Chapter

Choledochal Cysts

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Abstract

Choledochal cysts are congenital dilatations of the intra- and extrahepatic biliary tract that cause various pancreatic and hepatobiliary disorders. Pancreaticobiliary maljunction (PBM) results in choledochal cysts. PBM is a congenital pancreatic and bile duct juncture anomaly. It is widely accepted that the clinical presence of PBM is an etiological factor in the pathogenesis of biliary carcinogenesis in patients with choledochal cysts. For definitive diagnosis, ultrasonography sometimes shows the relationship with the biliary tract. If USG findings cannot rule out other causes, ideally MRI should be performed together with MRCP. CT may be the initial test for undiagnosed common bile duct malformations. In rare cases where conventional imaging results are uncertain, nuclear hepatobiliary iminodiacetic acid (HIDA) scanning enables the evaluation of radiological trace of involvement and accumulation in cystic structures associated with the biliary system. Todani added five anomalies and organized the most commonly used classification system. There are five subtypes. A type I cyst, A choledochal diverticulum (Todani type II), Choledochoceles (Todani type III), type IV cyst, Caroli disease (Todani type V). Surgical treatment should be based on the extent of biliary involvement based on the widely used Todani classification and anatomical findings and the presence or absence of PBM. The standard treatment in most CCs is the resection of the bile duct up to the lobar bifurcation. Residual postoperative intrapancreatic choledochal cyst may also lead to secondary carcinogenesis and associated morbidity. The localization of the pancreatic cyst is inside the head of the pancreas, close to the neck and to the left of the bile duct. Surgical treatment options include laparoscopic treatment. Its main advantages include excellent visualization and low blood loss.

Keywords: choledochal cysts, pancreaticobiliary maljunction, choledochal diverticulum, choledochoceles, caroli disease, cholangiocarcinoma

1. Introduction

Choledochal cysts are congenital dilatations of the intra- and extrahepatic biliary tract that cause various pancreatic and hepatobiliary disorders, collectively known as congenital cystic dilatation of the common bile duct [1]. Its total incidence is about 1/100,000–1/150,000 in the Western population and 1/1000 in the Asian population [2]. The male-to-female ratio was reported to be approximately 1:3 or 1:4 [3]. Since they do not contain an epithelial structure, they are not real cysts. They are choledochal malformations that can be detected by imaging and cover various morphological anomalies of the biliary ductal system [4–6]. The exact cause of choledochal

malformations is unclear. However, a number of theories were proposed about their pathogenesis, ranging from complete congenital anomaly to sequelae of multiorgan disease process [2]. Choledochal malformations are prone to complications such as cholestasis, pancreatitis, cholangitis, and cholelithiasis. Moreover, some of them, especially those associated with chronic inflammation, have a 50% higher probability of malignant transformation [2]. Choledochal malformations are mostly observed in the pediatric population. An estimated 25% of them are detected in the first year of life and after, while 35–55% by the age of 10 [2]. Up to 20% of them occur in adulthood and have a worse prognosis than in childhood [7, 8].

The similar incidence rates of choledochal cysts in the Western and Eastern cohorts are probably associated with similar etiology. Babbitt's theory is the most commonly proposed one. According to this theory, pancreaticobiliary maljunction (PBM) results in choledochal cysts. PBM is a congenital pancreatic and bile duct juncture anomaly caused by the union of the common duct and pancreatic and bile ducts outside the wall of the duodenum [9]. It is widely accepted that the clinical presence of PBM is an etiological factor in the pathogenesis of biliary carcinogenesis in patients with choledochal cysts. Here, the pancreatic duct and the bile duct are united at a position 1–2 cm proximal to the sphincter of Oddi [10–14]. The presence of pancreaticobiliary ductus junction anomaly allows reflux of pancreatic secretions into the biliary tree. This is the accepted theory for etiopathogenesis of cysts [15, 16]. The clinical presence of PBM is widely considered to be etiological in biliary carcinogenesis in patients with choledochal cysts [17–20]. PBM induces the reflux of pancreatic enzymes into the bile duct, which is thought to be involved in the formation of the bile duct cysts and the development of cancer within the biliary cyst [21]. Biliary cysts (BDC) have a 100–1000 times lower incidence rates in Western series [22–24]. Most of the studies with Western series reported BDC in adults, which had less association with PBM compared with Eastern series [25]. The common duct theory posits that the PBM above the sphincter of Oddi complex causes reflux of pancreatic and digestive enzymes into the bile duct. As a result, wall weakness and subsequent dilatation occur. Moreover, high levels of phospholipase A2 and trypsinogen in the bile of choledochal cyst patients were suggested to exacerbate inflammation and epithelial deterioration in the bile duct [26].

There is no consensus for the diagnosis of PBM, at least in Western countries. Diagnosis includes MRCP, ERCP, and functional criteria (measurement of intrabiliary amylase level). The diagnosis of PBM is based on two strict criteria: (1) The presence of conclusive evidence for the intraduodenal junction between the choledochal and the main pancreatic duct, and (2) the presence of an abnormal common duct longer than 10 mm. PBM is found in only 50–80% of choledochal cyst cases [27].

In a multicenter study conducted in France, 72.2% of patients with BDC were found to have PBM. While 66.8% of the patients were adults, 33.2% were children (<15 years old). The most common symptoms were abdominal pain (64.3%), jaundice (24.7%), cholangitis (24%), and pancreatitis (23.6%). At the same time, the most common types were C-P type I (57.2%), P-C type II (34.5%), and type III (complex type) (8.3%). The mean common duct length was 15.8–6.8 mm while children had shorter common ducts (5.3–13.8 mm). The common duct was longer than 6 mm in 97.6% of the patients and longer than 10 mm in 90.5%. Considering the abnormal common duct longer than 8 mm based on the diagnosis, the sensitivity and specificity rates were 97.6% and 80%, respectively, while the positive and negative predictive values were 99.2% and 57.1%, respectively. In the same study, the coexistence of the common duct longer than 8 mm and amylase values above 8000 UI/L were associated

