Abdominal Imaging

Published online: 15 February 2015

Abdom Imaging (2015) 40:1567–1580
DOI: 10.1007/s00261-015-0381-4

Imaging of choledochal cysts

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Abstract

Choledochal cysts are rare cystic dilatations of the intrahepatic and/or extrahepatic biliary tree, which may be mistaken for other cystic lesions if their characteristic features are not recognized. The etiology is unknown, and likely multifactorial, and it is uncertain whether they are congenital or acquired. Multiple imaging modalities can be used to diagnose choledochal cysts, including ultrasound, computed tomography, magnetic resonance (MR) cholangiopancreatography (MRCP), endoscopic retrograde cholangiopancreatography, and percutaneous transhepatic cholangiography. MRCP has replaced the more invasive techniques as the gold standard of diagnosis. In addition, MRCP is helpful in detecting an abnormal pancreaticobiliary junction, which is seen in the majority of choledochal cysts. Reaching a correct diagnosis is essential, given the associated risk of complications, including cholangitis, biliary strictures, stones, and malignancy, and accurately assessing the location and length of involvement is important for surgical planning. This review aims to familiarize radiologists with the different types of choledochal cysts and their imaging features according to the Todani classification.

Key words: Choledochal cyst—Todani classification—Imaging—MRCP

Choledochal cysts are rare congenital cystic dilatations of the intrahepatic and/or extrahepatic biliary tree. The diagnosis is made when other causes of biliary ductal dilatation are excluded, although no specific cut-off value for ductal dilatation has been reported [1]. Choledochal cysts are relatively uncommon in Western populations with a reported incidence of 1 in 100,000–150,000 live births [1–3]. Up to two-thirds of reported cases are in Asian countries where they are considerably more common, with incidence as high as 1 in 1000 in Japan [4]. Choledochal cysts occur more frequently in women [5, 6], leading to the classic notion that biliary cystic disease is a disease of female children, which presents with a clinical triad of jaundice, abdominal pain, and a palpable mass. However, nowadays this entity is being initially diagnosed in adults with increasing frequency, which in part may be due to the increased use of diagnostic imaging [2, 5, 7]. The classic clinical triad is less common in adult patients, who more often present with abdominal pain alone [3, 5] or nonspecific clinical symptoms [1]. It is important for radiologists to correctly diagnose choledochal cysts, given the risk of complications, including cholangitis, biliary strictures, stones, and malignancy. Accurate assessment of the location and length of involvement is also important to help plan the surgical procedure.

Etiology

The etiology and pathogenesis of choledochal cysts are unknown. The etiology is likely multifactorial and multiple processes have been hypothesized to contribute to the development of biliary cystic disease.

One theory is that choledochal cysts arise from a congenital malformation of the ductal plate [8]. This theory is related to the pathogenesis of Caroli disease in which ductal plate malformation results in intrahepatic biliary ductal dilatation.

A second hypothesis, which is the most common one, is that choledochal cysts are an acquired condition thought to arise from an abnormal pancreaticobiliary junction (APBJ). This is seen in 57% to 96% of choledochal cysts, but may also occur without biliary ductal dilatation [9, 10]. APBJ is defined as the union of the distal common bile duct and pancreatic duct outside the duodenal wall [11] (Fig. 1). Prior studies have suggested that the common channel length must be greater than

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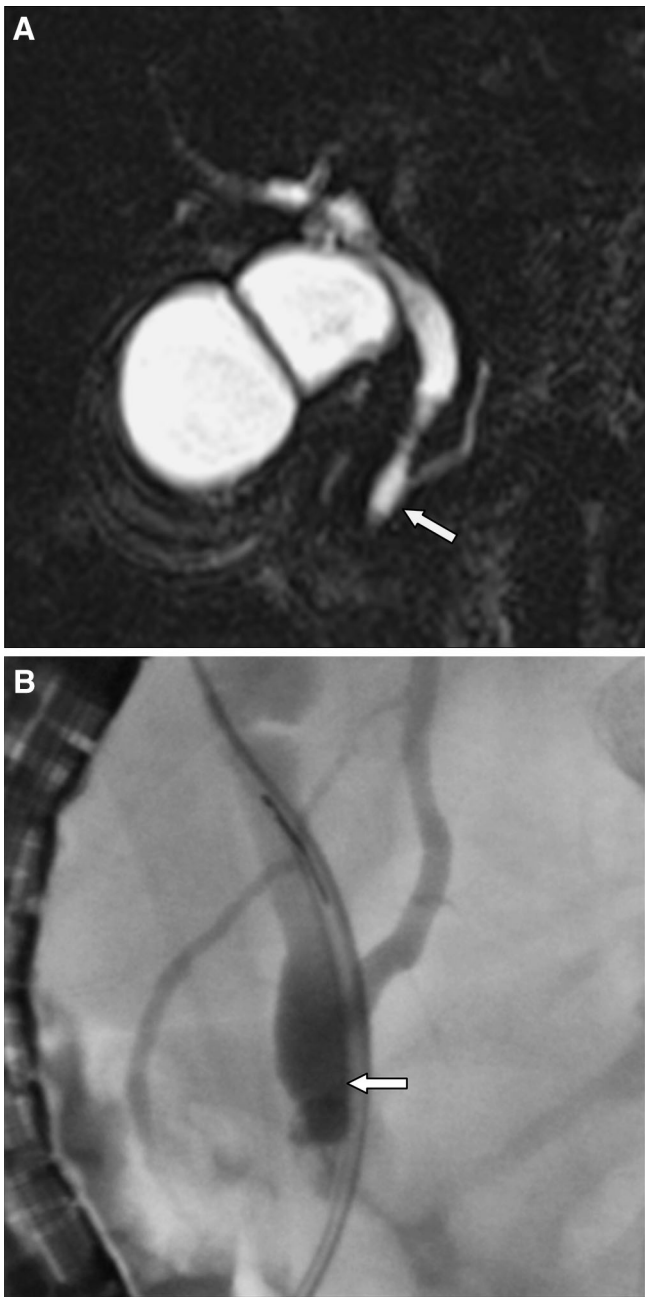


Fig. 1. Anomalous pancreaticobiliary junction (APBJ) in a 51-year-old woman. Coronal MIP reformatted 3D T2 MRCP (**A**) and ERCP (**B**) images show an extraduodenal insertion of the pancreatic duct into the common bile duct and a long common channel leading to the ampulla (*arrows*).

1.5 cm to be diagnosed as APBJ [12]. A normal pancreaticobiliary junction usually has an acute angle between the common bile duct and the pancreatic duct [1] and is located within the duodenal wall, where the sphincter of Oddi protects the biliary tree from reflux of pancreatic enzymes and bile [13]. APBJ is thought to be the result of embryologic failure of migration of the choledochopancreatic junction into the duodenal wall, causing formation of a common channel between the pancreatic and bile ducts. A common channel serves as the site for mixing of pancreatic enzymes and bile, which leads to chronic inflammation, weakening of the bile duct wall, and eventual biliary ductal dilatation [14]. In support of this theory, it has been found that higher levels of amylase in the bile duct of patients with choledochal cysts are associated with a younger age of onset of symptoms and higher grade of dysplasia at the time of diagnosis [14]. Other enzymes, such as phospholipase A2, trysinogen/trypsin, as well as byproducts of phospholipase activation such as lysolecithins, have also been detected in the bile ducts of patients with choledochal cysts [15, 16]. Higher pressures in the sphincter of Oddi have been documented in APBJ, which may promote reflux of pancreatic secretions. Chronic inflammation of the biliary tree from APBJ predisposes to stones and malignancy, including cholangiocarcinoma and gallbladder cancer [13]. One must note that APBJ is associated with increased risk of cholangitis and pancreatitis [17].

In addition, primary strictures of the common bile duct may also play a role in the development of choledochal cysts. The location, severity, and length of the stricture determine the type of choledochal cyst [18]. High-grade or long strictures cause cystic changes, while low-grade or short strictures cause a fusiform dilatation. These are most common with types IC and IVA choledochal cysts. Detection of these strictures pre-operatively is important because treating the choledochal cyst without addressing the stricture may lead to recurrent episodes of cholangitis. This mechanism has been underestimated, and is now believed to be more significant in the pathophysiology of choledochal cysts [18].

Imaging techniques

Multiple imaging modalities are used to evaluate choledochal cysts. Ultrasound is a good screening tool for evaluating biliary ductal dilatation, especially in children [19]. Computed tomography (CT) is widely used to assess for other causes of biliary ductal dilatation. However, for optimal evaluation of the biliary anatomy, ultrasound and CT are not enough, and cholangiography is necessary. Magnetic resonance (MR) cholangiopancreatography (MRCP) is considered the current gold standard for initial evaluation and diagnosis of choledochal cysts. MRCP techniques are able to accurately assess intra- and extrahepatic biliary anatomy, evaluate the pancreaticobiliary junction, and look for associated complications [20–22]. Endoscopic retrograde cholangiopancreatography (ERCP) and percutaneous transhepatic cholangiography (PTC) are invasive forms of cholangiography that may still be required to confirm diagnosis and demonstrate the APBJ.

