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Chapter

Pediatric Choledochal Cysts: Unknowns are Decreasing

Hasan Özkan Gezer

Abstract

Choledochal cysts (CCs) are congenital cystic dilatation of extrahepatic and/or intrahepatic bile ducts. CCs are more common in Asian population, the cause is still unknown. Although the etiology is controversial, the main elements in the natural historical emergence of the type I and type IV, which make up the majority of all types, have become clearer. The majority of CCs are diagnosed in childhood. Clinical presentation varies from jaundice in young patients to nonspecific abdominal pain in older, but morbidity increases with complications such as cholangitis, pancreatitis, perforation, hepatitis, liver failure, and malignancy in delayed diagnosed patients. MRCP is considered the current gold standard diagnostic modality that is able to accurately assess biliary anatomy. Although the treatment approach has been formed over the years, it still has not reached the last state. Eventually, the removal of the entire cyst and the reconstruction of the remaining biliary tract to drainage is the current treatment approach. But the dilemma is the way of reconstruction procedure (hepaticoduodenostomy or hepaticojejunostomy). All patients should be followed up for a long period of time, regardless of the surgery method.

Keywords: choledochal cyst, children, hepaticoduodenostomy, Roux-en-Y hepaticojejunostomy, common bile duct

1. Introduction

Choledochal cysts (CCs) are congenital dilatations of extrahepatic and/or intrahepatic bile ducts defined by Vater and Ezler in 1723. It is a rare biliary entity with an estimated incidence of 1:100–150,000 live births in Western countries. In the Asian population, the incidence can be as high as 1:1000 live births. CC is primarily a childhood disease—up to 80% of patients are diagnosed before 10 years of age. The original classification, first described by Alanso-Lej and colleagues in 1959, was changed in 1977 by Todani and colleagues that classified the CCs as five types. In addition, isolated cystic dilatation of the cystic canal was identified subsequently and proposed as type VI, apart from the revised Todani classification. Although the etiology is controversial, the main elements in the natural historical emergence of the type I and type IV, which make up the majority of all types, have become clearer. A common symptom is nonspecific abdominal pain in older children. When the cyst is complicated, the diagnosis is delayed, the treatment becomes complex, and the results are affected. External drainage (ED), internal drainage (ID), total cyst excision (CE) + hepaticoduodenostomy (HD), and total CE + hepaticojejunostomy (HJ) were defined according to historical development of treatment. Resection is considered that is necessary to prevent further complications and long-term sequel.

2. History

The anatomist Abraham Vater first described the normal anatomy of the bile ducts and the fusiform dilatation of the common bile duct (CBD) in 1723 [1]. Then, Doctor Halliday Douglas first described clinically CBD dilatation in 1852. Douglas had detected a large tenderness cystic mass on the right upper quadrant by physical examination of a 17-year-old girl who had an intermittent right-sided pain, obstructive jaundice, and fever complaints in her history. Despite performing external drainage promptly, she died within 1 month. Subsequently, Douglas detected a CC with her autopsy [2]. In 1894, British surgeon William Swain performed the first successful operation in a 17-year-old girl presented with CC, by anastomosing the jejunum to a giant CC. This patient had been reported 2 months later with no jaundice. In 1922, Golder McWhorter underwent hepaticoduodenostomy after excising the CC in a 49-year-old patient who had complaints since infancy. In 1959, Alanso-Lej and colleagues first published the series of CCs. In this publication, they reviewed 94 cases, published previously, together with their own 2 cases and classified the congenital cystic dilatations of the bile ducts anatomically for the first time [3]. In 1977, Todani and colleagues modified the classification of CCs according to cholangiographic images [4–8].

3. Incidence

CCs are more common in Asian populations with an incidence of 1 in 13,000, even 1:1000 in Japan [9], versus 1 in 100,000–150,000 in Western populations [10]. The reason for this Asian preponderance is still unknown [11]. Although predominately diagnosed in children, CCs are found with increasing frequency in adults such that adults comprise the majority of patients in recent series [12, 13], which in part may be due to the increased use of diagnostic imaging [14]. In both adult and children [15], females are higher risk for the disease with a nearly 4:1 female preponderance [10, 16]. Nearly, 80% of CCs are diagnosed in early infancy [10, 17].

4. Pathology

Grossly, CCs appear as a diffuse dilatation of the bile ducts [18]. In the congenital dilatations of the bile ducts, the cyst wall thickness is between 2 and 7 mm in diameter and usually involves an inflammatory reaction (80%) [18] that becomes severe after 10 years of age. Cysts, especially infected, are usually being adherent to the surrounding tissues. Bile ducts and columnar epithelium can be seen in the microscopic examination of the cyst wall. Choledochocele that is covered with duodenal mucosa appears different from other types of CCs according to epithelial histology. Liver biopsy findings usually vary with the age in patients with CC. Although, newborn liver is mostly normal, mild periportal fibrosis may be seen in older children. Varying degrees of histological hepatic changes severity may be seen in most patients with CC when it is diagnosed [19]. Higher degree of liver damage associates with the presence of an anomalous pancreaticobiliary ductal union (APBDU), more severe symptoms, type IVa CC, and younger age [19–21]. It has been observed by investigating liver biopsies that most of these changes resolve after surgical excision; however, preoperative portal fibrosis and central venous distension may remain stable or increase in severity [19]. It is considered that carcinoma of the cyst wall may develop due to recurrent chronic inflammation attacks.

However, it can also be seen in non-APBDU-associated CC patients. Additionally, the cancer can develop from anywhere such as the cyst wall, the gall bladder, or the common channel junction nearly pancreas, as a consequence of chronic inflammation due to cholangitis [22, 23].