with positive predictive and specificity rates of more than 90%. In terms of the association between PBM and Todani BDC types, there was a coexistence rate of 78.4% for type I and IVb, 16.6% for type II, 33.3% for type III, and 19% for type IVa. Although the relationship between type IV and type I BDC and PBM was not significant, the incidence of biliary cancer in type I and type IV patients with PBM was 83.3 and 25%, respectively. The relationship between PBM and BDC is similar in Asia and the West cohorts as 72% [22, 23]. Likewise, coexistence of PBM is more frequent in type IV and type I (69 and 78%, respectively), which is less in type II and III (16.6 and 33.3%, respectively) [9, 22–25]. However, there was no association between type V (Caroli disease) and PBM [28]. PBM type P-C subtype is more commonly found with BDC-type Ic (fusiform) while PBM-type C-P subtype is more common with BDC-type Ia. Therefore, in patients with slightly large choledochal (approximately 20 mm), the evidence of the presence of P-C-type PBM represents a key indication for the diagnosis of Todani BDC type Ic diagnosis. Then, complete resection is planned [29, 30]. Anatomical studies showed that 80–85% of adults have a short common duct, whose mean length was reported to be 4.6 mm ± 2.2 mm. In the last study, the mean common duct length was found to be 13.8 ± 5 in patients with suspected PBM. This also confirmed Kawisawa's notion of considering common duct length as the main morphological diagnostic tool for PBM [24]. However, there is no consensus for the cutoff value for the abnormally long common duct. This length is variable during infancy. More than 90% of adults and infants had a common duct length equal to or greater than 10 mm. Taking a cutoff value of 8 mm for the abnormal common duct length may indicate high precision for the diagnosis of PBM. Apart from morphological tools, functional methods using intrabiliary amylase measurement are easy to perform. In one study, intrabiliary mean amylase values were significantly higher in patients with PBM compared with those without PBM (> 50,000 versus <2000, respectively). A literature survey also showed that intrabiliary amylase value of >10,000 UI/L is an important functional tool to confirm the diagnosis of PBM [31]. Intrabiliary amylase concentration can be measured by the following methods: (1) by intraoperative sampling from the gallbladder during cholecystectomy prior to cholangiography or any other manipulation; (2) preoperatively through transhepatic fine-needle aspiration from the gallbladder; and (3) less frequently through bile aspiration during ERCP [32]. In the case of intrabiliary amylase level > 8000 UI/L and common duct length of >8 mm, a definitive diagnosis of PBM can be made with a sensitivity of 87% and a positive predictive value of 90%. However, if the intrabiliary amylase level is normal, the diagnosis of PBM cannot be completely ruled out. When the intrabiliary amylase level is normal and the common duct length is <8 mm, negative predictive value is close to 90%. Therefore, this could be an appropriate method to rule out PBM and then BDC diagnosis. Likewise, the level of amylase in the gallbladder is slightly higher than in the biliary tract, which may support the stagnation theory in carcinogenesis. These theories blamed pancreatic fluid reflux and its attack to biliary tract as the main culprit of cancer degeneration in especially dilated segments against the cholangiocarcinoma degeneration in the distal part of the biliary tree even when these observations were not directly related to Caroli disease, with or without PBM. In the presence of PBM, especially the Todani type I BDC, the incidence of biliary cancer seems to increase. Cholangiocarcinoma cancer degeneration develops 10 years earlier than those without PBM [33]. There is a high risk of cancer degeneration in patients who underwent cyst enterostomy due to BDC [34, 35]. The global cancer incidence of 8.3% in patients with PBM also including children observed in a Western cohort study was similar to what was reported in Eastern series, albeit at a slightly higher rate [22, 23].

Each entity has different imaging findings, diagnostic features, and surgical management. When it is not detected incidentally in prenatal imaging, it is frequently found in the right upper quadrant ultrasound examination performed for the symptoms and signs of pain, palpable mass, or cholestasis in childhood and young adults [2, 36]. Bile duct larger than 10 mm without obstruction in childhood is always associated with choledochal malformation [37]. For definitive diagnosis, ultrasonography sometimes shows the relationship with the biliary tract. If USG findings cannot rule out other causes, ideally MRI should be performed together with MRCP [36]. The T2-weighted technique routinely used in MRCP allows for a quality depiction of the hepatobiliary system [9]. MRI-MRCP with or without contrast enhancement is a method of choice in preoperative planning. Contrast application indicates malignant change and the choice of alternative approach [38]. CT may be the initial test for undiagnosed common bile duct malformations that are detected later in life or incidentally. The frequency of incidentally detected choledochal cysts in adults has increased from 10–36% due to the availability and convenience of CT [7]. The sensitivity of CT cholangiopancreatography in showing the common pancreatobiliary and pancreatic duct is 64% [39]. Due to concomitant sedation requirements, ERCP and percutaneous cholangiography carry independent risks and should be reserved for situations that are difficult, complicated, or contraindicated for MRI [40]. Diagnostic confirmation is more difficult in individuals with a cholecystectomy history. MRCP has been used more than ERCP or intraoperative cholangiography in the last 10 years for the diagnosis of PBM. MRCP is the most useful method for detecting PBM as it shows the junction of the common pancreatic duct and the common bile duct outside the duodenal wall even in patients with a clearly normal common duct. In the most recent series, the detection rate of PBM by MRCP is 82–100% [41]. Therefore, in patients with or without BDC, MRCP has been indeed used more frequently as the first-choice method than the endoscopic ultrasound or ERCP for focusing on the pancreatic head region [42]. In rare cases where conventional imaging results are uncertain, nuclear hepatobiliary iminodiacetic acid (HIDA) scanning enables the evaluation of radiological trace of involvement and accumulation in cystic structures associated with the biliary system. This is especially important in distinguishing of true choledochal malformation from imitations such as pancreatic pseudocyst and duodenal duplication cyst [43]. Combining preoperative and intraoperative imaging methods during surgical intervention planning reduces the need for preoperative and postoperative ERCP or percutaneous transhepatic cholangiography. Indeed, the addition of intraoperative cholangiopancreatography to preoperative MRCP, especially in pediatric patients, effectively determines intrahepatic biliary structures. This, in turn, may change surgical planning and diagnostic classification by reducing invasive preoperative imaging methods [44]. Ultrasound, MRI-MRCP with or without contrast enhancement, and CT are considered in evaluating complications and examining malignant transformation.

2. Clinical presentation and diagnosis

In adults, symptoms are usually nonspecific, vague abdominal pain being the most common [15]. When specific symptoms arise, they are typically of acute biliary tract and pancreatic origin [45]. In some patients, the classic symptom triad of abdominal pain, palpable abdominal mass, and jaundice occurs in only 25% of adults, while 85% of children have at least two features of the classic triad. Unexpected presentations

such as gastric outlet obstruction, cyst perforation, giant cystolithiasis, giant cyst, and mixed type were reported [46]. Although the frequency of emerging symptoms is similar in Western and Eastern populations, associated biliary conditions such as cholecystitis, cholangitis, and choledocholithiasis are more common at presentation in the Eastern population [20, 22]. Recently, ultrasound has been found to have high sensitivity in the examination of biliary tract diseases [47]. Prenatal diagnosis was also defined in some cases at the 15th week of gestation [48]. In the evaluation of intrapancreatic residual choledochal cyst, it is important that the soft tissue neoplasm is found within the cyst wall, and the wall has a uniform and smooth structure, which should be indicated in the imaging [49]. Peripancreatic lymph nodes are classified as abnormal growth in residual choledochal cysts to prevent carcinogenesis misdiagnosis [50]. In this context, spiral CT-type B has higher resolution than ultrasound and MRCP. All three diagnostic methods can be used together. Serum CA19-9 and carcinoembryonic antigen are listed in the routine preoperative examination of choledochal cyst and are used as important reference indices for the prediction of bile duct carcinogenesis [51]. Caroli syndrome may present with right upper quadrant pain or signs of portal hypertension. It is usually a childhood or young adult disease.

