal union. However, ERCP is the best of these methods because it allows selective demonstration of the pancreatic and bile ducts (5–7). Cholangiopancreatography is reviewed with specific attention to the union of the common bile duct and pancreatic duct, the angle of an anomalous pancreaticobiliary ductal union, and the length and shape of the common channel. This review is supplemented by the findings of several large, comprehensive studies on anomalous pancreaticobiliary ductal junctions (8–12).

## ■ CLINICAL MANIFESTATIONS AND PATHOLOGIC FEATURES

The prevalence of choledochal cysts is higher among female patients; in most series, 70%–84% of the patients are female (2,3). Abdominal pain, jaundice, and an abdominal mass are the classic triad of signs and symptoms. These signs and symptoms may be present in 19%–60% of cases, but generally they are present in fewer than one-third of cases (3,13–15). Choledochal cysts usually manifest clinically in one of two ways: Some patients have abdominal pain, whereas others have persistent jaundice or an abdominal mass. Some studies have suggested that the type of manifestation is related to the morphology of the dilated common bile duct (10,12). Abdominal pain is the most common finding in patients with fusiform dilatation, whereas patients with cystic dilatation usually have a palpable abdominal mass or jaundice. Intermittent jaundice and an abdominal mass are the most common findings in infants; in children and adults, there is usually a history of intermittent fever, vomiting, jaundice, and abdominal pain, which seem to be related to cholangitis and pancreatitis (2,3,12,16,17).

These signs and symptoms do not occur at all times in all patients. An occasional "attack" of signs or symptoms may occur during an otherwise clinically silent course of disease as a result of increased pancreaticobiliary ductal pressure along the common channel and consequent intermixture of bile and pancreatic juice, which in turn gives rise to irritation of the pancreatic and bile ducts (12). On the other hand, there are also instances in which no signs or symptoms occur in the presence of a clearly anomalous pancreaticobiliary ductal junction. This latter situation indicates that some other factor, such as stenosis or obstruction, is necessary for signs and symptoms to occur (12).

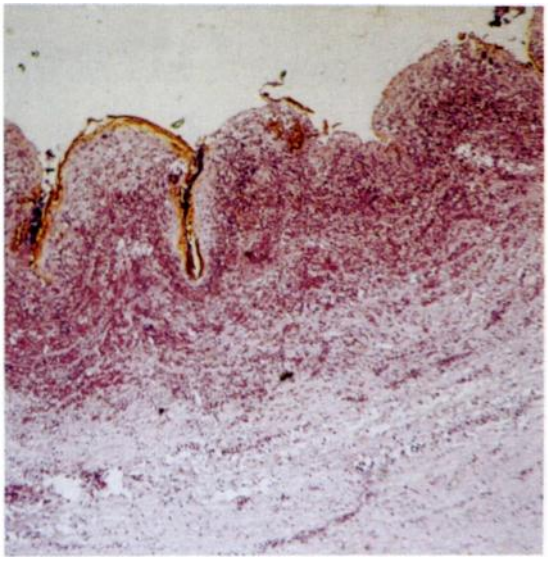


**Figure 1.** Histologic features of choledochal cysts. Photomicrograph (original magnification, ×40; hematoxylin-eosin stain) shows widespread ulceration and a denuded mucosa in the dilated common bile duct. Most of the thickened ductal wall consists of infiltrates of chronic inflammatory cells and dense fibrous connective tissue.

It is thought that a normal common bile duct has a single fibrous and columnar epithelial layer, with gland cells and muscle fibers making up the ductal wall (10,12,18). The inner lining of epithelium is easily denuded by irritation; histopathologic specimens from choledochal cysts show marked degeneration, as evidenced by denuded mucosa, thickening of the wall with fibrosis, and inflammatory cell infiltration (Fig 1).

## ■ PATHOGENESIS

Numerous pathologic, anatomic, and radiologic observations have stimulated various theories on the pathogenesis of this condition, but no theory has yet been able to explain the origin of this disorder satisfactorily. A theory that involves faulty epithelial proliferation and recanalization of the embryonic common bile duct was once widely accepted (2,3). However, in

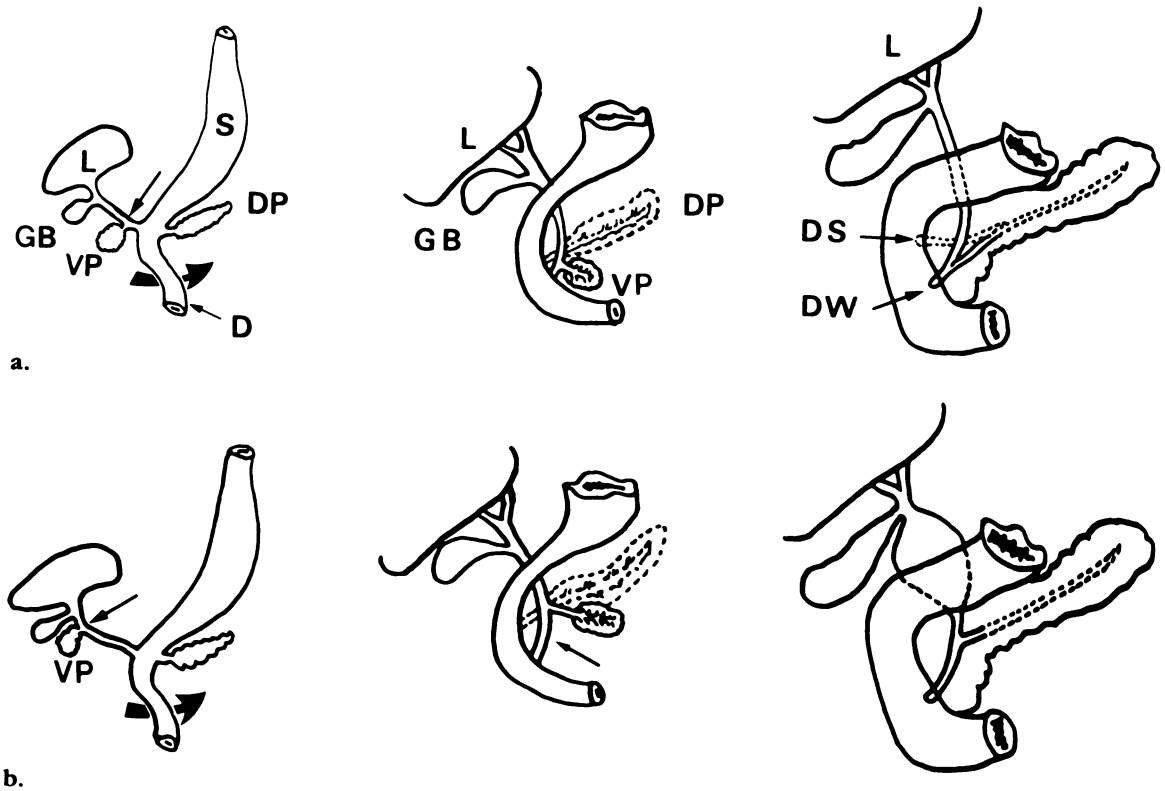



**Figure 2.** Development of the pancreaticobiliary ductal union. **(a)** Schemata show development of a normal pancreaticobiliary ductal union. *GB* = gallbladder, *L* = liver, *S* = stomach. Left: The dorsal (*DP*) and ventral (*VP*) pancreatic buds arise on opposite sides of the duodenum. The ventral bud eventually rotates (curved arrow) to within the "C loop" of the duodenum (*D*). The ventral bud originates from the common bile duct close to the duodenum, creating a short pancreaticobiliary duct (straight arrow). Middle: The dorsal (*DP*) and ventral (*VP*) pancreatic buds fuse. Right: The pancreatic ductal system fuses in the same manner as the pancreatic buds. The ventral duct and the distal part of the dorsal duct become the pancreatic duct (duct of Wirsung [*DW*]). The proximal part of the dorsal duct becomes the accessory pancreatic duct (duct of Santorini [*DS*]). (Adapted and reprinted, with permission, from references 4 and 19.) **(b)** Schemata show development of an anomalous pancreaticobiliary ductal union. Left: The ventral pancreatic bud (*VP*) originates in an abnormal position (nearer the liver) on the common bile duct (straight arrow). Curved arrow indicates direction in which the ventral bud will rotate. Middle: After rotation and fusion, an anomalously long pancreaticobiliary duct (arrow) is created. Right: The anomalous pancreaticobiliary ductal union leads to formation of a choledochal cyst. (Adapted and reprinted, with permission, from reference 20.)

recent years, attention has been drawn to the close association between anomalies of the pancreaticobiliary ductal junction and formation of choledochal cysts (4–12).

During normal development of the human pancreaticobiliary ductal system (Fig 2a), the ventral and dorsal pancreatic buds arise on opposite sides of the duodenum. Of the two pancreatic buds, the dorsal bud grows more rapidly. Eventually, the ventral bud rotates to within the C loop of the duodenum and becomes fused to the dorsal bud. The dorsal bud forms all of the pancreas except for the head portion, which is derived primarily from the ventral bud. The ductal system fuses in such a way that the ventral duct and the distal part of the dorsal duct become the pancreatic duct (duct of Wirsung). The proximal part of the dorsal duct becomes the accessory pancreatic duct (duct of Santorini), which may or may not persist into adult life (19).

