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Choledochal Cysts: Presentation, Clinical Differentiation, and Management

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Choledochal cysts (CC) are a rare congenital cystic dilation of the biliary tract, first described by Vater and Ezler in 1723.[1] They present primarily in female infants and young children and are more prevalent in East Asian populations. Although benign, CC can be associated with serious complications including malignant transformation, cholangitis, pancreatitis, and cholelithiasis.[2] We herein provide a state-of-the-art, evidence-based review of CC with particular emphasis on clinical differentiation and approach to management. A search of the available electronic databases, including MEDLINE/ Pubmed, using the term *choledochal cyst* as well as under the MeSH database subheading *choledochal cyst*, was performed. Criteria for inclusion included English articles (Fig. 1).

Incidence and epidemiology

Approximately 80% of CC are diagnosed in infants and young children within the first decade of life.[3,4] The incidence of CC ranges from 1 in 100,000 to 1 in 150,000 individuals in Western countries[5] to 1 in 13,000 individuals in Japan.[6] Choledochal cysts are 4 times more common in females.[2,7,8] Although the exact etiology is unknown, anomalous pancreaticobiliary duct union (APBDU) is seen in 30% to 70% of all CC where the common bile duct (CBD) and pancreatic duct junction occurs outside the duodenum, allowing reflux of pancreatic fluid into the biliary tree.[9-13] The exposure of biliary epithelium to digestive and caustic pancreatic enzymes may contribute to CC formation. In 1969, Babbitt[14] initially described APBDU, and it is believed to be secondary to arrest in migration of the choledochopancreatic junction into the duodenal wall, leading to a long common channel

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(Fig. 2).[15] A long common channel is defined as insertion of the CBD farther than 15 mm from the ampulla of Vater.[16] It occurs in less than 2% of the population,[16] although it is more commonly seen in pediatric CC patients. Eighty percent to 96% of pediatric CC are associated with APBDU.[2,13,17] In one series of 2,885 patients undergoing ERCP, nearly 90% of patients diagnosed with an APBDU had a CC.[16] Animal studies have given credence to this theory since iatrogenic APBDU in murine models demonstrated cystic dilatation of the CBD.[18,19] Amylase levels in the fluid contained in the gallbladder and CC are typically elevated in patients with APBDU.[13] Other pathophysiologic mechanistic hypotheses for CC include a weak bile duct wall, sustained increased intrabiliary pressure, inadequate autonomic innervations, sphincter of Oddi dysfunction, and distal obstruction of the CBD.[5,20,21]

Classification Alonso-Lej and colleagues[20] proposed the first CC classification in 1959. Komi and associates[11] later proposed a new CC classification according to the type of APBDU based on 2 unique features: a long common channel and the angle of the junction between the pancreatic duct and distal CBD as they converge on the sphincter of Oddi.[5] However, the most widely accepted classification was reported by Todani and colleagues[22] in 1977, derived from the original Alonso-Lej classification and based on the site of cystic change (Fig. 3). Five types of CC are described and classified: type I (80% to 90% of all CC), type II, type III, type IV (15% to 20% of all CC) and type V or Caroli's disease.[2,7,22-24]

Type I cysts typically appear as anechoic cystic lesions, which communicate with the biliary tract. A type I cyst can be associated with mild enlargement of the intrahepatic bile ducts secondary to biliary stasis (Fig. 4).[7] Further differentiation of type I cysts (1A, 1B, or 1C) is accomplished using ultrasound and cholangiography to evaluate the gallbladder relationship and cystic duct location. In type IA CC, the gallbladder arises from the choledochal cyst and a dilated extrahepatic biliary tree is seen while the intrahepatic ducts are normal in size and appearance.[7] Type IB CCs contain a mostly normal appearing extrahepatic biliary tree with an isolated dilatation of the most distal aspect of the CBD, with no evidence of pancreaticobiliary malunion.[5] A smooth fusiform dilatation of the common hepatic duct (CHD) and CBD along with pancreaticobiliary malunion is classified as type 1C CC.[25]

Type II cysts are true diverticula of the CBD and represent 2% of reported cases.[26] Type II cysts appear as anechoic cysts juxtaposed to the CBD with a normal appearing gallbladder and CHD (Fig. 5). Cholangiography demonstrates opacification of a true diverticulum arising from the CBD[7] and can resemble gallbladder duplication.[5,27]

Type III cysts, or choledochoceles, were initially described by Wheeler[28] in 1940. Type III cysts comprise 1% to 4% of CC and are characterized by their intraduodenal location at the pancreaticobiliary junction.[5,7,26,29] Although CC have a female predominance, choledochoceles are more evenly distributed between the sexes.[17,30] Type III cysts are also more likely to be diagnosed using ERCP and are managed primarily with endoscopic therapy.[17,31] Pancreatitis is commonly seen and biliary tract symptoms are less common.[17,32,33] Type III cysts are associated with a much lower incidence of malignant transformation (2.5%).[29,34,35] Additionally, APBDU is less commonly seen in choledochoceles in comparison with other types of CC, and patients are more likely to have

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undergone a previous cholecystectomy at the time of diagnosis.[10,17,36] In fact, given the distinct differences in presentation, clinical course, diagnosis, and pathophysiology, some authors argue that choledochoceles represent a different disease entity.[17,30,33]

Type IV CC can include both intrahepatic and extra-hepatic duct involvement. Type IV CC are subclassified into type IVA and type IVB. Type IVA CC dilatation extends from the CBD and CHD into the intrahepatic biliary tree (Fig. 6). Additionally, primary ductal stricture around the hepatic hilum is commonly seen.[5,25] Although intrahepatic biliary dilatation most commonly presents with bilobar involvement, dilatation of the left lobe is the second most common presentation.[37,38] Isolated dilatation of the right lobe is rarely seen.[38] By contrast, type IVB CC consists of multiple dilations of the extrahepatic biliary tree, classically described as a “string of beads,” with an uninvolved intrahepatic biliary tree.[5]

