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REVIEW ARTICLE

Choledochal cyst

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Abstract
Choledochal cyst (CC) or congenital biliary dilatation, has a skewed distribution with hereditary features that is far more common in East Asian females. CC is usually associated with pancreaticobiliary malunion (PBMU) forming a common channel. CC requires early definitive diagnosis, since there is a risk for malignancy occurring in the CC and/or intrahepatic bile ducts (IHBD). Complete CC excision and Roux-en-Y hepaticoenterostomy is required and can be performed by open or minimally invasive surgery with hepatojejunostomy the recommended procedure of choice. Principles of open surgical intervention form the basis of minimally invasive management with laparoscopy and robotic assistance. Current surgical management is associated with fewer early and late complications, such as hepaticoenterostomy anastomotic leakage, cholangitis, anastomosis stricture, and cholangiocarcinoma. Specific features of CC management at Juntendo include: intraoperative endoscopy of the common channel and IHBD for inspecting and clearing debris to significantly reduce post-operative pancreatitis or stone formation; near infra-red fluorescence with indocyanine green for visualizing tissue planes especially during minimally invasive surgery for CC; and a classification system for CC based on PBMU that overcomes inconsistencies between existing classification systems and clinical presentation.

Keywords Choledochal cyst · Congenital biliary dilatation · Hepaticojejunostomy · Hepaticoduodenostomy · Pancreaticobiliary maljunction · Laparoscopic cyst excision · Robotic-assisted cyst excision · Near-infrared fluorescence · Indocyanine green

Introduction

Cystic disease of the biliary tree was first described by Vater and Ezler in 1723 but its exact etiology has yet to be established, although understanding has progressed [1]. What is apparent is that its distribution is skewed with a distinct female preponderance and a reported incidence of 1 in 100,000–150,000 live births in the West, increasing to 1 in 1000 live births in East Asia, with two-thirds of these occurring in Japan [2]. There is also a hereditary component from familial incidence in siblings and twins in Japan [3].

Dilatation of the common bile duct (CBD) can present clinically at any age and is referred to as choledochal cyst (CC) in pediatric patients. Considered to be congenital, it can also arise secondary to stenosis of the distal CBD due to intrapancreatic tumor and has been induced experimentally, notably by Lewis Spitz who created CC in a fetal lamb model by ligating the distal bile duct [4]. Although benign, CC can be associated with cholangitis, pancreatitis, cholelithiasis and malignant transformation [5]; a common etiologic factor being some kind of physical stress that disrupts the integrity of the walls of the biliary tree. Nearly all CC patients have an anomalous junction of the pancreatic duct and the common bile duct (pancreaticobiliary malunion: PBMU). PBMU is an expression of disordered proximal biliary development and is associated with CC up to 90% of the cases [6] that appears to be associated with weakness of the wall of the CBD and obstruction distal to it. Distal stenosis of the CBD is closely associated with cystic dilatation and the site of stenosis is related to a PBMU [7]. The etiology of the stenosis is speculative but is considered to be vascular/ischemic in nature occurring in the fetus.

Anatomic features of CC include cystic choledochal dilatation (cystic CC), fusiform choledochal dilatation (fusiform CC), minimal/no choledochal dilatation ("forme fruste" CC) [8], PBMU, and intrahepatic bile duct (IHBD) dilatation

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with or without downstream stenosis; hepatic fibrosis is very uncommon as are cardiac defects, anorectal malformation, although there a case of CC associated with duodenal atresia has been reported [9]. Diagnosis and treatment of CC are straightforward but new technology has contributed to making surgical intervention for excising all malformed ductal tissue less invasive with enhanced accuracy/precision. Excellent outcome with low morbidity is expected after open, laparoscopic, or robot-assisted CC excision in the short- to mid-term. However, with longer follow-up, reports of complications increase, and careful extended lifelong follow-up should be considered mandatory especially for all cases with IHBD dilatation to monitor for potentially life-threatening postoperative sequelae.

CC classifications by Alonso-Lej et al. [10], Todani et al. [11] and Komi et al. [12] using anatomy and cholangiography of the hepatobiliary duct system with reference to disease entities and symptoms have been criticized for lacking consistency, so a classification based on the presence or absence of PBMU (Fig. 1) was developed to better reflect signs/symptoms and anatomy/structure and have greater international application both clinically and practically. Thus, CC with PBMU may be classified as: A: cystic dilatation of the common bile duct (CBD); B: fusiform dilatation of the CBD; and, C: forme fruste CC with minimal or no dilatation of the CBD or common channel syndrome [13] (Fig. 1), while CC without PBMU may be classified as: D: cystic diverticulum of the CBD; E: choledochocele (diverticulum of the distal CBD); and, F: polycystic segmental dilatation of IHBD (Caroli's disease).

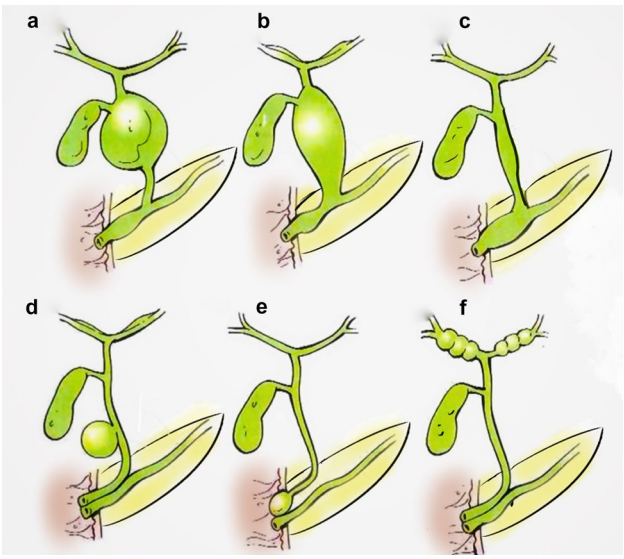

Fig. 1 Classification of choledochal cyst (CC) using the presence/absence of pancreaticobiliary malunion (PBMU). **a** Cystic choledochal dilatation (cystic CC). **b** Fusiform choledochal dilatation (fusiform CC). **c** Minimal/no choledochal dilatation ("forme fruste" CC). **d** Cystic diverticulum of the common bile duct. **e** Choledochocele (diverticulum of the distal common bile duct). **f** Intrahepatic bile duct dilatation alone (Caroli's disease). **a**, **b**, and **c** are associated with PBMU. **d**, **e**, and **f** are not associated with PBMU

This review will focus on CC with PBMU, i.e., cystic, fusiform, and forme fruste, based on experience of treating CC at the Department of Pediatric Surgery at Juntendo University Medical School, Tokyo, Japan the first independent pediatric surgery center established in Japan in 1968, and Hospital Regional de Alta Especialidad Materno-Infantil, Monterrey, Mexico.

Etiology

Anomalies of the pancreaticobiliary ductal system identified by cholangiography in association with CC prompted Babbit to hypothesize the long common channel theory in 1969 [14], where reflux of pancreatic enzymes resulted in subsequent dissolution of duct walls. High amylase content of fluid aspirated from dilated ducts in patients with CC supported his theory and a dilated common channel and anomalous pancreatic duct were also observed frequently and considered responsible for the formation of protein plugs or pancreatic stones, often associated with pancreatitis. Although Babbit stressed that pancreatic fluid was the most likely factor causing edema, weakness, and eventual fibrosis of the distal CBD [15], neonates are not capable of fully functional secretion of active pancreatic enzymes [16] and CC can actually be diagnosed antenatally as early as 15–20 week gestation [17], at which time the pancreas is definitely too immature to function [18]. While weakness of the duct wall secondary to refluxed pancreatic enzymes may be implicated in the clinical presentation of CC, the most likely etiology of CC is an anomalous choledochopancreatic duct junction combined with congenital stenosis both of which are associated with abnormal development of the ventral pancreatic duct and biliary duct system. PBMU gives rise to recurrent abdominal pain and/or recurrent pancreatitis with cystic, fusiform, or forme fruste CC.