5. Etiology

Despite the existence of numerous theories and laboratory works to explain the etiology of the disease, the exact etiology remains incompletely understood. Initial theories, put forward in this regard, were congenital weakness of the choledochal wall, distal obstruction, oligoganglionosis, and disturbances in the process of recanalization. However, more accepted theories have been produced parallel to the progress in radiological imaging methods [24, 25]. Today, there are two main theories that are widely accepted; (1) reflux of trypsin and other pancreatic enzymes to the bile ducts due to an APBDU; (2) obstruction of distal CBD [26–28]. The idea that the choledochal and pancreatic ducts' abnormality about joint and angle was first reviewed by Babbitt in 1969, and subsequently many number of studies, supporting this view, were made [22, 23, 29–32]. A normal pancreaticobiliary junction usually has an acute angle between the CBD and the pancreatic duct [33] and is located within the duodenal wall [34]. The common channel (distance from between the junction of the CBD and the pancreatic duct to ampulla Vater) length is 4 mm or less, normally. In patients with CCs, this distance (common channel length) increases 5–20 mm [22, 23, 35, 36] that makes the common channel longer [37]. Okada defined it as "common channel syndrome" [36, 38]. It is considered that an abnormal long common channel (especially >15 mm proximal to ampullary sphincter) [39, 40] causes the pancreatic duct communicating with the choledochal duct without the support of ampulla Vater's circular muscular layer (sphincter of Oddi) [41], which protects the biliary tree from reflux of pancreatic enzymes and bile [34]. Another observation is that the junction angle of the two ducts that should be acute normally, however, is close to 90° in these patients. It also causes the pancreatic fluids to flow into the CBD due to the higher pressure of pancreatic duct (**Figure 1**) [41]. Eventually, it is considered that APBDU has a tendency to cause reflux of the pancreatic enzyme into biliary tree with consequent biliary duct inflammation and increased duct pressure, leading to duct wall damage and cystic changes [42, 43]. In animal models of murine APBDU, this mechanism has also been demonstrated [44, 45]. APBDU is seen in up to 90% of patients with CC [25, 46], compared with 2% in the general population [10] and this seems to have important clinical implications. In a comparison of APBDU-associated CC versus non-APBDU-associated CC, APBDU-associated CC patients were significantly more likely to have evidence of pathologically confirmed inflammation including hepatitis, cholangitis, and pancreatitis [47, 48]. However, APBDU is not enough to explain the etiology of all CCs, such as diverticular cyst (type II), where the bile ducts were normal except cyst and the cyst is considered to be a sequel of an intrauterine CC rupture, such as meconium pseudocyst [28]. Additionally, type V CCs are possibly due to dysfunctional remodeling of the ductal plate during embryogenesis [40, 49].

In addition, it is considered that primary strictures of the CBD may also play a role in the development of CCs. The types of the CCs are determined according to the location, severity, and length of the stricture. Detection of these strictures preoperatively is important because treating the CC without addressing the stricture may lead to recurrent episodes of cholangitis. This mechanism has been under estimated and is now believed to be more significant in the pathophysiology of CCs [50].
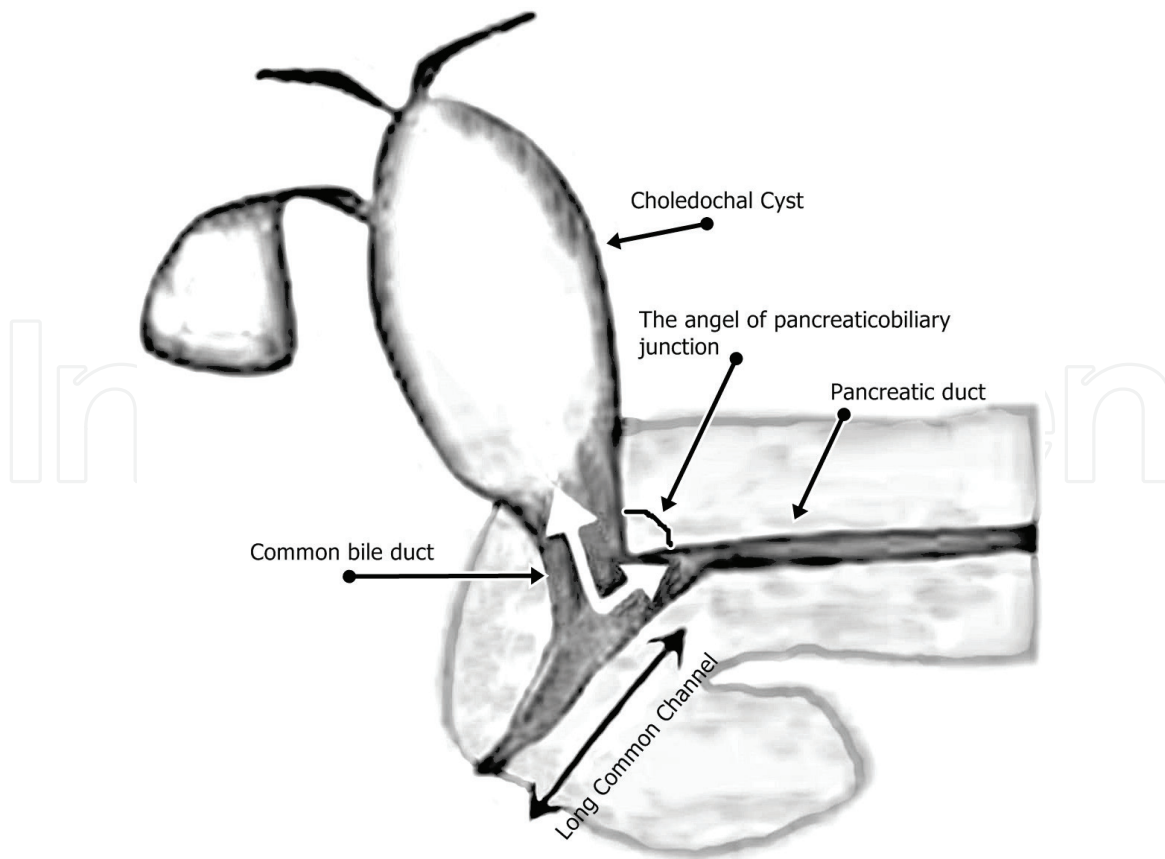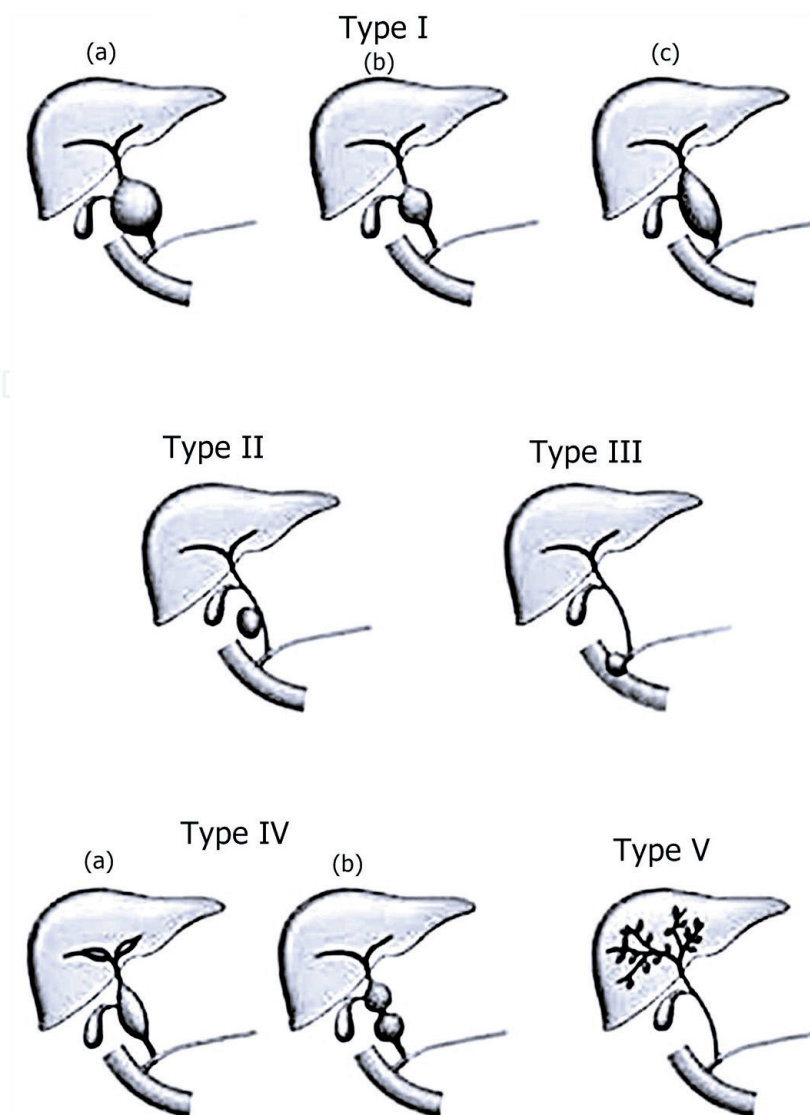
Figure 1.
Anomalous pancreaticobiliary union.