Finally, type V CC, or Caroli's disease, demonstrates intrahepatic saccular or fusiform dilatation with no underlying obstruction or extrahepatic biliary tree involvement (Fig. 7A, B).[7] Type V CCs are thought to arise from ductal plate malformation[15] and be associated with polycystic kidney disease,[39] an autosomal recessive inherited condition associated with mutation in PKD1 gene (Fig. 7C).[40] When type V CCs are accompanied with congenital hepatic fibrosis, it is termed Caroli's syndrome.[15] The enhancement of the portal vein surrounded by dilated intrahepatic bile ducts, or “central dot sign,” is highly suggestive of Caroli's disease and can easily be seen on magnetic resonance cholangiopancreatography (MRCP) or contrast-enhanced CT.[7,41,15] Contrast filling in well-defined intrahepatic cystic dilatations is pathognomic.[15]

Visser and colleagues[27] have challenged the modified Todani classification, stating that it combines multiple and different disease entities. In support of this, the investigators note the different clinical courses, management, and complication rates of the 5 types of CC. Specifically, Visser and colleagues[27] note the distinction of types I and IVA CC as arbitrary given that there is generally some intrahepatic duct involvement in both classes of CC. The authors further state that gallbladder-like diverticula, choledochoceles, and Caroli's disease are completely unrelated to CC and therefore propose abandoning the Todani classification and instead using descriptive terminology.

Clinical presentation

Choledochal cysts are usually diagnosed in childhood, although in utero and adult diagnosis is also common.[7,42] Common presentations include abdominal pain, jaundice, and right upper quadrant mass and are most commonly seen in pediatric patients.[24,43] Cholangitis, pancreatitis, portal hypertension, and liver function test abnormalities are common and are thought to be a result of ABPDU or stone obstruction.[23,24,43-47] Biliary amylase levels can be elevated in CC patients, and clinical features correlate with degree of elevation.[48-51] The classic triad of abdominal pain, right upper quadrant mass, and obstructive jaundice is mainly seen in the pediatric population, although still rare.[26,50,52]

There are distinct differences to the pattern of presentation in adults and children.[53] Specifically, adults are more likely to present with biliary or pancreatic symptoms and abdominal pain; children are more likely to present with an abdominal mass and

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jaundice.[2,23,24,53,54] Cystrupture is rare and typically is seen only in neonates and infants.[43,55] Adults with CC are more likely to have symptomatic gallstones (45% to 70% of patients)[43] or acute cholecystitis, both of which are attributed to biliary stasis.[5] As a result, adult patients with CC are more likely to have undergone previous biliary procedures including surgery and stenting.[23,24,43]

Associated congenital anomalies include double common bile duct, sclerosing cholangitis, congenital hepatic fibrosis, pancreatic cyst,[56] and annular pancreas.[26,57] In a nationwide study, congenital cardiac anomalies occurred in 31% of pediatric patients with CC and are most commonly manifested in infancy.[58]

Biliary malignancy is seen in 10% to 30% of CC.[59-62] Malignancy is rarely seen in pediatric CC; however, CC-associated biliary malignancy carries a dismal prognosis.[59,63,64] Histories of cholangitis and internal drainage procedures have both been associated with an increased risk of CC-related malignancy.[64]

Differential diagnosis includes biliary lithiasis, primary sclerosing cholangitis, pancreatic pseudocyst, biliary papillomatosis, and biliary hamartoma.[7] Biliary atresia (BA) is commonly associated with CC and must therefore be ruled out in neonatal obstructive jaundice.[7] More specifically, cystic biliary atresia (CBA), a subtype of BA, very closely resembles CC. Distinguishing between CBA and CC is critical because delayed therapy in CBA results in irreversible long-term sequelae.[65] Unlike BA, CC and CBA can typically be identified with prenatal ultrasound; however, these lesions are often all thought to be CC until surgical intervention.[66] However, CBA patients are symptomatic at earlier ages (less than 3 months old), and one-third of CBA patients develop liver failure or require liver transplantation.[65] On ultrasound, CBA cysts appear smaller, with less dilatation of the intrahepatic bile ducts and are associated with an atretic or elongated gallbladder.[67-69] This is in contrast to commonly seen intrahepatic duct dilation and a normal or distended gallbladder in CC.[69] Zhou and colleagues[68] identified sonographic detection of the triangular cord sign (a thickness of the echogenic anterior wall of the right portal vein just proximal to the right portal vein bifurcation) and the presence of biliary sludge as features suggestive of a diagnosis of CBA rather than CC. In their series, 11 of 12 CBA patients had a triangular cord sign vs none seen in the CC cohort.[68] Moreover, immunohistochemical analysis of CD56-stained liver biopsy specimens from CC and CBA patients showed no CD56 positivity and less hepatic fibrosis in CC group compared with varying levels of CD56 positive hepatocytes and increased hepatic fibrosis in all prenatally diagnosed CBA.[70,71]

Differentiating Caroli's disease from polycystic liver disease and primary sclerosing cholangitis can be difficult. Although similar in radiographic appearance, the cysts associated with polycystic liver disease do not communicate with the biliary tree, while primary sclerosing cholangitis is associated with a distal biliary obstruction and inflammatory bowel disease. The risk of neoplasia in Caroli's disease is less than 7%, but surgical management is usually indicated secondary to cholangitis and liver complications.[27,72,73]