Diagnosis

Dewbury [19] first reported the detection of prenatal CC using ultrasonography (US). Improvements in US technology and magnetic resonance imaging have resulted in choledochal dilatation and cysts as small as 1.0×1.8 cm being reported as early as 18–20 weeks of gestation [20, 21]. However, most CC detected on prenatal US are incidental and a definitive diagnosis of CC can be difficult on prenatal US alone because of similarities between dilatation/cysts and normal anatomy such as the fetal gallbladder and umbilical vein and similarities with other subhepatic cyst

morbidity such as duodenal duplication, parenchymal liver cysts at the porta hepatis, and most importantly cystic biliary atresia [22, 23]. Definitive diagnosis requires repeat US or magnetic resonance cholangiopancreatography (MRCP) (Fig. 2); MRCP after birth is particularly useful for documenting hepatopancreaticobiliary ductal anatomy including common channel and PBMU [24].

After birth, normal pigmented stools should be confirmed after meconium is passed and unconjugated jaundice levels monitored and confirmed to decline. At this stage, the most important concern is biliary patency and this can be confirmed either clinically, biochemically, or by radio-isotope scanning. Some surgeons recommend early surgery for prenatally detected CC, i.e., in the neonatal period, but most will defer for 3–6 months or until the infants weighs more than 5 kg. Nevertheless, in infants with biliary obstruction or infants who cannot be distinguished from cystic biliary atresia, early surgical intervention is mandatory.

Clinical presentation

CC should always be considered in the differential diagnosis of all pediatric patients with acute abdominal signs and symptoms. Clinical manifestations of CC differ according to age. Neonates and young infants may present with obstructive jaundice, acholic stools, and/or hepatomegaly resembling biliary atresia and may even have advanced liver fibrosis. However, on cholangiography, there is a patent communication with the duodenum and a well-developed IHBD tree [25]. Young infants may also present with an asymptomatic large upper abdominal mass without jaundice.

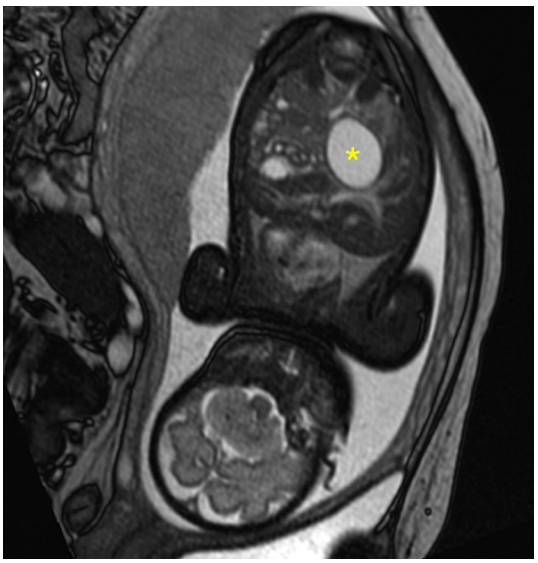

Fig. 2 Prenatal magnetic resonance imaging suggestive of a choledochal cyst (*) requires differentiation from cystic biliary atresia if the cyst is not large

Young children may present either with a right upper quadrant mass and intermittent jaundice due to biliary obstruction, usually with cystic CC, or with abdominal pain due to pancreatitis, which is characteristic of fusiform or forme fruste CC. Older children may present with the classical triad of pain, mass, and jaundice with fever and vomiting described in surgical textbooks but this mode of presentation is rarely seen today, although a report from 1995 [26] described that some 15% of all CC patients presented with the classical triad. When adolescents and adults present with CC, there is a much higher risk for premalignant and malignant histology (PMMH) in the CC, IHBD, and/or gallbladder compared with children [27]. Thus, all specimens after surgery for CC, especially from older children and adults, must have thorough histopathology for cholangiocarcinoma performed to ensure any suspicious tissue has been excised thoroughly.

Other modes of presentation include pancreatitis, the mechanism believed to be related to reflux of bile into the pancreatic duct, possibly precipitated by obstructive protein plugs in the common channel. This can occur even after CC excision if the common channel is not irrigated to remove debris and/or protein plugs by intraoperative endoscopy (IE) [28]. Spontaneous CC perforation can also occur at a rate of around 5% in most series [29–31]. CC perforation will present as an acute abdomen but is difficult to diagnose in infancy and bile peritonitis can be life-threatening.

Imaging studies and blood biochemistry

Currently, abdominal US is probably the best investigation available for assessing a patient suspected of having CC, even though it does not permit visualization of the entire pancreaticobiliary duct system and it is not sensitive enough to demonstrate an undilated common channel and pancreatic duct. US also clearly demonstrates IHBD dilatation and the state of the liver parenchyma. However, for accurate diagnosis and adequate surgical treatment, the CBD, IHBD, PBMU, other anomalies of the pancreatic duct, and presence of debris or protein plugs in the common channel should also be identified. Endoscopic retrograde cholangiopancreatography (ERCP) can accurately delineate the configuration of the pancreaticobiliary duct system in detail, and is unlikely to be replaced by other investigations when fine detail is required, preoperatively. However, ERCP is an invasive procedure requiring general anesthesia in infants and young children, so it is now rarely performed. MRCP provides excellent visualization of the pancreaticobiliary ducts in patients with CC allowing PBMU, IHBD dilatation, narrowing, dilatation and filling defects of the ducts to be detected with medium to high degrees of accuracy [25, 32]. MRCP is non-invasive, although sedation is required in infants and

young children, and it has replaced ERCP as a diagnostic tool (Fig. 3). Percutaneous transhepatic cholangiography is also available for patients with IHBD dilatation and severe jaundice, and endoscopic US [33] performed via the gastrointestinal tract can delineate the distal parts of the CBD and pancreatic duct successfully.

Intraoperative cholangiography has also largely been superseded by preoperative MRCP imaging, because it provides clear visualization of the entire biliopancreatic ductal system (Fig. 3a), including IHBD, CBD, pancreatic duct, and PBMU in detail. However, if a CC is huge, both preoperative MRCP (Fig. 3b) and intraoperative cholangiography via the gallbladder or directly via the CC would fail to visualize PBMU and the pancreatic duct, because they would be overlapped by the large CC. Thus, intraoperative cholangiography may be indicated for confirming anatomy at the porta hepatis, PBMU, and the pancreatic duct when performed independently from both the proximal and distal CBD after transecting the CC into proximal and distal parts. On rare occasions, a radio-isotope scan (99mTc-DISIDA) can be performed to show baseline liver function and impairment of biliary excretion as a marker for the progression of liver cirrhosis.

Biochemical liver function tests may be entirely normal or reflect hepatic functional deterioration due to biliary obstruction. Amylase and lipase levels are usually elevated during episodes of abdominal pain in fusiform CC or forme fruste CC, suggestive of active pancreatitis. Although rare, if blood clotting is prolonged secondary to chronic cholestasis, parenteral vitamin K should be prescribed.

Surgical intervention

Open surgery

The aim of surgery is complete excision of all affected/suspicious duct tissue because of high morbidity/risk for carcinoma associated with internal drainage, such as cystoduodenostomy, a common treatment used in the past. For cystic/fusiform CC this means bile flow is diverted to flow from dilated CBD away from contact with pancreatic juice in the pancreatic duct and proximal restoration of bile flow from the liver to the intestine. The procedure of choice for biliary reconstruction at Juntendo is Roux-en-Y hepaticojejunostomy (HJ), but hepaticoduodenostomy (HD) or other biliary reconstructions are preferred at other centers [34–38].

