Abstract

Congenital biliary dilatation (CBD) continues to baffle researchers and surgeons because its cause remains unknown. There is a female preponderance and higher incidence in Asia. It is often identified in childhood but some 20% may remain undiagnosed until adulthood. Prenatal diagnosis has increased in recent years accounting for 25% of new cases. Hypotheses for its etiology and pathogenesis include an anomalous junction between the pancreatic duct and the common bile duct, intrahepatic bile duct dilatation, and hepatic fibrosis. Because there is a risk for malignancy increasing with age and inadequate surgery, early excision with Roux-en-Y hepaticoenterostomy is recommended. Intraoperative endoscopy should be performed routinely to prevent postoperative bile stone formation by washing out any debris present. Early and late complications include leakage, cholangitis, anastomotic stricture, and cholangiocarcinoma. Laparoscopic treatment is safe with outcomes comparable to open surgery.

Keywords

Choledochal cyst • Hepaticojejunostomy • Hepaticoduodenostomy • Laparoscopic surgery • Cholangiocarcinoma • Pancreaticobiliary maljunction

Contents

Introduction ................................................. 2
Etiology ..................................................... 2
Embryology ............................................... 3
Anatomy ................................................... 3
Classification ............................................. 4
Clinical Presentation ...................................... 5
Diagnosis ................................................... 5
Treatment .................................................. 6
Incidental CBD ............................................ 7
Open Surgery ............................................ 7
Intraoperative Endoscopy ............................... 9
Postoperative Complications and Management ... 10
Malignancy ............................................... 11

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Introduction

In order to gain an understanding of biliary tract disease, it is necessary to study the basics of the development, anatomy, and function.

Congenital biliary dilatation (CBD) or dilatation of the common bile duct was first reported by Douglas in 1852 (Lane et al. 1999a). The condition, relatively rare in the West, occurs at an estimated incidence of 1 in 13,000 to 15,000 live births, but in the East it is far more common, with rates as high as 1 in 1000 live births in some parts of Japan. Female predominance is as high as 4:1 (Yamaguchi 1980). There is little doubt that CBD is a congenital lesion with a strong hereditary component, which may explain the higher incidence seen in Asia, and its familial occurrence in siblings and twins (Ando et al. 1996).

Anatomic features frequently associated with CBD include a cystic or fusiform dilatation of the common bile duct, an anomalous junction of the pancreatic duct and the common bile duct (pancreaticobiliary malunion: PBMU), intrahepatic bile duct (IHBD) dilatation with or without downstream stenosis, and various degrees of hepatic fibrosis. Early diagnosis and treatment is important as relationship with malignancy is documented. The risk of biliary cancer increases according to age (Mizuguchi et al. 2017).

Etiology

There are two known causal factors for the etiology of CBD – weakness of the wall of the common bile duct and obstruction distal to it. Distal stenosis is closely associated with cystic dilatation of the common bile duct, and the site of stenosis is related to an abnormal choledochopancreatic ductal junction (Miyano et al. 1979). Jona (1979) proposed that the pathogenesis of CBD associated with PBMU may be related to faulty budding of the primitive ventral pancreas (Jona et al. 1979). Wong and Lister (1981) conducted research on human fetuses and demonstrated that the choledochopancreatic junction lies outside the duodenal wall before the 8th week of gestation, whereupon it moves inward towards the duodenal lumen, suggesting that an anomalous junction may be caused by arrest of this migration (Wong and Lister 1981), while Tanaka (1993) proposed that regression of the terminal common bile duct and canalization of the ventral pancreatic duct caused by sinistral dislocation of the ventral pancreas are responsible for PBMU (Tanaka 1993).

Cholangiography has identified anomalies of the pancreaticobiliary ductal system in association with CBD, which may allow reflux of pancreatic enzymes and subsequent dissolution of duct walls. This is known as the long common channel theory and was first proposed by Babbit in 1969 (Babbitt 1969). This theory is further supported by the high amylase content of fluid aspirated from dilated ducts in patients with CBD. A dilated common channel and anomalous pancreatic duct are also frequently observed, which may be responsible for the formation of protein plugs or pancreatic stones, often associated with pancreatitis. Although Babbit stressed that pancreatic fluid is the most likely factor causing edema, weakness, and eventual fibrosis of the distal common bile duct, CBD can be diagnosed antenatally as early as 15–20 weeks’ gestation (Schroeder et al. 1989), at which time the pancreas is regarded to be too immature to function (Laitio et al. 1974), and even in neonates, enzymes secreted by the pancreas are not fully functional (Lebenthal and Lee 1980). While weakness of the duct wall secondary to refluxed pancreatic enzymes may be implicated, CBD is most likely to be caused by an anomalous choledocho-pancreatic duct junction combined with congenital stenosis, both of which are associated with abnormal development of the ventral pancreatic duct and biliary duct system. Komi (1992) suggests that PBMU can be divided into three types: a right angled union without accessory pancreatic duct, an acute angled union without accessory pancreatic duct, and an acute

angled union or right angled union with an accessory pancreatic duct (Komi et al. 1992). Another theory according to Davenport suggests that CBD is secondary to a reduced number of neurons and ganglions; as a result, congenital cysts were round because reduced ganglia induced distal obstruction with proximal dilatation while bile stasis was responsible for chronic inflammation (Davenport and Basu 2005). Histologically, different changes can be found in CBD, ranging from fibrous connective tissue in duct walls to inflammation of the mucosa and submucosa. The histological appearance of CBD is generally consistent although choledochoceles have duodenal mucosa and can resemble bile ducts in structure (Komi et al. 1986). Liver biopsies are seldom needed, and usually show inflammatory changes, central venous distention, and portal fibrosis. Postoperative liver biopsies taken after CBD resection may actually show worsening of portal fibrosis and central venous distention with little evidence of regression (Sugandhi et al. 2014; Soares et al. 2014). The possibility of incipient adenocarcinoma is of constant concern.