Hepatobiliary MR contrast agents allow for better visualization of the bile ducts than heavily T2-weighted

MRCP sequences because of their elimination through the biliary system, and can be used for contrast-enhanced MRCP in difficult cases to evaluate for communication between a cystic lesion and the biliary tree (Fig. 2). Currently used agents are gadobenate dimeglumine (Gd-BOPTA, MultiHance, Bracco Diagnostics, Princeton, NJ) and gadoxetate disodium (Gd-EOB-DTPA, Eovist/Primovist, Bayer Healthcare, Leverkusen, Germany).

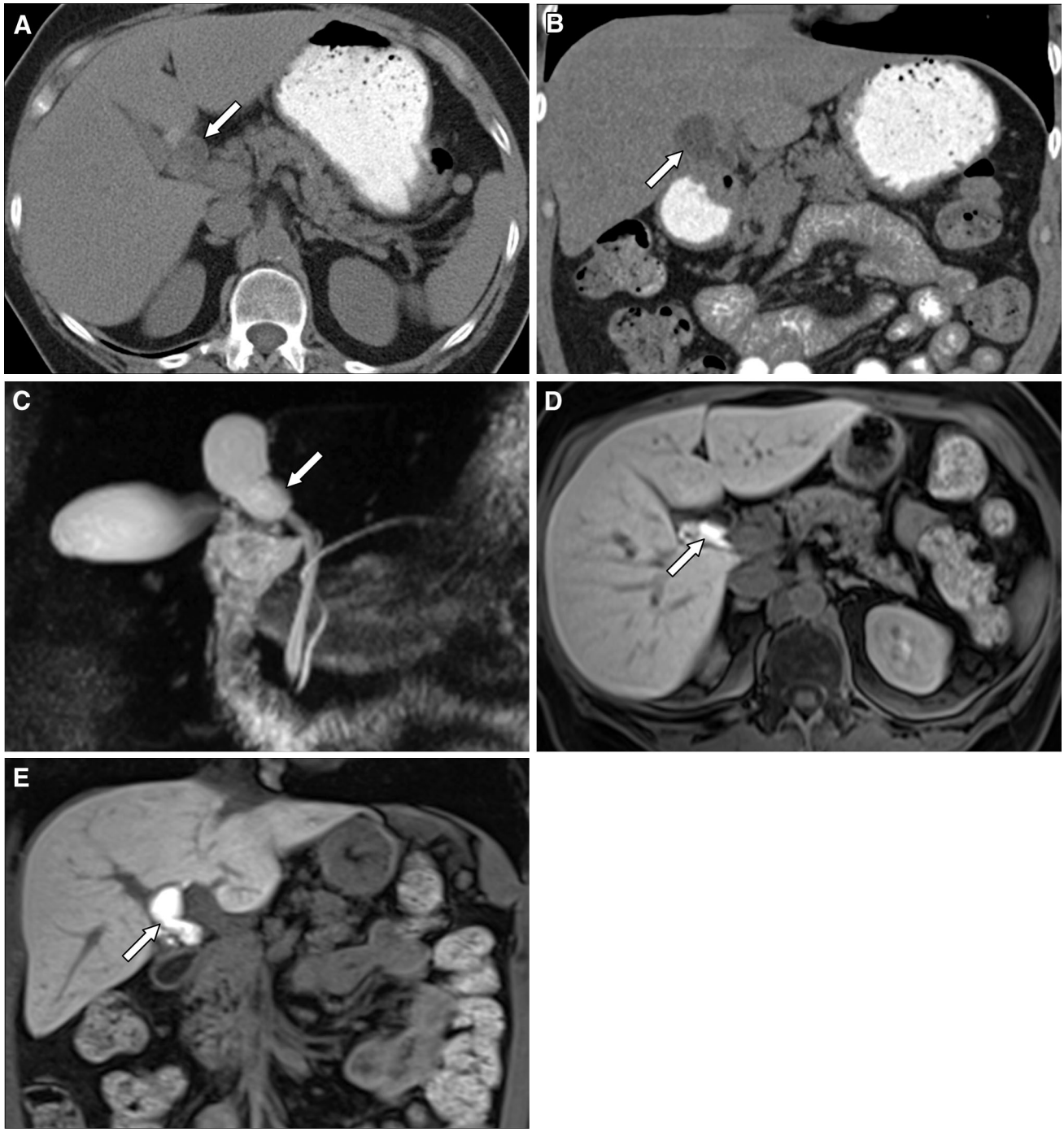


Fig. 2. Imaging techniques used to evaluate a cystic hilar lesion in a 72-year-old woman. Unenhanced axial (**A**) and coronal (**B**) CT images of the abdomen show a cystic lesion in the hilar region adjacent to the biliary tree without definitive communication (*arrows*). Coronal MIP reformatted 3D T2 MRCP (**C**) fails to show definite communication between the cystic lesion and the biliary tree. Contrast-enhanced axial (**D**) and coronal (**E**) MR images after Gd-EOB-DTPA administration during the hepatobiliary phase were helpful to show excretion into the biliary system and filling of the cystic lesion with contrast (*arrows*), proving direct communication between the cystic lesion and the biliary tree, consistent with a type II choledochal cyst.

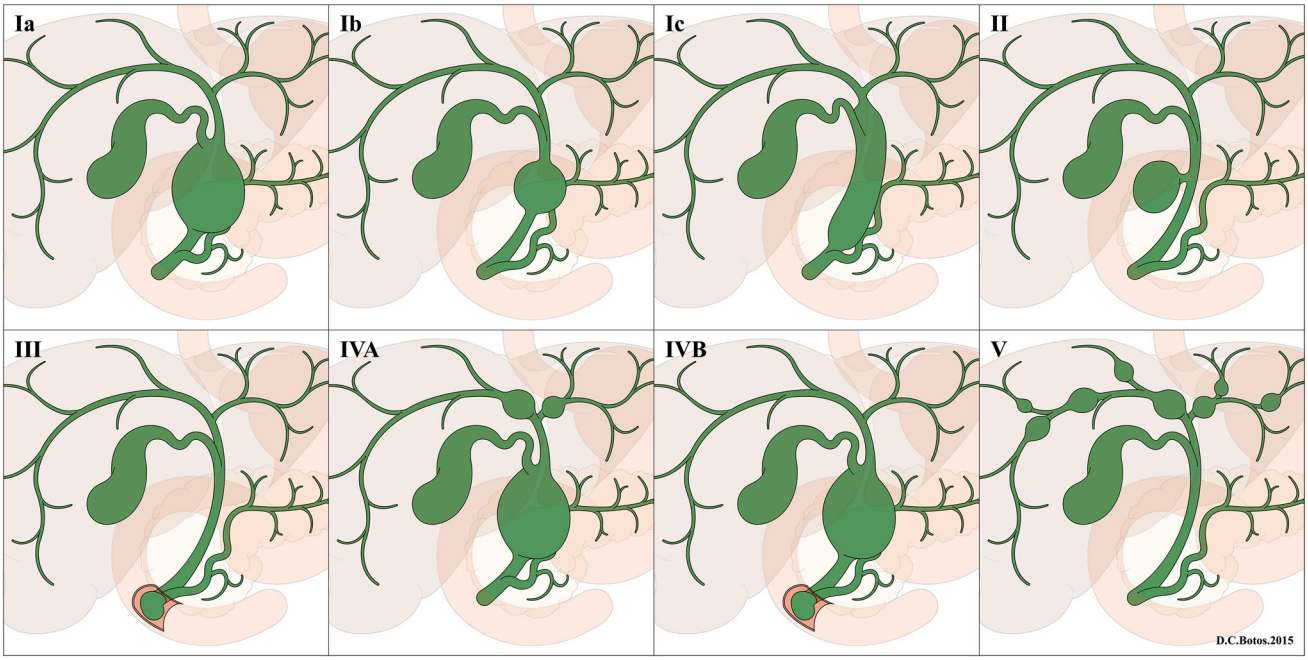


Fig. 3. Types of choledochal cysts according to the revised Todani classification.

They differ in the percentage of biliary elimination (5% for Gd-BOPTA and 50% for Gd-EOB-DTPA) and in the timing of the hepatobiliary phase (60–120 min for Gd-BOPTA and 10–60 min for Gd-EOB-DTPA) [23]. Optimal timing for hepatobiliary phase contrast-enhanced MRI is therefore 15 min after Gd-EOB-DTPA administration and 90 min after Gd-BOPTA administration [24]. However, in patients with liver impairment, such as secondary to cirrhosis or biliary obstruction, these agents may be problematic because of limited biliary excretion [25]. Performing contrast-enhanced MRCP may require modifications of the scan protocol, because hepatobiliary agents cause T2 shortening, and therefore conventional T2-weighted MRCP sequences must be acquired prior to administering the contrast agent.