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With the increased use of axial imaging, more CC are being diagnosed as incidental findings.[74] Choledochal cyst diagnosis is typically accomplished using multimodality imaging including ultrasound, CT, and MRI, including MRCP. Ultrasound is the most frequently used imaging modality given its low cost and accessibility, and has been shown to be reliable and cost effective as single modality imaging in the pediatric population.[54,75-78] A CBD measuring greater than 10 mm in an adult should alert the physician to the possibilities of cystic dilatation of the biliary tree or obstructive biliary lithiasis.[7] Importantly, intrahepatic biliary dilatation is an indication for further imaging in order to differentiate type I cysts from type IVA disease.[77] Additionally, a right upper quadrant cyst separate from the gallbladder is suggestive of CC disease.[26] Choledochal cyst diagnosis is further supported by the presence of a direct communication between the biliary tree and the cystic duct.[15,26] Thickening and irregularity of the CC wall suggests malignancy, and intraductal ultrasound has been shown to differentiate between early T-stage tumors arising within CC.[26,79] These criteria allow for the differentiation of CC disease and other right upper quadrant cystic entities, such as pancreatic pseudocyst, renal cyst, and hepatic cysts.[26] However, ultrasound fails to determine the cause of a dilated CBD in one-third of patients. Moreover, it is unable to accurately identify APBDU.[80] Endoscopic ultrasound has been shown to be safe and accurate in these instances, particularly in its ability to detect a long common channel and choledochoceles[81,82] although ERCP remains the gold standard for these diagnoses.[16,80]

Cholangiography, specifically ERCP and percutaneous transhepatic cholangiography, is the most sensitive technique to define the anatomy of the biliary system, but can be difficult to perform in the pediatric population given the need for general anesthesia, technical difficulty, and potential complications.[83,84] An ERCP allows for direct visualization of the pancreaticobiliary junction. In addition to its diagnostic yield, ERCP can be therapeutic by allowing biliary drainage and endoscopic sphincterotomy of choledochoceles.[85,86] Percutaneous transhepatic cholangiography also permits sensitive evaluation of the intrahepatic bile ducts, but sometimes can fail to adequately delineate the distal and intraduodenal portions of the CBD. Notably, both procedures are associated with potential complications, including bleeding, cholangitis, acute pancreatitis, and perforation.[5] As a result, noninvasive imaging with MRCP has gained popularity and is replacing direct cholangiography's diagnostic role in CC.[87,88]

Magnetic resonance cholangiopancreatography is noninvasive and does not require irradiation or oral or intravenous contrast.[89] Modern MRCP technology has removed the need for exaggerated breath holding techniques,[90] increasing its utility and accuracy in pediatric patients.[89] The MRCP is highly sensitive (70% to 100%) and specific (90% to 100%) in CC diagnosis and classification.[88,91] Moreover, it reliably identifies APBDU (particularly with the use of secretin[92,93]) as well as cholangiocarcinoma and choledocholithiasis with concurrent CCs.[7,88,90-92,94-96] Although MRCP is associated with lower cost and decreased morbidity,[92,94,97] it is limited in its ability to detect minor ductal abnormalities or small choledochoceles.[88] Magnetic resonance cholangiopancreatography cannot be used for therapeutic purposes; therefore its utility remains limited as a diagnostic tool.[88] Computed tomography is also commonly used and can help demonstrate important anatomic relationships for surgical planning.[26] Although ultrasound and CT have each have

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a sensitivity and specificity of more than 90% in the diagnosis of CC, MRI leads to improved delineation of the exact pathologic anatomy and therefore is generally the imaging technique of choice.[26,87]

Pathologic characteristics

Fibrosis of the cyst wall lined with columnar epithelium and lymphocytic infiltration is typical in pediatric CC; adult CC includes evidence of inflammation and hyperplasia.[43,98] Most CC show some degree of pathologic changes in the liver including portal fibrosis, central venous distention, parenchymal inflammation, and bile duct proliferation.[99] Except for portal fibrosis and central venous distention, these resolve after appropriate surgical management.[99] Other common findings across all classes of CC include acute and chronic mucosal inflammation, mucosal dysplasia, and few to no mucus-producing glands (Fig. 8).[100]

Interestingly, distinct differences exist in the histologic appearances of the different CC subtypes. Type I (and sometimes type IV) CC lack biliary mucosa; type II CC closely resemble gallbladder duplication. Type III cysts are lined by duodenal mucosa, while type V cysts can have extensive hepatic fibrosis.[31,101] Immunohistochemical analysis demonstrates an increasing rate of epithelial metaplasia and biliary intraepithelial neoplasia in the walls of CC with advancing age.[102,103] Concordantly, case studies repeatedly demonstrate an increased risk of malignant transformation with age: half of CC patients more than 50 years old have invasive biliary neoplasms vs less than 1% before the age of 10.[2,23,24,104,105] Although the incidence of harboring a malignancy at diagnosis of CC increases with age at diagnosis, the risk of developing a future malignancy in an existing benign CC during one's remaining lifetime likely decreases with advancing age.

Malignancy is most commonly associated with types I and IV cysts, while types II, III, and V CC have minimal neoplastic risk.[5,104] Carcinogenesis is thought to occur via multistep genetic events where early K-ras and p53 mutations are seen in more than 60% of CC-related carcinomas[79,106-108] followed by a late occurring DPC-4 gene inactivation.[107] Most reported cases of malignant transformation are cholangiocarcinoma; however, gallbladder carcinoma is identified in 10% to 25% of CC-related malignancies[5,10,24,27,54] (Table 1). The presence of an APBDU is thought to play a role in carcinogenesis and hepatocellular damage due to reflux of pancreatic contents into the bile duct.[15,99] Moreover, elevated biliary amylase in CC patients is associated with higher expression of inducible nitric oxide synthase (iNOS), implying a role for iNOS in CC mucosa hyperplasia and carcinogenesis.[109]