The association of CC with congenital anomalies remains ambiguous. Previous reports have demonstrated an association of pediatric CC and congenital anomalies. Murphy et al. reported in 2012 that screening for cardiac anomalies may be prudent in CC patients [51]. Other reports have postulated an association of CC with duodenal atresia, colonic atresia, gastroschisis, annular pancreas, and pancreatic cysts [10, 52–57].

6. Classification

CCs are first classified in 1959 by Alanso-Lej and colleagues [58]. The original classification identified four types of biliary cysts (types I–IV). In 1977, Todani and colleagues [8] modified this classification and added a fifth category of CC, type V biliary cysts or Carol disease (**Figure 2**) [10]. 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 [59].

Type I CC, most commonly seen, 80–90% of all CC, is a dilatation of the extrahepatic biliary tree. Importantly, the intrahepatic biliary tree is sometimes dilated secondarily due to biliary stasis. Type I cyst can be further subdivided into type Ia, Ib, and Ic cysts [10, 60]. Type Ia CCs are composed of the gallbladder arising directly from the CC, dilated extrahepatic biliary tree, and a nondilated intrahepatic tree. Type Ib CCs are focal segmental dilatation of the CBD and contain no evidence of APBDU [50, 60]. Finally, type Ic CCs are represented by a fusiform dilatation of the common hepatic duct and CBD in the presence of APBDU [10, 50, 60], and often also a low-grade stricture at the distal CBD [50]. In type I CC, usually, gallbladder is involved in cyst structure, and cyst extends from hepatic bifurcation to duodenum [61]. Most commonly, the ducts above (right, left, and

Figure 2.
Types of choledochal cysts, classified by Todani et al.

intrahepatic ducts) and below of the CC are not dilated [14], except type Ic. In type Ic, cyst extends continuously to the common hepatic duct or intrahepatic ducts [62]. Type I CCs, along with type IV cysts, have the highest risk of malignancy [10]. This is not surprising because both of them have extrahepatic involvement associated with APBDU [63].

Type II CCs (2% of all CCs) consist of a diverticular dilatation of the extrahepatic bile duct system and is considered true diverticulum. During the diagnostic cholangiography study, the diverticulum is filled with radiopaque substance and can be confused with the gall bladder duplication seen rarely [10].

Type III CCs (choledochoceles), 4% of all CCs, are characterized by distal (located at the pancreaticobiliary junction) CBD dilatation, confined to the wall of the duodenum and often bulging into the duodenal lumen [25]. Type III choledochal cysts are dissimilar to other types of CCs, with features such as appearing in both sexes equally and low malignancy incidence [10, 64]. Cysts are mostly not associated with APBDU. Because of all these characteristics, it has been suggested that type III CCs should not be classified as a type of CCs revised by Todani [25, 64, 65]. In addition, Ziegler et al. reported that choledochoceles occur more frequently in older male patients presenting with acute pancreatitis [64].

Type IV CCs, the second most common (15–20% of all), are multiple cysts which can involve both the intrahepatic and extrahepatic biliary trees. Type IV CC

can be further subdivided into type IVa and IVb cysts depending on intrahepatic involvement. Type IVa CC refers to multiple segmental communicating biliary dilatations located in the intra- and extrahepatic biliary tracts, and relative stricture at the junction that is used to distinguish the true type IVa CC from type I [11]. Type IVa CCs are usually associated with APBDU [50]. Type IVb CC refers to multiple extrahepatic biliary cysts without intrahepatic involvement [25]. Some recent studies have shown that intrahepatic ductal dilatations seen on preoperative imaging are thought to have been caused by distal obstruction and not true intrahepatic biliary duct disease [11]. Additionally, the question of "Is the distinguishing type I from type IVa really necessary preoperatively?" has not answered yet. Because distinguishing between types I and IVa CCs is controversial for some authors due to complete excision of the extrahepatic bile ducts, and intensive long-term follow-up still remains standard of care for both types [11].

Type V CC or Carol disease, added by Todani, is characterized by multifocal segmental intrahepatic biliary ductal dilatation [14] without the evidence of extrahepatic dilatation [66]. Caroli disease is uncommon, accounting for less than 10% of cases. Patients often present in adolescence or early adulthood with recurrent cholangitis, abdominal pain, or jaundice. However, they may present later with the sequel of portal hypertension and cirrhosis [67]. Renal abnormalities, such as medullary sponge kidney, autosomal dominant polycystic kidney disease, and medullary cystic disease can be seen in Caroli disease [49]. Some authors call Caroli disease as Caroli syndrome when congenital hepatic fibrosis is also seen, as in half of the patients [68, 69].

Type VI CC, isolated cystic dilatation of the cystic duct, is rare with only several case reports describing it. Although it is not officially part of the revised Todani classification, it has been proposed to be called type VI CC [70]. If the cyst emerges from the cystic duct near a level close to the CBD, it can be confused with type II CCs. In such cases, the relation of the cyst with the cystic duct should be thoroughly evaluated to differentiate them [59].

7. Clinical presentation

Type IC cysts are the earliest cysts that can be detected by 15-gestational week fetal ultrasonography [71, 72]. There are two clinical forms of disease: adult and infant. In infant form, symptoms such as obstructive jaundice, clay colored stools, and hepatomegaly make it difficult to distinguish from biliary atresia. Adult form of CCs is also congenital, although they usually remain silent until the age of 2 years. There are three main symptoms in the classical clinical triad: recurrent jaundice 69–75%, right upper quadrant pain 47–60%, and right upper quadrant mass 47–80%. But the classic triad only presents in 10% of cases (6–25%) [46, 73, 74].