Classification of choledochal cysts

The first classification scheme for choledochal cysts was proposed by Alonso-Lej in 1959. Alonso-Lej classified choledochal cysts into types I through III based on types of extrahepatic cysts [26]. In 1977, Todani et al. broadened the original classification system to include presentations involving intrahepatic biliary duct cysts [27], and have since revised the classification to account for the presence of APBJ [18, 28] (Fig. 3). Apart from types I–V included in the revised Todani classification, isolated cystic dilatation of the cystic duct has been described and suggested as type VI [29].

Type I choledochal cyst

Type I choledochal cysts make up the majority, approximately 50–90%, of choledochal cysts. They are characterized by fusiform or saccular dilatation of the extrahepatic bile ducts (Fig. 4). There are multiple subtypes: IA is cystic dilatation with associated APBJ; IB is segmental dilatation without APBJ; and IC is diffuse, fusiform dilatation with associated APBJ, and often also a low-grade stricture in the distal common bile duct [18]. Unlike biliary obstruction, type I choledochal cysts are generally not associated with significant intrahepatic biliary dilatation. The duct above and below the choledochal cyst is not dilated. The exception is type IC, which may extend continuously to the common hepatic duct or intrahepatic ducts. This extension into the intrahepatic ducts may be the result of elevated pressures in the choledochus, and may resolve soon after excision of the choledochal cyst [18]. Therefore, in these circumstances, differentiating between type IC and biliary obstruction may be difficult.

Type I choledochal cysts are surgically managed with complete cyst excision and Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy reconstruction [7, 9, 30].

Type II choledochal cyst (diverticulum)

Type II choledochal cysts are rare, comprising only 2–3% of all choledochal cysts. They manifest as an extrahepatic

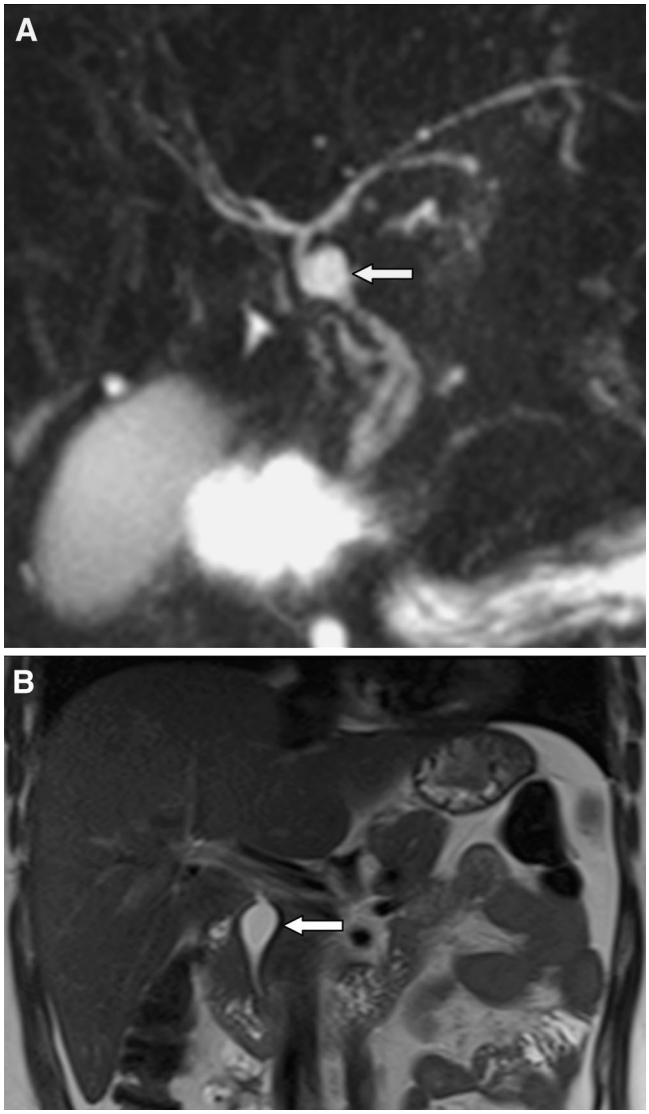


Fig. 4. Type I choledochal cyst (*arrows*). (**A**) Coronal MIP reformatted 3D T2 MRCP in a 65-year-old woman with a type IA choledochal cyst showing cystic dilatation of the common hepatic duct. (**B**) Coronal T2-weighted MR image in a 40-year-old woman with a type IB choledochal cyst demonstrates a segmental dilatation of the common bile duct.

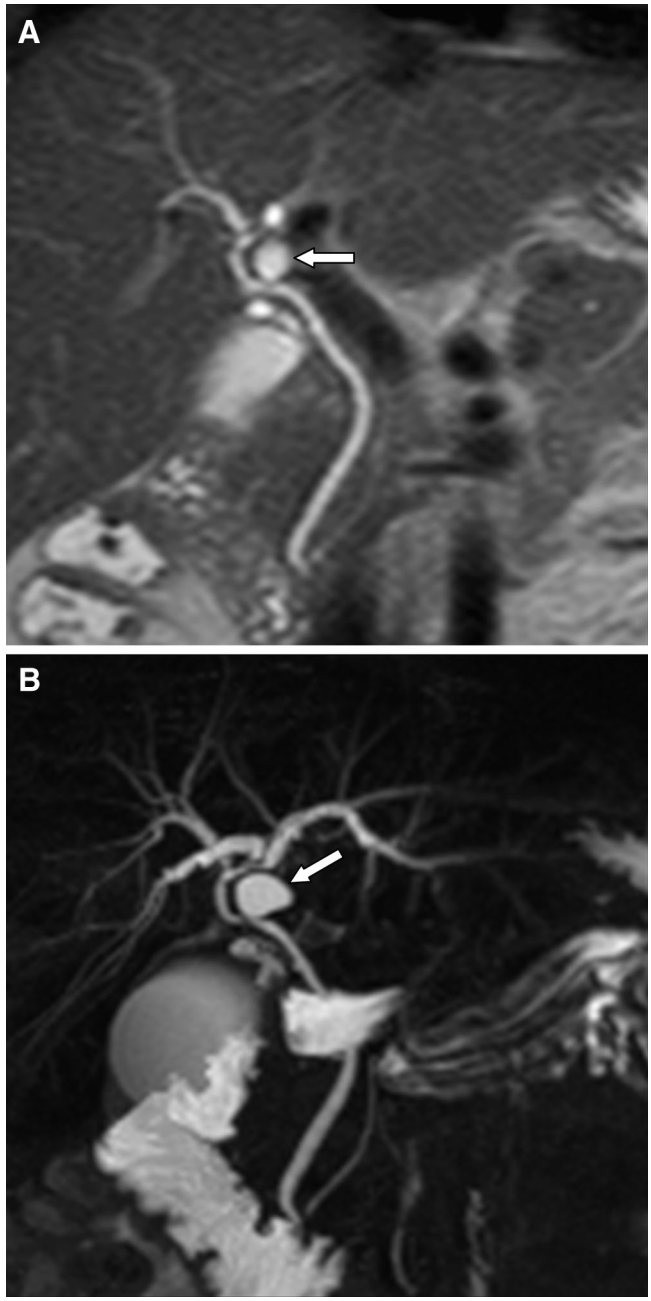


Fig. 5. Type II choledochal cyst in a 23-year-old woman. (**A**) Coronal T2-weighted and (**B**) coronal MIP reformatted 3D T2 MR images of the biliary tree show a diverticular outpouching from the common hepatic duct, consistent with a type II choledochal cyst.

biliary diverticulum without APBJ (Fig. 5). These can be seen in any portion of the extrahepatic bile ducts [27].

Surgical management includes complete cyst excision with Roux-en-Y hepaticojejunostomy or primary closure over a T-tube [9].

Type III choledochal cyst (choledochocele)

Type III choledochal cysts (or choledochoceles) are also uncommon, making up 1–5% of all choledochal cysts. They are characterized by dilatation of the intraduodenal part of the common bile duct, and are not associated with APBJ (Fig. 6). It has been suggested that choledochocele should be classified independently of the revised Todani classification scheme given differences in presentation and demographics [31]. Ziegler et al. reported that choledochoceles occur more frequently in older male patients presenting with acute pancreatitis [31].