Management

MacWorter[110] performed the first CC excision in 1924. Historically, CC management consisted of internal or external drainage procedures along with cholecystectomy.[22] However, this resulted in unacceptably high rates of infection, pancreatitis, cholangitis, cholangiocarcinoma, and recurrent stenosis.[52,111-114] Moreover, although benign, the risk of malignant transformation warrants complete and total excision whenever possible. Fetal and newborn diagnosis is associated with early progression to liver fibrosis, particularly in type

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IV CC.[24,115,116] In a randomized controlled clinical trial, Diao and colleagues[117] demonstrated that early CC excision (less than 1 month old) in prenatally diagnosed asymptomatic CC resulted in significantly less hepatic fibrosis and improved the rate of liver function normalization. Early excision is recommended.[60,115,116,118]

Type I and IV CC management consists of complete extrahepatic bile duct cyst excision down to the level of communication with the pancreatic duct, cholecystectomy, and restoration of bilioenteric continuity.[13,24,119,120] Care should be taken to not injure the pancreatic duct. The extent of liver resection in type IVA CC depends on the nature of the extrahepatic component of the CC. In some cases, excision of the extrahepatic duct alone is reasonable because intrahepatic duct dilatation typically resolves in 3 to 6 months.[121,122] However, biliary stricture, lithiasis, and reoperation rates are significantly higher in the extra-hepatic duct excision alone group when compared with these rates in patients undergoing concomitant extrahepatic duct and liver resection.[123] Accordingly, hepatectomy is warranted in type IVA cysts with a significant intrahepatic component likely to result in postoperative complications if not removed.[37,123,124]

Hepaticoduodenostomy and Roux-en-Y hepaticojejunostomy (RYHJ) bilioenteric reconstruction after type I and IV CC resection are both reported in the literature, but RYHJ is preferred. Hepaticoduodenostomy has been associated with increased rates of gastric cancer (due to bile reflux) and biliary cancer.[124,125] Moreover, a recent meta-analysis comparing RYHJ with hepaticoduodenostomy reported significantly more postoperative reflux and gastritis with hepaticoduodenostomy.[126] A wide anastomosis allowing free flow of bile into the intestine is imperative in order to avoid anastomotic stricture and bile reflux, and may prevent complications and carcinoma arising in the intrahepatic ducts after cyst excision.[127-129]

Patients who have previously undergone drainage procedures require resection of the cyst due to the continued risk of malignancy and recurrent symptoms.[27,52,130] Biliary ductal and vascular anomalies are seen in 15% and 22% of CC patients, respectively.[131] Preoperative MRCP allows for accurate delineation of these abnormalities and aids in surgical planning. Some groups advocate early transection of type I CC near its midpoint[132] and routine intraoperative endos-copy,[133,134] allowing for visualization of the hepatic and pancreatic ducts from within the cyst and significantly lower incidence of postoperative stone formation. In CC-associated chronic severe pancreatitis and atrophic pancreatic head due to APBDU, pancreaticoduodenectomy may be indicated.[124] Resection of complicated CC is associated with worse outcomes.[111,135] Therefore, severely ill CC patients may benefit from staged procedures consisting of external drainage followed by complete cyst excision and hepaticoenterostomy.[111,135,136]

Type II and III CC are associated with an extremely low risk of malignant transformation.[17,121,137] Diverticulectomy of type II CC followed by primary CBD closure at the diverticulum neck is usually sufficient. Appropriate management of small choledochoceles consists of endoscopic sphincterotomy.[7,32] Transduodenal excision may be considered for large choledochoceles with associated complications such as gastric outlet obstruction or pancreatitis.[7,31]

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Type V (Caroli's disease) management consists of liver resection or orthotopic liver transplant (OLT).[9,42] Localized or unilobar cystic disease is best managed with hepatic resection. Importantly, however, incomplete resection of cystic disease leads to poor long-term outcomes; therefore, an aggressive surgical approach is recommended.[72,138] Asymptomatic bilobar disease is typically managed nonoperatively, at which point aggressive surveillance for potential malignant transformation is warranted.[139,140] Although prophylactic OLT is not indicated, complicated bilobar Caroli's disease with cholangitis, portal hypertension, or suspicion of early malignant transformation is definitively best managed with OLT.[72,139,141-143] Both liver resection and OLT produce excellent and comparable long-term outcomes and survival rates.[72,138,140,141,143,144]

Minimally invasive resection of CC has gained popularity, particularly in the pediatric population.[145-148] Laparoscopic CC resection with RYHJ reconstruction has been shown to be safe with comparable outcomes to open resection in retrospective analyses.[148-157] Reported advantages of the laparoscopic approach include improved intraoperative visualization of deeper structures, decreased postoperative pain, shorter hospital stay, improved cosmetic result, and decreased postoperative ileus.[148,149,155,158] However, these cases remain reserved for highly specialized surgeons with a thorough understanding of hepatobiliary anatomy and minimally invasive techniques.[157] Finally, limited case series of robotic pediatric CC resection and reconstruction have been reported with acceptable outcomes, although more studies are needed before widespread acceptance and implementation of this technique.[147,159-161]

Outcomes and prognosis

Postoperative morbidity and mortality are typically very low in children,[23,24,124] while postoperative complications are more commonly seen in adult patients.[43,130,134,162] Late complications (greater than 30 days postoperatively) occur in up to 40% of adult patients and include anastomotic stricture, cancer, cholangitis, and cirrhosis.[2,43,119,124,129,134,163] Type IVA cysts are most commonly associated with complications after management including intrahepatic stones and anastomotic stricture.[43,124,164] Overall, CC resection has an excellent prognosis, with an 89% event-free rate and 5-year overall survival rates well over 90%.[50,165,166] However, the risk of biliary malignancy remains elevated even more than 15 years after CC excision, and CC-associated biliary malignancy is associated with extremely unfavorable outcomes, with a reported median survival of 6 to 21 months.[10,52,29,63,64,72,79,167] Therefore, long-term surveillance is warranted, particularly in instances with persistent intrahepatic biliary dilatation.[10,165] Typically this consists of regular biochemical evaluation and abdominal ultrasound or cross-sectional imaging.[3]

