

Retrospective Study of Choledochal Cyst: Clinical Presentation, Diagnosis and Treatment

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Abstract: Choledochal cyst is a rare congenital anomaly lead to dilatation of intrahepatic or extrahepatic bile duct or both. Much about etiology, pathophysiology and natural course of the disease are still on debate. Gastroenterologists, surgeons and radiologists alike still strive to optimize their roles in the management of choledochal cysts. Here we have analyzed 30 operated cases of cc in our KEM hospital. We found most common age group of presentation of cc in adult is 32yr-41yr with female preponderance. Most common presenting symptom is pain. Though various radiological modalities were used to diagnose it but MRCP is most sensitive and specific for cc. In our series type I cc is most common occurrence of (63.33%). Out of 30 patients, one had developed cholangiocarcinoma and Whipples procedure was done. All other patients had undergone roux.en y hepaticojejunostomy with cyst excision without any major complication. One mortality due to malignancy. So in view of risk of malignancy early cyst excision and internal drainage is treatment of choice.

Keywords: choledochal cyst, cholangiocarcinoma, MRCP

1. Introduction

Epidemiology- Though cystic disease of the biliary tree has been described since 1723, much about its etiology, pathophysiology, natural course and optimal treatment remains under debate. Almost 300 years later, gastroenterologists, surgeons and radiologists alike still strive to optimize their roles in the management of choledochal cysts. To that end, much has been written about this disease entity in multiple attempts to unravel the enigma.

Choledochal cysts (CCs) are rare medical conditions with an incidence in the western population of 1 in 100 000–150 000 live births, although the incidence has been reported to be as high as 1 in 13 500 births in the United States and 1 in 15 000 births in Australia^{2, 3}. Chledochal cyst (CC) or dilatation of common bile duct (CBD) was first reported by Douglas in 1852. The rate is remarkably higher in Asian populations with a reported incidence of 1 in 1000, and about two third of cases occur in Japan⁴. The reason for this Asian preponderance is still unclear. There is also an unexplained female: male preponderance, commonly reported as 4: 1 or 3: 1.¹⁻⁴ Distribution of the different types of CCs are as follows: 50%–80% are type I, 2% type II, 1.4%–4.5% type III, 15%–35% type iv and 20% type v.¹⁴⁴

Although the etiology is unknown, choledochal cyst likely to be congenital. BABBITTS THEORY⁶ of cysts caused by anomalous pancreaticobiliary duct junction (APBDJ) or pancreatobiliary malunion (PBMU) , considered as most accepted theory in pathogenesis. This theory postulates that the long common channel allows mixing of the pancreatic and biliary juices, which then activates pancreatic enzymes. These active enzymes cause inflammation and deterioration of the biliary duct wall, leading to dilation. Furthermore, greater pressures in the pancreatic duct can further dilate weak-walled cysts. Many

studies have measured the amylase level³ in CC bile, which is always higher in patients than in controls.

Diagnosis of choledochal cyst is made based on a disproportionate dilation of extrahepatic biliary duct, without ruling out the possibility of a tumour, stone, or inflammation as a cause of this dilation. Incidence of diagnosis of choledochal cyst is much higher in children (80%) than in adults (20%) ¹⁴⁵. Symptom triads are: abdominal pain, jaundice and abdominal mass.

Furthermore, hepato-biliary diseases in adults can conceal the primary condition. In addition to this, ultrasound, CT, MRI, endoscopic cholangiopancreatography (ERCP), transhepatic percutaneous cholangiography (PTC) guides us for a detailed examination in order to verify the diagnosis.

Surgical options in treatment of choledochal cyst are excision/removal of the cyst together with a part of bile duct achieved through Roux-en-Y hepatico-jejunostomy. Open method is most standard procedure.