Forme fruste CC is somewhat controversial with some surgeons performing cholecystectomy without excising the CBD as is commonly performed in adult cases; however, at Juntendo, pediatric cases are treated in the same way as cystic/fusiform CC cases, by excision of the CBD and the afore-mentioned diversion procedure, after several patients treated by cholecystectomy alone, elsewhere, developed recurrent postoperative pancreatitis necessitating further surgery to excise the CBD. Care is required when intrahepatic and/or intrapancreatic ductal anomalies may also be present. Examples include IHBD dilatation with downstream stenosis, debris in the IHBD, and protein plugs or stones in the common channel. The level of excision of the distal CBD within the pancreas is also important. Thus, at Juntendo, all pediatric CC with PBMU have complete CC excision with biliary reconstruction (HJ) and IE [39].

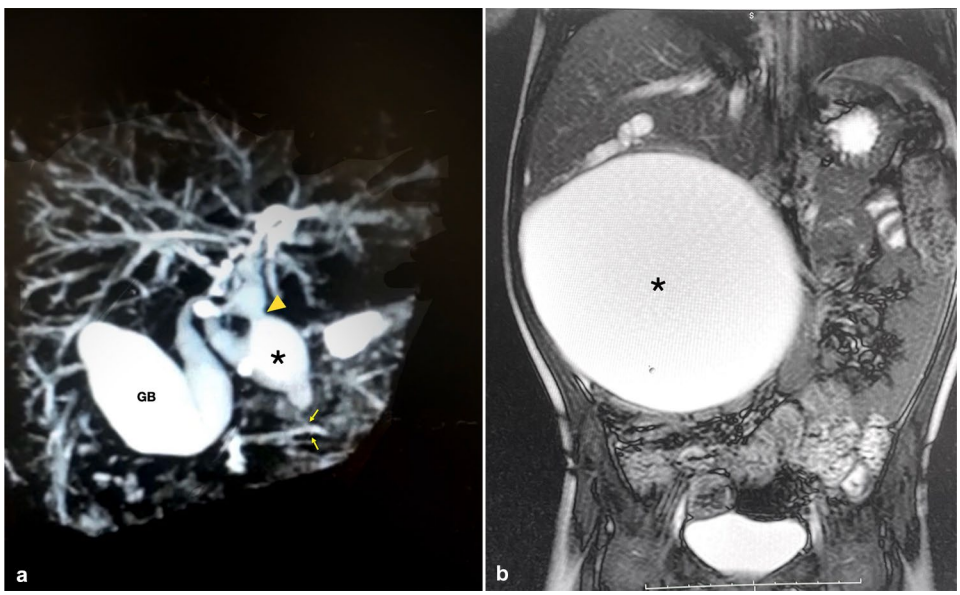

Fig. 3 **a** MRCP appearance of a choledochal cyst (*), intrahepatic ducts, common hepatic duct (arrowhead), and pancreaticobiliary duct system. Arrows indicate pancreaticobiliary malunion. GB indicates gallbladder. **b** Giant choledochal cyst (*)

Prior to commencing or during dissection of a cystic CC, the anterior wall of the cyst is incised transversely to expose the posterior wall for direct inspection from the inside (Fig. 4). By incising the anterior wall, the CC can be freed from surrounding tissues including the portal vein and hepatic artery more easily and safely than by dissecting without incising the anterior wall. This approach can even be used when a fusiform CC is severely adhesed to surrounding vital structures such as the portal vein and hepatic artery as a complication of repeated episodes of preoperative pancreatitis. In older children, there are usually more adhesions between a cystic CC and surrounding vital structures, compared with a cystic CC in infants or younger children. If a CC is extremely inflamed and adhesions are very dense, mucosectomy of the CC should be performed rather than full-thickness dissection to minimize injury to surrounding structures (Fig. 5).

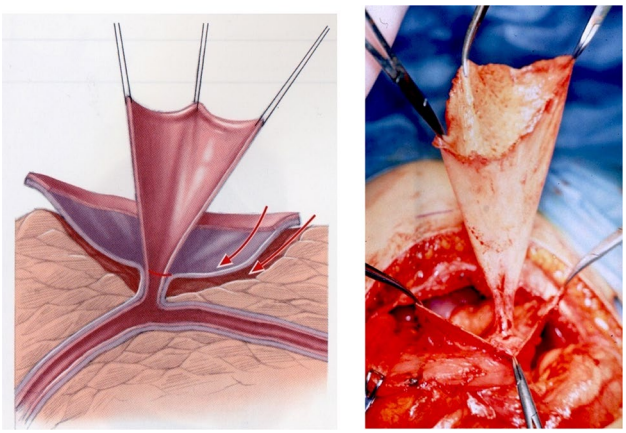

Fig. 5 Upper and lower arrows indicate the plane for mucosectomy and full-thickness dissection of a choledochal cyst, respectively. Intraoperative photo shows mucosectomy

Intraoperative endoscopy

Intraoperative endoscopy (IE) was originally performed to examine the hepatobiliary pancreatic duct system and remove any biliary debris/stones and protein plugs in the CC and dilated IHBD [40, 41]. IE has proven to be extremely effective for decreasing postoperative pancreatitis due to residual protein plugs in the common channel and postoperative stone formation in the IHBD on long-term follow-up (Fig. 6) [39, 41]. During open CC excision, a fine pediatric cystoscope is used to view the lumens of the common channel and IHBD directly [28, 41]. However, in cystic CC, the distal CBD is often so narrow that it cannot be identified and a fine pediatric cystoscope cannot be inserted. Such cystic cases have no past history of pancreatitis and IE is unnecessary as it is most unlikely that debris will be present in the common channel. During laparoscopic CC excision, IE is performed with a fine ureteroscope through the left paraumbilical trocar to inspect and remove debris from both the common channel and the IHBD. Further details of laparoscopic IE may be found under laparoscopic surgical management.

IE is also useful for determining the ideal level for excising a fusiform CC (Fig. 7). There is little likelihood of incomplete excision in cystic CC patients with a distal end that tapers strongly, in contrast to fusiform CC, where the distal CBD within the pancreas is still wide at the pancreaticobiliary duct junction, and the likelihood of incomplete excision is high. Essentially, the distal CBD should be resected as close as possible to the pancreaticobiliary ductal

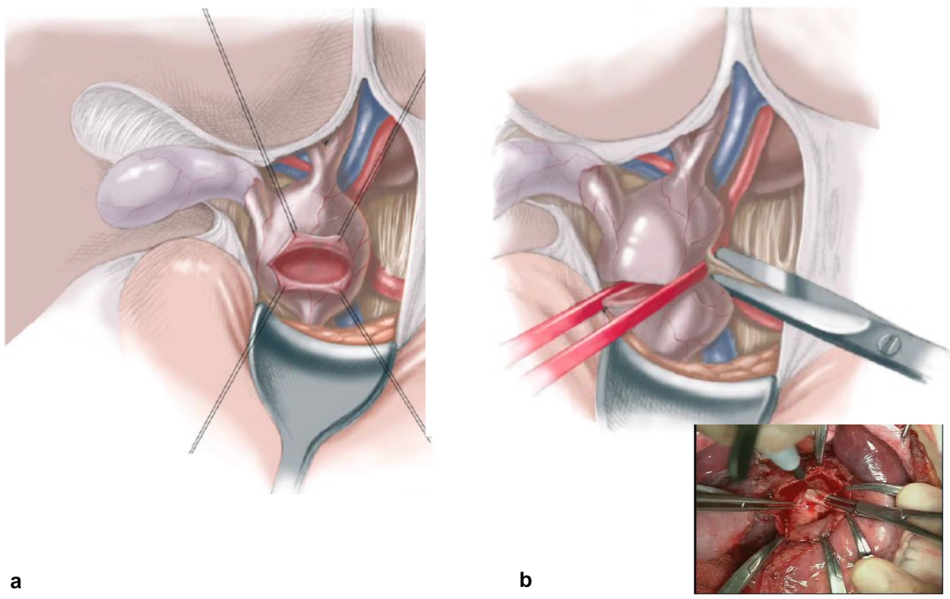

Fig. 4 Initial anterior wall incision (**a**) of a choledochal cyst (CC) allows the posterior wall of the cyst to be visible directly from the inside of the CC, facilitating safe dissection of the posterior wall of the cyst (**b**) and intraoperative photo (inset), preventing injury to surrounding vital structures, such as the portal vein and hepatic artery