Surgical management may be less invasive given the lower risk of malignancy, and may include ERCP with

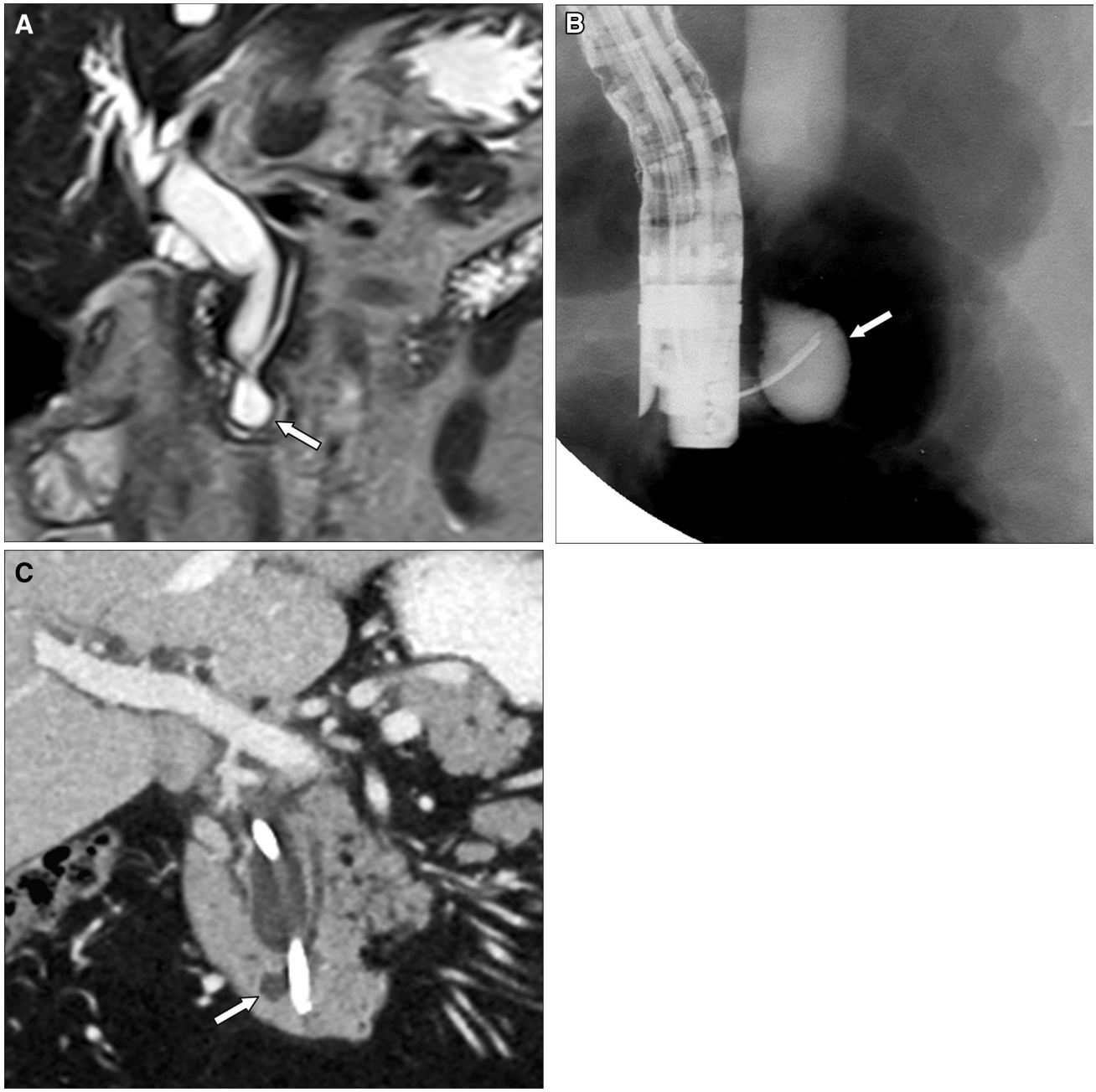


Fig. 6. Type III choledochal cyst in a 69-year-old woman who presented with biliary obstruction and intraperitoneal abscesses. Coronal T2-weighted MRI (**A**) and ERCP (**B**) images show a bulging of the common bile duct into the duodenum in the ampullary region from choledochocele (*arrows*), with a dilated common bile duct. Subsequent endoscopic biliary drainage was performed, evident on the coronal CT image (**C**).

sphincterotomy in cysts less than 2 cm or cyst excision with sphincteroplasty [7, 9].

Type IV choledochal cyst (multiple intrahepatic and extrahepatic choledochal cysts)

Type IV choledochal cysts are the second most common type, making up 30–40% of all choledochal cysts, the majority of which are type IVA. They are characterized by intra- and extrahepatic fusiform or cystic dilatation of the bile ducts. Type IVA choledochal cysts have intra- and extrahepatic biliary ductal dilatation, and are usually associated with APBJ (Figs. 7, 8). It can be further classified based on morphology of the extrahepatic and intrahepatic ducts into cystic–cystic, cystic–fusiform, and fusiform–fusiform dilatation [18]. Type IVA cysts may be associated with a stricture at the level of the hilum, which

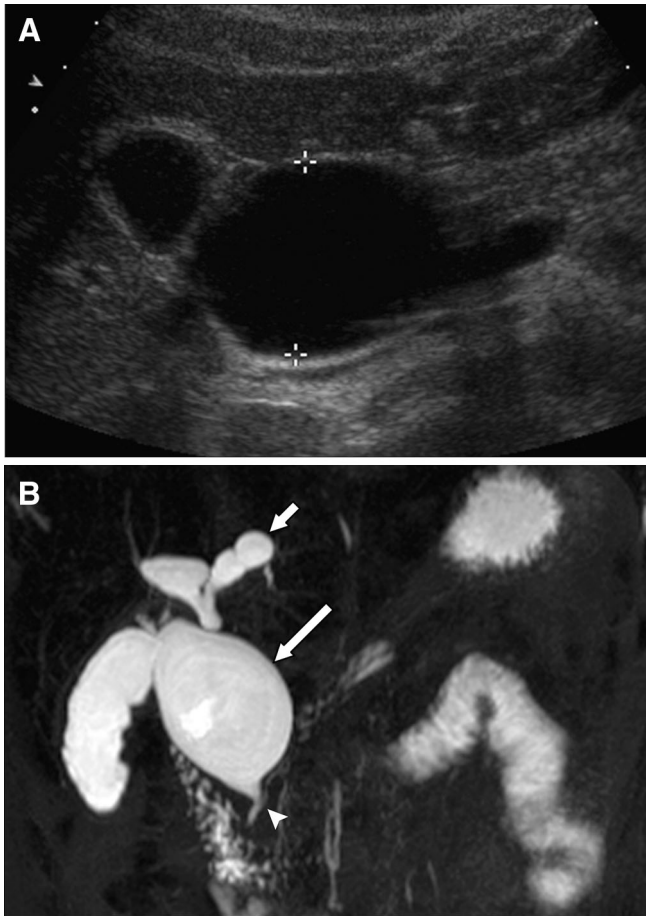


Fig. 7. Type IVA choledochal cyst in a 25-year-old woman with nausea and vomiting. (**A**) Longitudinal sonography and (**B**) coronal MIP reformatted 3D MRCP images demonstrate saccular intra- and extrahepatic biliary ductal dilatation (*arrows*). Also note the APBJ (*arrowhead*), typically seen in type IVA cysts.

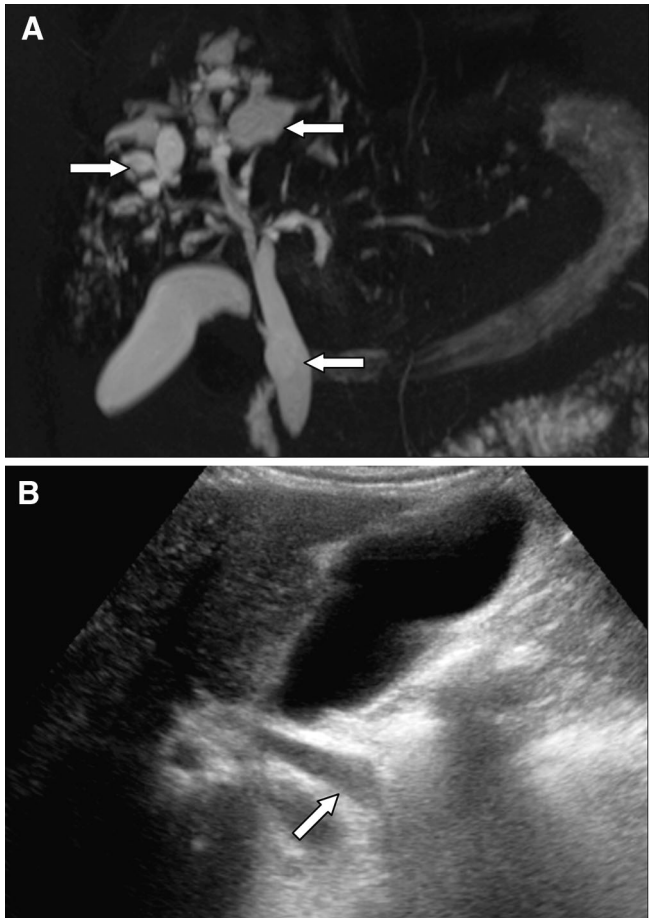


Fig. 8. Type IVA choledochal cyst in a 40-year-old man with abnormal liver function tests. (**A**) Coronal MIP reformatted 3D T2 MRCP and (**B**) transverse sonography image of the abdomen show multiple dilated intra- and extrahepatic biliary ducts (*arrows*). The patient required orthotopic liver transplantation 6 months after presentation.

may lead to its characteristic intrahepatic dilatation [18]. Looking for this hilar stricture may help differentiate fusiform type IVA cysts from type IC cysts, which may involve the intrahepatic ducts as well, only in a continuous fashion [18]. Type IVB choledochal cysts are rare, and appear as multiple extrahepatic cysts.