Abdominal pain is the most common symptom (93.8%) [75], especially in older patients and presents with colic pattern which has a variable interval time (between attacks) up to several years. When investigating the cause of CBD dilatation and differential diagnosis of unclear upper abdominal pain, jaundice, and pancreatitis in children, CCs must be considered [76]. Unfortunately, 29–62% of pediatric patients with CC have been reported that they have choledocholithiasis [77, 78], that is, distinguishing this two situation (CC-associated choledocholithiasis and -nonassociated choledocholithiasis) may be difficult. Choledocholithiasis can also lead to CBD dilatation which can be misdiagnosed as a CC [79].

In 1–2% of cases, especially in infants, CCs may present with rupture and biliary peritonitis prompting emergency biliary drainage [80, 81]. It is not a surprise in diagnosing pancreatitis in patients with CC, because of association of the presence of APBDU [10, 46, 82].

The risk for development of biliary carcinoma in the general population starts after the fourth decade and the incidence increase with age to 0.15% after the eighth decade. However, the risk for the development of carcinoma in patients with a CC starts in childhood and shows a significant increase with age. Interestingly, the age of biliary carcinoma development in patients who have undergone internal drainage without cyst excision has been reported to be 15 years earlier on average than patients who have never had surgery. This is thought to be associated with intestinal bacterial contamination and pancreatic enzymes added to biliary stasis [83]. The malignancy incidence in resected bile duct material has been reported as 7.5% for all age groups, and 0.4 and 11.4% for those under and over the age of 18, respectively. The incidence has been reported to gradually increase every decade to 38.2% over the age of 60, possibly related to chronic inflammation [5]. The incidence of a biliary malignancy development following CC excision is reported as 0.7–5.4%. The malignancy can arise from anywhere such as the porta hepatis, pancreas, or the intrahepatic bile ducts. The time to onset after primary surgical intervention is reported to be 1–34 years. The total excision of the cyst significantly decreases the probability of a malignancy although it does not eliminate it completely [84, 85].

8. Diagnostic evaluation

Ultimately, multimodality imaging techniques are often utilized including computed tomography (CT), magnetic resonance imaging (MRI), and/or endoscopic retrograde cholangiopancreatography (ERCP) to confirm the extent of ductal involvement or the presence of extrahepatic disease [25]. Frequently, further imaging techniques are used to differentiate type I CC from type IVa, in the presence of intrahepatic biliary dilatation [11]. A cyst, presenting in the porta hepatitis, separated from the gallbladder and continuing with enlarged biliary ducts can be shown by ultrasound (US). Additionally, fusiform dilatation of choledoch, intrahepatic biliary dilatation (60–80%), biliary stones, and state of liver parenchyma can be shown by US [86].

Other intraabdominal cysts, such as pancreatic pseudocysts, echinococcal cysts, or biliary cystadenomas should also be differentiated from CCs, whether the cyst has continuity with the biliary tree or not [40]. CT is not only useful for demonstrating continuity of the cyst with the biliary tree, but also demonstrates relation of the cyst with the surrounding structures and the presence of associated malignancy [87]. In order to correctly plan surgery, CT cholangiography can be used to identify the full anatomy of the biliary tree but unfortunately it has been reported to be less sensitive for imaging the pancreatic duct which is responsible for the reflux of contrast into the biliary ducts [87]. As it is well known today, the nephrohepatotoxicity of the contrast and the ionized radiation exposed are the restrictions of CT utilization in pediatric population (**Figure 3**) [62].

MRCP is noninvasive and highly sensitive (70–100%) and specific (90–100%) in the diagnosis of CCs [88, 89], so, is considered the current gold standard imaging even for initial evaluation [62]. Additionally, there is no irradiation, and modern scanners have alleviated the need for protracted breath-hold making it more amenable to the pediatric population [10, 90]. Although both ultrasound and CT are highly sensitive and specific in the diagnosis of CCs, MRCP can better identify the CCs subtypes and coexisting abnormalities [89]. For example, MRI can easily identify the pancreaticobiliary ductal anatomy, while ultrasound cannot accurately demonstrate the APBDU [89, 91]. Additionally, MRCP is preferred modality in the pediatric population due to invasive nature and inherent risks of endoscopic ultrasound and ERCP, despite their ability of detecting the abnormality of the common
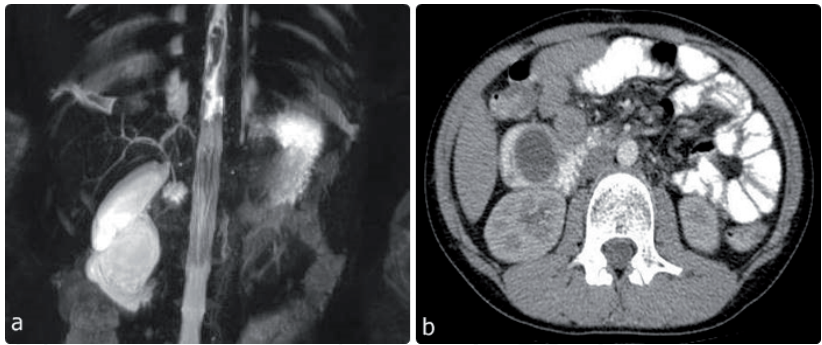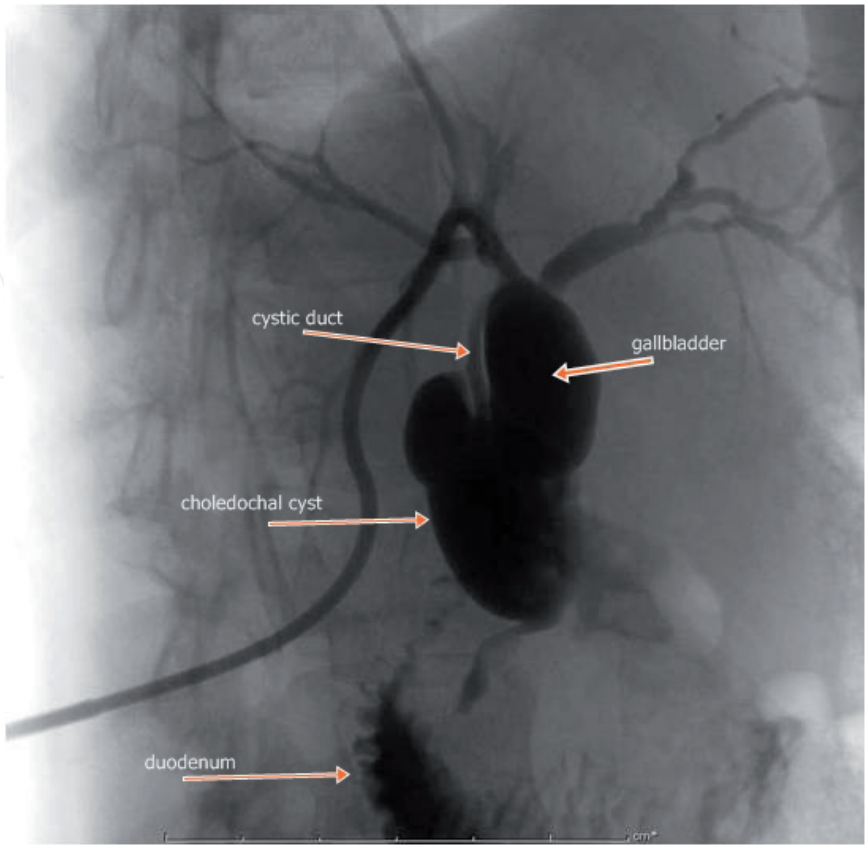
Figure 3.
Type III CC. (a) MRCP; (b) CT image.