**Embryology**

The extrahepatic and intrahepatic biliary systems arise from endoderm as separated units, around the 4th week of gestation, from the hepatic bud and divide in two. Hepatocytes, intrahepatic ducts, proximal extrahepatic ducts, and the gallbladder arise from the cranial part and the distal extrahepatic ducts arise from the caudal part (Ando 2010). Around the 5th week, the dorsal and ventral pancreatic buds appear. The main ventral pancreatic bud develops near the entry of the common bile duct in the duodenum; then as it rotates in a C-shape, the ventral pancreatic bud is carried dorsally with the bile duct and fuses with the dorsal pancreas, lying posteriorly to the dorsal pancreatic bud and fuse each other, giving rise to the uncinate process (Roskams and Desmet 2008). The ventral pancreas is connected by two ducts to the primitive hepatic duct. The left ventral duct disappears during development. The right ventral duct divides into two branches, superior and inferior; the former joins the dorsal pancreatic duct to form the main pancreatic duct, which merges with the common bile pancreatic duct, inserting into the duodenum as the ampulla of Vater (Baumann et al. 2008).

**Anatomy**

The biliary tract is the connection between the liver where bile is produced by hepatocytes and excreted into canaliculi that channel it into larger intrahepatic and extrahepatic ducts. Its action is to transport stored bile into the duodenum and is regulated by the sphincter of Oddi. The portal triad is comprised of inter lobular or terminal bile ducts, hepatic artery branches, and the hepatic portal vein. The bile ducts usually have a diameter of <100 μm. Bile drains from hepatocytes into septal then segmental bile ducts before reaching the left and right hepatic ducts. The left hepatic duct collects bile from segments II, III, and IV; the right hepatic duct collects bile from segments V, VI, VII, and VIII (Baumann et al. 2008). Segment I has its own drainage or drains into both right and left hepatic ducts in 78% of individuals. Variations in intrahepatic bile duct anatomy can be observed in as many as 60% of individuals. The right and left hepatic ducts join to form common hepatic duct. The left hepatic duct is usually longer than the right hepatic duct and lies outside the liver parenchyma, making it more accessible during surgery if needed. The gallbladder and cystic duct join the common hepatic duct to form the common bile duct; several variations can be found making it mandatory to have a good understanding of anatomy to prevent injuries. The common bile duct is divided into 4 portions: supra-duodenal, retroduodenal, pancreatic, and intraduodenal, and drains into the second part of the duodenum at the papilla of Vater. At this point, the pancreatic duct joins to form a common short channel (Blumgart and Hann 2008).
CBD has been classified by Alonso-Lej et al. (1959), Todani et al. (1978), and Komi et al. (1992) according to anatomy and cholangiography of the hepatobiliary duct system, but the most internationally reliable method would appear to be presence or absence of PBMU (Fig. 1). The vast majority of CBD is associated with PBMU and is commonly referred to as being cystic, fusiform, or forme fruste. Choledochal cyst is commonly associated with PBMU involving concurrent abnormalities of the common channel, pancreatic duct, and intrahepatic ducts. Choledochal cysts with PBMU are classified as: (A) cystic dilatation of the extrahepatic duct, (B) fusiform dilatation of the extrahepatic bile duct, and (C) forme fruste choledochal cyst without biliary dilatation. Choledochal cysts without PBMU are classified as: (D) cystic diverticulum of the common bile duct, (E) Choledochocele (diverticulum of the distal common bile duct), (F) Intrahepatic bile duct dilatation alone (Caroli’s disease) (Miyano et al. 2005).

Technical issues during surgery for cystic choledochal cyst are frequently found on the proximal side of the pathology occurring as a result of

Fig. 1 Classification of CBD according to pancreatobiliary malunion (PBMU). (a) Cystic dilatation of the extrahepatic bile duct. (b) Fusiform dilatation of the extrahepatic bile duct. (c) Forme fruste CBD without PBMU. (d) Cystic diverticulum of the common bile duct. (e) Choledochocele (diverticulum of the distal common bile duct). (f) Intrahepatic bile duct dilatation alone (Caroli’s disease) (From Miyano et al. (2005))
anatomic variants of the common hepatic duct, uncertainty in relation to the excision level of common hepatic duct, dilated IHBD, debris, and/or stenosis in the IHBD. In contrast, Technical issues during surgery for fusiform choledochal cyst most often arise on the distal side of the pathology and are due to uncertainty in relation to the excision level of the distal choledochus, debris in the common channel, and complicated PBMU.