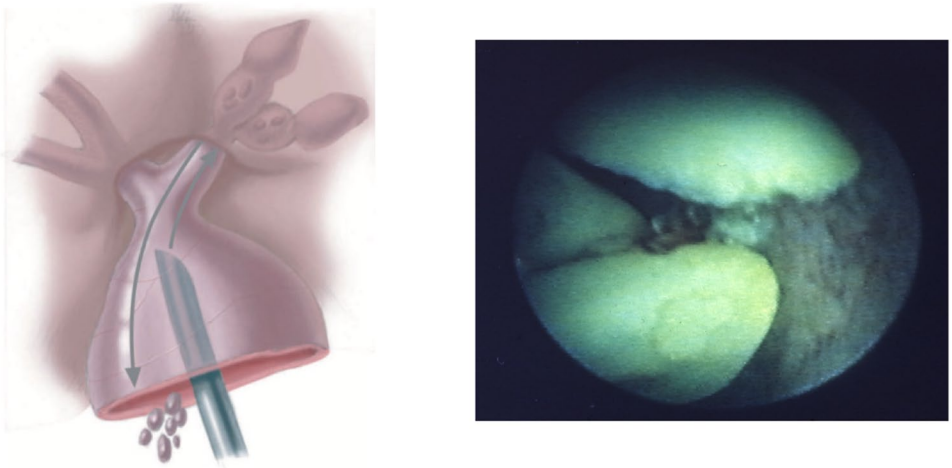

Fig. 6 Intraoperative endoscopy (IE) is useful for examining for the presence of debris and clearing the intrahepatic ducts of stones/debris. Diagram of IE (left); view of debris during IE (right)

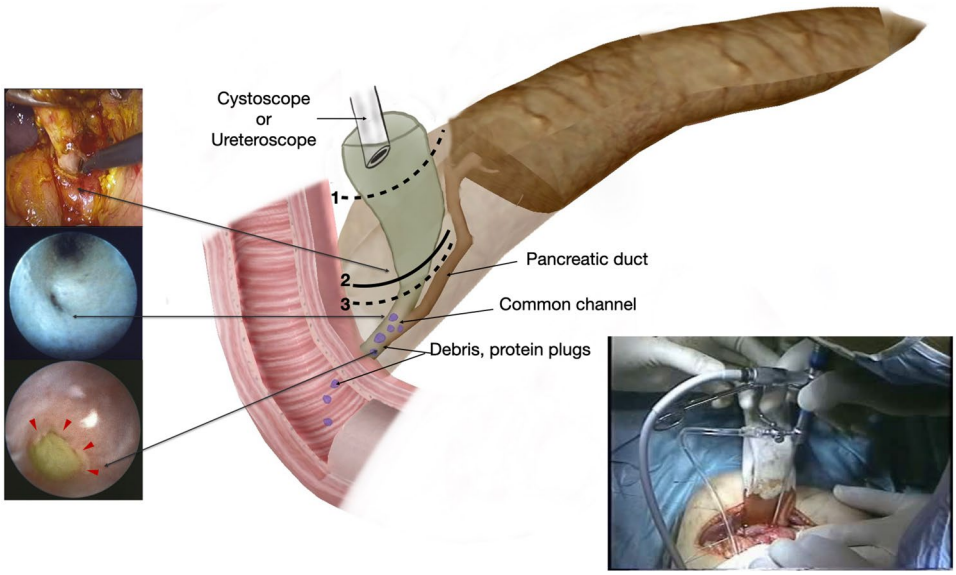

Fig. 7 Diagram of intraoperative endoscopy in a case of choledochal cyst with debris and protein plugs in the common channel indicating various levels for resecting the distal common bile duct. Once intraoperative endoscopy has cleared any debris or protein plugs, level 1 of resection has a risk for residual distal common bile duct and at level 3, there is risk for stenosing or injuring the pancreatic duct. Level 2 shows the ideal level of transection

junction along line 2 in Fig. 7, that is, just above the PBMU, so there is no residual distal CBD within the pancreas, preventing postoperative pancreatitis or stone formation in the residual distal CBD. If the distal CBD is resected along line 1 in Fig. 7, over time, the distal remnant in the pancreas may induce pancreatitis, stone formation, or malignancy. If the distal CBD is resected along line 3 in Fig. 7, that is, just above the pancreaticobiliary ductal junction, there is high risk for stenosis or injuring the pancreatic duct.

Intrahepatic bile duct dilatation

Dilatation of the peripheral portion of the IHBD in patients with CC has been reported to be associated with late complications, such as recurrent cholangitis and stone formation. The management of IHBD dilatation in the peripheral portion is difficult and can be managed by segmentectomy of the liver, intrahepatic cystoenterostomy, and balloon dilatation of the stenotic lesion causing IHBD dilatation at the time of CC excision. While these concerns are valid, the incidence of such late complications from extensive experience of CC patients at Juntendo is actually low, especially in younger children, especially after IE was introduced at the time of CC excision in 1986 [28]. Specific surgical intervention for IHBD dilatation, such as hepatectomy at the time of CC excision or endoscopic treatment of the stenosis in the peripheral portion of the IHBD should be reserved only for cases with massive dilatation of peripheral IHBD with severe downstream stenosis. In addition, from experience at Juntendo, hepaticoenterostomy (HJ or HD) at the hepatic hilum is indicated in specific cases only, such as in patients with dilated IHBD with stenosis in the common hepatic duct at the porta hepatis (Fig. 8) or adolescent patients with severe inflammation and fibrosis of the common hepatic duct [42]. Careful follow-up of all cases with IHBD dilatation is mandatory.

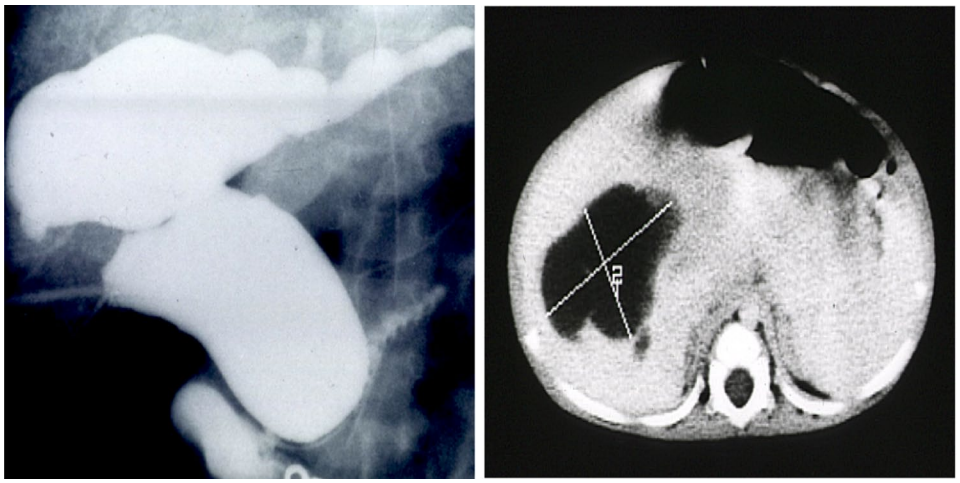

Fig. 8 Patient with grossly dilated IHBD at the porta hepatis seen on intraoperative cholangiography (left) and computerized tomography (right)

Hepaticojejunostomy versus hepaticoduodenostomy

Most surgeons use the established Roux-en-Y HJ technique, while others prefer HD, first advocated by Todani [34, 43]. 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 [44, 45]. HD is clearly a more "physiological" tension-free bile duct anastomosis following Kocherization of the duodenum, and quicker to perform. Although bile is delivered into the correct part of the intestine there is no form of valve and two-way flow of duodenal contents into the biliary tract may occur. Duodenogastric bile reflux into the stomach is recognizable on upper gastrointestinal endoscopy irrespective of symptoms [46, 47] and histopathology of biopsied gastric mucosa is typical of gastritis [25]. Similarly, reflux of duodenal contents into IHBD after HD has also been implicated as a potential cause for "sump syndrome" [48] which can be prevented by performing a Roux-en-Y HJ.