3. Classification of common bile duct malformations

It is important to categorize malformations for the purpose of appropriate management and risk classification. First in 1959, Alonso-Lej proposed a classification scheme that included four anomalies of the biliary tree. Later, in 1977, Todani added five anomalies and organized the most commonly used classification system [33, 52]. In the Todani classification, a type I cyst features fusiform or spherical dilatation in the entire extrahepatic bile duct. It is the most common type in both the Western and Eastern populations. It is observed in 65–84% of the Eastern cohort and 67–73% in the Western cohort. It is divided into three within itself: type Ia: diffuse cystic dilatation, type Ib: focal saccular cystic dilatation, and type Ic: diffuse fusiform dilatation.

Type Ib cystic dilatation typically results from the more distal common choledochal segment. Type II cyst, which involves the lateral wall of the common bile duct, is also called an extra-hepatic supraduodenal biliary diverticulum. Type III cyst choledochal is a cystic dilatation of the duodenal intramural segment of the common bile duct. It can also have a mass effect while protruding into the lumen of the duodenum. Todani identified type IV cyst also as multiple extrahepatic biliary cysts. It could be either isolated in extrahepatic bile duct (type IVb) or combined with multiple large intrahepatic biliary cysts (type IVa). Type V cyst, on the other hand, is known as Caroli disease. It is characterized by multiple large and small intrahepatic biliary dilatations. Coexistence of Caroli disease and congenital hepatic fibrosis is called Caroli syndrome. In 2004, Visser proposed a revised classification to facilitate the diagnosis and management of choledochal bile duct malformations [53]. Here, the spectrum of Todani type I and type IV anomalies was classified under the name of congenital choledochal cyst. Type II koledokal divertikül and type III koledokosel. Likewise, depending on the way it manifests, Caroli disease or syndrome was called type V. In the Visser's classification, congenital choledochal cysts (Todani types I and IV) consist of focal or diffuse extrahepatic bile duct dilatation with varying degrees of intrahepatic involvement. It is the most common type of malformation with incidence rates of approximately 1/100,000–1/150,000. Common duct syndrome is the most commonly considered cause. Other proposed causes include increased intraluminal

pressure due to narrow biliary stricture (stenosis), abnormal recanalization during organogenesis, reovirus infection, and insufficient ganglion cells in narrow bile duct part resulting in dilatation [8, 38].

4. Choledochal diverticulum

A choledochal diverticulum (Todani type II) is described as outpouching in the extrahepatic supraduodenal common bile duct. It is rare, has an incidence rate of less than 1/1,000,000 and accounts for approximately 2% of Todani-type malformations [2, 6]. It is a true diverticulum lined with biliary epithelium [2]. Histologically, it is similar to gallbladder duplication. It is associated with the common bile duct with a thin stalk. Increased intraluminal pressure as a result of sphincter of Oddi dysfunction was suggested as the cause.

5. Choledocele

It was first described by Courcy Wheeler in 1940 [54]. Choledochoceles (Todani type III) are poorly understood entities of the choledochal malformations. It is different from other choledochal cysts (CCs). True choledochoceles are the dilated segment of the common bile duct that prolapses or herniates into the small intestine [2, 55]. Their origin has long been the subject of debate. One hypothesis is that these cysts may be caused by an anomaly acquired by the ampulla of Vater's rudimentary lower embryonic bud or by dysfunction or obstruction of the sphincter of Oddi [56]. Although multiple etiological factors are considered, the most commonly blamed factor is inflammation after stone (calculus) passage or damage to the wall. It could be lined with biliary epithelium or ectopic mucosa of the intestine, or it may lie completely bare. It is very rare with an incidence rate of less than 1/1,000,000. It accounts for 1–4% of choledochal malformations [6]. Its differential diagnosis from submucosal duodenal lesions or periampullary duodenal duplication cysts is difficult. The diagnosis is typically made by cholangiography or endoscopic ultrasonography [57].

6. Caroli disease and syndrome

It was first defined by French gastroenterologists in 1958 as congenital multiple intrahepatic cystic dilatation pattern. Caroli disease (Todani type V) is a rare autosomal recessive disorder. It is the second most common type of malformation. It is observed at a ratio of 6–30% in the West and 18–19% in the East. It is the most common type of choledochal malformation with an incidence rate of approximately 1/500,000 in Western countries [2, 6]. Caroli disease is characterized by *in utero* malformation product of the ductal plate and congenital intrahepatic ectasia of the bile duct. When Caroli disease is observed together with congenital hepatic fibrosis, it is called Caroli syndrome [2, 6]. The intrahepatic biliary tract originates from monolayer cells around the portal branches, called the ductal plate [55]. These cell layers are structured to produce small ducts in the periphery and large bile ducts in the hilum. Ductal plate malformation at the level of the great bile duct results in Caroli disease [2, 58]. Imaging studies show varying size of intrahepatic saccular cystic structures associated with the biliary system. On MRI and CT, it appears as ectatic bile ducts

around enlarged portal branches. It is called the central dot sign, which is thought to be highly indicative for the diagnosis of Caroli disease [2]. Complications of Caroli disease include intraductal stone formation due to bile stasis and recurrent cholangitis [2, 6]. Besides causing intrahepatic cysts by blocking the large bile ducts, Caroli syndrome also affects peripheral bile ducts as a result of congenital hepatic fibrosis [2, 59]. Associated conditions include congenital choledochal cysts, cholangiocarcinoma (7–10% incidence), autosomal recessive polycystic kidney disease, and renal cystic diseases involving medullary sponge kidney [2]. Here, the association of renal and hepatic diseases is the common genetic locus (PKHD1 gene on 6p21 chromosomal region) [58].