The embryologic theory of anomalous pancreaticobiliary ductal formation relates the congenital aspect of choledochal cysts to faulty budding of the primitive pancreatic duct (Fig 2b). In this theory, the outpouching of the ventral pancreatic bud originates from a more proximal position (close to the liver); when this faulty ventral bud rotates and becomes the pancreatic duct, a long common channel that drains both the biliary and pancreatic systems is created, owing to the originally high position of the faulty ventral bud. This anomalous arrange-

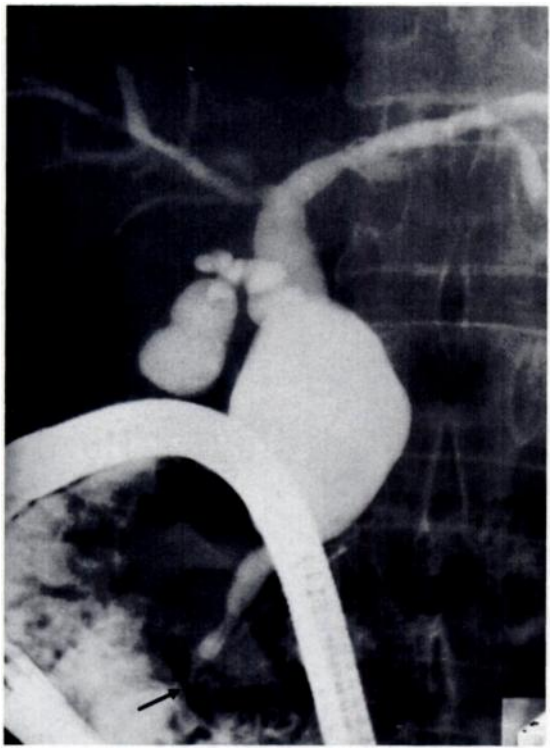


**Figure 3.** Choledochal cyst in a patient with a normal pancreaticobiliary ductal union. Cholangiogram shows fusiform dilatation of the common bile duct, yet the common bile duct and pancreatic duct form a normal common channel (arrow).

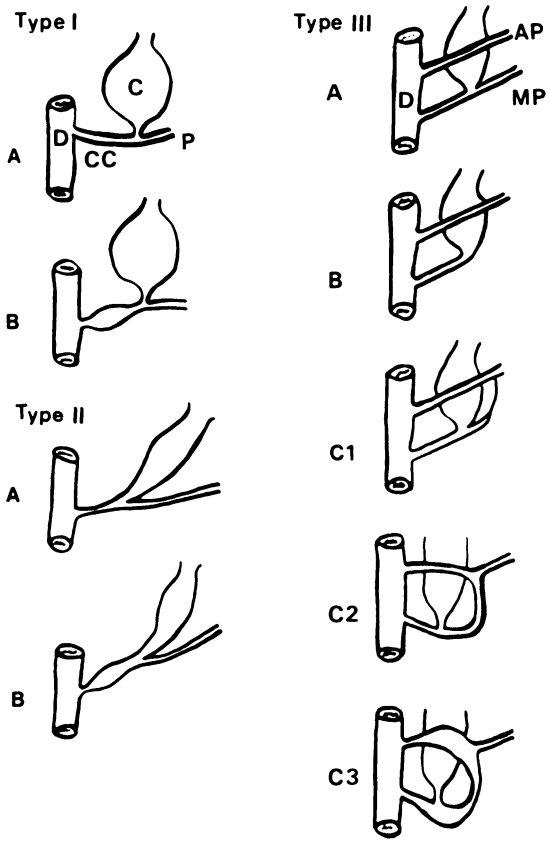



**Figure 4.** The system of Komi et al (23) for classification of anomalous pancreaticobiliary ductal junctions. *AP* = accessory pancreatic duct, *C* = choledochal cyst, *CC* = common channel, *D* = duodenum, *MP* = main pancreatic duct, *P* = pancreatic duct. (Adapted and reprinted, with permission, from reference 23.)

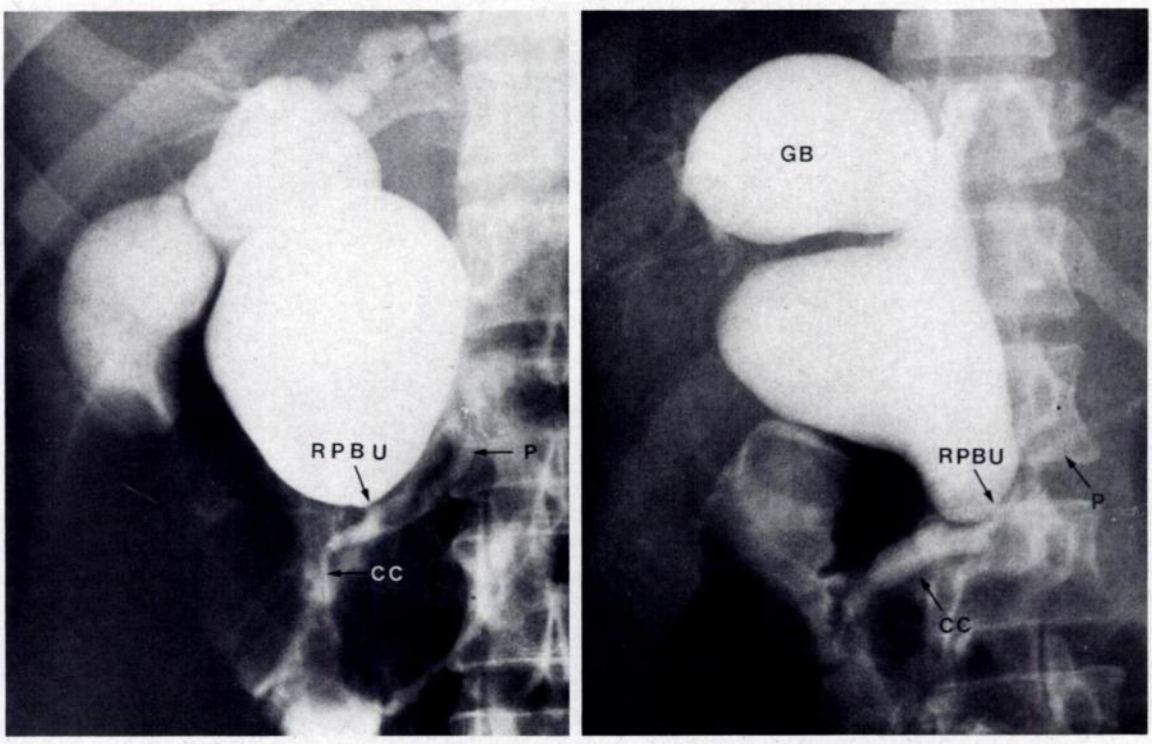
ment of the pancreaticobiliary ductal system is generally located far from the sphincter of Oddi (20).

Since the maximum pressure in the pancreatic duct exceeds that in the biliary duct by two to three times, this pressure difference allows flow of pancreatic juice into the bile ducts if no sphincter is present. Repeated bouts of cholangitis due to free reflux of pancreatic juice cause inflammation, mucosal destruction, and eventual fibrosis. Destruction of the common bile duct wall combined with distal stenosis of the wall because of fibrosis lead to dilatation of the common bile duct (4,20).

There is evidence to support this theory: High levels of pancreatic amylase have been detected in choledochal cysts (9,10,12,18,21). In addition, in an experimental model, cylindric dilatation of the common bile duct was induced by prolonged exposure to pancreatic juice (18,22). On the basis of these clinical and experimental observations, it is conceivable that choledochal cysts are an acquired condition that results from chronic pancreatic juice reflux due to a congenital anomalous pancreaticobiliary ductal union. However, the theory of malformation of the pancreaticobiliary ductal junction does not explain the occurrence of choledochal cysts in the presence of a normal pancreaticobiliary ductal union (9) (Fig 3).

## ■ CHOLANGIOPANCREATOGRAPHIC OBSERVATIONS RELATED TO AN ANOMALOUS PANCREATICOBILIARY DUCTAL UNION

The common bile duct and pancreatic duct normally unite within the sphincter of Oddi to form a common channel, termed the *normal pancreaticobiliary duct*, which opens into the middle portion of the duodenum. The normal



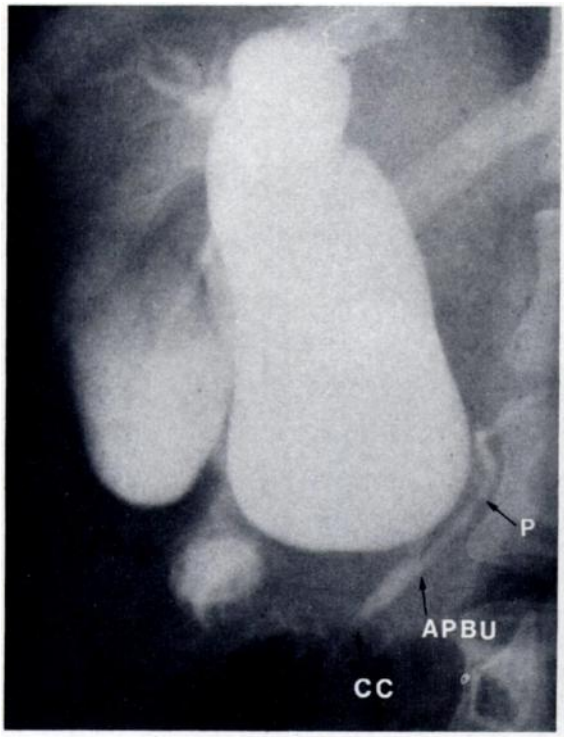

a. b.

**Figure 5.** Type I anomalous pancreaticobiliary ductal junctions. *GB* = gallbladder, *P* = pancreatic duct, *RPBU* = right-angled pancreaticobiliary ductal union. **(a)** ERCP image shows cystic dilatation of the common bile duct and a slender common channel (*CC*). This is a type IA union. **(b)** ERCP image shows eccentric cystic dilatation of the common bile duct and a long, ectatic common channel (*CC*). This is a type IB union.