Caroli’s disease is characterized by segmental dilatation of large intrahepatic ducts, involving one or several segments, without an obstructive cause (Caroli et al. 1958). In up to 40% of cases, dilatation is confined to the left lobe of the liver. The more serious form, Caroli’s syndrome, associated with peripheral fibrosis, portal hypertension, and liver failure, has an association with mutations in PKHD1, a gene that is linked to adult recessive polycystic kidney disease, as well as jaundice secondary to obstruction of extrahepatic bile ducts by bile plugs or stones (Desmet 1992). Differential diagnoses include primary sclerosing cholangitis and secondary bile duct dilatation due to distal obstruction. Treatment varies according to the presence of cholangitis or bile duct obstruction. In mild cases, conservative management with antibiotics for cholangitis is usually necessary; surgical intervention is indicated for bile drainage but can include liver resection to reduce the risk for recurrent cholangitis or cholangiocarcinoma. Liver transplantation should be considered for Caroli’s syndrome (Okada et al. 2002).

Clinical Presentation

CBD should always be considered in the differential diagnosis of a child with abdominal signs and symptoms. CBD can present at any age, but approximately half become symptomatic in infancy. Neonatal cases have been uncommon. Clinical manifestations of CBD differ according to age. Neonates and young infants may present with obstructive jaundice, acholic stools, and hepatomegaly resembling biliary atresia (BA) and may even have advanced liver fibrosis, but on cholangiography, there is a patent communication with the duodenum and a well-developed IHBD tree. Young infants may also present with a large upper abdominal mass without jaundice. Young children may present either with a right upper quadrant mass and intermittent jaundice due to biliary obstruction, usually seen in patients with cystic CBD, or with abdominal pain due to pancreatitis, which is characteristic of fusiform or forme fruste CBD. Older children may present with the classical triad originally described by Alonso-Lej in 1959, consisting of pain, mass, and jaundice with fever and vomiting. This “classic” triad is usually only present in 20–30% of patients. Pain characteristically mimics that of recurrent pancreatitis, with or without raised serum amylase. In adolescence and adulthood, CBD is usually misdiagnosed for many years as being cholelithiasis or cholecystolithiasis, both of which are secondary to bile stasis, cirrhosis leading to portal hypertension, upper gastrointestinal bleeding, and splenomegaly; hepatic abscess and biliary carcinoma are known complications. Spontaneous CBD perforation can occur in up to 12% of patients presenting with acute abdomen or septic shock.

Diagnosis

Currently, abdominal ultrasonography (US) is probably the best screening method available even though it does not permit visualization of the entire duct system and it is not sensitive enough to demonstrate an undilated common channel and pancreatic duct. It is a low-cost, non-invasive imaging study, accurate enough to detect a cystic mass in the upper right quadrant that can be distinguished from gall bladder pathology or pathology related to the porta hepatitis. In addition, it can differentiate readily between hepatic cysts, liver abscess, pancreatic pseudocysts, omental cysts, or duodenal duplications. In patients suspected of having CBD, US can also demonstrate IHBD dilatation and the state of the parenchyma of the liver. However, for thorough diagnosis, the extrahepatic bile duct, intrahepatic ducts, anomalies of the pancreatic duct, and
PBMU should also be identified. Increased diameter of extrahepatic bile duct can be identified, requiring further investigation if needed. Normal common bile duct diameter in infants measures up to 2 mm and in children up to 3.5 mm (Hernanz-Schulman et al. 1995); in adolescents and young adults, up to 6 mm is considered normal; however, clinical correlation is mandatory. Endoscopic retrograde cholangiopancreatography (ERCP) can accurately delineate the configuration of the pancreaticobiliary duct system and is unlikely to be replaced by other investigations, especially in cases where fine detail is required preoperatively. However, ERCP is an invasive procedure unsuitable for repeated use and is contraindicated during acute pancreatitis. Nevertheless, it is performed routinely in many centers in Japan, with reasonable success (Iinuma et al. 2000).

Magnetic resonance cholangiopancreatography (MRCP) provides excellent visualization of the pancreaticobiliary ducts in patients with CBD, allowing narrowing, dilatation, and filling defects of the ducts to be detected with medium to high degrees of accuracy (Yamataka et al. 1997a; Shimizu et al. 2000; Saito et al. 2016) (Fig. 2). MRCP is also noninvasive and can be used to visualize the duct system upstream to an obstruction or area of stenosis, and has replaced ERCP as a diagnostic tool. Recently, better MRCP equipment has been developed, rendering three-dimensional imaging useful for delineating anatomy. However, in children less than 3 years old, the pancreaticobiliary duct system may not be visualized clearly because of their small caliber. Percutaneous transhepatic cholangiography is also available, especially for patients with IHBD dilatation and severe jaundice, and intraductal US (Kamisawa et al. 2011) can delineate the distal parts of the common bile duct and pancreatic duct successfully.

The importance of intraoperative cholangiography (IOC) in the modern era is controversial (Saito et al. 2016). If preoperative MRCP imaging can allow clear visualization of the entire biliopancreatic ductal system, including the intra- and extrahepatic bile ducts and pancreatic duct in detail, then IOC is unnecessary. Furthermore, if CBD is too extensive, IOC via the gall-bladder or directly via the common bile duct is ineffective and must be replaced by selective IOC of the IHBD and distal common bile duct during excision. Technetium-99m di-isopropylphenylcarbamoylmethylimidodiacetic acid scintigraphy scan is not used routinely; however, it can be useful to distinguish biliary atresia from CBD in a jaundiced child, if suspected. It is also helpful in cases of subacute or chronic cyst perforation showing localized accumulation or peritoneal spillage of radiotracer and as a follow-up investigation. Abdominal computed tomography (CT) is frequently not necessary and involves radiation exposure. However, it is useful for assessing the severity and extent of pancreatitis and/or malignancy. It is superior to US for visualizing IHBD, the distal bile duct, and the pancreas. CT is also valuable for defining the status of surrounding structures such as the portal vein and hepatic artery (Jablońska 2012).