7. Complications

Complications of common bile duct malformations range from cholestasis with stone formation to recurrent cholangitis, pancreatitis, biliary and hepatic fibrosis, and malignant transformation (cholangiocarcinoma). Cholestasis is the result of external compression of a large extrahepatic cystic malformation causing a regional mass effect or inadequate prolonged biliary drainage by the malformed biliary tract [60]. Stone formation can occur anywhere along the affected biliary system. Recurrent cholangitis and cholecystitis are probably multifactorial. The reasons blamed for this are static lithogenic bile salts as well as chronic inflammation, reflux of pancreatic enzymes, and intestinal bacterial reflux [2]. Regardless of the underlying cause, recurrent cholangitis attacks lead to fibrosis in the cyst wall, ductal sclerosis, and an increased risk of malignant degeneration [6]. Hepatic fibrosis may develop when chronic inflammation affects the intrahepatic biliary system as in congenital choledochal cysts (Todani type IVa) and Caroli disease (Todani type V). Malignant transformation is the best known and feared complication of choledochal malformations. The thesis that chronic inflammation in the malformation-affected biliary system results in cellular dysplasia is assumed to be correct. Chronic inflammation destroys protective mucin-producing epithelial cells. This effect is also enhanced by carcinogen products such as bile salts metabolized by *Escherichia coli*.

The risk of developing malignancy is between 6% and 30% in patients with choledochal cysts. This risk is low in childhood (<1%) but increases by 30–40% at the age of more than 50 years [45, 61]. The malignancy of choledochal cysts originates from both the gallbladder and the choledochal. In a large-scale Japanese study with patients who had choledochal cysts and developed cancer, 62.3% had gallbladder cancer, 32.1% bile duct cancer, and 4.7% had both, which was consistent with other reports [15, 62]. Gallbladder cancer is found in 5% of choledochal cyst patients with PBM [63]. An evaluation of the adult cohort revealed that this is usually observed in the sixth or seventh decade of life [64]. With the cumulative effect of biliary stasis and chronic inflammation itself, the risk of malignancy increases with age [2, 7]. Cholangiocarcinoma develops in a small subgroup of patients (0.7–3%), especially in type I and IV, after surgical resection [18, 22, 65, 66]. This shows that the risk of malignancy does not completely regress after resection in this group of patients. The etiology of this residual malignancy is unclear. Many studies performed in the East and West do not have follow-up periods long enough to determine persistent risk because it takes more than three decades to occur [67, 68]. However, currently, there is a 6% risk of malignancy after complete cyst resection, while there is a 33% risk of malignancy after incomplete cyst resection [69]. The most common type of cancer is

adenocarcinoma, followed by anaplastic carcinoma, undifferentiated carcinoma, and squamous cell carcinoma [70]. About 70% of malignancies originate from the cystic wall. Cholangiocarcinoma in adults and rhabdomyocarcinoma or adenocarcinoma in children are frequently observed. Twenty-four percent of them originate from the gallbladder. The remaining 6% are hepatocellular carcinoma and pancreatic carcinoma [7, 70, 71]. Intrahepatic cysts have a low malignant potential depending on the extent of the involved area and the extent of inflammation and hepatic parenchymal atrophy [20, 72, 73]. Among the different types of choledochal malformations, extrahepatic choledochal cysts (Todani types I and IV) have the highest risk of developing cancer. They are followed by intrahepatic choledochal cysts (Todani type IVa), Caroli disease (Todani type V), choledochal diverticulum (Todani type II), and choledochocele (Todani type III). A surveillance strategy is proposed for patients treated primarily for cyst types I and IV and unresected type V using annual liver function tests, Ca 19–9 measurement and biannual ultrasound assessment for 20 years post cyst resection, with biannual liver function testing, Ca 19–9 measurement and 3-yearly ultrasound assessment thereafter [74]. it has been opined that long- term follow-up strategies might not be associated with a better prognosis. One report suggested that follow-up with regular clinic reviews alone does not affect the resectability of cholangiocarcinoma [75].

8. Surgical treatment

Surgical treatment should be based on the extent of biliary involvement based on the widely used Todani classification and anatomical findings and the presence or absence of PBM [76]. The treatment goal for choledochal malformations is to eliminate the risk of malignancy and to treat complications. Surgical treatment depends on the localization and size of the cyst rather than the morphological features [53, 55, 72]. The standard treatment in most CCs is the resection of the bile duct up to the lobar bifurcation, specifically toward the pancreatic parenchyma close to the pancreatic duct junction. Biliary tract continuity is provided by roux-en-Y hepaticojejunostomy (HJ), hepaticoduodenostomy (HD), or jejunal interposition [64, 77, 78]. Hepaticoduodenostomy presents an effective approach to biliary reconstruction following surgical interventions for choledochal cysts. It presents a faster alternative to hepa ticojejunostomy in both operative times and length of stay, with similar rates of complications, including the feared cholangitis [79]. Proximal cyst excision is a non-precisely programmable procedure that needs attention to preserve the portal vein and hepatic artery. Although extrahepatic congenital choledochal cysts usually require complete excision of the entire extrahepatic biliary tree using cholecystectomy and hepaticojejunostomy, more conservative surgical options can be used if there is limited involvement in the distal bile duct, and the interventional approach is based on a 3-cm-size threshold [5, 60]. When the cyst extends into the pancreas, its management is critical and sometimes challenging. During recurrent cholangitis attacks, adhesions in the tissues surrounding the portal vein and pancreas could develop. For these reasons, complete excision may not be possible due to peritoneal infection, bleeding, and the risk of postoperative pancreatic leak [80–82]. The residual intrapancreatic portion of the choledochal cyst hosts multiple pathological changes such as pancreatitis, secondary calculus in the bile duct, and even carcinogenesis [83, 84]. Acute pancreatitis is often secondary to the intrapancreatic portion of the postoperative residual choledochal cyst [85]. Residual postoperative intrapancreatic choledochal cyst may also lead to secondary carcinogenesis and associated

morbidity. Some previous reports showed that stone formation not only stimulates cancer, but also some carcinogenic factors may exist in the residual intrapancreatic part of the choledochal cyst [86]. These findings indicated the necessity of surgical excision of asymptomatic residual choledochal cysts. Intrapancreatic choledochal cysts carry a 0.7–6.0% risk of malignant transformation [5]. There has been no consensus so far on whether excision of the intrapancreatic common bile duct cyst is necessary if there are no clinical symptoms. At the same time, the optimal timing of surgery is unclear. The proximal and distal endpoint of the biliary malformation including involvement of the ampulla of Vater expanding into the pancreatic head should be determined preoperatively [72]. Generally, the localization of the pancreatic cyst is inside the head of the pancreas, close to the neck and to the left of the bile duct [87]. Investigation and differentiation of a residual intrapancreatic choledochal cyst should be started from the back and right side of the pancreatic head. Then, it is gradually deepened into the left field. During this procedure, the direction of the bile duct is confirmed at intervals by fine needle aspiration and finger control [88]. The separation should be close to the bile duct wall. When the lumen terminates abruptly near the end of the bile duct, attention should be paid to the junction of the pancreatic and bile ducts. The diagnosis of a residual intrapancreatic cyst is relatively easy with type-B ultrasonography, CT, and magnetic resonance cholangiopancreatography (MRCP) [89]. Preoperative MRCP examination is crucial to prevent pancreatic duct injury during surgery. It clearly shows the specific anatomy of the pancreatobiliary junction [90]. Different cholangiopancreatography angles of duct reveal the angle of the cholangio-pancreatic duct junction, the rough flow direction to the main pancreatic duct, and the length of the stenosed portion of the lower portion of the bile duct [91].