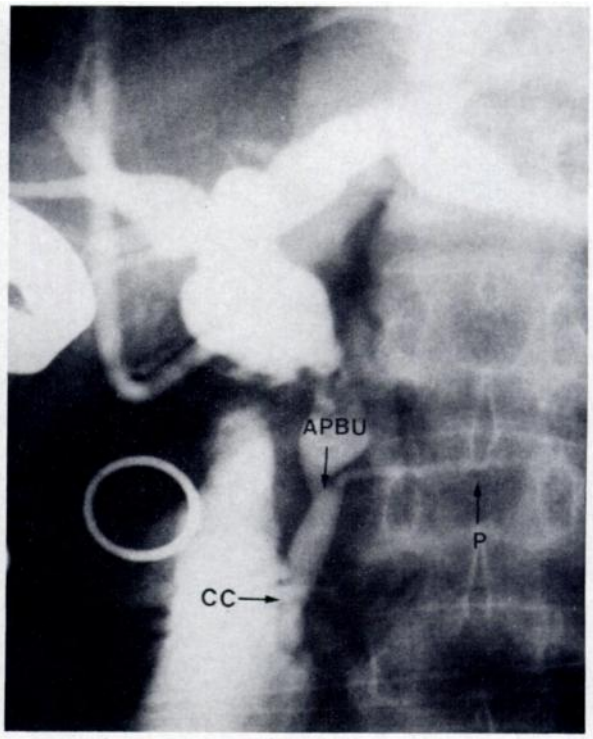
length of the common channel is 0.2–1.0 cm (average, 0.5 cm), and the diameter is usually less than that of the normal pancreatic duct (3–5 mm in normal adults) because the common channel is encircled by sphincter muscle fibers (5,8,9,11).

An anomalous pancreaticobiliary ductal union has two features that are relevant to formation of choledochal cysts (21). One is that the union of the pancreatic duct and common bile duct is located far from the duodenum, creating a long common channel. The second feature is the angle of this junction. Normally, the pancreatic duct and the distal part of the common bile duct form an acute angle as they converge and are enclosed by the sphincter of Oddi (20). Some investigators (23) classify anomalous pancreaticobiliary ductal junctions into three types according to the angle of ductal union: (*a*) a right-angled union without an accessory pancreatic duct, (*b*) an acute-angled union without an accessory pancreatic duct, and (*c*) a right- or acute-angled union with an accessory pancreatic duct. These types are subdivided according to the shape of the common channel.

A scheme for classifying anomalous pancreaticobiliary ductal junctions is outlined in Figure 4. In a type I union, a narrowed common bile duct joins the pancreatic duct at a right angle; a type IA union has a slender common channel (Fig 5a), whereas a type IB union has a short or long ectatic common channel (Fig 5b). In a type II union, the pancreatic duct joins the

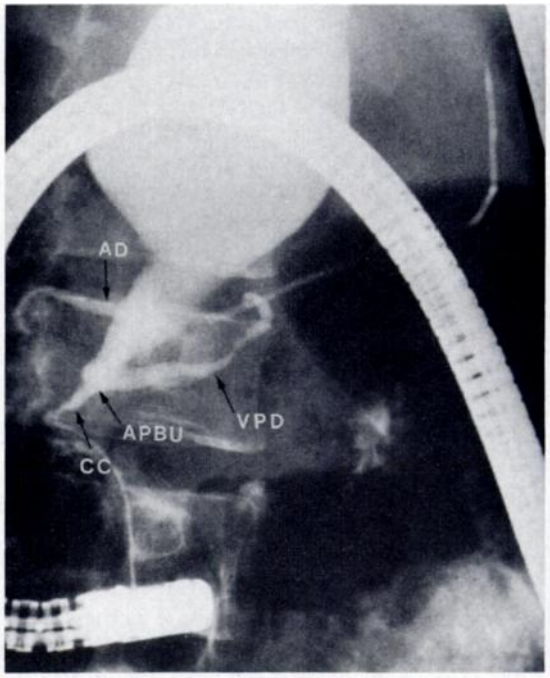



6a.




6b.

**Figures 6, 7.** **(6)** Type II anomalous pancreaticobiliary ductal junctions. *APBU* = acute-angled pancreaticobiliary ductal union, *P* = pancreatic duct. **(a)** ERCP image shows fusiform dilatation of the common bile duct and a slender common channel (*CC*). This is a type IIA union. **(b)** Intraoperative cholangiogram shows a long, ectatic common channel (*CC*). This is a type IIB union. **(7)** Type III anomalous pancreaticobiliary ductal union. ERCP image shows proximal cystic and distal fusiform dilatation of the common bile duct, an acute-angled pancreaticobiliary ductal union (*APBU*), and a short and slender common channel (*CC*). There is also fusion of the ventral pancreatic duct (*VPD*) and the accessory duct (*AD*). This is a type IIIC2 union.




7.

common bile duct at an acute angle; a type IIA union has a slender common channel (Fig 6a), whereas a type IIB union has a short or long ectatic common channel (Fig 6b). A type III union is complicated by a patent accessory pancreatic duct. This type is subclassified as follows: A type IIIA union is a classic pancreas divisum. A type IIIB union is a pancreas divisum without a pancreatic duct. A type IIIC union has a patent accessory pancreatic duct with an intricate network and is further classified as (*a*) a type IIIC1 union, which is characterized by a tiny communicating duct between the main and accessory ducts; (*b*) a type IIIC2 union, which is characterized by a communicating duct with the same caliber as the main and accessory ducts (Fig 7); and (*c*) a type IIIC3 union, which is characterized by total or partial dilatation of the ductal system.

The reasons for classifying anomalous pancreaticobiliary ductal junctions are because the shape of the choledochal cyst—cystic or fusiform—may be influenced by the angle of the

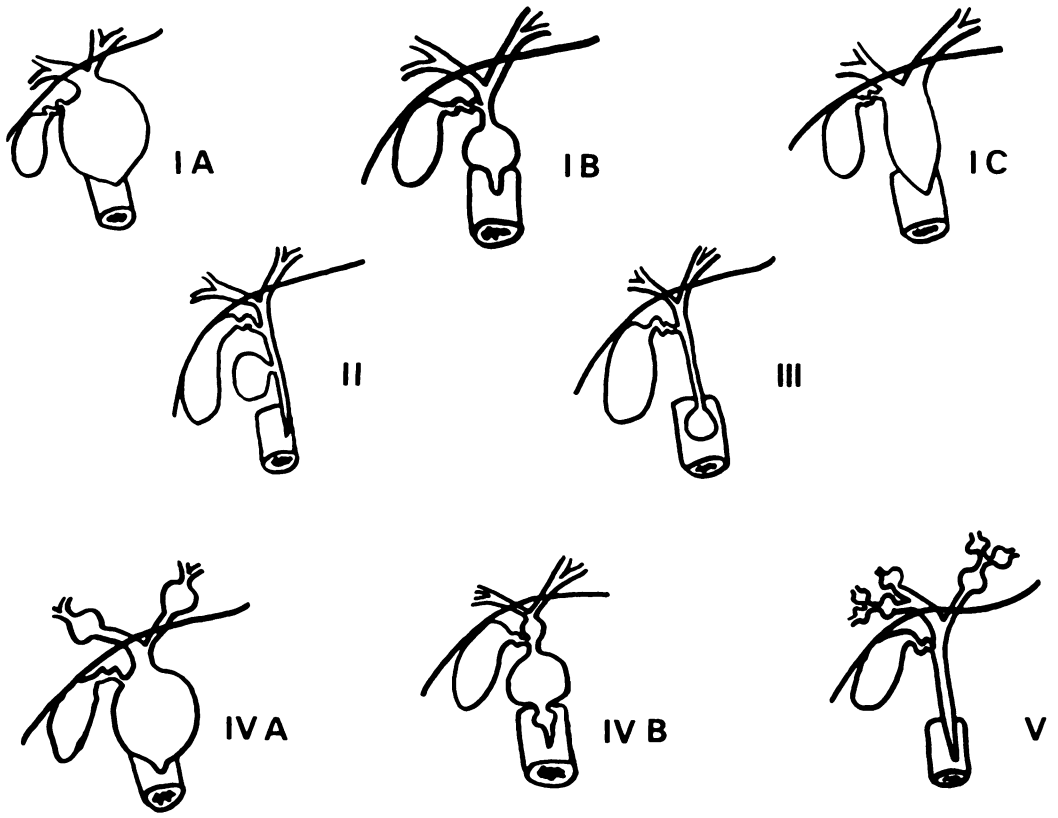
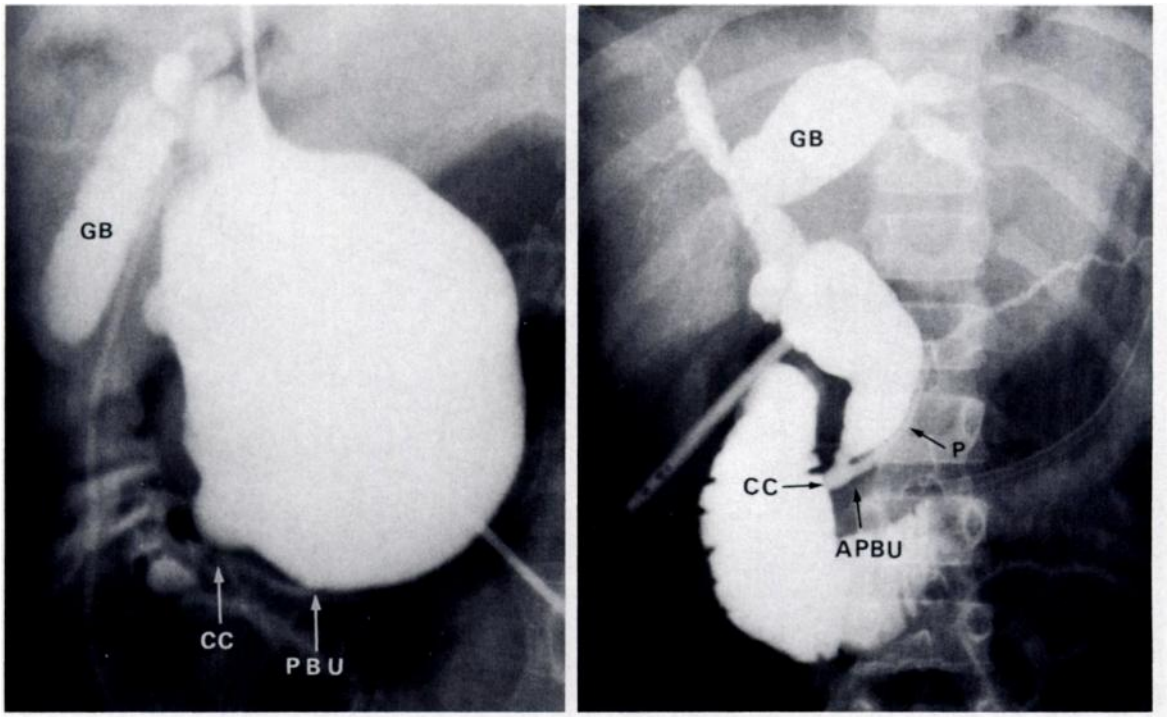