In recent years, CBD has been detected with increasing frequency during routine prenatal US at as early as 15 weeks gestation, resulting in an increasing incidence of CBD, particularly in
neonates (Howell et al. 1983; Elrad et al. 1985). In a recent series, some 20% of patients were detected either neonatally or antenatally, and interestingly, the ratio of cystic to fusiform-type CBD neonatally or antenatally was 20:1, in contrast to an overall ratio of 5:3 (Lane et al. 1999b), which further supports the hypothesis that cystic dilation of the common bile duct is a prenatal event and fusiform dilation begins after birth. Overall, up to 60% of CBD are diagnosed in childhood and up to 20% can remain undiagnosed until adulthood.

**Treatment**

Complete cyst excision (CE) with Roux-en-Y hepaticoenterostomy (HE) is the definitive treatment of choice for CBD because of the high morbidity and high risk for carcinoma associated with internal drainage, a commonly used treatment in the past. Some of the procedures that had been performed are only of historical interest only and include: external drainage, internal drainage operations including choledochocystogastrostomy, choledochocystoduodenostomy with/without gastrostomy, choledochocystojejunostomy, hepaticocholedochostomy, hepaticoduodenostomy, intrahepatic cystoduodenostomy at the porta hepatis, and intrahepatic cystojejunostomy at the porta hepatis. Recently, more attention is being given to the treatment of conditions affecting the intrahepatic and intrapancreatic ducts, such as IHBD dilatation, focal stenosis, debris in the IHBD, and protein plugs or stones in the common channel. The optimum levels for transecting the common hepatic duct and excising the intrapancreatic duct will affect the incidence of complications and prognosis. In cases of Caroli’s disease, if cystic lesions are confined to one lobe of the liver, hepatic lobectomy should be considered, but if multiple lobes are involved with evidence of progressive hepatic fibrosis, only palliative measures or liver transplantation may be possible. In choledochocele, the transduodenal unroofing with sphinteroplasty of the pancreatic or common bile duct is the procedure of choice (Lobeck et al. 2016).

**Incidental CBD**

Incidental CBD identified on routine antenatal US should be observed during the antenatal and neonatal periods. Some pediatric surgeons recommend CE soon after diagnosis (Burnweit et al. 1996; Suita et al. 1999), while others believe there is no need for hasty excision if jaundice is not present (Miyano and Yamataka 1997). In the absence of protocols for management, neonates with incidental CBD should receive standard conservative medical management and nutritional support and with thorough assessment should they become symptomatic. Early surgery provides the opportunity to exclude BA, relieve obstructive jaundice if present, prevent fibrosis (Burnweit et al. 1996), reduce the risk for cholangitis by clearing debris, as well as prevent cyst perforation and malignancy (Howell et al. 1983; She et al. 2009). However, in complicated neonatal cases presenting with severe cholangitis, poor general condition, or huge dilated CBD, external biliary drainage is recommended by either percutaneous transhepatic cholangio drainage or direct percutaneous cyst drainage. Subsequently, delayed primary CE may be performed 3–6 months later. In cases of biliary peritonitis secondary to CBD perforation, either external biliary drainage or CE is recommended. Outcome of surgery in neonates is generally excellent (Suita et al. 1999).

**Open Surgery**

CE with biliary reconstruction to avoid two-way reflux of bile and pancreatic secretions is the standard procedure of choice (Liuming et al. 2011) because of the high morbidity and high risk for carcinoma associated with internal drainage, a commonly used treatment in the past. While the essentials of management of CBD are the same, treatment in early infancy has unique aspects that must be considered in relation to the risks of surgery itself in a small patient with immature physiology and immunology, while CE in older children and adults is much more difficult than in younger children because of complications associated with repeated episodes of
chronic inflammation. Full-thickness CE is easier in neonates and young infants, because the wall of the dilated common bile duct is not inflamed and there are few adhesions to surrounding structures, such as the portal vein and hepatic artery (Filler and Stringel 1980; Somasundaram et al. 1985). Adhesions are usually denser with cystic CBD than forme fruste CBD, especially in older children; in adolescents and adults, they can be severe. Thus, mucosectomy is rarely indicated in neonates because there is usually little inflammation present. Aspiration of the cyst prior to dissection makes surgery easier if the cyst is large.

Specifically, the cyst should be incised in the distal portion transversely close to the duodenum, because anomalous right and left hepatic duct openings may exist outside the porta hepatis in addition to the true openings in the distal part of the cyst. By exposing the posterior wall directly from the inside, dissection of the portal triad is facilitated. If the cyst is extremely inflamed and adhesions are very dense, mucosectomy of the cyst (Fig. 3) should be performed rather than attempting full-thickness dissection to minimize the risk for injuring the portal vein and hepatic artery and also to prevent the residual epithelium of the distal portion of the cyst from undergoing malignant transformation. Otherwise, the cyst is transected after careful circumferential dissection from the hepatic artery and portal vein. The distal common bile duct should be resected as close as possible to the pancreaticobiliary ductal junction. If the distal common bile duct is resected along line 1 (Fig. 4) over time, a cyst will re-form around the distal remnant in the pancreas, causing recurrent pancreatitis, stone formation, or malignancy. However, if the distal duct is resected along line 3 (Fig. 4), that is, just above the pancreaticobiliary ductal junction, there is no residual duct within the pancreas and a cyst is unlikely to re-form. Finally, the common hepatic duct is transected at the level of distinct caliber change, ensuring there is enough left for HE. If CBD is fusiform with no distinct caliber change, the cyst should be excised just above the choledochopancreatic junction, and the stump double-sutured, ligated, and transected. The distal bile duct within the pancreas must also be excised entirely. In cystic CBD, the distal bile duct is often so narrow that it cannot be identified and the likelihood of incomplete