Because of the high risk of malignancy arising from inadequate resection of the mucosa, any residual area should be excised and always followed up regularly. It is difficult to distinguish pathological changes in choledochal cysts from the inflammatory bile duct. The extent of the excision is mainly based on radiological and intraoperative findings. Surgical treatment is ideally adopted for type I CC. In Todani type IV with intrahepatic and extrahepatic cysts, extrahepatic cysts should be definitively excised. If intrahepatic cysts are limited, biliary-enteric reconstruction is performed along with partial hepatectomy. Especially in patients with bilobar involvement and with diffuse intrahepatic dilatation associated with complications such as stones, cholangitis, and biliary cirrhosis, liver transplantation should be considered [92, 93]. In the distal management of choledochal cysts, if there is no relationship between the choledochal cyst and the pancreatic duct in the preoperative imaging, the distal cyst is excised at a distance of about 5 mm at the junction with the pancreatic duct of the visible bile duct, and the stump is sutured [94]. If the choledochal cyst extends to the junction of the pancreatic duct, the same procedure is adopted. Sometimes, the insufficiency of the ampulla of Vater is observed along with a cylindrical cyst. The distal end of the choledochal cyst opens directly into the duodenum with the insufficient ampulla, and the pancreatic duct appears to be attached to the cyst. In this case, the cylindrical cyst and papilla are excised, and duodenum mucosa and pancreatic duct are subjected to ductoplasty as a modified procedure of local excision of early noninvasive adenocarcinoma and benign lesions in the papilla. Secondary infections may require antibiotics, drainage, and sometimes surgery. Partial hepatectomy and liver transplantation are disputable. However, it is successful in the presence of diffuse intrahepatic ductal dilatation, stenosis, and multiple intraluminal stones [72]. If resection is not performed, long-term follow-up of intrahepatic biliary cysts is required [20, 72]. Despite their low malignant potential, choledochal diverticula

(Todani type II) are prophylactically excised to prevent the sequelae of compression effect on adjacent structures [55]. Usually, these cysts are ligated at the neck and excised without the need for reconstruction of bile duct [12]. However, sometimes extrahepatic bile duct excision may be required. In this case, sometimes patients with tight adhesion of the diverticulum to the extra- or intrahepatic biliary tract due to inflammation may be encountered. Releasing the diverticulum from the bile duct could be technically challenging [95]. The isolated segment is removed, and primary repair is performed via T-tube with a low recurrence rate without the need for cholecystectomy [60, 96]. Choledochoceles, which are very similar to choledochal diverticulum, have a low malignant potential of approximately 2.5% throughout life [5]. If they cause biliary or intestinal obstruction, they are excised [6, 55, 56]. Due to the low risk of malignancy, unroofing (endoscopic or transduodenal sphincteroplasty) or transduodenal excision (in large cysts) is the optimal treatment. Endoscopic monitoring should be considered for younger patients treated with sphincterotomy. The prognosis of Caroli disease or syndrome depends on the extent of involvement, but is generally poor. Localized disease is treated with prophylactic hepatic lobectomy. Recurrent and life-threatening cholangitis, liver failure, cirrhosis, portal hypertension, or malignant disease requires orthotopic liver transplantation [97, 98]. Surgical treatment options include laparoscopic treatment. Its main advantages include excellent visualization and low blood loss [99]. In addition, it has better postoperative recovery, less surgical trauma and postoperative pain, less abdominal wall trauma, less cavity drainage time, reduced postoperative paralytic ileus time, and shorter hospital stay. The overall complication and mortality rates are also lower compared with the series involving treatment with open surgery [100, 101]. Recent report supports HD as an effective alternative to the conventional Roux-en-Y HJ reconstruction in laparoscopic excision of choledochal cyst in children [102]. Exposure of the anastomotic site and IHBD is very difficult to achieve by conventional endoscopy. DBE provides a direct view of these sites and enables diagnostic assessment and minimally invasive therapy to be performed simultaneously. It will replace more invasive treatments such as percutaneous transhepatic intervention and surgical procedures [103].

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References

[1] Liu Y, Yao X, Li S, et al. Comparison of therapeutic effects of laparoscopic and open operation for congenital choledochal cysts in adults. Gastroenterology Research and Practice. 2014;**2014**:670260

[2] Singham J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 1. Classification and pathogenesis. Canadian Journal of Surgery. 2009;**52**:434-440

[3] Liu CL, Fan ST, Lo CM, et al. Choledochal cysts in adults. Archives of Surgery. 2002;**137**:465-468

[4] Levy AD, Rohrmann CA. Biliary cystic disease. Current Problems in Diagnostic Radiology. 2003;**32**:233-263

[5] Singham J, Yoshida EM, Scudamore CH. Choledochal cysts. Part 3. Management. Canadian Journal of Surgery. 2010;**53**:51-56

[6] Soares KC, Arnaoutakis DJ, Kamel I, et al. Choledochal cysts: Presentation, clinical differentiation, and management. Journal of the American College of Surgeons. 2014;**219**: 1167-1180

[7] Sastry AV, Abbadessa B, Wayne MG, Steele JG, Cooperman AM. What is the incidence of biliary carcinoma in choledochal cysts, when do they develop, and how should it affect management? World Journal of Surgery. 2015;**39**:487-492

[8] Katabathina VS, Kapalczynski W, Dasyam AK, Anaya-Baez V, Menias CO. Adult choledochal cysts: Current update on classification, pathogenesis, and cross-sectional imaging findings. Abdominal Imaging. 2015;**40**:1971-1981

[9] Kimura K, Ohto M, Ono T, Tsuchiya Y, Saisho H, Kawamura K, et al. Congenital cystic dilatation of the common bile duct: Relationship to anomalous pancreaticobiliary ductal union. AJR. American Journal of Roentgenology. 1977;**128**(4):571-577