channel [89]. MRCP has also been shown to be as effective as intraoperative cholangiography in planning surgery [18]. In addition, lower cost and morbidity compared to other imaging/diagnostic modalities, and reliability for detecting abnormalities associated with CCs such as cholangiocarcinoma and choledocholithiasis, are some of the favorable features of MRCP [89, 92]. Unfortunately, as seen often in patients with CCs, intraductal air, debris, stones, or protein plugs can interfere with the signal and alter visualization (**Figure 4**) [93].

Although it is highly sensitive, invasiveness and associated risks including cholangitis, bleeding, pancreatitis, and perforation makes percutaneous transhepatic cholangiography (PTC) or ERCP utilization less frequently [94]. Moreover, PTC and ERCP can be technically challenging and require general anesthesia in the pediatric population. A lot of surgeons are finding the use of diagnostic ERCP and PTC in CCs unnecessary due to advantages of both MRCP and perioperative cholangiography (performed in nearly almost patients and give highly detailed information about

Figure 4.
PTC reveals the detailed anatomy of the biliary tracts and associated CC.

biliary anatomy) [10, 95, 96]. Contemporary, ERCP should only be performed in cases where the appropriate diagnosis cannot be made by other less-invasive examinations, or when therapeutic performance (complications such as cholangitis or biliary stone obstruction [97, 98] and stabilization of the patients with preparing them to the next definitive surgery) is required (**Figure 4**) [39, 99, 100].

Another method that is not preferred now for diagnostic purposes is hepatobiliary scintigraphy. Although it is possible to do it with all IDA variants, DISIDA is the best. The radio isotopic substance is collected in the liver and is normally thrown into the biliary tract, but accumulates in the cyst space in patients with CC [101, 102].

8.1 The differential diagnosis

There are many diseases including biliary atresia, infectious hepatitis, embryonal hepatic rhabdomyosarcoma, biliary lithiasis, pancreatitis, biliary hamartoma in the differential diagnosis of CCs, especially biliary atresia that is one of the two causes of neonatal obstructive jaundice in neonatal period [10]. Differentiating cystic biliary atresia (CBA), a subtype of biliary atresia and has an entirely different treatment approach, from CCs is particularly difficult. Therefore, prompt accurate diagnosis is critical [103]. While earlier presentation (<months of age), smaller cysts with less dilatation of the intrahepatic biliary system, and an atretic gallbladder with irregular and hypoplastic biliary radicles that is seen on ultrasound and cholangiography are characteristics of CBA patients [10, 20, 104, 105]; a dilated gallbladder communicated with the cyst in addition with a dilated intrahepatic biliary tree is mostly a determiner to infantile CC [104].

It is still important to differentiate biliary rhabdomyosarcoma, a rare soft tissue tumor that affects only 1% of children, from CC [106, 107]. In the presence of a mass or intraductal growth that causes obstructive jaundice, the possible diagnosis should return in favor of rhabdomyosarcoma in children and prompt evaluation is necessary [107].

"Children with CBD dilatation did not differ significantly in clinical characteristics compared with children who had obstructive CBD dilatation" said Oh and colleagues [78] by evaluating the cholangiographic characteristics of 85 children with CBD dilatation to differentiate obstructive and congenital CBD dilatation. Indeed, it can be difficult to distinguish them. Therefore, in the pediatric population with dilated biliary trees, ruling out a distal biliary obstruction that causes secondary biliary dilatation is prudent and essential [18].

As noted above, type I CC may present with intrahepatic biliary dilatation secondary to biliary stasis, thus resembling a type IVa CC. Some authors consider that this distinction is critical given the therapeutic implications and the need to include hepatic resection (in the case of type IVa CC) in addition to extrahepatic biliary tree excision [18], whereas the others consider that the need preoperatively distinguishing between type I and IVa CCs is controversial because for both, complete excision of the extrahepatic bile duct and intensive long-term follow-up remains standard of care [11].

9. Management

The treatment time of antenatal diagnosed of CCs has been still a matter of debate. Some reports say that they can be operated within 2–6 weeks, even if they are asymptomatic, due to a potential complication risk of cysts, whereas, the others suggest that they can be followed-up for a time of period with US and regular monitoring of liver functions [108, 109].

Surgical treatment of CCs should be performed electively except complicated ones such as cyst perforation. Children, who have acute problems related to liver and pancreas, should be initially received appropriate medical treatment to remove inflammation and associated obstruction at the pancreaticobiliary system before surgery [110].

In the first half of this century, treatment methods such as cyst aspiration, marsupialization, and external drainage (cholecystostomy, tube drainage) had been used more extensively in the surgical treatment of CCs. And as expected, high mortality and morbidity rates had been detected in patient's follow-up period. Surgical methods such as partial cyst excision and cystoduodenostomy were defined between 1920 and 1930 [111, 112]. Especially, cystoduodenostomy has been the preferred method by many surgeons until the early 1970s [113, 114] when the long-term morbidity was detected as higher (30–50%) [7]. Therefore, in those years, Roux-en-Y cystojejunostomy had identified with the idea of preventing the reflux of the duodenal contents into the bile ducts [7, 30, 115]. Indeed, the cholangitis had been significantly reduced with this method, but not completely eliminated [28]. After soon, it had been demonstrated that anastomosis with a large stoma, as possible as (at least 4 cm) is more important for protecting reflux-related cholangitis attacks than which intestinal segment it is performed (duodenum or jejunum) [116]. The recognition of the cancer development from the left cystic wall and Babbitt's APBDU theory made the idea of cyst excision popular soon after. It had been reported that carcinoma develops after the internal drainage procedures at a frequency of 2.5–17.5% several years later as a consequence of chronic inflammation of the cyst wall. Therefore, cyst excision had gain popularity in a very short time [116].