Surgical management should include complete extrahepatic cystic excision with biliary-enteric anastomosis [32], because of the potential for development of malignancy, however complete resection is not always possible. If only the left hepatic lobe is involved, left hepatic lobectomy should also be performed. If there is diffuse intrahepatic disease, intrahepatic cyst drainage with bilateral stents is recommended for alleviation of biliary stasis and associated complications [9].

Type V choledochal cyst (Caroli disease)

Type V choledochal cysts, otherwise known as Caroli disease, are characterized by multifocal segmental intrahepatic biliary ductal dilatation (Fig. 9). Caroli disease is uncommon, accounting for less than 10% of cases. Unlike the other types, this is a known congenital ductal plate malformation, affecting women more than men, with presumed autosomal recessive inheritance [33]. Patients often present in adolescence or early adulthood with recurrent cholangitis, abdominal pain, or jaundice. However, they may present later with the sequelae of portal hypertension and cirrhosis. Many complications are due to bile stasis, including recurrent cholangitis, hepatic abscesses, stones, and intrahepatic biliary stric-

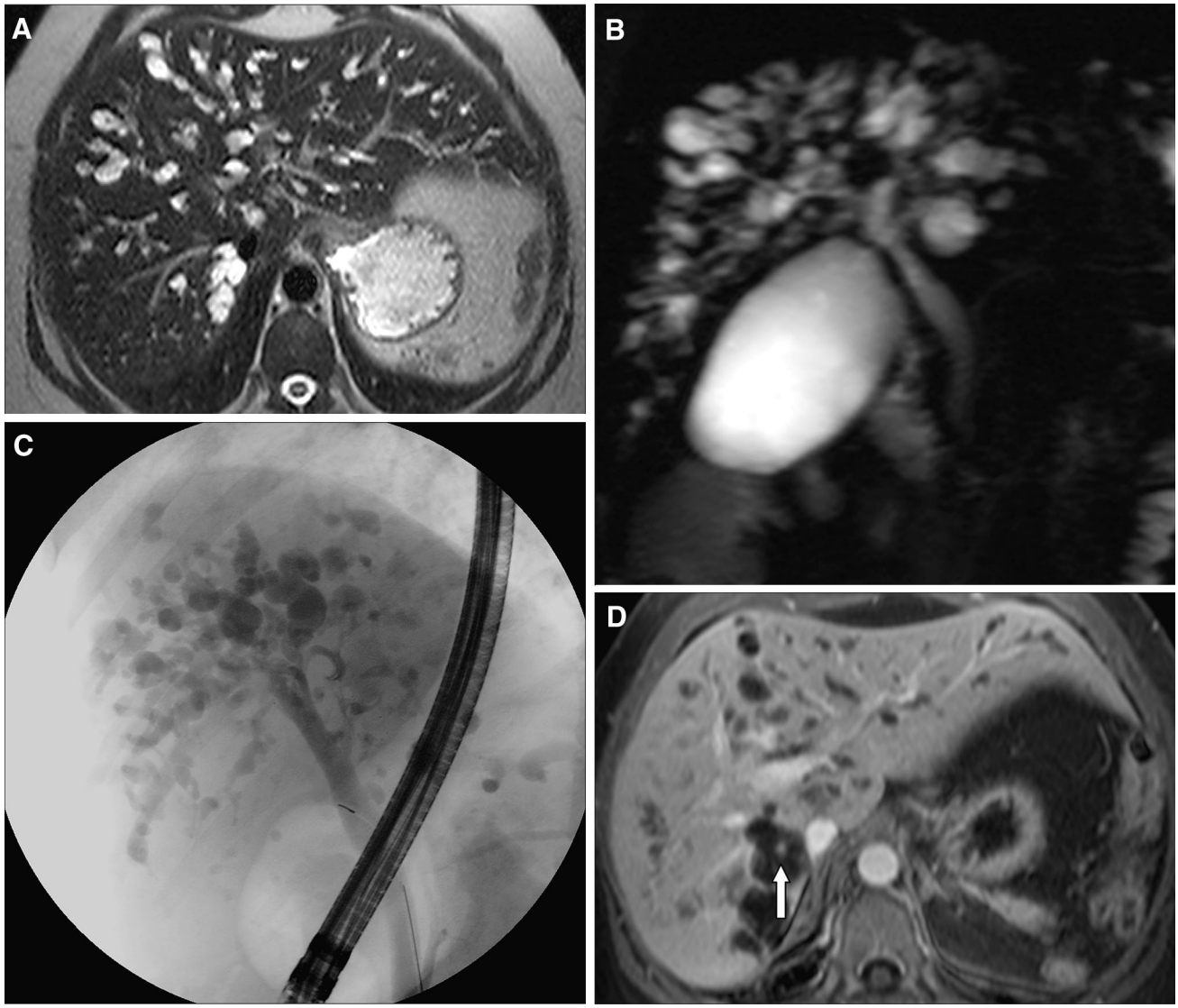


Fig. 9. Type V choledochal cyst in a 37-year-old woman. (**A**) Axial T2-weighted MR image of the abdomen shows cystic dilatation of the intrahepatic bile duct, consistent with Caroli disease. (**B**) Coronal thick-slab T2 MRCP, and (**C**) ERCP images of the abdomen better show the communication with the biliary tree. Axial contrast-enhanced T1-weighted fat saturated MR image (**D**) shows the classic "central dot" sign, evident as an enhancing portal vein within a dilated intrahepatic duct (*arrow*).

tures. However, some patients eventually develop hepatic fibrosis or even malignancy [34]. When congenital hepatic fibrosis is also present, as is the case in 50% of patients [7], some refer to this entity as Caroli syndrome [35] (Fig. 10). Often, Caroli disease is associated with renal abnormalities, such as medullary sponge kidney, autosomal dominant polycystic kidney disease (Fig. 11), and medullary cystic disease [36, 37]. Caroli disease is characterized by saccular or fusiform dilatation of the intrahepatic bile ducts, which can be diffuse or focal. Diagnosing Caroli disease on CT can be challenging because the appearance is of multiple low-density lesions in the liver that can be misdiagnosed as hepatic cysts or hamartomas. ERCP and hepatobiliary phase contrast-enhanced MRI confirm communication with the biliary tree, which is the key to the diagnosis [33]. On MRCP, Caroli disease appears as multiple T2 hyperintense cystic dilatations of the intrahepatic bile ducts [38, 39]. A classic, but not specific, imaging feature, seen on CT, MR and sometimes even ultrasound, is the "central dot" sign, seen as an enhancing dot that represents the portal vein radicle and the adjacent hepatic artery branch, surrounded by the dilated bile ducts [12, 40] (Fig. 9). The same structures can sometimes appear as linear bulging