[10] Funabiki T, Matsubara T, MiyakawaS, IshiharaS. Pancreaticobiliary maljunction and carcinogenesis to biliary and pancreatic malignancy. Langenbeck's Archives of Surgery. 2009;**394**:159-169

[11] Nicholl M, Pitt HA, Wolf P, Cooney J, Kalayoglu M, Shilyansky J, et al. Choledochal cysts in western adults: Complexities compared to children. Journal of Gastrointestinal Surgery. 2004;**8**:245-252

[12] Babbitt DP. Congenital choledochal cysts: New etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Annales de Radiologie (Paris). 1969;**12**:231-240

[13] García Cano J, Godoy MA, Morillas Ariño J, Pérez García JI. Pancreatobiliary maljunction. Not only an eastern disease. Anales de Medicina Interna. 2007;**24**:384-386

[14] Kamisawa T, Honda G, Kurata M, Tokura M, Tsuruta K. Pancreatobiliary disorders associated with pancreaticobiliary maljunction. Digestive Surgery. 2010;**27**:100-104

[15] Bhavsar MS, Vora HB, Giriyappa VH. Choledochal cysts: A review of literature. Saudi Journal of Gastroenterology. 2012;**18**:230-236

[16] Lipsett PA, Pitt HA, Colombani PM, et al. Choledochal cyst disease. A

changing pattern of presentation. Annals of Surgery. 1994;**220**:644-652

[17] Shi LB, Peng SY, Meng XK, Peng CH, Liu YB, Chen XP, et al. Diagnosis and treatment of congenital choledochal cyst: 20 years' experience in China. World Journal of Gastroenterology. 2001;**7**:732-734

[18] Soares KC, Kim Y, Spolverato G, Maithel S, Bauer TW, Marques H, et al. Presentation and clinical outcomes of Choledochal cysts in children and adults: A multi-institutional analysis. JAMA Surgery. 2015;**150**:577-584

[19] Kinoshita H, Nagata E, Hirohashi K, Sakai K, Kobayashi Y. Carcinoma of the gallbladder with an anomalous connection between the choledochus and the pancreatic duct. Report of 10 cases and review of the literature in Japan. Cancer. 1984;**54**:762-769

[20] He XD, Wang L, Liu W, Liu Q, Qu Q, Li BL, et al. The risk of carcinogenesis in congenital choledochal cyst patients: An analysis of 214 cases. Annals of Hepatology. 2014;**13**:819-826

[21] Baumann R, Uettwiller H, Duclos B, Jouin H, Kerschen A, Adloff M, et al. Congenital cystic dilatation of the common bile duct, anomaly of the biliopancreatic junction and cancer of the bile ducts. Gastroentérologie Clinique et Biologique. 1987;**11**(12):849-855

[22] Lee SE, Jang JY, Lee YJ, et al. Choledochal cyst and associated malignant tumors in adults: A multicenter survey in South Korea. Archives of Surgery. 2011;**146**(10):1178-1184

[23] Tashiro S, Imaizumi T, Ohkawa H, Okada A, Katoh T, Kawaharada Y, et al. Pancreaticobiliary maljunction: Retrospective and nationwide survey in Japan. Journal of Hepato-Biliary-Pancreatic Surgery. 2003;**10**(5):345-351

[24] Kamisawa T, Ando H, Suyama M, Shimada M, Morine Y, Shi-Mada H, et al. Japanese clinical practice guidelines for pancreaticobiliary maljunction. Journal of Gastroenterology. 2012;**7**(7):731-759

[25] Komi N, Udaka H, Ikeda N, Kashiwagi Y. Congenital dilatation of the biliary tract; new classification and study with particular reference to anomalous arrangement of the pancreaticobiliary ducts. Gastroenterologia Japonica. 1977;**12**(4):293-304

[26] Okada A, Hasegawa T, Oguchi Y, Nakamura T. Recent advances in pathophysiology and surgical treatment of congenital dilatation of the bile duct. Journal of Hepato-Biliary-Pancreatic Surgery. 2002;**9**:342-351

[27] Imazu M, Iwai N, Tokiwa K, Shimotake T, Kimura O, Ono S. Factors of biliary carcinogenesis in choledochal cysts. European Journal of Pediatric Surgery. 2001;**11**:24-27

[28] Mabrut JY, Kianmanesh R, Nuzzo G, Castaing D, Boudjema K, Le'toublon C, et al. Surgical management of congenital intrahepatic bile duct dilatation, Caroli's disease and syndrome: Long-term results of the French Association of Surgery Multicenter study. Annals of Surgery. 2013;**258**(5):713-721

[29] Okada A. Forme fruste choledochal cyst. Journal of Pediatric Surgery. 1986;**21**(4):383

[30] Shimotakahara A, Yamataka A, Kobayashi H, Okada Y, Yanai T, Lane GJ, et al. Forme fruste choledochal cyst: Long-term follow-up with special reference to surgical technique. Journal of Pediatric Surgery. 2003;**38**(12):1833-1836

[31] Kamisawa T, Anjiki H, Egawa N, Kurata M, Honda G, Tsuruta K. Diagnosis and clinical implications of pancreatobiliary reflux. World Journal of Gastroenterology. 2008;**14**(43):6622-6626

[32] Misra SP, Dwivedi M. Pancreaticobiliary ductal union. Gut. 1990;**31**(10):1144-1149

[33] Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. American Journal of Surgery. 1977;**134**(2):263-269

[34] Todani T, Watanabe Y, Toki A, et al. Carcinoma related to choledochal cysts with internal drainage operations. Surgery, Gynecology and Obstetrics. 1987;**164**:61-64

[35] Flanigan DP. Biliary carcinoma associated with biliary cysts. Cancer. 1977;**40**:880-883

[36] Soares KC, Goldstein SD, Ghaseb MA, Kamel I, Hackam DJ, Pawlik TM. Pediatric choledochal cysts: Diagnosis and current management. Pediatric Surgery International. 2017;**33**:637-650

[37] Rozel C, Garel L, Rypens F, et al. Imaging of biliary disorders in children. Pediatric Radiology. 2011;**41**:208-220

[38] Lewis VA, Adam SZ, Nikolaidis P, et al. Imaging of choledochal cysts. Abdominal Imaging. 2015;**40**:1567-1580

[39] Park DH, Kim MH, Lee SK, Lee SS, Choi JS, Lee YS, et al. Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts? Gastrointestinal Endoscopy. 2005;**62**:360-366

[40] Park KB, Auh YH, Kim JH, et al. Diagnostic pit falls in the cholangiographic diagnosis of choledochoceles: Cholangiographic quality and its effect on visualization. Abdominal Imaging. 2001;**26**:48-54