**Figure 8.** The system of Todani et al (1) for classification of choledochal cysts.

anomalous ductal union and because the clinical manifestations tend to be different (9,10,12, 23). A right-angled union is associated with cystic dilatation of the common bile duct; patients with this type of union have a palpable mass or jaundice as the main sign of disease. In contrast, an acute-angled union is associated with fusiform dilatation of the common bile duct; patients with such dilatation usually have abdominal pain, and an erroneous diagnosis of acute pancreatitis is occasionally made (9,12). Moreover, type IB, IIB, and III junctions, which are associated with a dilated common channel and an accessory pancreatic duct, are frequently complicated by relapsing pancreatitis that leads to chronic pancreatitis (23). Thus, it is important to note the type of anomalous pancreaticobiliary ductal union and the presence of a dilated common channel or an accessory pancreatic duct.

## ■ CLASSIFICATION OF CHOLEDOCHAL CYSTS

The widely accepted classification of choledochal cysts devised by Todani et al (1) (Fig 8) is based on the cholangiographic morphology, location, and number of intrahepatic and extrahepatic bile duct cysts. This classification leads to some confusion because it includes Caroli disease (multiple cystic dilatation of the intrahepatic ducts without dilatation of the common bile duct). However, when one considers the relationship between possible underlying anomalous development of the pancreaticobiliary ductal union and the resultant dilatation of the common bile duct, Caroli disease is seen to be a distinct disease entity. Similarly, choledochocele seems to be an anatomic variation rather

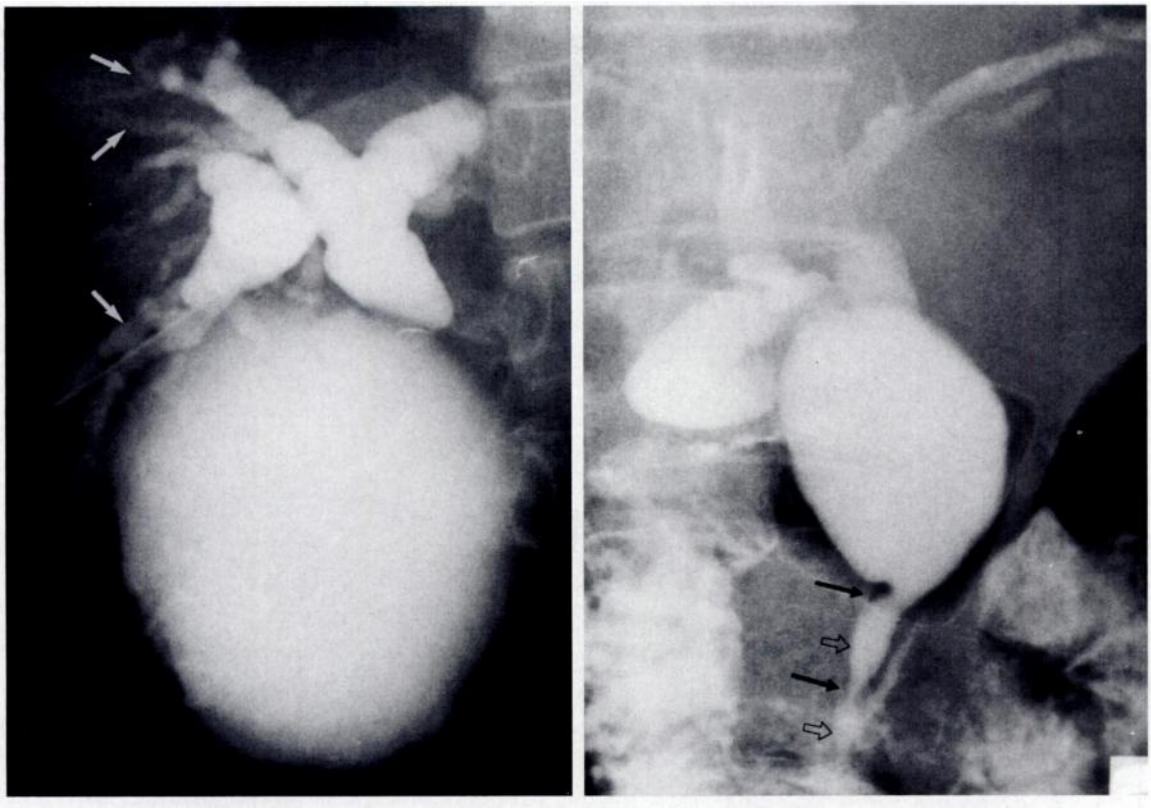



9. 10.

**Figures 9, 10.** **(9)** Type IA choledochal cyst. Intraoperative cholangiogram shows cystic dilatation of the entire common bile duct. The gallbladder (*GB*) arises from the cyst. The pancreaticobiliary ductal union (*PBU*) is right angled, and a long and stenotic common channel (*CC*) is seen. **(10)** Type IC choledochal cyst. Intraoperative cholangiogram shows fusiform dilatation of the common bile duct and diffuse, cylindric dilatation of the common hepatic duct but no dilatation of the intrahepatic ducts. The gallbladder (*GB*) arises from the dilated common bile duct. *APBU* = acute-angled pancreaticobiliary ductal union, *CC* = common channel, *P* = pancreatic duct.

than representing true dilatation of the common bile duct. However, many authors include Caroli disease and choledochocele in the spectrum of choledochal cysts (2,3,5,13,14,24).

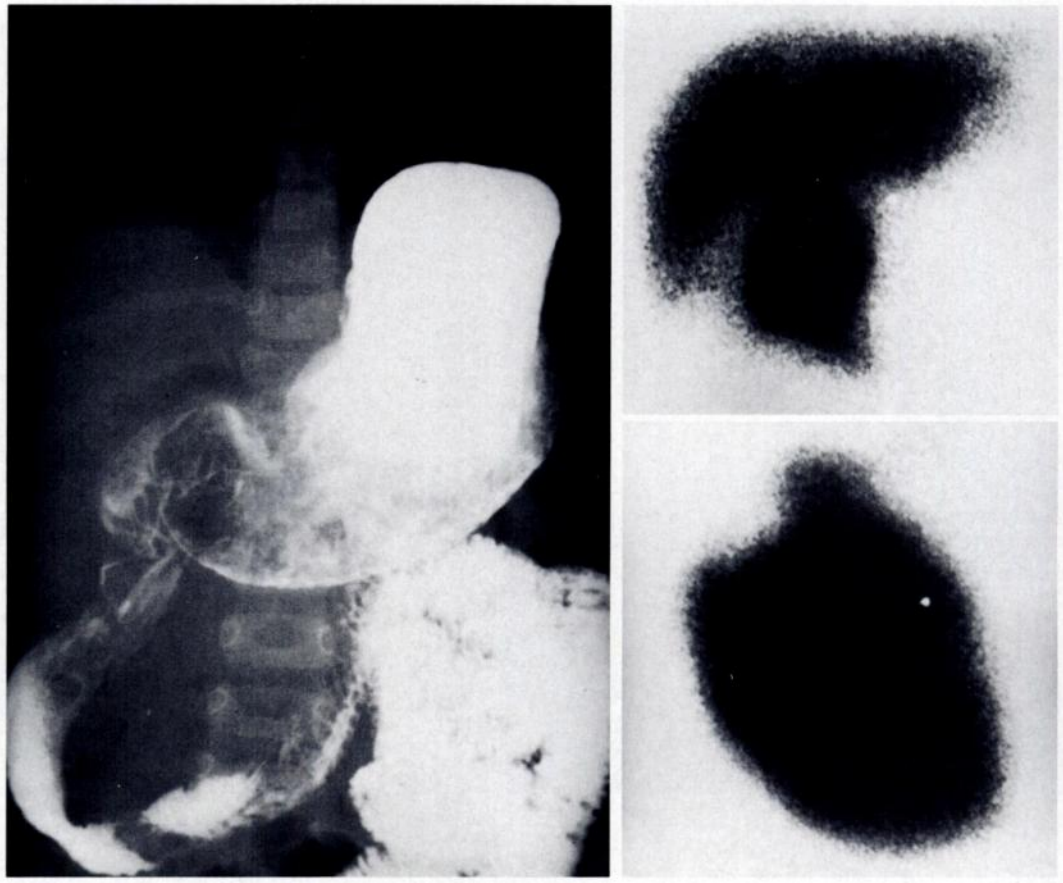
In the classification of Todani et al (1), a type I cyst is the most common form, present in 80%–90% of cases. Type I cysts are further classified according to the shape of the affected segment. A type IA cyst involves dilatation of the common bile duct, with marked dilatation of part or all of the extrahepatic biliary tree (Fig 9). The gallbladder commonly arises from the cyst, and the intrahepatic biliary tree is normal. A type IB cyst involves focal, segmental dilatation of the common bile duct, usually of the most distal part of the duct. A normal segment of common bile duct is present between the cyst and the cystic duct. A type IC cyst involves fusiform dilatation of the common bile duct, along with diffuse, cylindric dilatation of the common hepatic duct and common bile duct (Fig 10). The gallbladder arises from the dilated common bile duct, and the intrahepatic biliary system is not dilated. A type II cyst is a choledochal diverticulum. A type III cyst is a



11. 12.