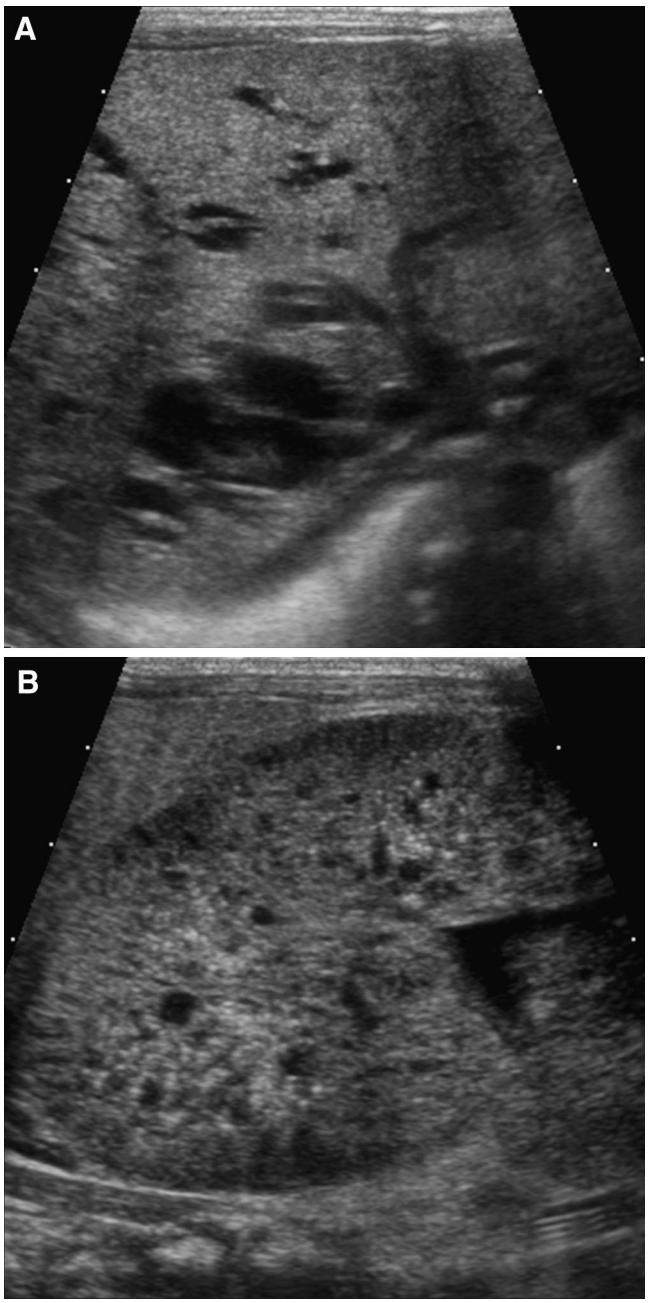


Fig. 10. Caroli syndrome in a full-term infant who initially presented with oligohydramnios and was found to have autosomal recessive polycystic kidney disease (ARPKD) on prenatal ultrasound. (**A**) Transverse sonogram of the liver shows intrahepatic biliary ductal dilatation. (**B**) Transverse sonogram of the right kidney demonstrates a hyperechoic, enlarged kidney with multiple tiny cysts, consistent with ARPKD.

in the wall of saccular bile ducts [33]. An unusual appearance of Caroli disease is focal dilatation of intrahepatic bile ducts, which can be difficult to distinguish from cholangiocarcinoma [41] (Fig. 12).

Management of Caroli disease is difficult when there is diffuse intrahepatic involvement. Initially, a conservative approach is favored with cystic drainage, stone removal, and antibiotics [5]. If there is focal hepatic involvement, a hepatic lobectomy may be performed. Eventually, liver transplantation is the definitive treatment for diffuse hepatic involvement [42].

Type VI choledochal cyst

Isolated cystic dilatation of the cystic duct is rare, with only several case reports describing it [43]. It has been suggested to include it as type VI choledochal cyst, although it is not officially part of the revised Todani classification [29]. When the cyst originates in the cystic duct close to the insertion into the common bile duct, it may be confused with type II choledochal cysts. Close evaluation for communication with the cystic duct will help differentiate them. Although often asymptomatic, patients may present with pain or similar clinical symptoms to choledocholithiasis or Mirizzi syndrome [44, 45].

Surgical treatment includes cholecystectomy with cyst excision [44], usually by laparotomy, although a laparoscopic approach has also been shown to be safe when the cyst does not involve the connection between the cystic duct and the common bile duct [46].

Complications

Choledochal cysts can have multiple complications, often due to biliary stasis, including biliary stones, cholangitis, and pancreatitis. Biliary stones (Fig. 13) will best be seen with MRCP as T2 hypointense filling defects within a dilated T2 hyperintense duct. However, sometimes, the normal T2 hyperintense signal of the dilated bile duct will not be seen because of multiple low signal stones filling the dilated duct [47]. Pigment stones (Fig. 14) may demonstrate intrinsic T1 hyperintense signal. Cirrhosis is a late complication in type IV and V choledochal cysts and results from long-standing biliary stasis.

Choledochal cysts are also associated with an increased risk of biliary malignancy, with a reported incidence of approximately 10–30% that increases with age [1, 6, 48]. However, most studies that evaluated the risk of malignancy of choledochal cysts were performed mainly in symptomatic patients. With widespread use of imaging in recent years, many asymptomatic patients are being diagnosed, and it is possible that the risk of malignancy was overestimated in early studies. Malignancies include cholangiocarcinoma, squamous cell carcinoma, sarcoma and gallbladder cancer [5, 9, 48]. The most common malignancy seen with choledochal cysts is cholangiocarcinoma. Malignancy is most commonly seen with types I and IVA choledochal cysts and

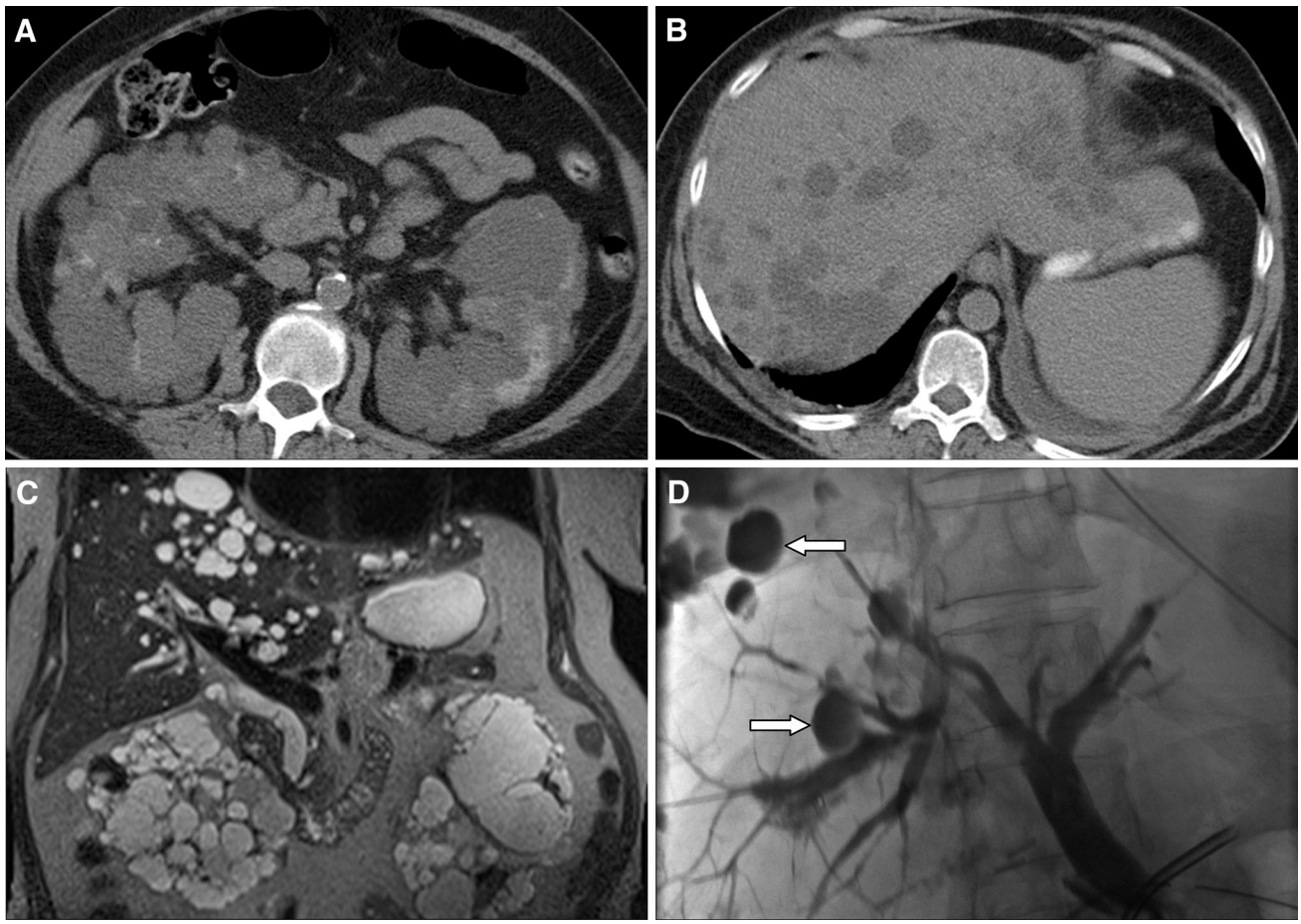


Fig. 11. Caroli disease in a 58-year-old woman with autosomal dominant polycystic kidney disease. (**A**) Unenhanced axial CT image through the kidneys shows enlarged kidneys with multiple cysts, some of which are hyperdense from hemorrhage or proteinaceous debris. (**B**) Unenhanced axial CT image of the liver shows multiple cystic lesions in the liver. (**C**) Coronal T2-weighted MR image demonstrates the multiple cystic lesions in the liver and kidneys. (**D**) Percutaneous transhepatic cholangiographic image shows that the multiple cystic lesions in the liver communicate with the biliary tree (*arrows*), consistent with intrahepatic biliary ductal dilatation from Caroli disease.