[41] Irie H, Honda H, Jimi M, Yokohata K, Chijiiwa K, Kuroiwa T, et al. Value of MR cholangiopancreatography in evaluating choledochal cysts. AJR. American Journal of Roentgenology. 1998;**171**:1381-1385

[42] Huang CT, Lee HC, Chen WT, Jiang CB, Shih SL, Yeung CY. Usefulness of magnetic resonance cholangiopancreatography in pancreatobiliary abnormalities in pediatric patients. Pediatrics and Neonatology. 2011;**52**(6):332-336

[43] Han BK, Babcock DS, Gelfand MH. Choledochal cyst with bile duct dilatation: Sonography and 99mTc IDA cholescintigraphy. AJR. 1981;**136**:1075-1079

[44] Saito T, Terui K, Mitsunaga T, Nakata M, Yoshida H. Significance of imaging modalities for preoperative evaluation of the pancreaticobiliary system in surgery for pediatric choledochal cyst. Journal of Hepato-Biliary-Pancreatic Sciences. 2016;**23**:347-352

[45] Edil BH, Cameron JL, Reddy S, Lum Y, Lipsett PA, Nathan H, et al. Choledochal cyst disease in children and adults: A 30-year single-institution experience. Journal of the American College of Surgeons. 2008;**206**:1000-1005; discussion 1005-8

[46] Gupta N, Gupta V, Noushif M, et al. Unusual presentations of choledochal cyst: Case series and review of literature. The Indian Journal of Surgery. 2015;**77**:1318-1322

[47] Forny DN, Ferrante SM, Silveira VG, Siviero I, Chagas VL, Méio IB. Choledochal cyst in childhood: Review of 30 cases. Revista do Colégio Brasileiro de Cirurgiões. 2014;**41**(5):331-335

[48] Subramony R, Kittisarapong N, Barata I, Nelson M. Choledochal cyst mimicking gallbladder with stones in a six-year-old with right-sided abdominal pain. The Western Journal of Emergency Medicine. 2015;**16**(4):568-571

[49] Shindo K, Aishima S, Ohuchida K, et al. Podoplanin expression in the cyst wall correlates with the progression of intraductal papillary mucinous neoplasm. Virchows Archiv. 2014;**465**:265-273

[50] Gadelhak N, Shehta A, Hamed H. Diagnosis and management of choledochal cyst: 20 years of single center experience. World Journal of Gastroenterology. 2014;**20**:7061-7066

[51] Chen W, Liu Q, Tan SY, et al. Association between carcinoembryonic antigen, carbohydrate antigen 19-9 and body mass index in colorectal cancer patients. Molecular and Clinical Oncology. 2013;**1**:879-886

[52] Alonso-Lej F, Rever WB, Pessagno DJ. Congenital choledochal cyst, with a report of 2, and an analysis of 94, cases. International Abstracts of Surgery. 1959;**108**:1-30

[53] Visser BC, Suh I, Way LW, Kang SM. Congenital choledochal cysts in adults. Archives of Surgery. 1960;**2004**(139):855-860; discussion, 860-862

[54] de Courcy Wheeler WI. An unusual case of obstruction to the common bile-duct (choledochocele?). BJS. 1940;**27**:446-448

[55] Martin RF. Biliary cysts: A review and simplified classification scheme. The Surgical Clinics of North America. 2015;**94**:219-232

[56] Law R, Topazian M. Diagnosis and treatment of choledochoceles. Clinical Gastroenterology and Hepatology. 2014;**12**:196-203

[57] Scholz FJ, Carrera GF, Larsen CR. The choledochocele: Correlation of radiological, clinical and pathological findings. Radiology. 1976;**118**:25-28

[58] Sato Y, Ren XS, Nakanuma Y. Caroli's disease: Current knowledge of its biliary pathogenesis obtained from an orthologous rat model. Internaticonal Journal of Hepatology. 2012;**2012**:1-10

[59] Bismuth H, Krissat J. Choledochal cystic malignancies. Annals of Oncology. 1999;**10**(suppl 4):94-98

[60] Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. Journal of Hepato-Biliary-Pancreatic Surgery. 2003;**10**:352-359

[61] Søreide K, Søreide JA. Bile duct cyst as precursor to biliary tract cancer. Annals of Surgical Oncology. 2007;**14**:1200-1211

[62] Mori H, Iida H, Maehira H, Kitamura N, Shimizu T, Tani M. Synchronous primary gallbladder and pancreatic cancer associated with congenital biliary dilatation and pancreaticobiliary maljunction. Surgical Case Reports. 2017;**3**:113

[63] Chijiiwa K, Kimura H, Tanaka M. Malignant potential of the gallbladder in patients with anomalous pancreaticobiliary ductal junction. The difference in risk between patients with and without choledochal cyst. International Surgery. 1995;**80**:61-64

[64] Ishibashi H, Shimada M, Kamisawa T, Fujii H, Hamada Y, Kubota M, et al. Japanese clinical practice guidelines for congenital biliary dilatation. Journal of Hepato-Biliary-Pancreatic Sciences. 2017;**24**:1-16

[65] Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. Journal of Hepato-Biliary-Pancreatic Surgery. 1999;**6**:207-212

[66] Ng WT. Bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. Surgery. 2000;**128**:492-494

[67] Takeshita N, Ota T, Yamamoto M. Forty-year experience with flow-diversion surgery for patients with congenital choledochal cysts with pancreaticobiliary maljunction at a single institution. Annals of Surgery. 2011;**254**:1050-1053

[68] Kobayashi S, Asano T, Yamasaki M, Kenmochi T, Nakagohri T, Ochiai T. Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. Surgery. 1999;**126**:939-944

[69] Liu YB, Wang JW, Devkota KR, Ji ZL, Li JT, Wang XA, et al. Congenital choledochal cysts in adults: Twenty-five-year experience. Chinese Medical Journal. 2007;**120**:1404-1407

[70] Fieber SS, Nance FC. Choledochal cyst and neoplasm: A comprehensive review of 106 cases and presentation of two original cases. The American Surgeon. 1997;**63**:982-987

[71] Todani T, Tabuchi K, Watanabe Y, Kobayashi T. Carcinoma arising in the wall of congenital bile duct cysts. Cancer. 1979;**44**:1134-1141

[72] Ronnekleiv-Kelly SM, Soares KC, Ejaz A, Pawlik TM. Management of choledochal cysts. Current Opinion in Gastroenterology. 2016;**32**:225-231

[73] Ohashi T, Wakai T, Kubota M, et al. Risk of subsequent biliary malignancy in patients undergoing cyst excision for congenital choledochal cysts: Biliary malignancy after cyst excision. Journal of Gastroenterology and Hepatology. 2013;**28**:243-247