**Figures 11, 12.** **(11)** Type IVA choledochal cyst. PTC image shows marked cystic dilatation of the common bile duct and of the intrahepatic bile ducts. The peripheral intrahepatic ducts have a beaded appearance (arrows). **(12)** Type IVB choledochal cyst. ERCP image shows marked cystic dilatation of the proximal part of the common bile duct and areas of lesser dilatation (open arrows) in the distal part of the duct. The segments of common bile duct between the cysts are stenotic (solid arrows). In this patient, no evidence of an anomalous pancreaticobiliary ductal union was noted.

choledochocele that involves only the intraduodenal portion of the common bile duct; this is analogous to the shape of a ureterocele. Type IV cysts are subclassified as follows: A type IVA cyst involves dilatation of the intrahepatic and extrahepatic bile ducts. Cholangiography shows gross cystic dilatation of the extrahepatic biliary tree, with extension of the cystic dilatation into the intrahepatic biliary tree (Fig 11). The intrahepatic dilatation may affect multiple segments, be smooth and fusiform, or be irregular. A type IVB cyst also involves dilatation of multiple segments but is confined to the extrahepatic bile duct. Type IVB cysts are much less common than type IVA cysts. Cholangiography shows multiple segmental dilatation of the common bile duct (Fig 12). The intrahepatic biliary tree is normal. A type V cyst (Caroli disease) involves dilatation of one or several segments of the intrahepatic bile ducts.



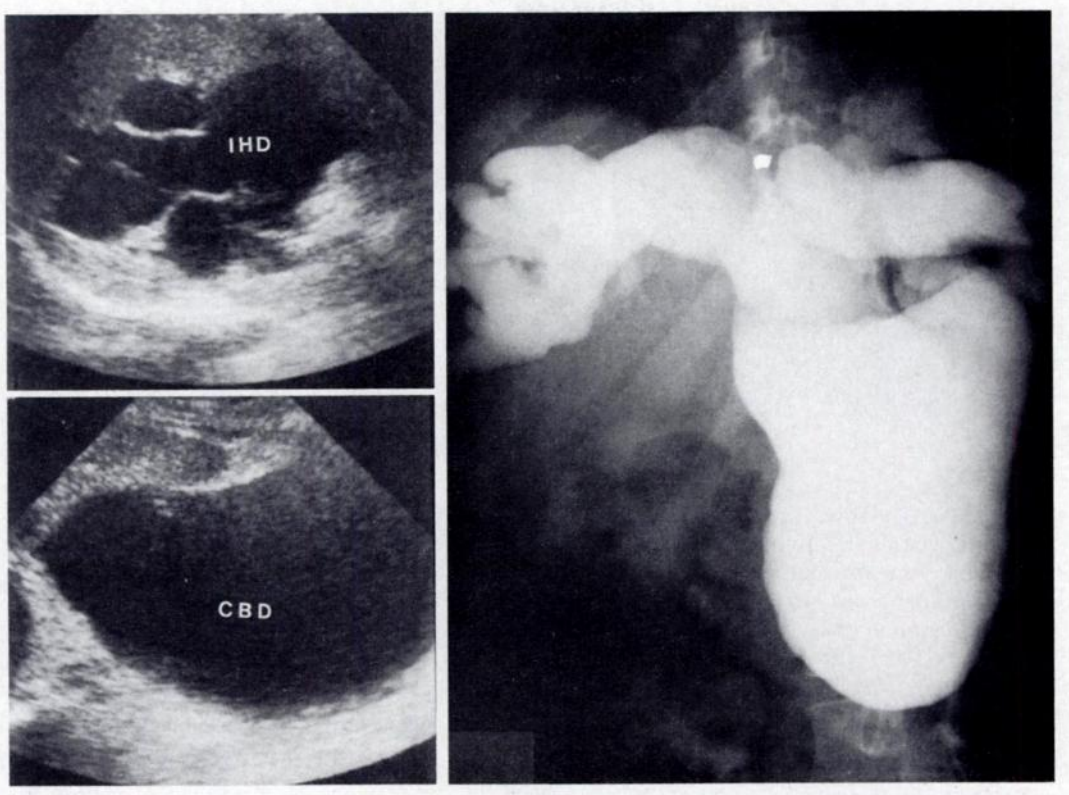
a. b.

**Figure 13.** Choledochal cyst in a 5-year-old girl with a palpable abdominal mass. **(a)** Anterior view from a barium-enhanced upper gastrointestinal radiographic examination shows widening and displacement of the duodenal C loop by a huge mass. **(b)** Technetium-99m disofenin (diisopropyliminodiacetic acid [DISIDA]) hepatobiliary scintigrams obtained at 30 minutes (top) and 2 hours later (bottom) allow confirmation of the presence of cystic dilatation of the common bile duct. The radionuclide stagnates in the markedly dilated common bile duct, and radionuclide activity in the bowel is not evident.

## ■ IMAGING FEATURES

In the past, barium studies were used in patients with large choledochal cysts, but this method is no longer used for evaluation of choledochal cysts (25,26). Upper gastrointestinal series radiography shows nonspecific displacement of the stomach and duodenum in patients with choledochal cysts. On the anteroposterior view, the duodenal C loop is widened and displaced downward; on the lateral view, the duodenum is displaced forward (Fig 13).

US is regarded as the best initial method of evaluating any patient with jaundice, regardless of age (27–29). It is capable of demonstrating the entire intrahepatic and extrahepatic biliary

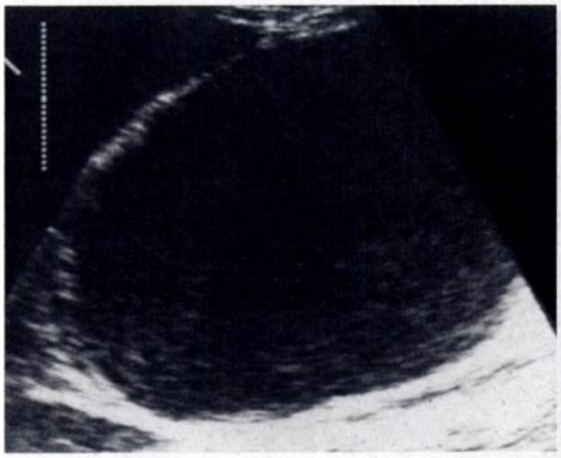



a. b.

**Figure 14.** Type IVA choledochal cyst in a 20-year-old woman with jaundice and abdominal pain. **(a)** Oblique sonograms obtained at (top) and below (bottom) the porta hepatis show marked cystic and fusiform dilatation of the intrahepatic ducts (*IHD*) and common bile duct (*CBD*). **(b)** PTC image shows marked dilatation of the intrahepatic ducts and common bile duct.

tree. Sonography shows a choledochal cyst as a characteristic cystic or fusiform structure in the porta hepatis, separate from the gallbladder, and may show communication with a dilated common hepatic duct or the intrahepatic ducts (Fig 14).

US and CT are both helpful in showing cystic masses in relation to the pancreatic head and porta hepatis. The size, extent, and cystic character of a choledochal cyst are clearly seen with both methods, but CT is considered to be more accurate in demonstrating the intrahepatic biliary tree and the status of the distal part of the common bile duct, which may be obscured by bowel gas on sonograms (14). However, when



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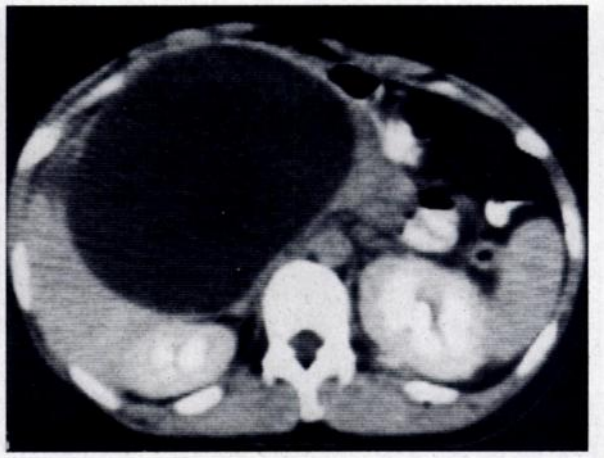