CONCLUSIONS

Choledochal cysts are a rare disease entity, more commonly seen in Asian populations. Given no contra-indication in patient performance status, most CC warrant resection in order to avoid future malignancies and future complications.[129] The risk reduction for development of future malignancy is variable depending on patient age and type of CC. Management includes total cyst excision and bilioenteric reconstruction performed by

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hepatobiliary specialists. Choledochoceles represent a different spectrum of presentation and management and may differ in pathogenesis compared with other types of CC. The rarity of this disease complicates development of a unified management approach. Regardless of CC subclass, appropriate therapy results in acceptable outcomes and complication rates. Although malignancy is rare, CC resection does not reduce it to baseline levels, so long-term surveillance is indicated given the increased likelihood of developing postexcision biliary malignancy.

Abbreviations and Acronyms

APBDU	anomalous pancreaticobiliary duct union
BA	biliary atresia
CBA	cystic biliary atresia
CBD	common bile duct
CC	choledochal cyst
CHD	common hepatic duct
MRCP	magnetic resonance cholangiopancreatography
OLT	orthotopic liver transplant
RYHJ	Roux en Y hepaticojejunostomy

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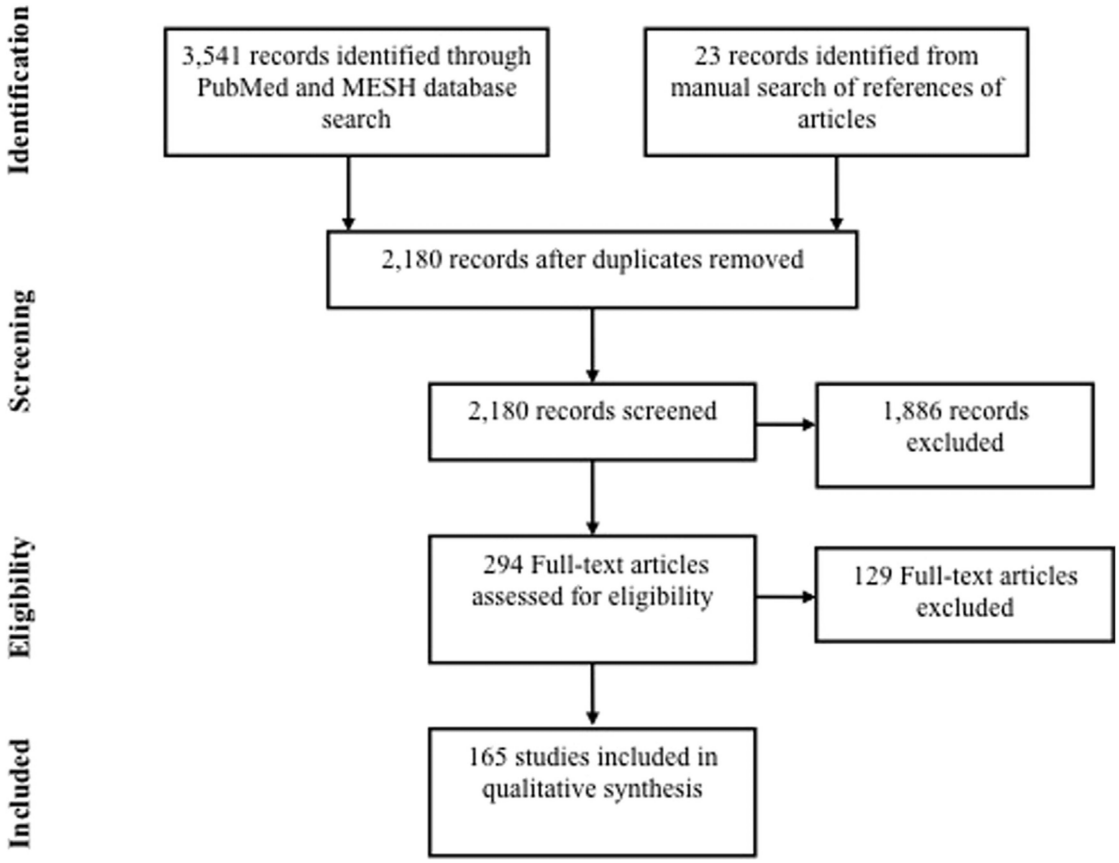

Figure 1.

A Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) diagram defining the method of inclusion and exclusion for studies used.

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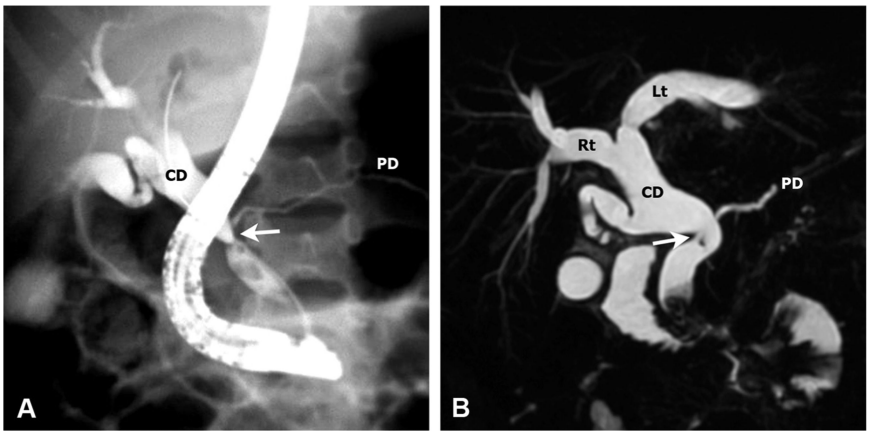

Figure 2.