To summarize, while HD is an easier, quicker procedure that allows bile to enter the duodenum directly which is more physiological, postoperative cholangitis and bile gastritis are known complications with risk for mucosal damage and possible malignant change. As evidence, Todani et al. [49] reported a patient who underwent cyst excision and HD at 13 months and developed hilar bile duct carcinoma 18 years later. Inflammation of the bile duct mucosa was thought to be related to the reflux of duodenal contents (including activated pancreatic enzymes) into IHBD though the anastomosis which prompted them to abandon HD in favor of HJ. In fact, HD appears to be associated with complications similar to those that arise secondary to internal drainage for CC and transduodenal sphincteroplasty for cholelithiasis, two procedures documented to develop biliary carcinoma. Consequently, HD is no longer routinely used for biliary reconstruction at Juntendo, and Todani himself changed from HD to HJ [49]. Laparoscopy has revived some interest in HD, but overall, HJ tends to be preferred for HE and is recommended for biliary reconstruction in children requiring CC excision, because of complications of reflux, such as "sump syndrome" [48] and duodenogastric bile reflux [44–47] mentioned in association with HD.

Hepatojejunostomy anastomosis

End-to-end anastomosis is recommended if the ratio between the diameters of the common hepatic duct and the proximal Roux-en-Y jejunum lumen is less than or equal to 1 (common hepatic duct) to 2.5 (jejunum) (Fig. 9a). If the gap in diameters is considerable, end-to-side anastomosis is unavoidable, but should be performed as close as possible to the closed end of the duodenal limb (Fig. 9a) to avoid creating a blind pouch that can increase as the child grows (Fig. 9b). Such blind pouches have been documented as causing adhesive bowel obstruction, bile stasis, and stone formation [39]. Bile stasis in the blind pouch is also known to cause intrahepatic stone formation, especially if IHBD dilatation is also present. 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 (Fig. 9a) and ensure that the jejunojejunostomy does not become T-shaped (Fig. 9b).

Customizing the Roux loop

When the length of the Roux-en-Y jejunal limb is determined arbitrarily, e.g., as 30, 40, 50, or 60 cm long, the Roux loop can become unnecessarily long as the infant/young child grows, contributing to redundancy and tortuosity of

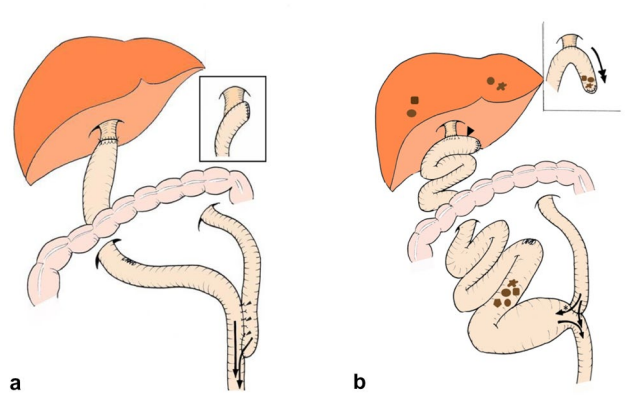

Fig. 9 **a** Appropriate Roux-en-Y hepaticojejunostomy (HJ). Customizing the length of the Roux-en-Y limb and end-to-end anastomosis is recommended. If end-to-side anastomosis is unavoidable, it should be performed as close as possible to the closed end (inset) to prevent elongation as the child grows. Approximation of the native jejunum and distal Roux-en-Y limb is indicated by small arrowheads and the long arrows indicate how small bowel contents may flow without reflux. **b** Inappropriate Roux-en-Y HJ. If the HJ is NOT close to the closed end, because the Roux-en-Y limb is NOT customized (arrowhead), elongation (double headed arrow in the inset) may occur with growth, causing redundancy and tortuosity that contributes to bile stasis and debris/stone formation (square, circle, star) in the elongated blind pouch, the Roux limb and/or intrahepatic ducts, as shown. As there is no approximation between the native jejunum and the distal Roux-en-Y limb, the jejunojejunostomy is T-shaped and jejunal contents may reflux into the Roux-en-Y limb (arrow with asterisk)

the Roux-en-Y jejunal limb later in life that can contribute to bile stasis (Fig. 9b). At Juntendo, the length of the Roux-en-Y limb is customized by identifying the ligament of Treitz and dividing the jejunum 15 cm distal to the ligament to create a Roux-en-Y jejunal loop that is brought up to be some 3 cm above the xiphoid process on the anterior abdominal wall so the Roux-en-Y jejunojejunostomy fits naturally in the splenic flexure after the jejunojejunostomy anastomosis is completed. This customization ensures there will be no redundancy of the Roux-en-Y limb (Fig. 9a).

Transecting the common hepatic duct

The common hepatic duct is usually transected at the level of distinct caliber change. Because residual CC mucosa has been implicated in malignant transformation, care must be taken to excise it completely, especially in older children. However, if the lumen of the common hepatic duct at the HJ anastomosis is 4 mm or less in younger patients with cystic CC, a Carrel patch with 2–3 mm of CC wall can be used to increase the diameter at the anastomosis to 6–7 mm as an option, because a larger lumen is easier to anastomose technically. A study of CC specimens from young pediatric patients followed-up for some 20 years found no evidence of premalignant lesions in excised CC specimens and no evidence for current malignant transformation [27].

Minimally invasive surgery

Laparoscopy

Hepato-biliary-pancreatic anatomy can be complicated and some procedures are often considered too complicated to be performed using minimally invasive surgery (MIS). However, the general concepts are exactly the same as for open CC excision; in other words, the above mentioned technique including IE can be performed as described but using laparoscopy or robotic assistance. While MIS can be cumbersome and time consuming, extra trocars improve maneuverability and with experience, MIS excision can approach open excision technically with far less surgical stress allowing patients to recover faster and go home earlier.

Patient/port positioning and initial preparation

The patient is positioned at the foot of the operating table in a reverse Trendelenburg position; vascular access is secured, the patient sedated, an orogastric tube is placed, and a urinary catheter is inserted. General anesthesia is induced conventionally. The operating surgeon stands at the patient's feet, an assistant with a laparoscope stands on the left side of the operating surgeon, and another assistant on the right side. The monitor is placed at the head of the operating table facing the surgeon. A GelPOINT® mini access platform (Applied Medical, Rancho Santa Margarita, USA) is inserted in a 2 cm long umbilical incision and used to introduce a 30° 10 mm laparoscope into the abdomen. Carbon dioxide pneumoperitoneum is established by gradual insufflation at 0.5 L/min increasing to 1.0 L/min resulting in a pressure of 10–12 mmHg maintained at 0.5–1.0 L/min. Three 5 mm trocars are inserted in the right upper quadrant, left paraumbilical area, and left upper quadrant, respectively.