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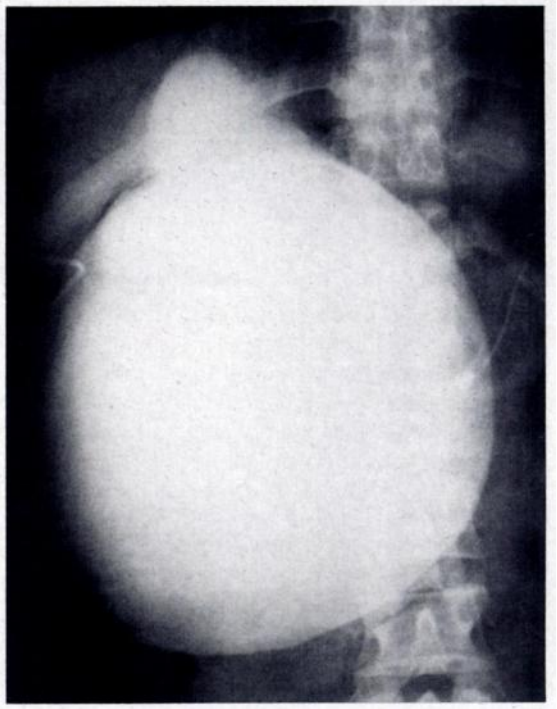
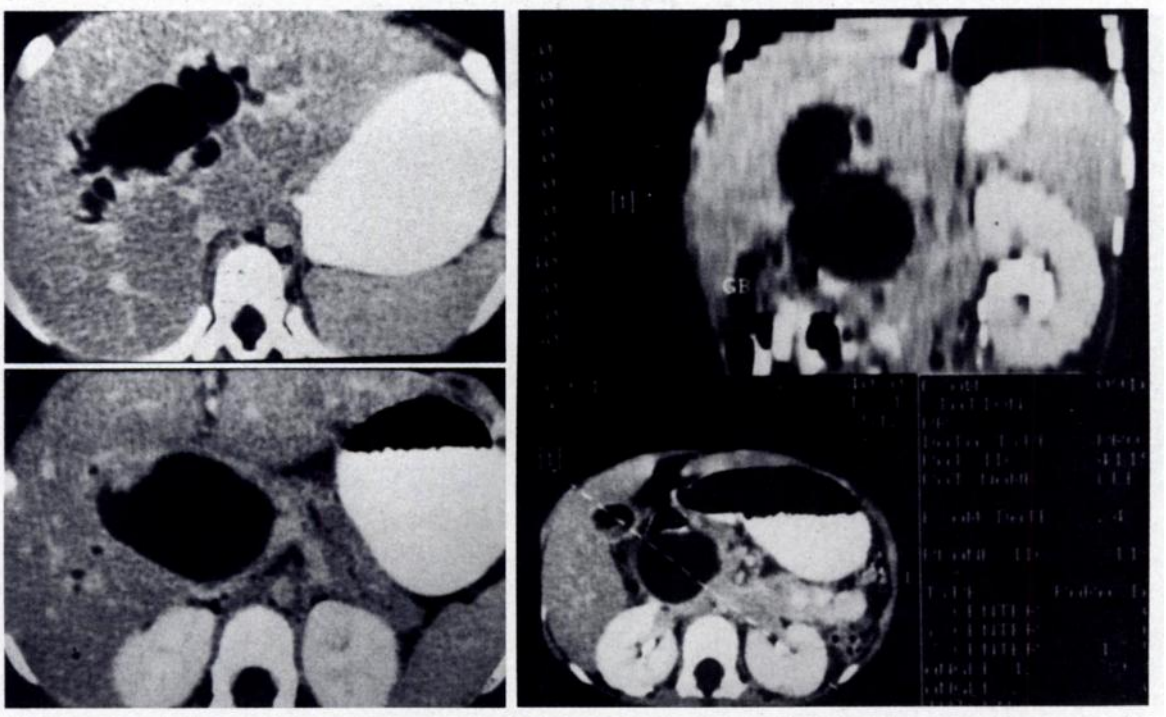


c.

**Figure 15.** Choledochal cyst with no evidence of intrahepatic ductal dilatation in a 27-year-old woman who was 7 months pregnant and had an 8-day history of abdominal pain and a palpable mass in the right upper quadrant. **(a)** Transverse sonogram shows a large cyst in the subhepatic area. **(b)** Contrast material–enhanced abdominal CT scan shows a well-demarcated cystic mass. Because dilatation of the intrahepatic biliary tree was not seen, the mass was not expected to have a biliary origin. **(c)** Intraoperative cholangiogram shows gross cystic dilatation of the common bile duct. The cyst contained about 2,000 mL of thick bile.

a cyst is round and markedly dilated with no evidence of intrahepatic ductal dilatation, exact diagnosis is difficult with both CT and US; in such cases, biliary origin often cannot be determined (Fig 15). Abdominal cystic lesions such as mesenteric, omental, ovarian, renal, adrenal, and hepatic cysts; gastrointestinal duplication; hydronephrotic kidneys; and pancreatic pseudocysts are the main differential diagnoses when a huge choledochal cyst lacks intrahepatic involvement at CT (30).

CT and US are valuable in demonstrating choledochal cysts, but the biliary origin of such cysts can be confirmed with hepatobiliary scintigraphy. Tc-99m disofenin scanning demonstrates early uptake of the agent by hepatocytes followed by excretion of the radionuclide into the biliary system and accumulation and stasis of the radionuclide in the dilated ducts and the choledochal cyst (27,31). On scintigrams, a choledochal cyst appears as a group of photopenic areas in the porta hepatis, which represent the dilated intrahepatic biliary tree and which are gradually filled by the radionuclide on later images (Fig 16). Stasis of the agent within the dilated bile ducts may cause nonvisualization of radionuclide bowel activity (29) (Figs 13b, 16b).

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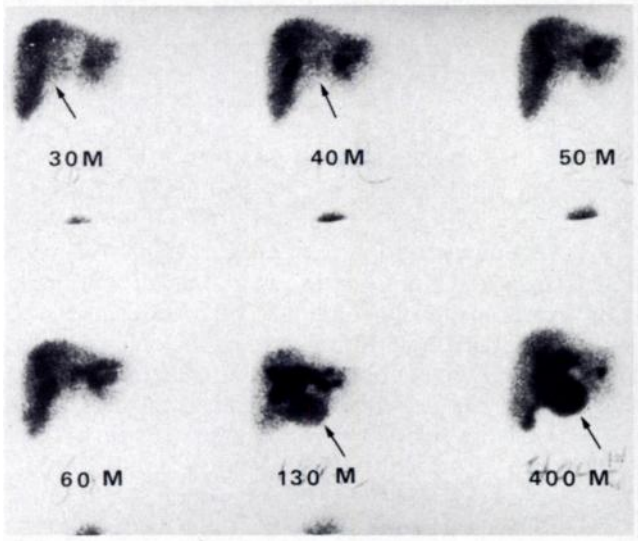


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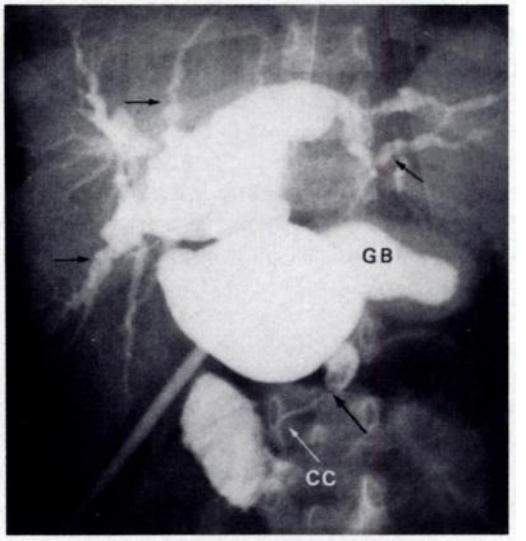
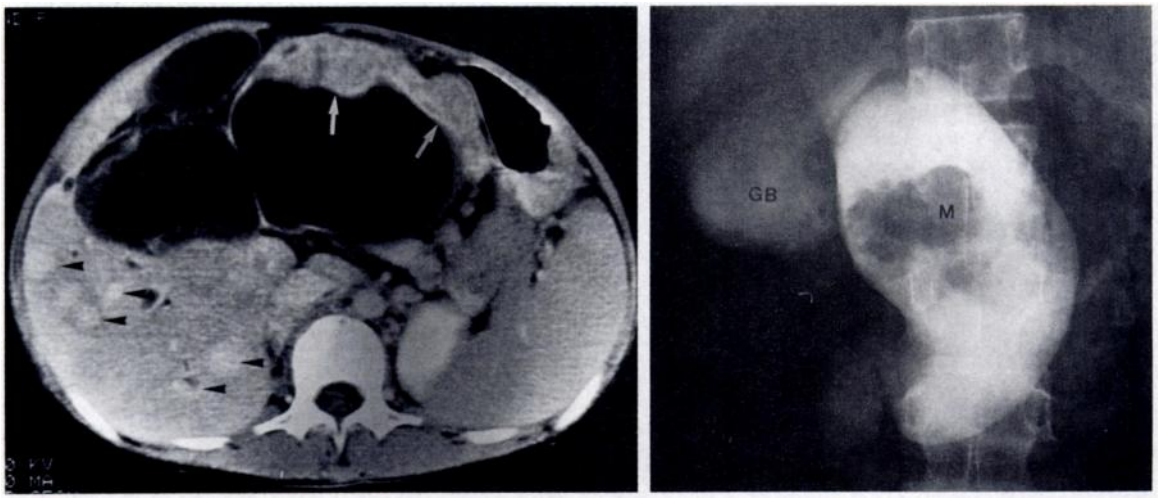



c.

**Figure 16.** Type IVA choledochal cyst in a 3-year-old girl with abdominal pain and jaundice. **(a)** Contrast-enhanced CT scans show marked dilatation of the intrahepatic (top left) and extrahepatic (bottom left) biliary tree, which is well demonstrated on the coronal reconstruction image (right). *GB* = gallbladder. **(b)** Serial hepatobiliary scintigrams demonstrate a lobulated "cold" area (eg, one with less activity than in surrounding tissue) in the porta hepatis (arrows) that is filled with radionuclide on the later images. *M* = minutes after injection. **(c)** Intraoperative cholangiogram shows cystic dilatation of the intra- and extrahepatic bile ducts. The intrahepatic bile ducts have a beaded appearance (small black arrows). The pancreaticobiliary ductal union (long black arrow) is right angled, and the common channel (*CC*) is long and stenotic. *GB* = gallbladder.




a. b.

**Figure 17.** Bile duct carcinoma within a choledochal cyst in an 18-year-old woman. **(a)** Contrast-enhanced CT scan shows a choledochal cyst and a lobulated, enhanced intraluminal mass (arrows). Liver metastases are seen (arrowheads). **(b)** ERCP image shows the mass (*M*) within the choledochal cyst as an area of irregular defects. *GB* = gallbladder. (Courtesy of Yong Ho Auh, MD, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea.)

## ■ COMPLICATIONS AND ASSOCIATED ANOMALIES

The most common complication associated with a choledochal cyst is stones in the gallbladder, within the cyst, in the dilated intrahepatic biliary tree, or in the pancreatic duct (3). The second most common complication is a malignant tumor. Common bile duct carcinoma and gallbladder carcinoma are the major malignancies (3,32–35) (Fig 17). The risk of developing cancer seems to be related to bile stasis and contact between epithelium and bile. It may also be related to reflux of pancreatic juice into the biliary tree, which gives rise to chronic irritation and metaplasia (36). In addition, the risk of malignancy appears to be related to the age of the patient; there is a higher risk with increasing age. Although excision of the cyst eliminates a potential source of carcinoma, it does not exclude the possibility of cancer developing in the intrahepatic ducts. Long-term observation is therefore indicated in all patients with choledochal cysts (36–38).