Common channel in a 4-year-old girl. (A) ERCP image revealing dilated intra- and extrahepatic ducts. Notice that the pancreatic duct (PD) drains (arrow) into the mid common duct (CD). (B) MRCP in the coronal oblique plane showing better delineation of the insertion point (arrow) of the pancreatic duct. The right (Rt) and left (Lt) intrahepatic ducts are also well visualized. Notice debris in the distal common duct.

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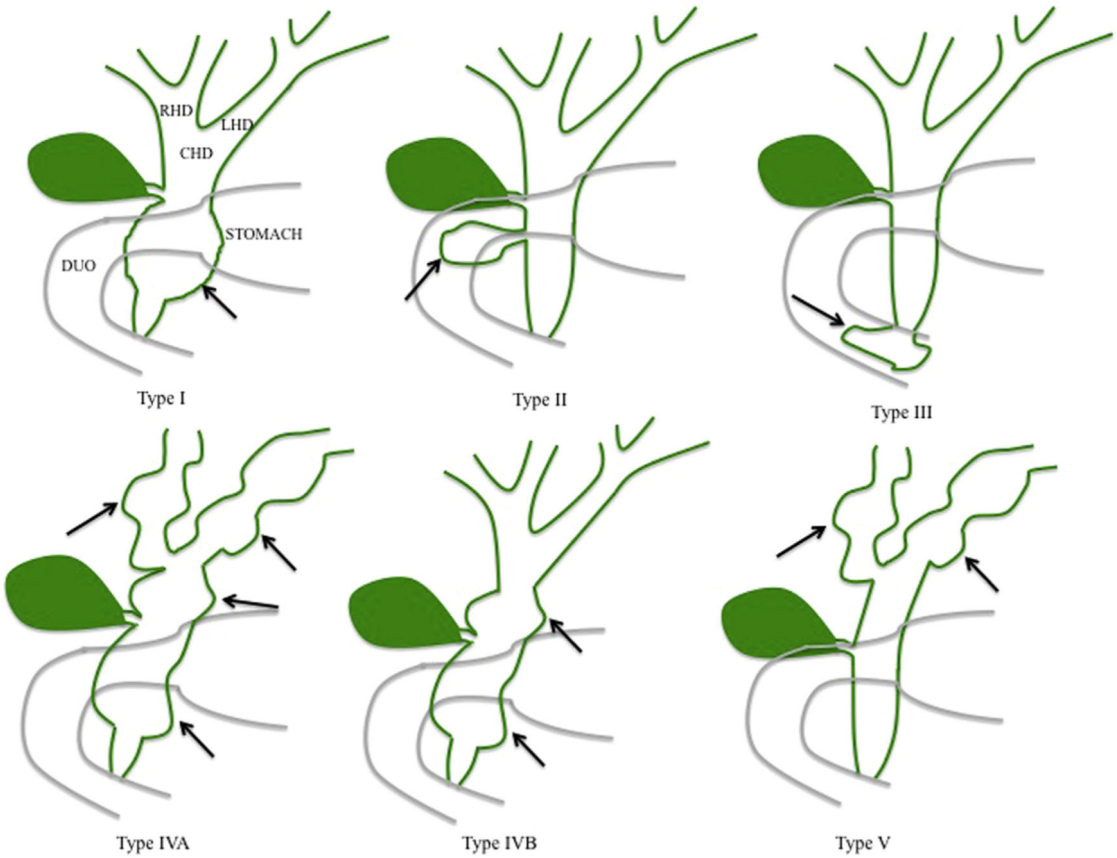

Figure 3.

Classification of choledochal cysts (CC). Type I cysts are fusiform dilatations of the common bile duct (CBD). Type II cysts are true diverticula of the CBD and type III CC (choledochoceles) are intraduodenal dilations of the common channel. Type IVA CC consist of multiple intrahepatic and extrahepatic biliary dilatations, while type IVB CC have extra-hepatic biliary dilatation with a normal intrahepatic biliary tree. Type V CC, or Caroli's disease, consist of cystic dilation of the intrahepatic biliary tree. RHD right hepatic duct, LHD left hepatic duct, CHD common hepatic duct, DUO duodenum.

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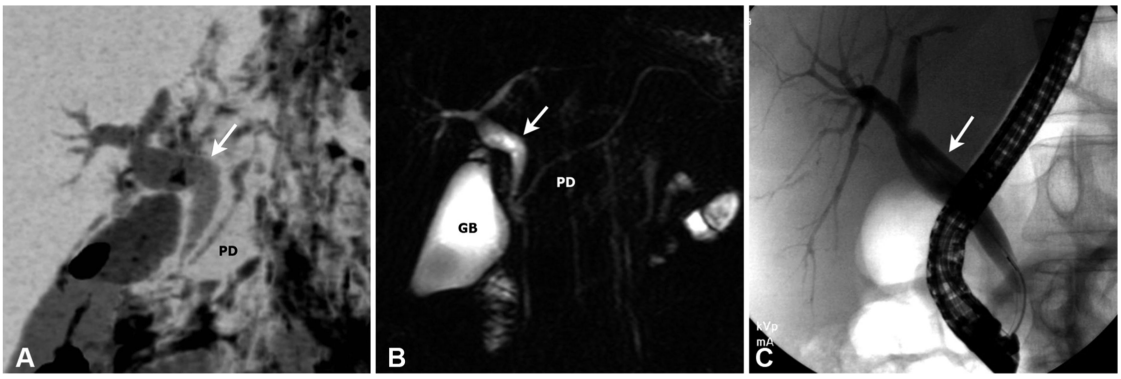

Figure 4.