![Fig. 3 Distal mucosectomy](Image)

![Fig. 4 Diagram of IE in a case of CBD with debris and protein plugs in the distal bile duct and recommended level of CE. Once IE has cleared any debris or protein plugs present CE is performed. If excised at Level 1, there is risk for residual cyst and at Level 3, there is risk for injuring the pancreatic duct. Level 2 is the ideal level for excision (From Miyano et al. 2000)](Image)
excision is virtually nil, in contrast to forme fruste CBD where the distal common bile duct is still wide at the pancreaticobiliary duct junction, and the likelihood of incomplete excision is high.

Basically, the only difference between operative procedures available is the type of biliary reconstruction performed. Although most surgeons use a Roux-en-Y hepaticojejunostomy (HJ), some (Todani et al. 1981) recommend a wide anastomosis at the level of the hepatic hilum, while others prefer hepaticoduodenostomy (HD). Good outcome with low early morbidity is to be expected irrespective of technique, but complications develop more often in the long term if dilated IHBD are present, and postoperative duodenogastric bile reflux appears to complicate HD although operative time and hospitalization are shorter for HD compared with HJ (Shimotakahara et al. 2005; Narayanan et al. 2013). Furthermore, a recent meta-analysis compared HJ with HD and reported significantly more postoperative reflux and gastritis with HD (Soares et al. 2014; Narayanan et al. 2013).

HE at the hepatic hilum is indicated in specific cases only, such as in patients with dilated IHBD with stenosis in the common hepatic duct or adolescent patients with severe inflammation of the common hepatic duct. HE at the hepatic hilum is more difficult than conventional HE, particularly in neonates and young infants without IHBD dilatation, and valved jejunal interposition HD is a complicated procedure.

End-to-end anastomosis of the jejunum to the upper remnant of the common bile duct is recommended if the ratio between the diameters of the common bile duct and the proximal Roux-en-Y jejunum is less than or equal to 1 (common hepatic duct) to 2.5 (jejunum) (Fig. 5). If the common bile duct is too small, then end-to-side anastomosis is unavoidable and the anastomosis should be as close as possible to the closed end of the duodenal limb (Fig. 5). If an end-to-side anastomosis is performed far from the closed end of the proximal jejunum, a blind pouch can develop as the child grows and such a blind pouch has been documented to be associated with adhesive bowel obstruction, bile stasis, and stone formation (Miyano et al. 1996) (Fig. 6a). Bile stasis in the blind pouch is also known to cause intrahepatic stone formation, especially if IHBD dilatation is also present. Figure 6b shows an ideal Roux-en-Y anastomosis without any redundant limb. The native jejunum should also be secured side-to-side to the Roux-en-Y jejunal limb from the ligament of Treitz for about 8 cm proximal to the end-to-side anastomosis to ensure smooth flow of bile and bowel contents distally, ensuring the jejunojejunostomy does not become T-shaped.

Severe dilatation of the IHBD can be managed by segmentectomy of the liver, intrahepatic cystoenterostomy, or balloon dilatation of a stenotic lesion at the time of CE. Stricture of the IHBD at the hepatic hilum can be treated by intrahepatic ductoplasty and cystojejunostomy or HJ at the hepatic hilum (Miyano et al. 1995). Stenosis of the papilla of Vater, stricture of the pancreatic duct, protein plugs, and septate common channel have been reported (Yamataka et al. 2000; Kaneko et al. 1997). If stenosis of the major papilla with a dilated common channel is identified, a transduodenal papilloplasty or endoscopic papilloplasty should be performed (Yamataka et al. 2000).

**Intraoperative Endoscopy**

Intraoperative endoscopy (IE) initially performed to examine the duct system and remove any biliary debris/stones and protein plugs in the common channel and dilated IHBD is now also used for determining the ideal level of resection. Briefly, a pediatric cystoscope or fine fiberscope with a flush channel is used to view the pancreatic and biliary duct systems directly at the time of CE (Yamataka et al. 2000; Takahashi et al. 2010). In younger patients, a neonatal cystoscope or a fine flexible scope (1.9–2.0 mm) with a flush channel may be required. IE is extremely effective. On long-term follow-up, the incidence of postoperative stone formation in postoperative CBD patients who had IE was lower than reported in the literature (Takahashi et al. 2010).
Postoperative Complications and Management

Satisfactory surgical outcome with low morbidity in the short- to mid-term is to be expected after CE. Surgical outcome is better, and early morbidity is lower in younger children than in older children. IE would appear to reduce complications (Yamataka et al. 1997b). Patients with IHBD dilatation, dilatation of the remaining distal bile duct, pancreatic duct, and common channel require more extensive follow-up, because chronic inflammation could contribute to stone formation and malignant change in the long-term.