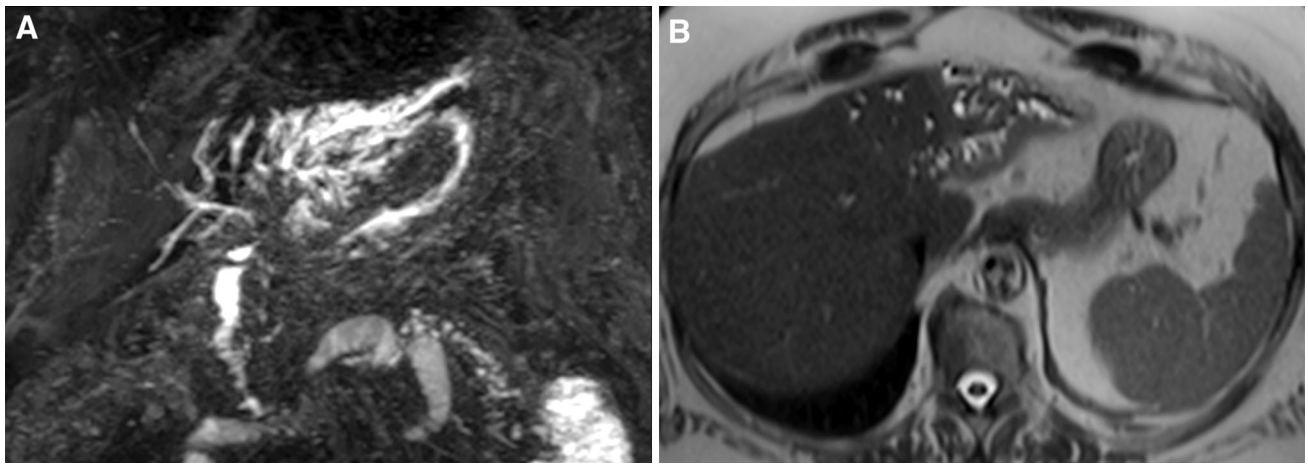


Fig. 12. Focal Caroli disease in a 69-year-old woman. (**A**) Coronal MIP reformatted 3D T2 MRCP and (**B**) axial T2-weighted MR images of the abdomen show moderate focal intrahepatic biliary ductal dilatation in the left hepatic lobe. The patient eventually underwent a left hepatic lobectomy with Roux-en-Y hepaticojejunostomy.

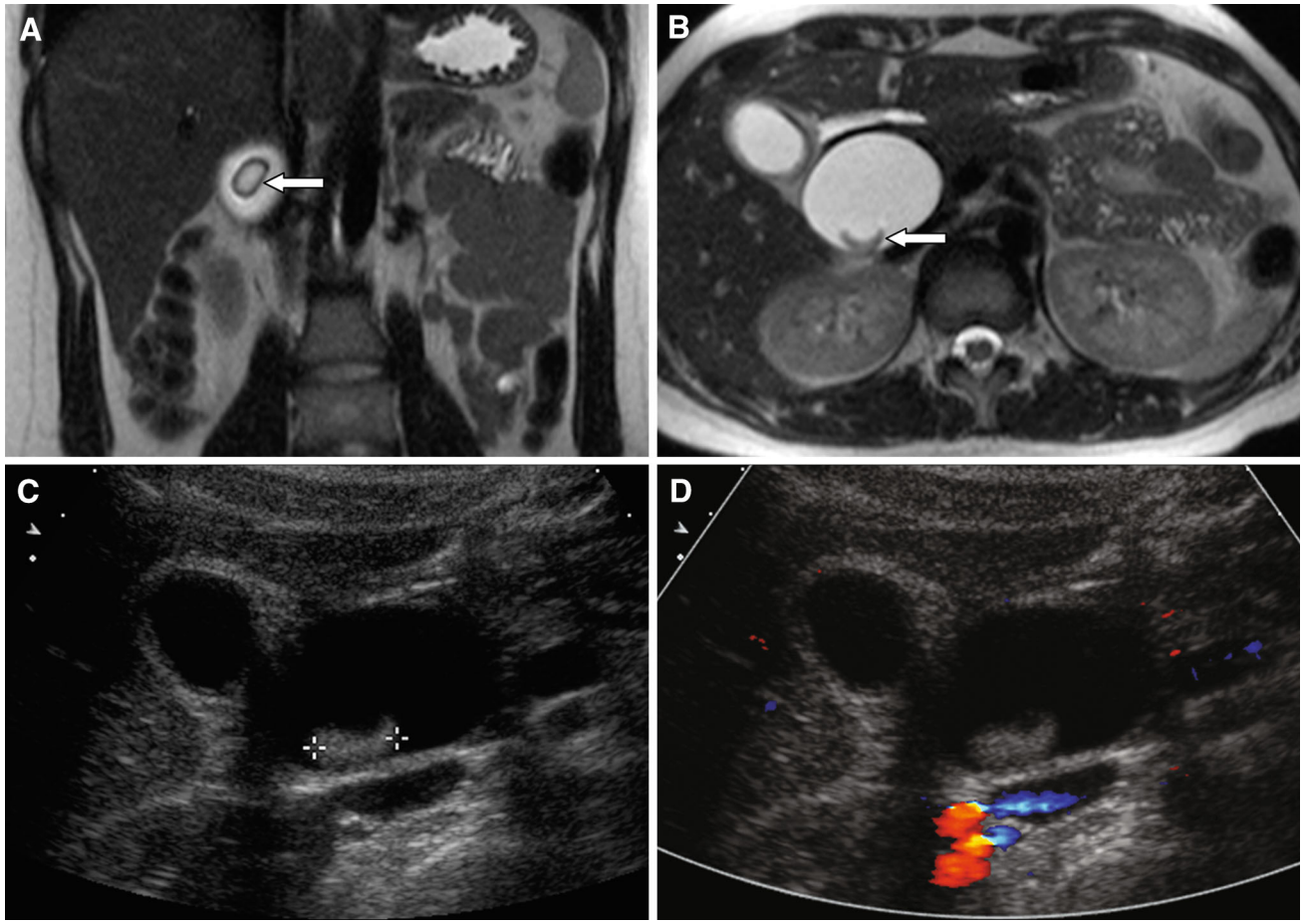
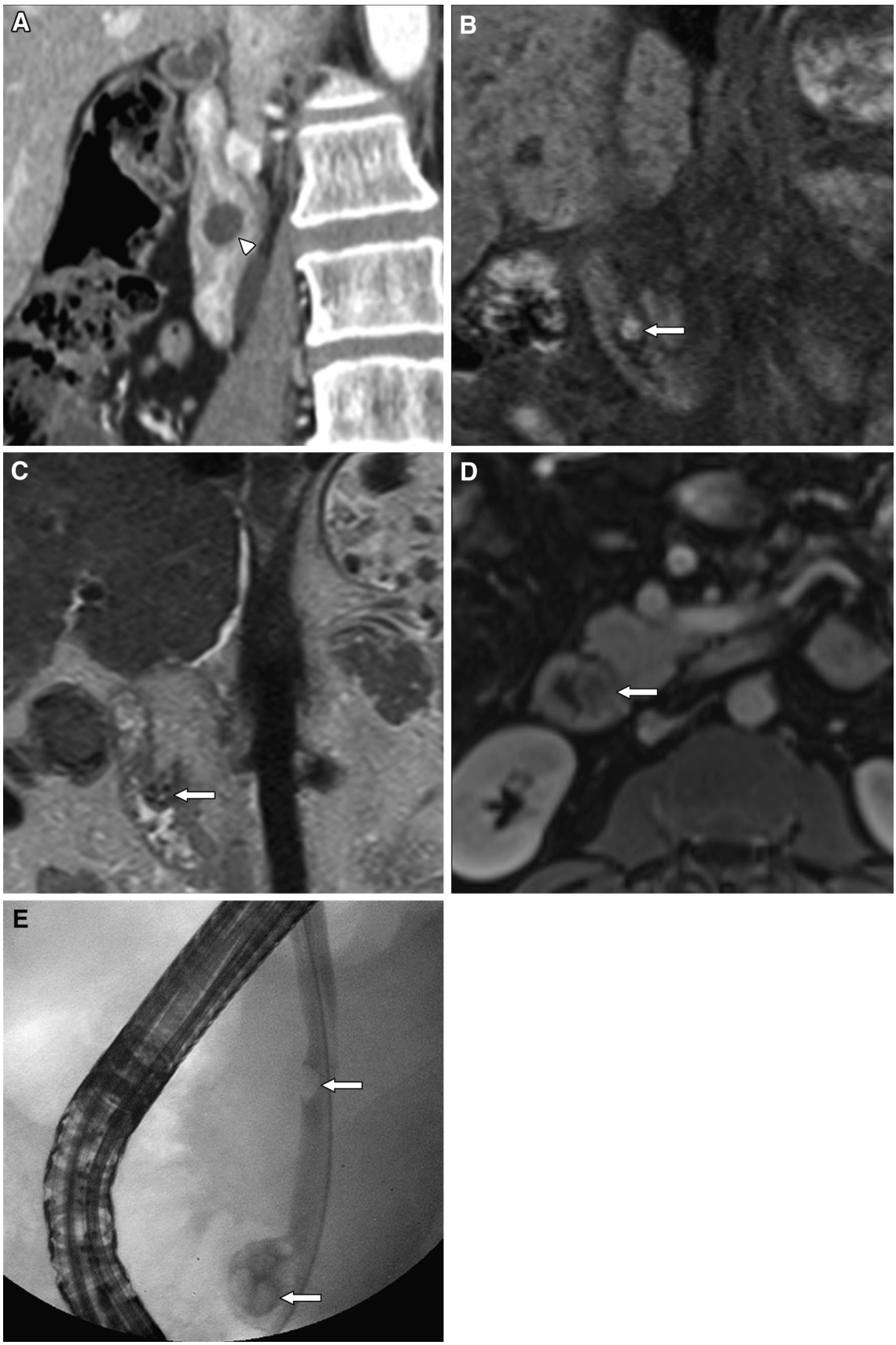