Type I in a 53-year-old woman. (A) Thick slab (15 mm) minimum intensity projection CT image in the coronal oblique plane. There is diffuse dilatation of the common duct (arrow) consistent with type I choledochal cyst. The pancreatic duct (PD) is normal. (B) MRCP in the coronal oblique plane demonstrating similar findings. The gallbladder (GB) is also visualized. (C) ERCP image confirming diffuse dilatation of the common duct (arrow).

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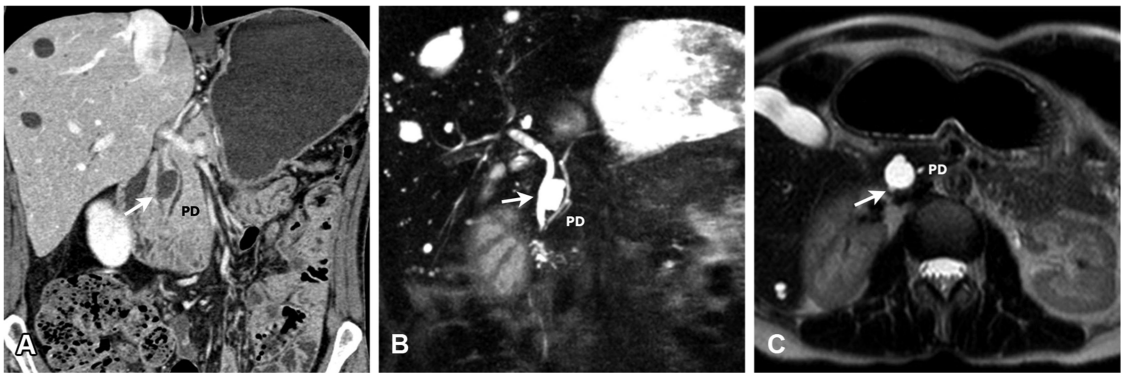

Figure 5.

Type II in a 61-year-old female. (A) Thick slab (5mm) coronal reconstruction of CT image in the portal venous phase. Notice focal saccular outpouching in the distal common duct (arrow) consistent with Type II choledochal cyst. Notice that the pancreatic duct (PD) is draped around the cystic lesion which was originally mistaken for IPMN communicating with the pancreatic duct. (B) MRCP in the coronal oblique plane demonstrating the communication between the cyst and the distal common duct (arrow). No communication between the cyst and the pancreatic duct was visualized. (C) Axial MRCP image confirming the communication between the cyst and the distal common duct (arrow). The pancreatic duct (PD) does not communicate with the cyst.

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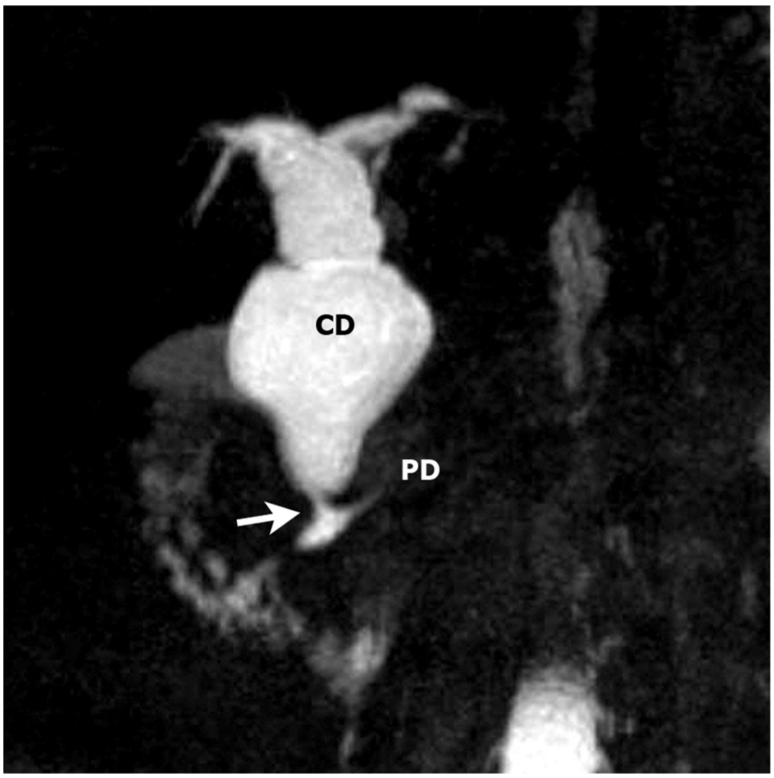

Figure 6.

Type IV in a 54-year-old woman. MRCP in the coronal plane shows multilobulated dilatation of the common duct (CD) with a short common channel noted inferiorly (arrow). Notice mild saccular dilatation of the intrahepatic right and left ducts. The pancreatic duct (PD) is not dilated.

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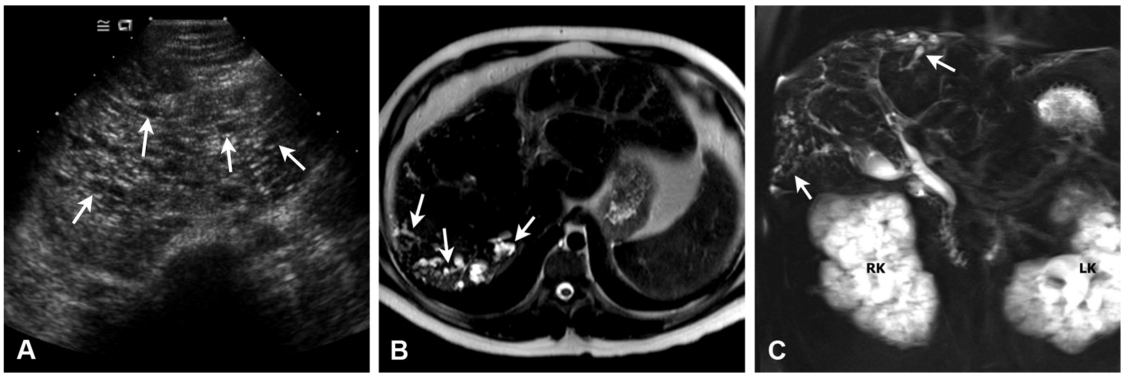

Figure 7.