In infants and neonates, a choledochal cyst may occasionally manifest as bile peritonitis secondary to rupture of the cyst (10,17). The signs and symptoms are abdominal distention, fever, pain, and vomiting. The pathogenesis of spontaneous perforation of choledochal cysts is unknown, but underperfusion with acute inflammation of the dilated ductal wall and mural weakness caused by reflux of pancreatic juice are two possible factors (10). CT and US of ruptured cysts show free intraperitoneal fluid and a dilated common bile duct with or without dilatation of the intrahepatic ducts (Fig 18). Operative cholangiography allows confirmation of the presence of a perforation.

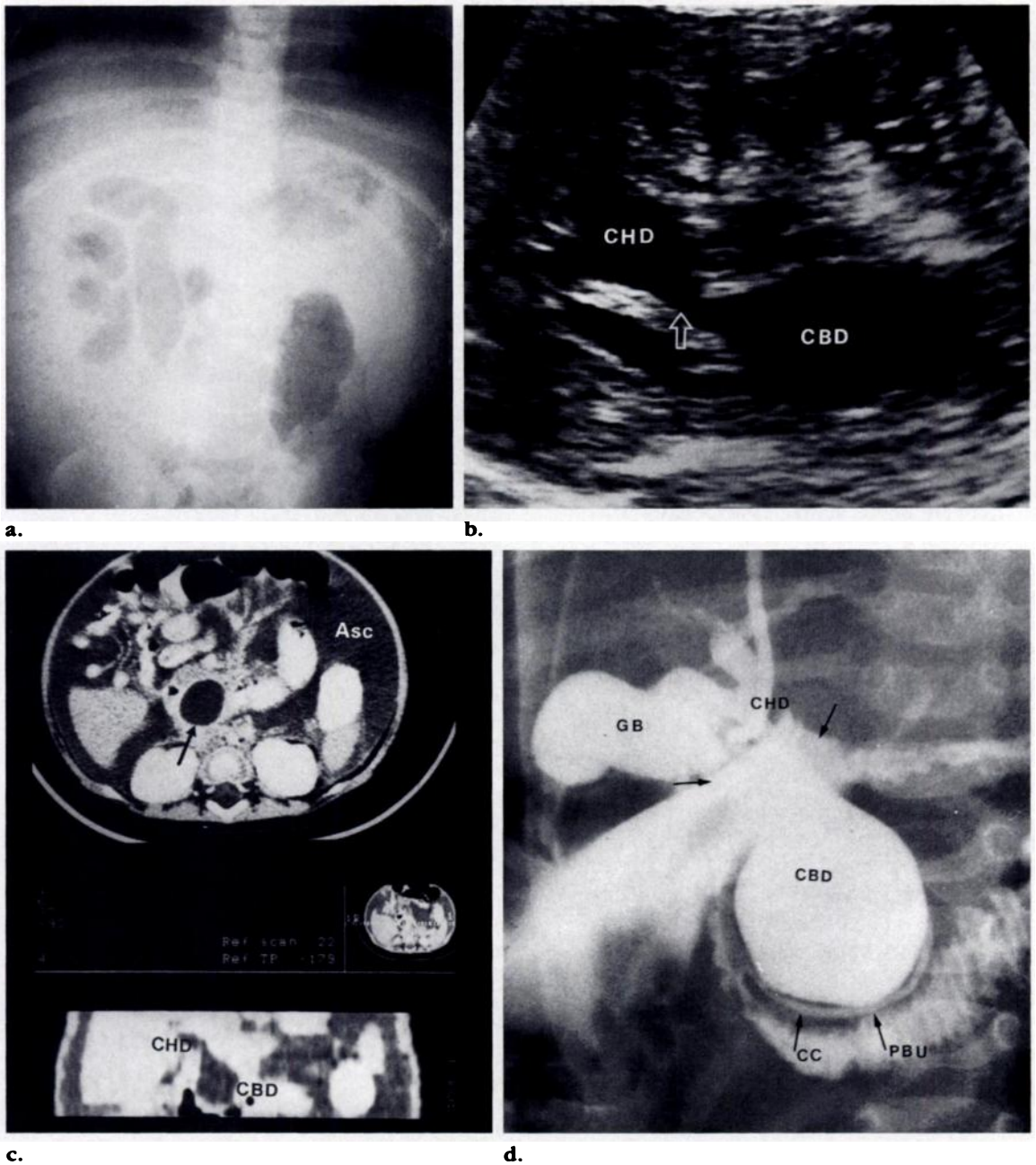



**Figure 18.** Rupture of a choledochal cyst in a 7-month-old female infant with a 7-day history of progressive abdominal distention. **(a)** Supine abdominal radiograph shows a distended abdomen with increased haziness and a floating appearance of the bowel, which are compatible with the presence of ascites. **(b)** Oblique sonogram obtained at the porta hepatis shows a dilated common hepatic duct (*CHD*) and common bile duct (*CBD*) with a figure eight appearance. Note the stenosis (arrow) at the junction of the ducts. **(c)** Top: CT scan shows ascites (*Asc*) and dilatation of the distal part of the common bile duct (arrow). Bottom: Coronal reconstruction image shows bile duct dilatation with an appearance similar to that on the sonogram **(b)**. *CBD* = common bile duct, *CHD* = common hepatic duct. **(d)** Cholangiogram obtained during surgery performed immediately after abdominal tapping demonstrated bilious ascites shows spillage of contrast medium (top two arrows) from the junction of the common hepatic duct (*CHD*) and common bile duct (*CBD*). The presence of a choledochal cyst that had spontaneously ruptured was confirmed. Cystic dilatation of the common bile duct and a right-angled pancreaticobiliary ductal union (*PBU*) are seen. The common channel (*CC*) is long and stenotic. *GB* = gallbladder.

The association of pancreatitis with choledochal cysts, although infrequent, is well recognized (9,10,12,16,18). The pathophysiology of pancreatitis is related to the anomalous pancreaticobiliary ductal union with a long common channel and ductal stones. In such cases, obstruction of the pancreatic duct or the common channel by a stone or reflux of bile into the pancreatic duct through the dilated common channel can precipitate pancreatitis (5,12,16) (Fig 19). Other rare complications are cholangitis, which is occasionally associated with pregnancy and development of liver abscesses (2,3,39) (Fig 15). Some patients have portal hypertension due to compression of the portal vein by a cyst or secondary to biliary cirrhosis (13). These patients may experience abdominal distention due to ascites (Fig 20).

There are many anomalies associated with choledochal cysts. In neonates and young infants, a choledochal cyst may be associated with biliary atresia (40,41); when distal biliary atresia is present, the patient typically has obstructive jaundice and acholic stools. Combined use of sonography, which allows identification of the choledochal cyst, and hepatobiliary scintigraphy, which allows determination of the distal obstruction, may suggest the diagnosis of a choledochal cyst with biliary atresia. In such cases, sonography shows a cystic structure at the porta hepatis that is separate from the gallbladder. Nonvisualization of radionuclide activity in the bowel during delayed hepatobiliary scintigraphy is consistent with distal biliary atresia (Fig 21). Associated intrahepatic biliary atresia is rare; however, intraoperative cholangiography should be performed to check for intrahepatic atresia because there is a specific treatment for this entity (the Kasai procedure)