Contemporarily, the definitive treatment for CCs are total excision that has become preferred management strategy over the internal drainage procedures (choledochocystoduodenostomy or choledochocystojejunostomy), which have an only historical value today despite they had been used as a treatment method in the past although caused high morbidity (probably because of not relieving biliary stasis sufficiently) [117, 118]. Furthermore, only complete resection can fully decrease the risk of malignant degeneration: a critical point in the pediatric population with a large number of expectant life years. The general aim is to remove the cyst completely and restore biliary enteric drainage either into the duodenum by hepaticoduodenostomy (HD) or jejunum by Roux-en-Y hepaticojejunostomy (RYHJ), although specific approaches for types vary minimally. Surgical intervention should be elective and patients should be medically optimized prior to operative intervention. İf patient has a cholangitis or pancreatitis attack preoperatively, the infection should be adequately treated with broad-spectrum intravenous antibiotics or biliary decompression if needed [118].

Surgery for CC disease can be performed open or laparoscopically based on patient characteristics and surgeon preference.

HD and RYHJ are the two most commonly utilized techniques of reconstruction [119], although other replacement conduits such as jejunal interposition HD, valved jejunal interposition HD, nonrefluxing biliary appendicoduodenostomy, hepaticoenterostomy, and wide hilar hepaticojejunostomy have been reported [120–125]. HD has been favored by some groups [126, 127] but most series suggests significantly more bile reflux compared with RYHJ [121], which is currently the most commonly utilized reconstruction.

9.1 Open surgery

In all cases, cholangiography should be performed initially to obtain detailed anatomical information about the intra- and extrahepatic bile ducts, irrespective of

preoperative examinations. Dissection of extrahepatic bile ducts starts from the gall-bladder. The terminal end of the cyst opening to the duodenum should be isolated, clamped, cut, and transfixed, firstly. Some surgeons suggest that dissection should be continued until the appearance of pancreatic ducts, while the others not suggest. Additionally, some surgeons taking into account that dissection toward the lower end of the cyst may cause inevitable unplanned pancreatic duct injury that pancre-aticoduodenectomy requirement should be in your mind, although very rarely [75]. After the distal portion of the cyst is ligated and cut, the posterior wall is dissected from the surface of the portal vein. In cases of marked inflammation, the cyst may be excised by leaving the posterior wall on the portal vein. The dissection should go on till the hepatic hilus. The best strategy to obtain a wide anastomosis stoma is to make a hepatic dissection more proximally until the left hepatic duct is seen. Although all parts of CCs need to be removed, sometimes residual proximal cyst walls can be left to facilitate biliary anastomosis [75]. Dilated bile ducts should be irrigated with hepa-rinized saline to clear the gallstones before anastomosis. After the cyst is excised, one of the hepaticoenterostomy methods, such as hepaticojejunostomy, HD, jejunal interposition HD, valved jejunal interposition HD, nonrefluxing biliary appendico-duodenostomy, hepaticoenterostomy, and wide hilar hepaticojejunostomy [120–125] is performed for biliary reconstruction. In RYHJ, 40-cm jejunal loop replaces to the hepatic hilus. In RYHJ surgery, to avoid the elongation of a blind pouch as the child grows, an end-to-end anastomosis of the jejunum to the CBD is recommended if technically possible [128]. If an end-to-side anastomosis is required (in some cases, the bile duct is too small), it should be as close as possible to the closed end of the jejunal limb. Additionally, although it is not possible to predetermine the length of the Roux limb, it should be appropriate to the child's overall bowel length consider-ing future growth. In HD, anastomosis is performed between the duodenum second part and the bile duct. The duodenum was mobilized to a limit. The duodenum is anchored to the liver at porta to avoid tension on the anastomosis (**Figure** 5).

The intraabdominal drain, kept in Morison's pouch, may be removed on the seventh postoperative day [124].
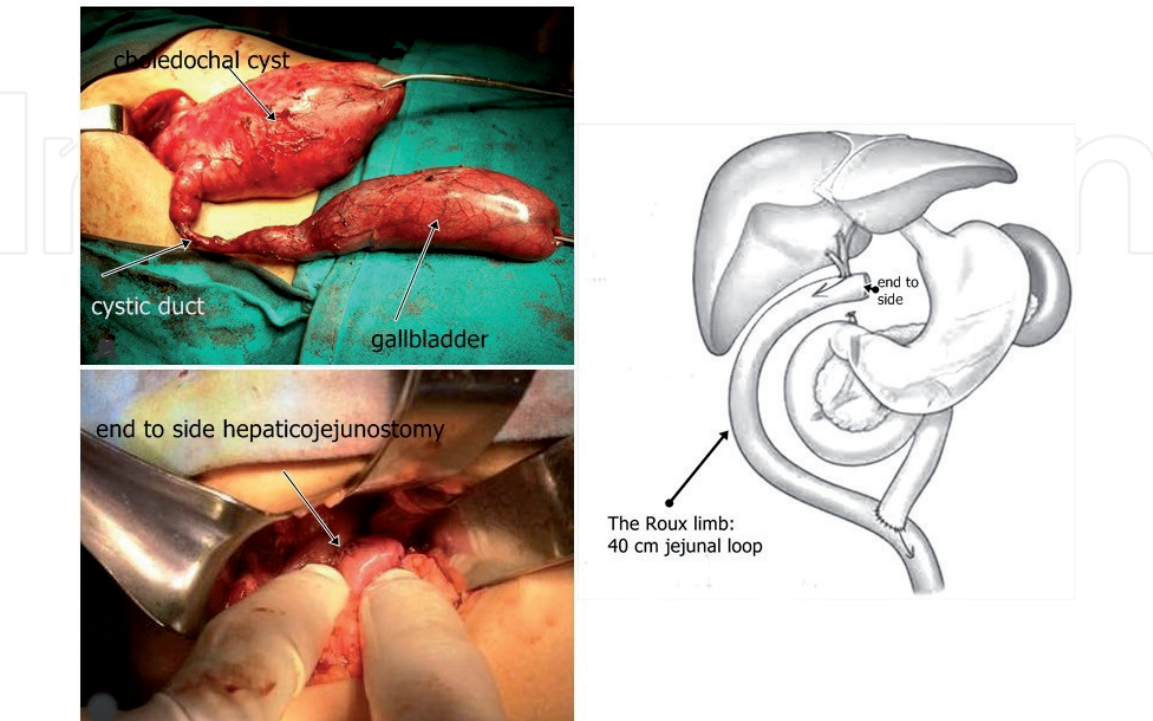
Figure 5.
Intraoperative pictures and drawn cartoon showing of the procedure.

9.2 Laparoscopic surgery

Laparoscopic treatment of choledochal cysts was first described in 1995 [129] and demonstrated that it could be performed in children as young as 3 months [130] and as small as 6 kg [131]. As with most surgical diseases, longer operating time and shorter hospital stay [132] were comparable with open surgical approaches, and in the absence of cholangitis or pancreatitis, it becomes more suitable treatment [132]. While four or five ports are typically used in the traditional laparoscopic approach [130, 133], the use of single-port laparoscopy [134] and robotic surgical system [135] has also been reported. In a prospective randomized study of 121 children undergoing laparoscopic cyst excision with RYHJ, routine postoperative drainage has been shown to be unnecessary [136].