Type V in a 27-year-old man. (A) Transverse ultrasound view of the liver demonstrating numerous anechoic lesions (arrows) scattered throughout the liver parenchyma. Ductal communication could not be detected. (B) MRCP in the axial plane demonstrating numerous small cysts, predominantly in the right lobe of the liver. These cysts are communicating with the intrahepatic bile ducts, which appear beaded (arrows). (C) MRCP image in the coronal plane showing communications between the cysts and the intrahepatic ducts (arrows). The kidneys are bright bilaterally (RK and LK) due to the presence of bilateral cystic renal disease.

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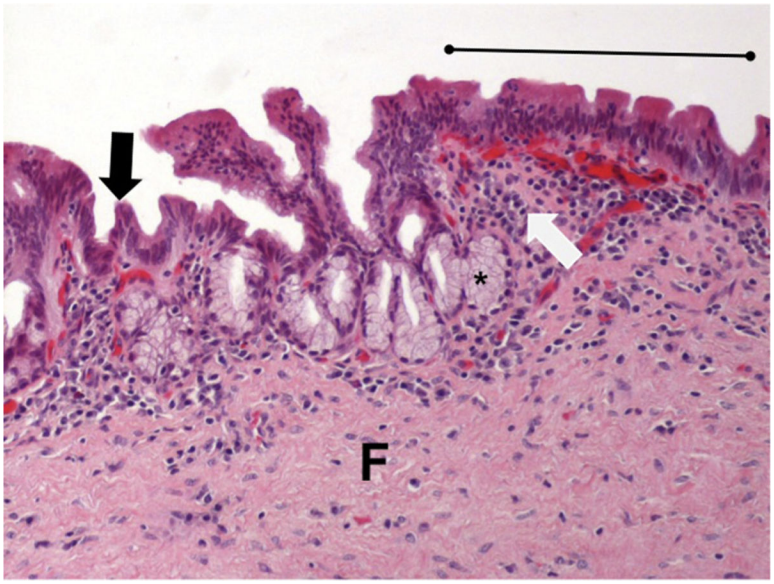

Figure 8.

Typical common duct (CD) cyst histology consists of relatively flat (line segment) or papillary columnar epithelium on type of a fibrous (F) wall. Chronic inflammation (white arrow), pyloric metaplasia (*), and reactive atypical epithelium (black arrow) are present in this example.

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Table 1

Stratification and Malignancies in Recent Case Series of Choledochal Cysts

First author	n	Types of choledochal cysts, n (%)	Malignancies encountered, n (%)
Gong 2012[42]	221; adult 221	Type I, 168 (76); type II, , 3 (1.4); type III, 3 (1.4); type IV, 26 (11.8); type V, 21 (9.4)	24 (10.9)
Lee 2011[10]	808; adult 808	Type I, 499 (61.8); type II, 7 (0.9); type III, 4 (0.5); type IV, 217 (26.96); type V, 5 (0.6); unspecified, 76 (9.4)	80 (9.9); cholangiocarcinoma, 40 (50); gallbladder cancer, 35 (43.8); synchronous gallbladder, cholangiocarcinoma, 2 (2.5); periampullary cancer, 3 (3.8)
Cho 2011[163]	204; adult 204	Type I, 116 (56.9); type II, 1 (0.5); type III, 0; type IV, 86 (42.1); type V, 1 (0.5)	20 (9.8); cholangiocarcinoma, 9 (45); gallbladder cancer, 11 (55)
Edil 2008[54]	92; children, 19; adult, 73	Type I (67.4); children, 15; adult, 45 Type II (6.7); children, 2; adult, 4 Type III (4.5); adult, 4 Type IV (19.1); children, 2; adult, 15 Type V (2.2); adult, 2	5 (5.6); cholangiocarcinoma, 3 (60); gallbladder cancer, 1 (20); embryonal rhabdomyosarcoma, 1 (20)
Singham 2007[75]	70; children, 19; adult, 51	Type I (42.9); children, 13; adult, 17 Type II (4.3), adult, 3 Type III (1.4), adult, 1 Type IV (48.6); children, 6; adult, 28 Type V (2.9), adult, 2	4 (5.7); cholangiocarcinoma, 4
Jesudason 2006[166]	57; adults, 57	Type I, 41 (72); type II, 0; type III, 0; type IV, 15 (26.3); type V, 1 (1.7)	None reported
Wiseman 2005[3]	51, adult, 51	Type I, 17 (33); type II, 3 (6); type III, 2 (3.9); type IV, 28 (54.8); type V, 2 (3.9)	4 (7.8)
Nicholl 2004[23]	57; children, 26; adult, 31	Type I, 41 (72); type II, 0; type III, 0; type IV, 10 (17.5); type V, 6 (10.5)	6 (10.5); cholangiocarcinoma, 5 (83.3); gallbladder cancer, 1 (16.7)
Lipsett 1994[24]	43; children, 11; adult, 32	Type I, 22 (51.2); children, 5; adult, 17 Type II, 1 (2.3); adult, 1 Type III, 2 (4.7); adult, 2 Type IV, 17 (39.5); children, 6; adult, 11 Type V, 1 (2.3); adult, 1	3 (7); cholangiocarcinoma, 2; gallbladder cancer, 1