Choledochal cyst dissection

Adequate exposure of the porta hepatis is achieved with a pair of Babcock forceps inserted through the left upper quadrant (left subcostal port in the anterior axillary line), to grasp and elevate the gallbladder to allow the CC to be dissected free from surrounding structures, such as the portal vein and hepatic artery. Dissection of the CC 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 (Fig. 10a), then to the distal portion. To prepare for excision, the cystic artery is identified

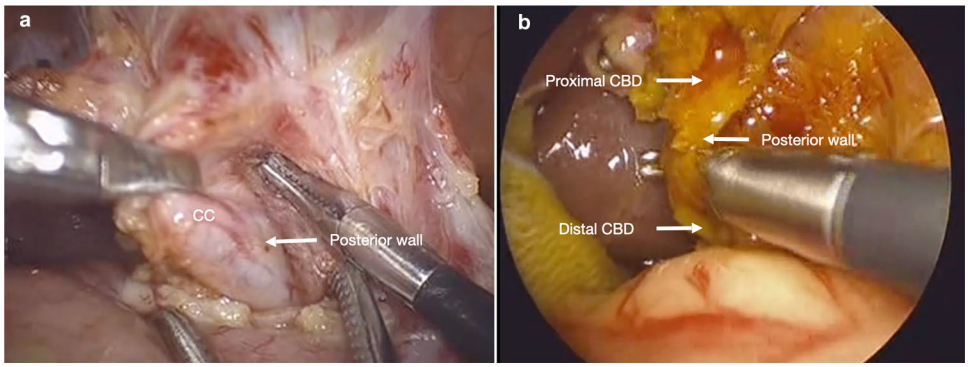

Fig. 10 Exposure of the posterior wall of the common bile duct (CBD). **a** Posterior wall of the CBD is dissected under laparoscopic control. **b** Anterior wall can be incised first, allowing visualization of the posterior wall under direct vision to facilitate safer dissection, as in the open technique

and divided with a Ligasure (Valley lab, Boulder, CO, USA) device. To excise the posterior wall of the cyst, the anterior wall is incised first to improve exposure of the posterior wall as described in the open CC excision section (Fig. 10b). This is used even in fusiform CC cases, especially if repeated episodes of preoperative pancreatitis have caused dense adhesions between the common bile duct and surrounding vital structures.

After the CC is freed, the distal part is divided at an appropriate level of the pancreaticobiliary junction and the stump is ligated with an endoloop. The proximal CC is excised at the level of the common hepatic duct, and HJ is completed. If the common hepatic duct is narrow, i.e., less than 5 mm, a 5 mm diameter Carrel patch can be considered for HJ [27]. Cholecystectomy is then performed. The excised CC and gallbladder are extracted through the umbilicus wound.

Intraoperative endoscopy

IE is performed by introducing a fine ureteroscope through an additional 3.9 mm trocar inserted in the left epigastrium and its tip is inserted into the common channel through the distal cyst under laparoscopic guidance, Fig. 7 [50]. As mentioned in the open CC excision section, IE facilitates transecting the common hepatic bile duct and the distal CBD by allowing exact levels to be determined, accurately (Fig. 7).

Extracorporeal transumbilical jejunal Roux-en-Y

HJ is described as it is the procedure of choice. The ligament of Treitz is identified and jejunum 15 cm distal from the ligament is exteriorized by extending the umbilical port incision, and a Roux-en-Y jejunojejunostomy is performed extracorporeally with customization and approximation of the Roux limb as described in the open CC excision section. An antimesenteric enterotomy, approximately 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 (Fig. 11). When creating the 5 mm enterotomy, a scalpel should be used (Fig. 11) instead of monopolar diathermy and bleeding points coagulated with bipolar diathermy, because considerable lateral thermal energy may be emitted during cutting with monopolar diathermy during laparoscopy and injure the bowel wall around the enterotomy, contributing to postoperative anastomosis leakage reported by many centers [51–55]. A maneuver developed at Juntendo facilitates suturing during laparoscopic HJ

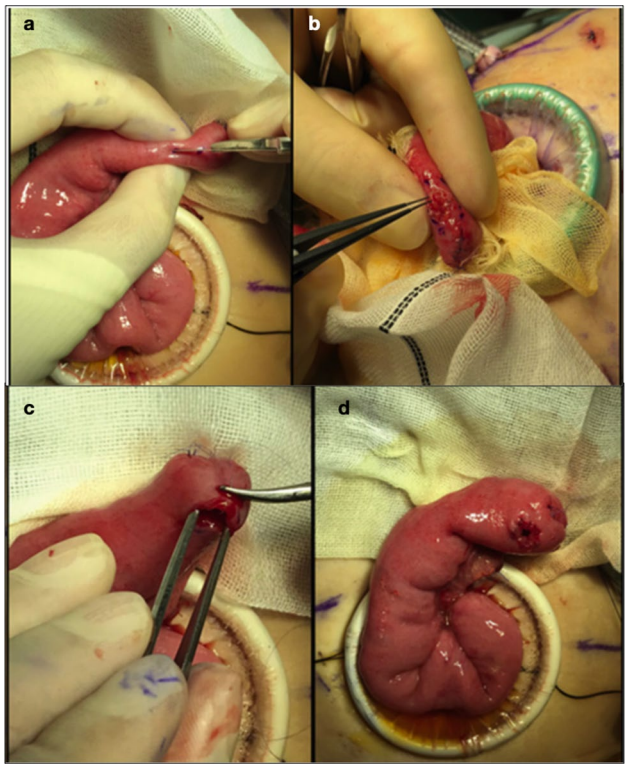

Fig. 11 Eversion of the mucosa of the enterotomy of the Roux-en-Y jejunum for laparoscopic hepaticojejunostomy. **a** Closed end of the jejunum is incised with a scalpel. **b** Bleeding points are coagulated with bipolar diathermy. **c** Mucosa at the incision is everted circumferentially using 7/0 absorbable sutures. **d** Everted mucosa of the jejunum

anastomosis by everting the mucosa of the Roux-en-Y-loop jejunum (Fig. 11) and the common hepatic duct mucosa circumferentially using 7/0 absorbable sutures and anastomosing securely to the common hepatic duct using 5/0 or 6/0 absorbable sutures [56]. This allows the lumen and mucosa on the jejunal side to be distinguished between (Fig. 11). Finally, the Roux-en-Y limb is returned to the abdominal cavity and the jejunal limb is passed through a retrocolic window to lie without tension at the porta hepatis.

Hepaticojejunostomy

Two additional trocars, one 3.9 mm lateral right subcostal and one 5 mm between the lateral right subcostal and right upper quadrant trocars, respectively, are required for the HJ anastomosis. End-to-side HJ is performed placing the back row first using interrupted 5/0 or 6/0 PDS sutures for neonates and 4/0 or 5/0 PDS sutures for older children; a needle holder in the right hand, the 5 mm port for the scope, and the 3.9 mm subcostal port as a needle receiver in the left hand. Both the right and left edge sutures are exteriorized and used as traction sutures during anastomosis of the anterior row to facilitate accuracy (Fig. 12) especially when the HJ anastomosis diameter is less than 5 mm.

Near-infrared fluorescence imaging with indocyanine green

Near-infrared fluorescence (NIRF) imaging with indocyanine green (ICG) improves intra-operative visualization of anatomic structures during pediatric MIS. As an aid to visualizing vascularity, it can be used for guiding resection and dissection during laparoscopic and robotic procedures [57].

Use of NIRF requires the injection of a specific Food and Drug Administration approved dye (ICG) that was initially used to study hepatic and cardiac function in humans [58, 59]. ICG is almost entirely metabolized by the liver and excreted into the bile in approximately 8 min and fluorescence can be detected to a maximum depth of 10 mm. ICG enhanced fluorescence has proven to be valuable during MIS for CC excision, especially when a CC is large (Fig. 13).