Complications documented in a review of 240 (200 children; 40 adults) CBD cases (Yamataka et al. 1997b) include ascending cholangitis, intrapancreatic terminal common bile duct calculi, pancreatitis, and bowel obstruction, requiring revision hepaticoenterostomy, percutaneous transhepatic cholangioscopic lithotomy, excision of the residual intrapancreatic terminal common bile duct, endoscopic sphincterotomy, pancreaticojejunostomy, or laparotomy (Table 1). Overall incidence of complications was 9% (Yamataka et al. 1997b). In the group of patients who had CE by the time they were 5 years old, none of the abovementioned complications were documented, indicating that early diagnosis followed by CE and IE would appear to be the best approach to achieve surgical cure and prevent postoperative complications.

Fig. 5 (a) End-to-end anastomosis during HJ is recommended to prevent elongation of the blind pouch if the ratio between the diameters of the common hepatic duct and the proximal Roux-en-Y jejunum at the proposed site of anastomosis is less than or equal to 1:2.5 (common hepatic duct:jejunum). (b) If end-to-side anastomosis is unavoidable, the common hepatic duct should be anastomosed as close as possible to the closed end of the blind pouch so there will be no blind pouch at the HJ anastomosis site; if an end-to-side anastomosis is performed far from the closed end of the blind pouch, the blind pouch will grow as the child grows and an elongated blind pouch can contribute to bile stasis in the pouch and IHBD (especially if they are dilated) leading to stone formation.
Malignancy

Since an association between malignancy and CBD was first reported in 1944, more than 100 cases of cholangiocarcinoma have been reported in CBD cases. Histopathologic findings compiled from excised CBD specimens include erosion of bile duct mucosa, epithelial desquamation, papillary hyperplasia with regenerative atypia, bile duct mucosa dysplasia, and presence of metaplastic changes such as mucous cells, goblet cells, and Paneth cells. There is a tendency for hyperplasia and metaplasia to increase with age. Cholangiocarcinoma has been documented in more than ten patients younger than 20 years at the time of initial surgery for CBD; the youngest case reported being a 3-year-old boy (Saikusa et al. 2009). In addition, gallbladder carcinoma has been reported in 10–25% of CBD cases (Soares et al. 2014; Edil et al. 2008). There are also case reports of malignancies arising from the intrapancreatic terminal choledochus, hepaticojejunostomy anastomosis, and IHBD after primary excision. Todani et al. (2002) reported a case of biliary carcinoma that developed 19 years after CE and HD, which was attributed to

**Table 1** Complications after cyst excision in children versus adults

<table>
<thead>
<tr>
<th>Incidence/200 children</th>
<th>Complications</th>
<th>Incidence/40 adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>Ascending cholangitis</td>
<td>9</td>
</tr>
<tr>
<td>3</td>
<td>Intrahepatic bile duct stones</td>
<td>5</td>
</tr>
<tr>
<td>3a</td>
<td>Intrapancreatic terminal choledochus calculi</td>
<td>1</td>
</tr>
<tr>
<td>1</td>
<td>Pancreatic duct calculus</td>
<td>1</td>
</tr>
<tr>
<td>1a</td>
<td>Stones in the blind pouch of an end-to-side Roux-en-Y HJ</td>
<td>0</td>
</tr>
<tr>
<td>9b</td>
<td>Bowel obstruction</td>
<td>3c</td>
</tr>
<tr>
<td>0</td>
<td>Cholangiocarcinoma</td>
<td>2</td>
</tr>
<tr>
<td>0</td>
<td>Liver dysfunction</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>Pancreatitis</td>
<td>5</td>
</tr>
<tr>
<td>25(18)</td>
<td>Total</td>
<td>27(17)</td>
</tr>
</tbody>
</table>

*Note: Numbers in parentheses indicate number of patients. Thus, 18 children and 17 adults had 25 and 27 complications, respectively.*

From Yamataka et al. (1997b) with permission

a One patient with intrapancreatic terminal choledochus calculi also had a stone in the blind pouch of an end-to-side hepaticojejunostomy

b Adhesions in six and intussusceptions in three

c Adhesions in all three

**Fig. 6** (a) Inadequate Roux-en-Y HJ reconstruction. Note: HJ is far from the closed end of the blind pouch (arrowhead). Double headed arrow in the inset: Elongation of the blind pouch. **Arrow with an asterisk**: Reflux of jejunal contents into the Roux-en-Y limb through a T-shaped Roux-en-Y jejunoojunalostomy. (b) Appropriate Roux-en-Y HJ reconstruction. **Arrowheads**: Approximated native jejunum and distal Roux-en-Y limb. **Arrows**: Smooth flow without reflux of small bowel contents (From Yamataka et al. 2003)
pancreatic enzymes being activated by the presence of bile and enterokinase in intestinal secretions. In fact, HD appears to be associated with complications similar to those that arise secondary to internal drainage for CBD and transduodenal sphincteroplasty for cholelithiasis, two procedures documented to develop biliary carcinoma. Consequently, HD is no longer routinely used for biliary reconstruction.

Malignant lesions are usually associated with abnormal epithelium and can involve K-ras gene and p53 mutations in up to 60% of carcinomas in CBD patients followed by late inactivation of the DPC-4 gene, especially in biliary tract adenocarcinomas (Tomono et al. 1996; Shimotake et al. 2003). Vigilant follow-up and awareness of the risk for malignancy cannot be overemphasized.