9.3 The optimal technique for biliary reconstruction

The most commonly performed operations for biliary reconstruction after complete surgical resection of CCs are RYHJ or HD [124]. There is a debate regarding the optimal technique for biliary reconstruction [124]. RYHJ is considered as an ideal technique for the repair of CC, but HD has gained wide acceptance and favored by many surgeons open as well as laparoscopically because of its advantages over hepaticojejunostomy. HD is more physiologic, but theoretically, the closeness of hepaticoenterostomy to stomach makes HD to have greater chance of cholangitis and bile gastritis, but in a meta-analyzed study [137], it has been shown that while the incidence of bile gastritis after HD is even higher when examined endoscopically, interestingly, there is no difference of cholangitis between HD and RYHJ. Additionally, HD is simpler to perform and associated with fewer complications such as adhesive bowel obstruction, anastomotic leakage, and peptic ulcer as compared to RYHJ [138]. HD requires less operative time, allows faster recovery of bowel function, and produces fewer complications requiring reoperation [139]. If there is an anastomotic stricture following HD, it can be easily managed by endoscopy as against hepaticojejunostomy [124]. But, when the diameter of the common hepatic duct more than 10 mm that lets duodenal contents more likely to reflux easily into the intrahepatic bile ducts through the HD anastomosis or when the intrahepatic biliary dilatation is present that lets refluxed duodenal contents remain longer in the intrahepatic bile ducts, HD is not recommended because of higher risk of cholangitis or anastomotic stricture formation [124]. Some studies have demonstrated high incidence of secondary bile reflux proven by endoscopy after HD [121]. Recently, a patient with hilary bile duct carcinoma, who was performed HD for the biliary reconstruction at the age of 13 months, has been reported in the 19 years follow-up after the primary cyst excision. Reflux of duodenal contents (including activated pancreatic enzymes) into the intrahepatic bile ducts through the HD anastomosis is thought to be hazardous to the bile duct mucosa in this patient [124]. Adhesive bowel obstruction is seen with a higher incidence in RYHJ that comprises a Roux-en-Y jejuna limb and two anastomoses, compared with HD. Cholangitis, peptic ulcer, fat malabsorption, diarrhea, and malnutrition are the other complications [126]. A significant incidence of long-term complications requiring reoperation such as anastomosis stenosis has been observed with the follow-up studies of patients who underwent hepaticojejunostomy after cyst excision [138, 140], and a wide hilar hepaticojejunostomy extending into the left hepatic duct is advocated for the way to prevent it [125].

9.4 Treatment specifically for types

Treatment of type I CC includes excision of the extrahepatic biliary tract, cholecystectomy, and reconstruction of the biliary system. If the duct is dilated at the

distal margin, the mucosa may be left behind to prevent damage to the pancreatico-biliary system and can be striped. Infrequently, because of recurrent episodes of the cholangitis, the cyst wall may densely adherent to the portal vein, precluding safe resection [141]. In such cases, resection of the anterior wall with careful fulguration of the mucosa of the posterior wall can be performed [141]. Hepatic bifurcation is carefully evaluated for stricture and inflammation before performing anastomosis during proximal transection. If one of them is seen, more proximal transection should be considered [18].

For type II CCs, mostly, diverticulectomy or simple cyst excision is enough for the treatment. Primary or over a T-tube closure can be performed, and reconstruction is occasionally required if there is significant luminal narrowing [18].

One of the methods such as endoscopic sphincterotomy, sphincteroplasty, sphincteroplasty with cyst excision, or pancreaticoduodenectomy may be used to manage pediatric patients with type III CCs (choledochoceles) [10, 25]. Various reports denote adequate symptom control with this approach [142, 143]; however, long-term follow-up is lacking. Cysts not amenable to endoscopic intervention may benefit from lateral duodenotomy with sphincteroplasty and unroofing or marsupi-alization of the cavity [18].

Type IV CC is approached differently based on the presence or absence and location of intrahepatic disease [15]. Type IVb cysts are treated in the same fashion as type I. Management for IVa disease differs due to the presence of intra- as well as extrahepatic involvement, as well as the presence of functional liver disease. Of foremost importance is the characterization of actual type IVa as opposed to type I with upstream ductal dilatation due to stasis and functional obstruction [50]. If the dilatation is anatomic and isolated (limited; i.e., left hemiliver), partial hepa-tectomy with reconstruction to the remaining hepatic ducts may be warranted due to the ongoing risk of malignant transformation in the intrahepatic biliary system [85]. However, not all patients are appropriate candidates for partial hepatectomy [144]. Those patients with obvious dilatations and stenosis of intrahepatic ducts, intrahepatic duct stones, or parenchymal atrophy may benefit from hepatectomy [144]. If hepatectomy is planned concomitantly with extrahepatic duct excision, the distribution should allow removal of all disease (of the vast majority of the severe disease) with adequate future liver remnant [15]. If the pattern is more diffuse or imaging is inconclusive, treatment in a type I paradigm with close postoperative surveillance to follow intrahepatic ducts has been utilized [145]. This approach is justified by studies demonstrating that patients who progress to malignancy most commonly develop extrahepatic cholangiocarcinoma or gallblad-der cancer (approximately 85% of malignancy), whereas intrahepatic cholangio-carcinoma rarely occurs [144]. It is reported that the intrahepatic component has actually resolved in 3–6 months with adequate drainage [146]. To differentiate type IVa from type I, while in adults, preoperative percutaneous biliary drainage to decompress the intrahepatic biliary ductal system has been advocated [147], in children, this practice has not been reported probability due to the difficulty in maintaining the external tube. Although long-term results are not known, intrahe-patic cystojejunostomy, in addition to hepaticojejunostomy, has been described as a way of preventing liver resection in type IVa cysts [148]. Complete extrahepatic excision with hepaticoenterostomy and drainage of the remaining cyst externally or internally should adequately ameliorate biliary stasis in the presence of bilobar unresectable intrahepatic cyst [18].

Management of patients with Caroli's disease can be particularly difficult given the location of the cysts and frequent necessity for surgery (considerable potential for cholangitis, liver complications, and biliary cirrhosis; moderate potential for neoplasia (7%)) [15]. In Caroli disease, intrahepatic cysts can be seen as limited

disease restricted to a single segment/lobe or diffuse disease involving the entire intrahepatic biliary tree. If the patient has not developed cirrhosis and portal hypertension, the unilobar cystic disease should be treated with anatomic hepatectomy and biliary enteric bypass. However, bilobar disease should be treated with symptom-directed nonoperative treatment methods as including litholytic agents such as ursodiol, antibiotics, and percutaneous drainage if possible. Close follow-up is required for malignant transformation. Although there is no identification for prophylactically orthotopic liver transplantation in the treatment of the disease, it should be kept in mind for the choice of the treatment in patients who have diffuse symptomatic disease with cirrhosis or portal hypertension [149].