Minimally invasive surgery

Robotic assistance

The da Vinci surgical system (Intuitive Surgical, Sunnyvale, CA) is particularly useful for HJ. The robotic platform overcomes many of the limitations encountered during laparoscopic surgery; in fact, suturing during anastomosis and knot tying are much easier (Fig. 14) than during laparoscopic surgery, because the operating surgeon is more comfortable

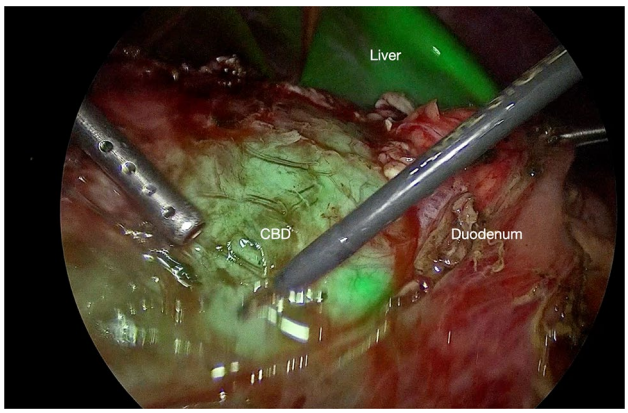

Fig. 13 ICG-enhanced fluorescence improves visualization during laparoscopy and increases safety

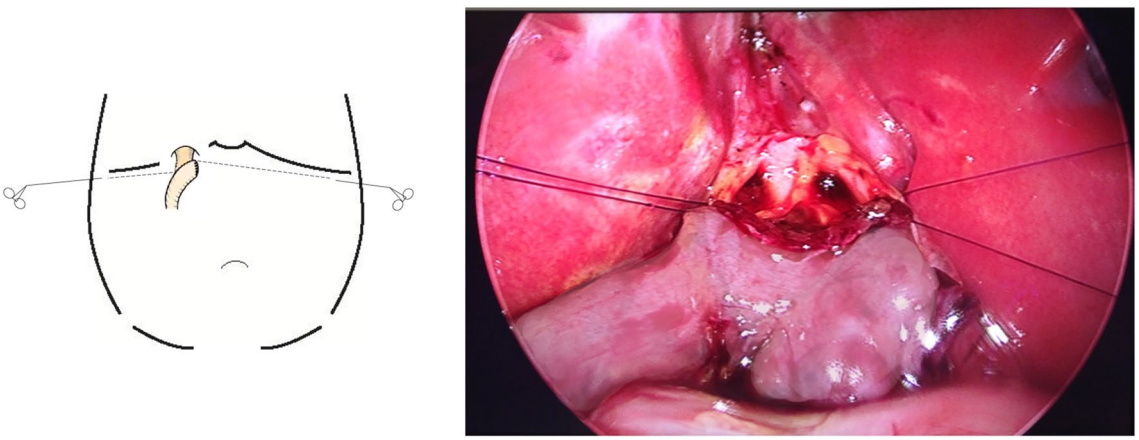

Fig. 12 Traction sutures during hepaticojejunostomy anastomosis. Both right and left edge sutures are exteriorized and used to apply traction during anastomosis to facilitate accuracy, especially when the HJ is less than 5 mm diameter

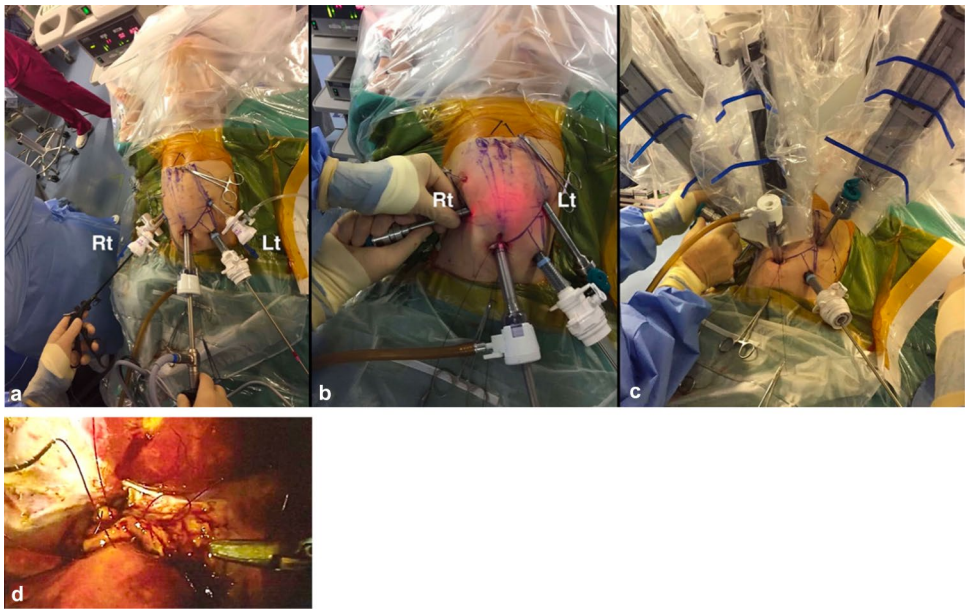

Fig. 14 Robotic-assisted hepaticojejunostomy. **a** Trocar positions for conventional laparoscopy. **b** Changing from laparoscopic to da Vinci trocars. The left (Lt) trocar has been replaced and the right (Rt) trocar is being replaced. **c** Docking of the da Vinci arms to da Vinci trocars. **d** Robotic-assisted hepaticojejunostomy

with no ergonomic issues to hinder performance. After the Roux-en-Y limb is brought up to the hepatic hilum for HJ, laparoscopic trocars are replaced with da Vinci trocars (Fig. 14). Robotic-assisted HJ can then be performed with interrupted sutures 5/0 or 6/0 absorbable sutures with all knots tied intracorporeally.

Outcomes

Open surgery

A review of 295 children aged of 15 years or less who had open CC excision performed between 1964 and 2022 reported that the mean age at onset was 3.2 years; age at initial onset of symptoms including abdominal pain and jaundice was 5 years or less in most ($n=238/295$) and between 6 and 15 years in the remainder ($n=57/295$). Mean age at CC excision was 4.2 years. Mean follow-up was 29.1 years. Treatment was primary CC excision ($n=256$), CC excision converted from intestinal drainage ($n=5$), and CC excision after other interventions, such as percutaneous transhepatic cholangiodrainage, T-tube drainage, and cholecystectomy ($n=34$). Surgical procedures performed were Roux-en-Y HJ ($n=275$), standard HD ($n=19$), and jejunal interposition HD ($n=1$). 165 children had IE performed. There was no operative mortality. Complications ($n=25$) included ascending cholangitis, intrapancreatic terminal choledochus calculi, pancreatitis, and bowel obstruction occurred in 20 children (6.7%) and 15 required surgical intervention (revision of HE, percutaneous transhepatic cholangioscopic lithotomy, excision of residual intrapancreatic terminal choledochus, endoscopic sphincterotomy, pancreaticojejunostomy, or laparotomy for bowel obstruction). In patients who underwent CC excision at the age of 5 years or less, there were no major complications, such as the intrahepatic stone formation, intrapancreatic terminal choledochus calculi, and anastomotic stricture of the HE. None of the165 children who had IE developed stones, anastomotic strictures, or cholangitis. Carcinoma in situ and dysplasia were identified in the excised CC in 2 children in whom the age at CC excision was 3 years and 1 year, respectively.

Laparoscopic surgery

Experience of 93 laparoscopic CC excisions by the authors at Juntendo and Monterrey between 2009 and 2022 is summarized. Cases requiring conversion to open laparotomy ($n=2$) and mini-laparotomy ($n=2$) were excluded, leaving 89 subjects; 64 females and 25 males. CC were fusiform ($n=46$), or cystic ($n=43$); forme fruste ($n=0$). Mean age (range) at surgery was 4.1 (0.2–14.1) years, and mean weight (range) at surgery was 16.9 (3.0–48.0) kg. 22 had IHBD dilatation. There were no intraoperative complications. Estimated mean blood loss was minimal at 15.1 mL. HJ diameters were: 6–9 mm (29/46 fusiform; 29/43 cystic) or more than 10 mm: (17/46 fusiform; 14/43 cystic).

IE involved both the common channel and IHBD ($n=54$; 32 fusiform, 22 cystic) and the IHBD alone ($n=35$) alone, because the ureteroscope could not be advanced into the distal intrapancreatic CBD and common channel. Protein plugs were present in the common channel in all 32 fusiform cases; massive ($n=9$), moderate ($n=18$), minimal ($n=5$) and cleared successfully by irrigation with normal saline from the side channel of the ureteroscope. There were no debris in the IHBD of the 18 who had IE of both the

common channel and IHBD. Debris were present in all 35 cases who had IE of the IHBD alone; moderate ($n=11$), minimal ($n=24$).