In most reported literature, an assisted mini-incision was needed, and studies reporting total laparoscopic Roux-en-Y cholangiojejunostomy (TLRCJS) are rare. The goal of this study¹⁴⁶ was to investigate how to treat hepatic portal bile duct diseases and perform jejunojejunostomy and cholangiojejunostomy totally laparoscopically . They evaluated the feasibility of TLRCJS in treating biliary tract diseases. It was concluded that TLRCJS is the best and first choice for patients with biliary tract diseases that need biliary-jejunal anastomosis¹⁴⁶. But it is essential that the surgeon has proficiency in laparoscopic surgeries.

Recently, a telemanipulative robotic surgical system was introduced, providing laparoscopic instruments with wrist-arm technology and 3-dimensional visualization of the operative field. A case of robot-assisted total excision of a choledochal cyst type I and biliary reconstruction in a 14-

year-old girl¹⁴⁸. No intraoperative complications or technical problems were encountered. An intra-abdominal collection occurred and was successfully treated with continuous percutaneous drainage. At one-year follow-up, she is doing well without evidence of recurrent cholangitis¹⁴⁸. This study report the feasibility and safety of robot-assisted laparoscopic resection of a type I choledochal cyst in a child. Compared to total laparoscopic surgery, the robot-assisted technique facilitates the most difficult part of the procedure, namely the creation of the hepaticojejunostomy anastomosis. Further experience is needed to properly evaluate the advantages and applicability of this approach, especially in the paediatric patient.

This is a retrospective study of 5 years operated cases of choledochal cyst: its clinical presentation, diagnosis and management and their outcome in our KEM Hospital, Mumbai, India. The purposes are:

- To identify ways of improving and maintaining the quality of care for patients.
- To assist in the continuing education of surgeons.
- To help make the most of resources available for provision of surgical devices.

2. Aims and Objectives

Aims

Retrospective study of operated cases of choledochal cyst: clinical presentation, diagnosis and treatment in a tertiary centre.

Objectives

1. To identify cases of choledochal cyst in adult (>12years).
2. To study their various clinical presentations.
3. To study their various diagnostic modalities.
4. To study their management and outcome.

3. Review of Literature

Classification Choledochal Cyst

Alonso-Lej and colleagues⁵ proposed the first classification system for CCs in 1959, describing 3 types of bile duct dilation, which has gained wide acceptance. Todani and colleagues⁶ expanded this system in 1977 to include the occurrence of intrahepatic and multiple cysts, and this modified classification is now most commonly used by clinicians.

Type-I cysts have subsequently been subclassified into 3 types. Type IA shows marked cystic dilation of the entire extrahepatic biliary tree, with sparing of the intrahepatic ducts. The cystic duct and the gallbladder arise from the dilated common bile duct (CBD). Type IB is defined by focal, segmental dilation of the extrahepatic bile duct. Although by definition the cyst can arise from anywhere within the extrahepatic biliary tree, it is most commonly distal, with the cystic duct branching off a normal CBD.

The biliary tree proximal to the gallbladder is usually normal. Type-IC cysts are smooth fusiform dilations of the entire extrahepatic bile duct, usually extending from the pancreaticobiliary junction to the intrahepatic biliary tree.⁷

Type-II cysts are discrete diverticuli of the extrahepatic duct with a narrow stalk connection to the CBD.

Type-III cysts are also called choledochoceles owing to their similarity in morphology, and postulated etiology, to ureteroceles. They consist of dilation of the distal CBD that is confined to the wall of the duodenum, and often bulge into the duodenal lumen. Although the outer lining of the cyst is always lined by duodenal mucosa, the inner lining can either be duodenal or biliary epithelium.⁸

Sarris and colleagues⁹ have further subdivided choledochoceles into 5 types based on the cysts' relations to the ampulla of Vater and the pancreatic duct. Although this system identifies the different configurations in which choledochoceles occur, the presentation and management of all subtypes are identical. Thus further characterizing type-III cysts into their subclassifications has not gained popularity among clinicians.