Fig. 13. 25-year-old woman with known type IVA choledochal cyst presenting with choledochal stones. Coronal (**A**) and axial (**B**) T2-weighted images of the biliary tree show a T2 hypointense filling defect within the dilated extrahepatic bile duct (*arrows*), concerning for a stone. (**C**) Transverse sonogram and (**D**) color Doppler image show a dependant, echogenic lesion in the extrahepatic bile duct without flow, confirming the diagnosis.

Caroli disease [48, 49]. Patients will often have nonspecific symptoms, such as abdominal pain, jaundice, fever, weight loss, or a palpable mass.

Imaging features concerning for cholangiocarcinoma include an irregularly thickened wall with or without an enhancing mass [48]. Contrast enhancement is necessary to differentiate benign from malignant wall thickening [48, 50]. Papillary adenocarcinoma (Fig. 15) presents with either a single or multiple enhancing papillary nodules. Gallbladder malignancy presents as heterogeneously enhancing, irregular gallbladder wall thickening [48]. MRI can help assess the location and extent of tumor invasion, including lymph node involvement and other distant metastases.

Given the risk of malignancy, long-term follow-up imaging is required for surveillance. The 5-year survival rate for patients with choledochal cysts complicated by malignancy is high, up to 55% in patients with cholangiocarcinoma [51, 52]. Even after complete excision, patients are at higher risk for malignancy than the general population [53]. Close follow-up is recommended to assess for early malignancy.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◀ **Fig. 14.** Type III choledochal cyst with pigment stones in a 46-year-old woman. (**A**) Contrast-enhanced coronal CT of the duodenum shows a cystic lesion in the region of the ampulla (*arrowhead*). Coronal T1-weighted fat saturated (**B**) and coronal T2-weighted (**C**) MR images show multiple T1 hyperintense and T2 hypointense foci in the cystic lesion in the ampulla (*arrows*). (**D**) Contrast-enhanced axial T1-weighted fat saturated MR image shows no associated enhancement (*arrow*). (**E**) ERCP image shows communication of this cystic lesion with the common bile duct with multiple filling defects in the common bile duct and choledochocele (*arrows*) consistent with stones.

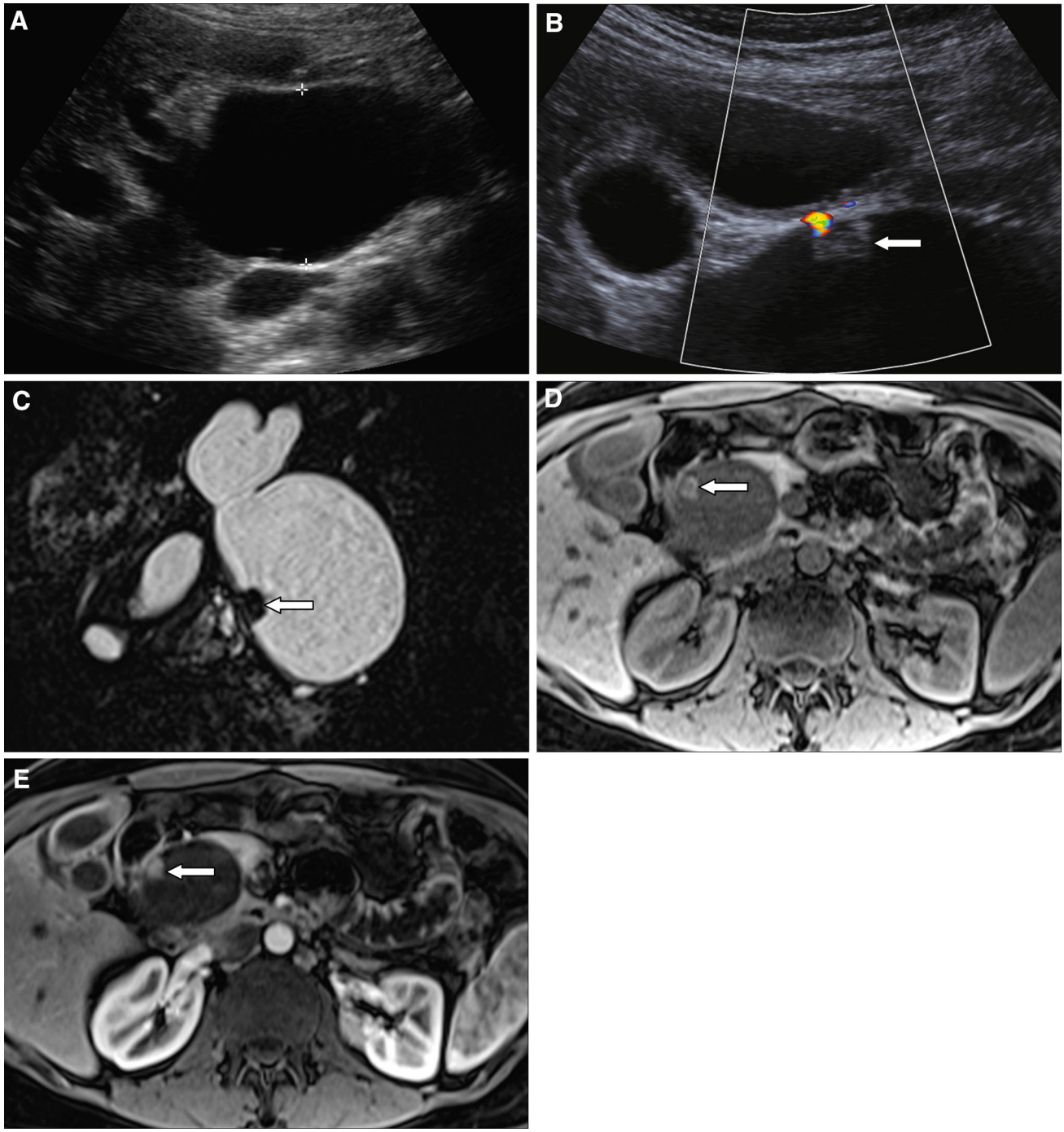

Fig. 15. Type IVA choledochal cyst with papillary carcinoma in a 48-year-old woman. (**A**) Transverse sonographic image of the biliary tree shows intra- and extrahepatic biliary ductal dilatation. (**B**) Transverse sonographic image of the dilated extrahepatic bile duct shows a non-dependent echogenic lesion projecting into the bile duct lumen with associated flow (*arrow*). (**C**) Coronal T2-weighted MR image of the biliary tree shows saccular dilatation of the extrahepatic bile duct with a T2 hypointense lesion along the duct wall (*arrow*). Unenhanced (**D**) and contrast-enhanced (**E**) axial T1-weighted fat saturated MR images show enhancement in the lesion (*arrow*), concerning for malignancy.

Summary

Because they are uncommon, radiologists may not recognize the characteristic imaging features of choledochal cysts, and mistake them for other cystic lesions. Accurate diagnosis and characterization are important, especially for subtypes that can undergo malignant transformation. Imaging diagnosis has markedly improved, and a diagnosis can be reached in most cases using MRCP instead of invasive procedures that were once the reference standard.

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