[74] Koea J, O'Grady M, Agraval J, Srinivasa S. Defining an optimal surveillance strategy for patients following choledochal cyst resection: Results of a systematic review. ANZ Journal of Surgery. 2022;**92**(6):1356-1364

[75] Ng DW, Chiow AK, Poh WT, Tan SS. Metachronous cholangiocarcinoma 13 years post resection of choledochal cyst-is long-term follow-up useful?: A case study and review of the literature. Surgical Case Reports. 2016;**2**:60

[76] Le RB, Gagnière J, Filaire L, et al. Pancreaticobiliary maljunction and choledochal cysts: From embryogenesis to therapeutics aspects. Surgical and Radiologic Anatomy. 2016;**38**:1053-1060

[77] Moslim MA, Takahashi H, Seifarth FG, Walsh RM, Morris-Stiff G. Choledochal cyst disease in a Western center: A 30-year experience. Journal of Gastrointestinal Surgery. 2016;**20**:1453-1463

[78] Kim Y, Hyun JJ, Lee JM, Lee HS, Kim CD. Anomalous union of the pancreaticobiliary duct without choledochal cyst: Is cholecystectomy alone sufficient? Langenbeck's Archives of Surgery. 2014;**399**:1071-1076

[79] Gonzalez DEH, Medrano AR, De Leon SUV. Biliary reconstruction after choledochal cyst resection: A systematic review and meta-analysis on hepaticojejunostomy

vs hepaticoduodenostomy. Pediatric Surgery International. 2021;**37**(10):1313-1322

[80] Saluja SS, Nayeem M, Sharma BC, et al. Management of choledochal cysts and their complications. The American Surgeon. 2012;**78**:284-290

[81] Jordan PH, Goss JA, Rosenberg WR, et al. Some considerations for management of choledochal cysts. American Journal of Surgery. 2004;**187**:790-795

[82] Cho MJ, Hwang S, Lee YJ, et al. Surgical experience of 204 cases of adult choledochal cyst disease over 14 years. World Journal of Surgery. 2011;**35**:1094-1102

[83] Wang WJ, Lv ZD, Yang ZC, et al. Prrx1 enhanced the migration ability of breast cancer cell by initiating epithelial-mesenchymal transition. Cancer Cell Research. 2017;**4**:362-367

[84] Chen TT, Liu YQ, Lin BD, et al. Effects of RNA interference of the eno1 gene on the malignant biological behaviors of gastric cell lines. Cancer Cell Research. 2017;**4**:326-333

[85] Ando H, Kaneko K, Ito T, et al. Complete excision of the intrapancreatic portion of choledochal cysts. Journal of the American College of Surgeons. 1996;**183**:317-321

[86] Fan F et al. Clinical significance of intrapancreatic choledochal cyst excision in surgical management of type I choledochal cyst. The Journal of International Medical Research. Mar 2018;**46**(3):1221-1229

[87] Ryu M, Takayama W, Watanabe K, et al. Ventral pancreatic resection for adenoma and low-grade malignancies of the head of the pancreas. Surgery Today. 1996;**26**:476-481

[88] Kolb A, Kleeff J, Frohlich B, et al. Resection of the intrapancreatic bile duct preserving the pancreas. Journal of Hepato-Biliary-Pancreatic Surgery. 2009;**16**:31-34

[89] Shimizu T, Suzuki R, Yamashiro Y, et al. Magnetic resonance cholangiopancreatography in assessing the cause of acute pancreatitis in children. Pancreas. 2001;**22**:196-199

[90] Yoon JH. Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the cystic duct: Report of three cases. The British Journal of Radiology. 2011;**84**:e18-e22

[91] Jara H, Barish MA, Yucel EK, et al. MR hydrography: Theory and practice of static fluid imaging. AJR. American Journal of Roentgenology. 1998;**170**:873-882

[92] Xia HT, Dong JH, Yang T, Zeng JP, Liang B. Extrahepatic cyst excision and partial hepatectomy for Todani type IV-A cysts. Digestive and Liver Disease. 2014;**46**:1025-1030

[93] Cerwenka H. Bile duct cyst in adults: Interventional treatment, resection, or transplantation? World Journal of Gastroenterology. 2013;**19**:5207-5211

[94] Xia HT, Yang T, Liang B, et al. Treatment and outcomes of adults with remnant intrapancreatic choledochal cysts. Surgery. 2016;**159**:418-425

[95] Ouaïssi M, Kianmanesh R, Belghiti J, Ragot E, Mentha G, Adham M, et al. Gigot JF; working Group of the French Surgical Association. Todani type II congenital bile duct cyst: European multicenter study of the French surgical association and literature review. Annals of Surgery. 2015;**262**:130-138

[96] Wiseman K, Buczkowski AK, Chung SW, FrancoEur J, Schaeffer D,

Scudamore CH. Epidemiology, presentation, diagnosis, and outcomes of choledochal cysts in adults in an urban environment. American Journal of Surgery. 2005;**189**:527-531; discussion, 531

[97] Bockhorn M, Malagó M, Lang H, et al. The role of surgery in Caroli's disease. Journal of the American College of Surgeons. 2006;**202**:928-932

[98] Habib S, Shakil O, Couto OF, et al. Caroli's disease and orthotopic liver transplantation. Liver Transplantation. 2006;**12**:416-421

[99] Liuming H, Hongwu Z, Gang L, Jun J, Wenying H, Wong KK, et al. The effect of laparoscopic excision vs open excision in children with choledochal cyst: A midterm follow-up study. Journal of Pediatric Surgery. 2011;**46**(4):662-665

[100] Liem NT, Pham HD, Vu HM. Is the laparoscopic operation as safe as open operation for choledochal cyst in children? Journal of Laparoendoscopic & Advanced Surgical Techniques. Part A. 2011;**21**(4):367-370

[101] Wang B, Feng Q, Mao JX, Liu L, Wong KK. Early experience with laparoscopic excision of choledochal cyst in 41 children. Journal of Pediatric Surgery. 2012;**47**(12):2175-2178

[102] Yeung F, Fung ACH, Chung PHY, Wong KKY. Short-term and long-term outcomes after roux- en-Y hepaticojejunostomy versus hepaticoduodenostomy following laparoscopic excision of choledochal cyst in children. Surgical Endoscopy. May 2020;**34**(5):2172-2177

[103] Mukai M, Kaji T, Masuya R. Long-term outcomes of surgery for choledochal cysts: A single-institution study focusing on follow-up and late complications. Surgery Today. 2018;**48**(9):835-840