Although all are well after a mean follow-up of 7.8 years (range 2 months to 13.8 years) with cosmetically esthetic wounds, there have been 7 postoperative complications: pancreatitis (onset 8 months postoperatively from newly formed 3 × 3 mm debris that formed after thorough clearance of all protein plugs/debris seen on IE by irrigation; treatment was conservative medical management without relapses); pancreatic pseudocyst (onset 14 days after surgery, 2 × 2 cm pseudocyst with abdominal pain that formed after CC excision, the pseudocyst gradually resolved spontaneously); duodenal obstruction (the third part of the duodenum was found to be compressed by the Roux-en-Y limb that had been fixed inadequately to the colonic mesentery at exploratory laparoscopy in a cystic CC case, sutures between the Roux-en-Y limb and the colonic mesentery were released laparoscopically, postoperative recovery was uneventful): anastomotic leak (treated by mini-laparotomy); bile leak (onset immediately after surgery at the HJ treated conservatively); internal hernia (onset 5 years after CC excision at the Roux-en-Y reconstruction site, treated by mini-laparotomy); and HJ anastomosis stricture (onset 5 years after CC excision treated successfully by redo laparoscopy).

Results of mid- to long-term follow-up of laparoscopic CC excisions published recently [60–62] indicate that experienced laparoscopic surgeons can achieve results as good as those for open surgery with no significant differences in bile leakage or wound infections. However, while laparoscopic CC excision and Roux-en-Y reconstruction may be safe and effective in the hands of skilled laparoscopic surgeons, HJ anastomosis remains extremely challenging, technically.

Robotic-assisted surgery

There have been 22 robotic-assisted CC excisions performed at Juntendo between 2017 and 2022. Of these, 17 were female and 5 were male. CC were fusiform ($n=15$), cystic ($n=6$), and forme fruste ($n=1$). Mean age (range) at surgery was 5.9 (1.4–14.3) years, and mean weight (range) at surgery was 19.7 (9.0–47.8) kg. 14 patients had IHBD dilatation. All had Roux-en-Y HJ for biliary reconstruction. HJ diameters were 5 mm or less in 6 cases, 6–9 mm in 8 cases: more than 10 mm in 8 cases. The first 14 cases had hybrid surgery; laparoscopic cyst excision and robotic-assisted HJ anastomosis. There were no intraoperative operative complications. Only one postoperative complication was identified on routine follow-up but the patient was asymptomatic. A forme fruste CC case had stone formation in non-dilated IHBD at the porta hepatis probably due to stenosis at the HJ anastomosis. At surgery, the diameter at the anastomosis was 4 mm, and the excised CBD and the common hepatic duct used for HJ were found to be severely inflamed with denuded mucosa due to severe life-threatening pancreatitis, preoperatively.

A comparative study conducted at Juntendo between HE anastomoses performed with robotic assistance and conventional laparoscopy demonstrated that robotic assistance was superior providing further evidence that robotic sutures are more precise and easier to execute, thereby ensuring secure sutures, compared with laparoscopic sutures [63, 64]. Interestingly, for robotic assistance and hybrid laparoscopic/robotic assistance surgery, total anastomotic time is shorter than for a comparable laparoscopic HJ, although time per suture during the anastomosis itself is similar, while the coefficient of variation for time to taken place one suture during robotic-assisted HJ is shorter than for laparoscopic HJ [63].

During 2023, Juntendo began robotic-assisted dissection for CC excision, but robotic assistance cannot be applied for dissecting CC in neonates and small infants because of space constraints. If pediatric patients are large enough, robotic assistance improves the overall experience of CC excision. Robotic-assisted CC excision will gain popularity for treating children because of the obvious advantages of robotic surgery on recovery and outcome. NIRF with ICG, available with the da Vinci surgical system can be used to enhances visualization of tissue planes and facilitates dissection. A current disadvantage of robotic surgery is that it is far more expensive than laparoscopic surgery.

Malignancy

Histopathologic findings compiled from excised CC specimens include erosion of bile duct mucosa, epithelial desquamation, papillary hyperplasia with regenerative atypia, bile duct mucosa dysplasia, and presence of metaplastic changes. Malignancies arising during follow-up after CC excision have been reported in both children and adults with no typical time frame for malignancy to develop [65–69]; cases have been identified after short (up to 5 years) [70], mid-term (up to 10 years) [66] or long-term (up to 34 years) [71] follow-up. However, occurrence of biliary tract cancer during follow-up after CC excision can be as high as 11.4% in adults over the age of 18 years with a median age for diagnosis of 42 years (principally cholangiocarcinoma and gallbladder cancer) compared with children [72], and is uncommon in young children aged less than 5 years at the time of CC excision [49, 65, 73–76].

Hyperplasia identified in excised CC specimens is not considered premalignant and increases with age; a notable finding suggestive of a potential risk for malignancy [77] probably as a consequence of irritation caused by a mixture of bile and pancreatic juice associated with PBMU inducing abnormal cell transformation that initiates a cascade of

Table 1 Japanese Study Group on Pancreatico Biliary Maljunction Registry data (2008–2020) [78]. Malignancies in excised specimens (CBD, gallbladder, and IHBD)

	Registered patients ($n=1572$)		
	Children (0–14 yo) ($n=678$)	Adults (15–90 yo) ($n=894$)	
Incidental malignancies at diagnosis			
(−)	676 (99.7%)	681(76.2%)	
(+)	2 (0.3%)	213 (23.8%)	
Malignancies			
Gallbladder			
Gallbladder only (*n*)	0	155	
Gallbladder and CBD (*n*)	0	10[a]	
Total (*n*)	0	165 (18.4%)	$p<0.01$
CBD			
CBD only (*n*)	2	27	
CBD and IHBD (*n*)	0	14	
CBD and gallbladder (*n*)	0	10[a]	
Total (*n*)	2 (0.3%)	51 (5.7%)	$p<0.01$
IHBD only			
Total (*n*)	0	7	

CBD common bile duct, *IHBD* intrahepatic bile duct, *yo* years old

[a]same patients

histopathologic changes starting with benign changes that evolve through borderline malignant dysplasia and carcinoma-in-situ, to malignancy over time, suggesting that older children may eventually have higher risk for abnormal histopathology because of PBMU.

Latest Japanese Study Group on Pancreatico-biliary Maljunction (JSGPM) registry data for 2008–2020 [78] shows that biliary tract cancer was identified in excised CC specimens in 2/678 (0.3%) children who had CC excision when aged 14 years or less and in 51/894 (5.7%) of adults who had CC excision when aged 15 years or more [78] (Table 1) ($p<0.01$), demonstrating an increase with age that reflects increased risk for biliary tract cancer in CC cases with age [78–82]. JSGPM also reported that biliary tract cancer was also identified in gallbladders excised from 165/894 (18.4%) adults, but none in gallbladders excised from 678 children ($p<0.01$) [78]. Of note is that of 51 adults with malignancy identified in excised CC specimens, 10 cases (19.6%) also had malignancy in the gall bladder, and 14 cases (27.5%) also had malignancy in the IHBD. On the contrary, of 2 children with malignancy identified in excised CC specimens, there was no malignancy in the gallbladder or IHBD reported. Post-CC excision malignancy that developed at the site of the HJ anastomosis in adults, years after CC excision performed in childhood has been reported in 2 cases. One had CC excision when 2 years and was diagnosed with biliary tract cancer of an extrahepatic bile duct when 22 years postulated to arise from the common hepatic duct at the hepaticoenterostomy anastomosis [82] and the other had CC excision when 7 years and was diagnosed with biliary tract cancer at the HJ anastomosis when 32 years [76]. Because of these two reported cases, thorough routine follow-up, protocolized and performed regularly with a high index of suspicion, is mandatory long-term in all children after CC excision.

Author contributions Joel Cazares, Hiroyuki Koga, Atsuyuki Yamataka reviewed the manuscript.

Declarations

Conflict of interest The authors have no conflicts of interest to declare.

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