9.5 Follow-up

Patients should be monitored every 6 months during the postoperative 3 years and then annually. On initial follow-up, while all patients should be evaluated with complete blood count, liver function tests and abdominal US, on subsequent follow-up, investigations are done only in symptomatic ones. Long-term follow-up can be made by visits, telephonic conversations, and postal inquiry [124].

10. Outcomes and results

Resection of pediatric CC is generally well tolerated [18]. Despite recent advances in surgical techniques and perioperative management, short- and long-term complications are not rare in children, while they are more common in adults [6]. Complications such as recurrent cholangitis attacks, malignant transformation, intracystic or intrahepatic gallstone formation, cirrhosis development, and pancreatitis are common in patients who are not operated on. Complications such as anastomotic leakage, gastrointestinal or intraabdominal bleeding, acute pancreatitis, pancreatic leakage, wound infection, wound dehiscence, intraabdominal infection/abscess, intussusception can be seen in early postoperative period defined as short-term complications [150]. Most early complications can be treated conservatively [151].

However, most series are without early mortality and report rates of acute complications including wound infections from 0 to 17%, without significant difference between infants and children [46, 132].

Surgical inexperience and severe inflammation are often implicated in the development of anastomotic bile leakage [151]. The diagnosis of bile leakage is difficult and delayed in some cases due to nonspecific symptoms [151]. It may not always be possible to differentiate with the imaging findings of US and CT because not all of the intraabdominal fluid collection after surgery is associated with bile leakage, and so, this late diagnosis may result in mortality due to septicemia and septic shock [151]. However, it is reported that MRCP can be used to diagnose and accurately localize the site of bile leakage noninvasively [7, 151]. Bile leaks in the hepaticooenterostomy line can self-limited within a few weeks if they can be drained externally. If the bowel movement is sufficient during this period, the child can be fed by enterally. If the extracted bile is given back to the stomach with NG catheter, electrolyte losses can also be prevented. The bilirubin level of the child may remain high due to edema in the anastomosis line within the first 2 weeks, even if the operation has been successfully performed. If this takes longer, biliary tree, even anastomosis, can be evaluated with PTC [110]. Reoperation is considered only after the failure of conservative treatment [151]. The leakage can be repaired by a circumferential buried suture around the anastomotic site, peritoneal lavage, and effective drainage [151].

Gastrointestinal bleeding may be due to hepaticojejunostomy or stress ulcer.

Acute pancreatitis occurs in patients with CC, both preoperatively and due to injury of the pancreatic tissue during distal dissection of the cyst or to edema in the distal part of the pancreatic duct related postoperatively (4.2%) [151]. Therefore, it has been recovered with a conservative treatment for a short time period. Some reports say that CC excision without ligation of the distal stenotic stump decreases the incidence of pancreatic duct injury [75]. A probe inserted into the pancreatic duct through a duodenotomy may help to prevent pancreatic duct injury in difficult cases [75]. Additionally, Urushihara et al. [152] consider that using bipolar electro cautery to scrape pancreatic tissue away from the bile duct wall during the dissection of the intrapancreatic part of the bile duct causes minimal bleeding and enables clear identification of the narrow part of the CBD [151]. Eventually, complete resection of the distal portion of the cyst, removal of debris and protein plugs in the long common channel and pancreatic duct, and correction of anomalous arrangement of the pancreaticobiliary duct junction are essential to minimize pancreatic complications after the operations [151]. The pancreatic fistula occurs because of not closing the distal choledoch well after the cyst excision or injured pancreatic duct during the dissection. An external drainage of 3–4 weeks allows the fistulas closing.

Late/long-term complications (5–15%) [151] include anastomotic stricture, cholangitis, hepatolithiasis, ileus, cirrhosis, and malignancy. Benign anastomotic stricture with recurrent cholangitis is less common than in adults but is still seen in many as 10–25% of patients and can be associated with both intrahepatic and bile duct stone formation [117].

After intraabdominal surgery, small bowel obstructions, mostly due to adhesions, are common [154]. Patients should be closely monitored for any possible clinical deterioration [154]. If there is no improvement after 48 h of follow-up, there is a high risk of bowel resection due to bowel necrosis despite it has not been clearly defined [156]. Furthermore, it has been defined that during biliary reconstruction, the length and placement of the Roux loop is very important in adhesive bowel obstruction developing postoperatively [156].

In terms of anastomotic stricture, improvement of surgical skills, preservation of blood supply, no or mild inflammation cyst wall, and construction of wide (larger than 1 cm) and tension-free stoma are key factors to reduce anastomosis-related complication [138]. There should be no delay in surgical or endoscopic intervention once biliary obstruction develops postoperatively, but a great deal of planning and a thoughtful workup are required [151]. Kim et al. [155] reported that PTC with stone removal and balloon dilatation was useful in patients with anastomotic stricture. However, some investigators consider that recurrent anastomotic strictures may occur due to fibrosis even after balloon dilatation with PTCS, and repeated cholangitis may cause multiple intrahepatic biliary strictures, recurrent hepatic stones, and development of biliary carcinoma [152, 153]. Hence, especially in young patients, it is recommended that revision of the hepaticojejunostomy followed by ductoplasty, to create a wide stoma for sufficient bile drainage [152, 153].

Rigorous long-term follow-up after pediatric CC resection is limited, but the risk of biliary carcinoma (cholangiocarcinoma, squamous cell carcinoma, sarcoma, gallbladder cancer [12, 157], most often cholangiocarcinoma) clearly remains elevated even after CC excision compared to the general population [18]. The malignancy risk is considered to increase with age at surgery, and the cumulative biliary malignancy risk 25 years after primary surgery has been reported to be as high as 11% [75]. Malignant disease has been noted in up to 14% 0f patients after CC resection as a child [46]. In fact, cancer is the most frequent cause of late mortality in pediatric CC series [18]. Even after complete excision, patients are at higher risk for malignancy than general population [158]. Continued surveillance is, therefore,

strongly recommended, though it is not known whether there are risk factors such as retained portion of cyst or not [18]. In those with known malignancy, oncologic principles should apply; patients who can undergo safe resection with negative margins are appropriate for operation [15]. Resection may include hepatectomy with regional lymphadenectomy, extirpation of extrahepatic bile ducts with regional lymphadenectomy (and cholecystectomy), or pancreaticoduodenectomy [15]. However, the 5-year survival rate for patients with CCs complicated by malignancy is high, up to 55% in patients with cholangiocarcinoma [159].

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