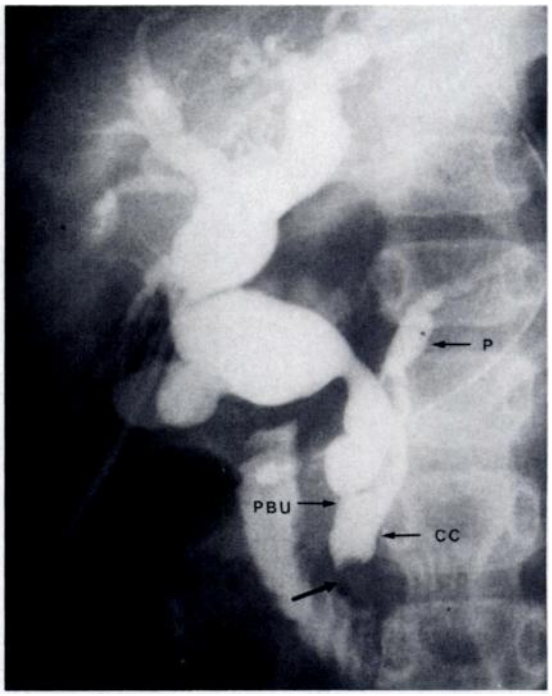
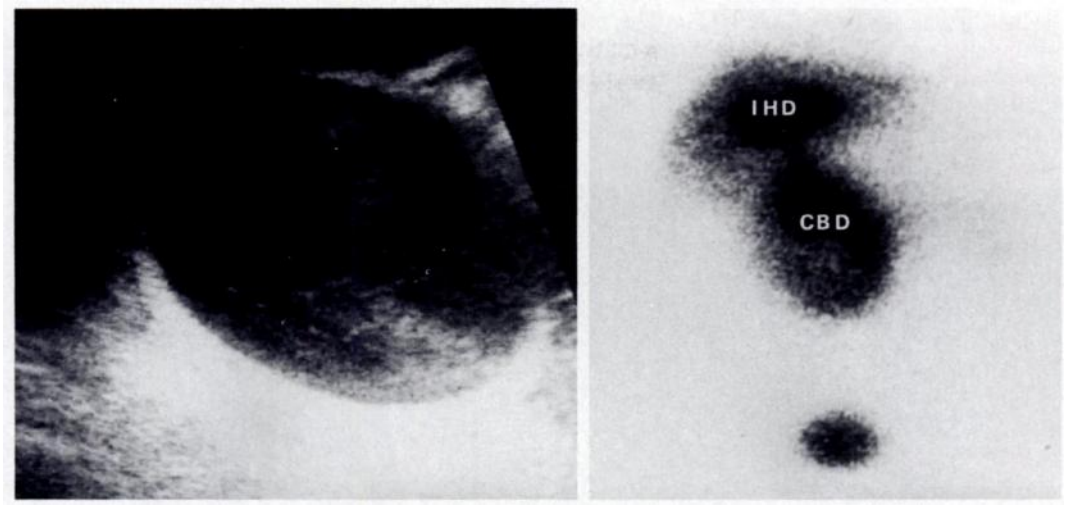
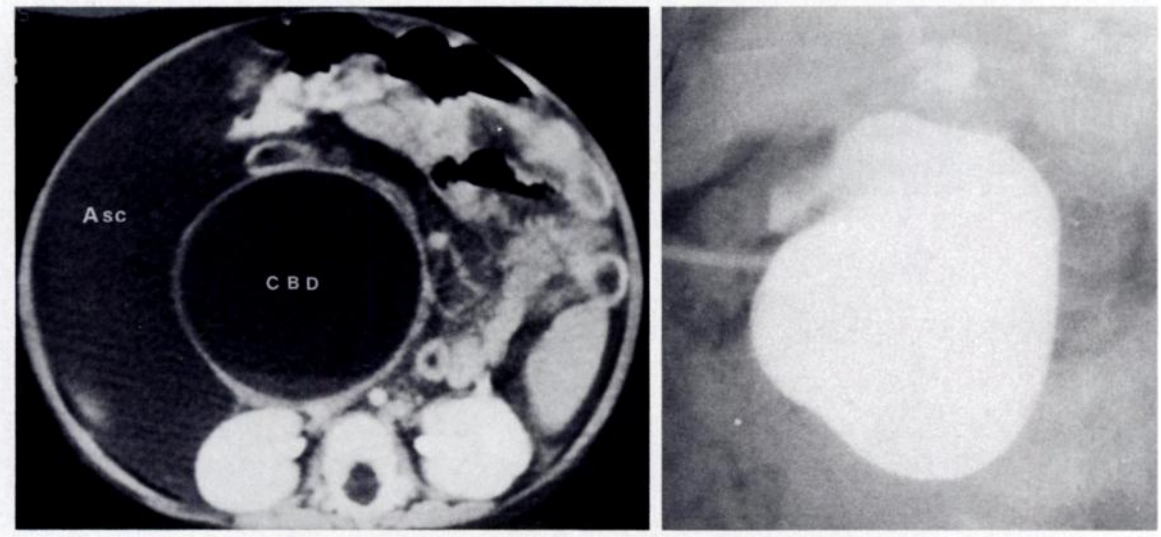



**Figure 19.** Choledochal cyst complicated by pancreatitis in a 10-year-old boy with recurrent cramping abdominal pain. Intraoperative cholangiogram shows fusiform dilatation of the intra- and extrahepatic bile ducts. The pancreaticobiliary ductal union (*PBU*) is acute angled. The common channel (*CC*) is ectatic and obstructed by a stone (arrow), resulting in dilatation of the pancreatic duct (*P*).



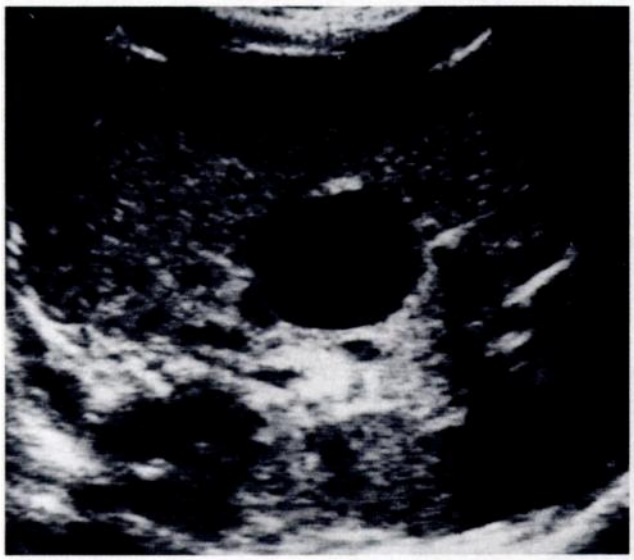

a. b.



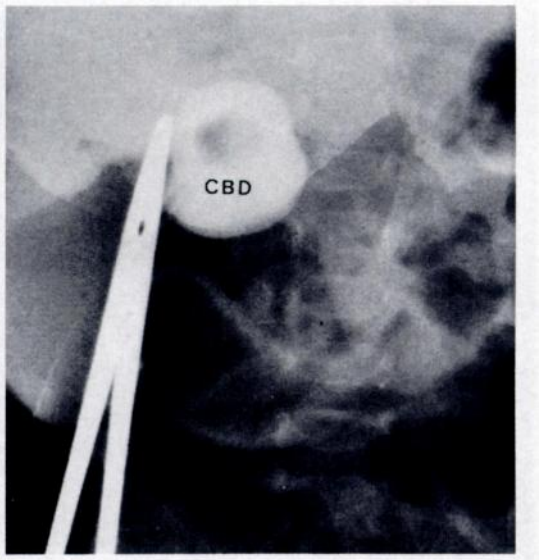

c. d.

**Figure 20.** Choledochal cyst in a 6-month-old female infant with abdominal distention. **(a)** Oblique sonogram obtained at a subhepatic level shows a huge cyst. **(b)** Hepatobiliary scintigram shows accumulation of radionuclide in the dilated intrahepatic ducts (*IHD*) and common bile duct (*CBD*). **(c)** Contrast-enhanced abdominal CT scan shows a large amount of ascites (*Asc*) and cystic dilatation of the common bile duct (*CBD*). **(d)** Intraoperative cholangiogram shows a choledochal cyst. No evidence of perforation of the cyst was noted. The cause of the ascites was hepatic dysfunction due to liver cirrhosis.

**Figure 21.** Choledochal cyst associated with intrahepatic and extrahepatic biliary atresia in a 1-month-old male infant with persistent jaundice. **(a)** Transverse sonogram obtained at the porta hepatis shows a round cyst. **(b)** Serial hepatobiliary scintigrams show no excretion of radionuclide into the biliary tree and no bowel activity. *H* = hours after injection, *M* = minutes after injection. **(c)** Intraoperative cholangiogram shows cystic dilatation of the common bile duct (*CBD*) with distal atresia. No filling with contrast medium is seen proximal to the dilated common bile duct.



a.

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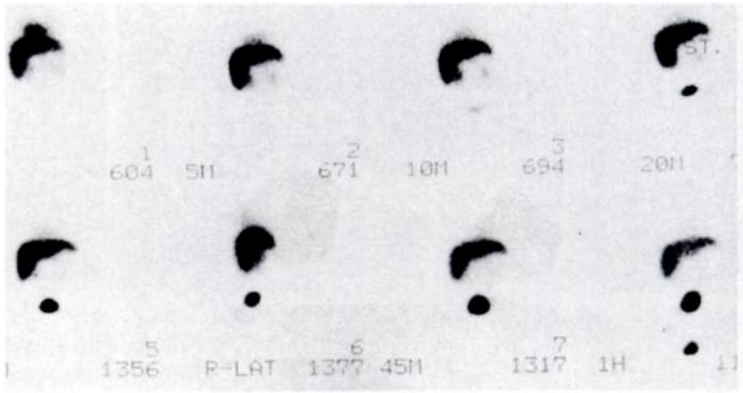


b.

(41). Congenital hepatic fibrosis may be associated with a type V cyst (Caroli disease) (26,29).

## ■ TREATMENT

Choledochal cysts are treated surgically. Although there is some controversy as to which procedure or type of anastomosis produces the best results, many investigators recommend excision of the cyst and Roux-en-Y hepaticojejunostomy as the best treatment for a choledochal cyst (2,3,38,39,42). The cyst should be completely resected to prevent ascending cholangitis, cystolithiasis, and malignant changes. Roux-en-Y hepaticojejunostomy is the procedure of choice for reconstruction of the common bile duct in cases of anomalies of the pancreaticobiliary ductal union (3).

## ■ SUMMARY

Diagnosis of choledochal cysts is not always straightforward. The classic signs and symptoms of abdominal pain, jaundice, and an abdominal mass occur in less than one-third of cases. The shape of the cyst may influence the clinical manifestations: cystic dilatation is associated with an abdominal mass or jaundice, whereas fusiform dilatation is associated with abdominal pain. The theory that choledochal cysts are related to an anomalous pancreaticobiliary ductal union is supported by cholangiopancreatographic findings. The type of anomalous pancreaticobiliary ductal union may

affect the shape of the cyst: a right-angled pancreaticobiliary ductal union is associated with cystic dilatation of the common bile duct, whereas an acute-angled union is associated with fusiform dilatation of the duct. The shape of the common channel can affect the clinical outcome; patients with a long, ectatic common channel are prone to develop pancreatitis.

US allows confirmation of the diagnosis of a choledochal cyst; however, the extent of intrahepatic involvement is more accurately demonstrated with CT. Diagnosis of choledochal cysts is facilitated with hepatobiliary scintigraphy, which shows whether a cystic structure communicates with the biliary system. In neonates and young infants, biliary atresia may be associated with a choledochal cyst. Absence of radionuclide activity in the bowel on hepatobiliary scintigrams and the presence of a cystic mass in the porta hepatis on sonograms suggest obstruction of the distal part of the common bile duct. Cholangiopancreatography is required in such cases to confirm the presence of a choledochal cyst, as well as to demonstrate the anatomy of the dilated biliary tree, the anomalous pancreaticobiliary ductal union, and the accessory pancreatic duct for correct operative planning. Accurate demonstration of an anomalous pancreaticobiliary ductal union is achieved with ERCP and PTC, but ERCP is the more reliable method because it is less invasive and consistently demonstrates the pancreatic duct by means of direct retrograde cannulation.

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