**Laparoscopic Treatment**

Minimally invasive surgery using laparoscopy is now widely used for treating CBD. The general concepts are the same as for open CE; in other words, laparoscopic CE (lapCE) is performed according to the principles of our open CE and includes IE (retitled intralaparoscopic endoscopy) as a routine procedure.

LapCE is performed under general anesthesia, with the patient positioned at the foot of the operating table in a reverse Trendelenburg position, the surgeon at the patient’s feet, an assistant with a scope at the surgeon’s left, and another assistant on the surgeon’s right (Fig. 7). An open Hasson technique through a supraumbilical incision is used for a 5 or 10 mm, 30 or 45 degree laparoscope, and carbon dioxide pneumoperitoneum is established at a pressure of 10–12 mmHg. Three additional 5 mm trocars are inserted in the right upper quadrant, left paraumbilical area, and left upper quadrant (Fig. 8). Adequate exposure is achieved by elevating the liver by introducing a percutaneous stay-suture just below the xiphoid process to snare the falciform ligament and retract the liver. To expose the porta hepatis, a pair of Babcock forceps is inserted through the left subcostal port in the anterior axillary line, to grasp and elevate the gallbladder to allow the CBD to be dissected free from surrounding structures, such as the portal vein and hepatic artery. To excise the posterior wall of the cyst, the anterior wall is incised first to improve exposure of the posterior wall, as in the open technique (Fig. 9). This step facilitates dissection especially if there is chronic inflammation and dense adhesions which tend to be worse in cystic CBD cases.

IE is performed during lapCE by introducing a fine ureteroscope through an additional 3.9 mm trocar in the left epigastrum (Fig. 8) and its tip is inserted into the common channel through the distal cyst under laparoscopic guidance to remove protein plugs or debris in the same way as IE is performed during open CE (Fig. 10). A fine long ureteroscope can also be inserted through the left paraumbilical trocar for intralaparoscopic hepatic bile duct endoscopy to remove debris in the IHBD, if present (Fig. 11). In cystic CBD, the distal end tapers and is often very narrow, and occasionally, the orifice of the distal end opening into the common channel cannot be identified. In such cases where even the finest ureteroscope cannot be inserted, IE cannot be performed, but debris in the common channel is also rare. Such patients usually do not develop pancreatitis but can present with jaundice with or without cholangitis and/or liver dysfunction. In general, IE is performed in all cases unless the ureteroscope cannot be inserted smoothly into the common channel from the distal part of the CBD (Miyano et al. 2011). During lapCE, it is tempting to examine the duct system with the laparoscope instead of changing to an ureteroscope, but inspection and irrigation are not possible without a constant flow of saline to expand the lumen, and while flexible scopes do have side channels, they are only designed for flushing, or introducing instruments and are not suitable for IE.

To prepare for excision, the cystic artery is identified and divided. Dissection of the cystic artery is initiated by removing the adjacent peritoneum using monopolar electrocautery and a Maryland dissector to establish a plane of dissection, beginning on the anterior wall and continuing to the medial and lateral sides, and then to the distal portion. The exact level of transection of the distal
common bile duct is easier to determine if the oriﬁce of the pancreatic duct in the common channel can be identiﬁed, as in the open technique (Fig. 4). After the CBD is freed, the distal part is divided as close as possible to the pancreati-cobiliary junction, and the stump is ligated with an endoloop. The proximal part is excised leaving 10 mm of common hepatic duct. Cholecystectomy is then performed.

A segment of proximal jejunum distal to the duodenojejunal ﬂexure is then exteriorized by extending the umbilical port incision, and a Roux-en-Y jejunoojejunostomy is constructed extracorporeally. The length of the limb is determined by placing the jejunoojejunostomy at the

![Diagram showing patient positioning and surgical setup](image.png)

**Fig. 7** The patient is initially positioned at the foot of the operating table, with the surgeon at the patient’s feet. An assistant with a scope stands on the surgeon’s left and an assistant on the right. *Note the different positions for the monitor and the surgeon when it is time for HJ.
umbilicus and bringing the distal end of the limb to be 3 cm above the xiphoid process. The exteriorized jejunum is returned to the abdominal cavity after the Roux-en-Y jejunal limb is approximated to the native jejunum for 8 cm cranially. The closed end of the jejunal limb is brought up via a retrocolic window to the porta hepatis. Then, an antimesentric enterotomy, 5 mm in length, for the HJ anastomosis, is created near the closed end of the Roux-en-Y limb to allow the common hepatic duct to be anastomosed as close as possible to the closed end of the blind pouch. When creating the 5 mm enterotomy, monopolar diathermy should be avoided because it appears to emit considerable lateral thermal energy which could injure the bowel wall around the enterotomy, which could be the cause of postoperative anastomosis leakage reported by many centers (Lee et al. 2009; Hong et al. 2008; Srimurthy and Ramesh 2006; Ure et al. 2005; Nguyen Thanh et al. 2010). The edges of the enterotomy are apposed temporarily with two 7/0 PDS sutures to prevent spillage of bowel contents. These sutures are cut at the time of HJ anastomosis. Two additional trocars, lateral right subcostal, and between the lateral right subcostal and right upper quadrant trocars are required for the HJ anastomosis using 5/0 absorbable sutures (Fig. 8). End-to-side HJ is performed using interrupted 5/0 or 6/0 absorbable sutures with the right upper quadrant port as a needle holder in the right hand, the 5 mm port for the scope, and the 3 mm subcostal port as a needle receiver in the left hand. Both the right and the left edge sutures are exteriorized and are used as traction sutures during anastomosis of the anterior wall to facilitate accuracy (Fig. 12), especially when the HJ anastomosis diameter is less than 9 mm. A tube drain is inserted in the pouch of