Type-IV cysts are multiple in nature and are further subdivided based on intrahepatic duct development. Type-IVA cysts are multiple intrahepatic and extrahepatic dilations. The intrahepatic duct dilation can be cystic, fusiform or irregular.⁷ Todani and colleagues¹⁰ have recommended further description of type-IVA cysts as cystic-cystic, cystic-fusiform or fusiform-fusiform to better delineate the nature of their intrahepatic and extrahepatic morphologies. Type-IVB cysts refer to multiple dilations of the extrahepatic biliary tree only, described radiographically as either a "string of beads" or "bunch of grapes" appearance.⁷

Type-V CCs refer to "Caroli disease", also known as "communicating cavernous ectasia", which is multiple saccular or cystic dilation of the intrahepatic bile ducts. Simple Caroli disease is isolated biliary dilation, whereas Caroli syndrome is cystic disease associated with congenital hepatic fibrosis.¹¹ Some authors have described Caroli disease with associated extra hepatic CC, but the distinction between this and type-IVA cysts is unclear. Levy and colleagues⁷ state that saccular dilation of the intrahepatic bile ducts and diffuse fusiform extra hepatic bile duct dilation less than 3 cm marks Caroli disease as separate from type-IVA cysts.¹² Figure 1 shows the different types of CCs.

Lilly and colleagues¹³ described an entity that they called "form fruste" CCs. Patients with these cysts present with typical symptoms of abdominal pain and obstructive jaundice, without bile duct dilation, but exhibiting an abnormal pancreaticobiliary duct junction. These patients have the same symptoms, histological evidence of inflammation and malignancy potential as those with CCs, and so some authors believe they should be included within the spectrum of disease.¹⁴⁻¹⁶

Kaneyama and colleagues¹⁷ described 4 patients, an incidence of 1.1% in that series, with a combination of type-I and type-II cysts. Intraoperatively, all 4 patients were morphologically identical, with a fusiform type-IC cyst with a type-II diverticulum arising from the middle portion of the cyst and the cystic duct draining into the right side of the diverticulum. The authors suggested that this may be a new clinical subtype. Four cases have also been reported of diverticular cysts of the cystic duct, which the authors suggested might be another subtype.¹⁸ The question arises, however, whether this is just a variant type-II cyst. Visser and colleagues¹⁹ recently challenged the traditional classification system, stating that it grouped together separate disease entities, marked by differing etiologies, natural courses, surgical options and complication profiles. They also contended that type-I and type-IVA cysts are simply variations of the same disease, as in their experience all type-I cysts had some element of intrahepatic dilatation, and the degree of intrahepatic dilatation defining one type versus the other was arbitrary. They advocated using descriptive nomenclature instead of the traditional alpha -numeric classification, and this has been supported by subsequent authors.²⁰

Pathogenesis

Pathogenesis of CC also supporting this theory are animal studies in which both ligation of the common bile duct and surgical creation of APBDJ lead to cystic dilation of the biliary tree in canine and murine models.³⁶ Administration of secretin, which increases pancreatic secretion, has been shown to dilate the CBD and gallbladder in patients with CC, whereas controls showed duodenal filling only. This demonstrates pancreaticobiliary reflux in these patients.³⁷ As described previously, the existence of form fruste CC supports the belief that APBDJ is related to the pathogenesis, symptoms and complications of overt CC.⁸ Skeptics of this theory call it into question because only 50%–80% of CCs are associated with APBDJ, and immature neonatal acini do not make sufficient pancreatic enzymes to explain antenatally diagnosed CC.^{39, 40}

Counterarguments by supporters of Babbitt's theory state that long common channels are arbitrarily defined in terms of length, with wide variation in measured length based on imaging modality and angles.⁴¹ In fact, different authors have defined a long common channel as anywhere from 10 to 45 mm. Therefore, APBDJ and a common channel may in fact exist in a much larger proportion of patients with CC, but may be under estimated owing to unrealistic long common channel definitions or inadequate imaging methods.