**Fig. 8** Trocar positions for laparoscopic excision. Note there is a 5 mm trocar in the epigastrium for the ureteroscope and additional 3 and 5 mm trocars (square brackets) on the right required for HJ. Numbers indicate trocar sizes

**Fig. 9** Excision of the posterior wall of the CBD under laparoscopic control
Winslow. The resected cyst and gallbladder are extracted through the umbilical wound, trocars are removed, and all wounds are closed.

**Outcomes**

As of mid-2014, the authors had performed 143 Roux-en-Y HJ with IE for CBD. Anastomoses were end-to-end in 85 cases and end-to-side in 58 cases. All cases are well at the time of writing without any complications after a mean follow-up period of 14 years (range: 4 months to 29 years). One hundred and fifteen of these cases had open CE and 28 had lapCE. Two lapCE cases required conversion to open CE because laparotomy and minilaparotomy were required, respectively. Thus, 117 cases had open CE and 26 cases had lapCE.

A summary of these 26 lapCE treated between 2009 and 2014 follows. Mean age at surgery was 5 years (range: 1.0–14.2), and mean weight was 16.5 kg (range: 8.0–47.0). There were 19 females and 7 males. CBD was fusiform in 19 and cystic in 7. Five patients had IHBD dilatation. Of the 19 fusiform CBD cases, 16 had HJ diameters of

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**Fig. 10** Under laparoscopic guidance, the tip of a fine ureterscope (E) has been inserted into the common channel through the distal CBD to remove protein plugs. The inset shows the IE field.

**Fig. 11** A fine ureteroscope (E) has been inserted into the IHBD to perform IE and delineate the transition zone into right and left hepatic ducts. Note the clamped cystic duct (CD). The inset shows the IE field.
6–9 mm, while the remaining 3 fusiform CBD cases and 7 cystic CBD cases had HJ diameters over 10 mm. IE of both the common channel and IHBD were performed in 16 cases (all fusiform); the remaining 10 had IE for IHBD only because the ureteroscope could not be inserted into the intrapancreatic choledochus or the common channel. During IE of the common channel, all 16 fusiform CBD cases had protein plugs removed successfully by irrigation with normal saline from the side channel of the ureteroscope (massive protein plugs in 4, moderate in 10, minimal in 2). Debris were present in the IHBD in 13/26 CBD cases (moderate 6, minimal 7); the remaining 13 had no debris. There were no intraoperative complications. Mean blood loss was minimal at 20 mL. Although all are well after a mean follow-up of 2.8 years (range: 1 months–5.2 years) with cosmetically attractive wounds, there were three postoperative complications. The first complication was pancreatitis developing 8 months postoperatively in a case with massive protein plugs probably due to residual fine debris, 3 mm × 3 mm in size, even though we thought all debris had been removed successfully during IE. Pancreatitis was treated conservatively with no sequelae. The second complication was duodenal obstruction in a cystic CBD case. At exploratory laparoscopy, the third part of the duodenum was found to be compressed by the Roux limb that had been inadequately fixed to the colonic mesentery. Once the sutures between the Roux limb and colonic mesentery were released laparoscopically, the postoperative course was uneventful. The third complication was anastomotic leakage that was treated by minilaparotomy.

**Conclusion and Future Directions**

CBD is a rare pathology that affects all ethnic groups but with higher incidence in Asian populations. Proper diagnosis and early treatment play important roles in preventing serious complications and malignancy; follow-up evaluation is necessary as progression of hepatic fibrosis or biliary cirrhosis may become problematic years after curative surgery is performed (Ishibashi et al. 2017). In 2014, the first congress of biliary atresia and related diseases (BARD) was held in Berlin with a committee of experts that discussed CBD, in an effort to share experience and unify management.

CE with Roux-en-Y HJ is considered the treatment of choice in both children and adults with CBD. IE at the time of CE effectively prevents common postoperative complications. Roux-en-Y HJ in children is different from that in adults, since the Roux-en-Y limb or blind pouch can elongate as the child grows.

Lately, laparoscopic treatment for CBD has gained popularity among pediatric surgeons; however, long-term follow-up results have not been collated; experienced laparoscopic surgeons would appear to obtain results as good as those for open surgery. While lapCE involves the use of extra trocars and takes longer, the extra trocars allow us to reproduce the maneuverability of open CE while preserving the primary benefits of minimally invasive surgery, namely, improved...
wound cosmesis, less requirement for analgesia, and earlier discharge from hospital. With experience, lapCE is as safe as open excision and if combined with IE, is the procedure of choice for treating children with CBD. Robot-assisted resection for CBD has already been reported and as a consequence, surgeons must familiarize themselves with new techniques as technology advances.

Cross-References

▶ Antenatal Diagnosis
▶ Biliary Atresia
▶ Clinical Research and Evidence Based Pediatric Surgery
▶ Disease of Gallbladder
▶ Embryology of Congenital Malformations
▶ Fetal Counseling for Congenital Malformations
▶ Innovations in Minimal Invasive Surgery

References