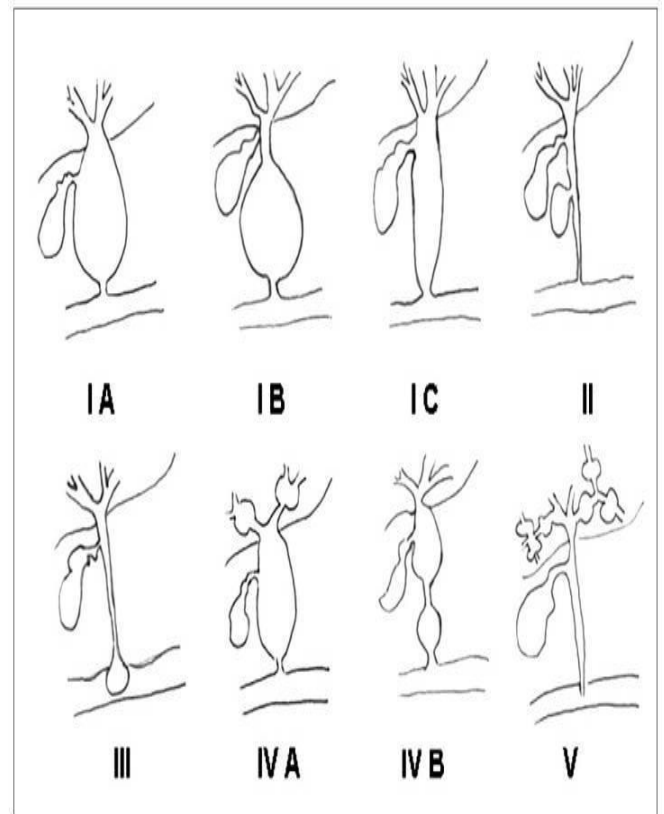


Figure 1.4 Choledochal cyst classifications

- [A] Type-Ia cystic dilation of the extra hepatic duct.
- [B] Type-Ib focal segmental dilation of the extra hepatic duct.
- [C] Type-Ic fusiform dilation of the entire extra hepatic bile duct.
- [D] Type-II simple diverticula of the common bile duct.
- [E] Type-III cyst/ choledochocoele distal intramural dilation of the common bile duct within the duodenal wall.
- [F] Type-IVa combined intrahepatic and extra hepatic duct dilation.
- [G] Type-IVb multiple extra hepatic bile duct dilations.
- [H] Type-V/Caroli disease multiple intrahepatic bile duct dilation.

Long common channel as any pancreatic biliary junction that lies outside of the duodenal wall and thus could result in pancreatic biliary reflux and mixing. There is a theory that CCs are instead purely congenital in nature.^{42, 43} This theory states that embryologic over proliferation of epithelial cells results in dilation during the cannulation period of development.

Davenport and Basu⁴⁴ noted that all neonatal CCs they reviewed were cystic in nature, and pathologically had fewer neurons and ganglions. Their theory was that round cysts are congenital in nature, with distal obstruction due to aganglionosis and proximal dilation (similar to Hirschsprung disease). In this case, chronic inflammation and symptoms occur owing to biliary stasis within the dilation rather than pancreatic reflux. They believe that fusiform dilations are acquired lesions due to APBDJ.⁴⁴

Ohkawa and colleagues⁴⁵ discovered that elastin fibres in the biliary tree do not develop until 1 year of age. They assert that increased neonatal tendency for round dilation is due to APBDJ and increased pressure within the bile duct, which yields round dilation before 1 year of age with the absence of elastin and fusiform dilation after the age of 1 year.⁴⁴⁻⁴⁶ Contradicting this is Xieijong's observation that neonatal CCs are round, whereas cysts associated with biliary atresia are fusiform, suggesting that round lesions are congenital and fusiform dilations are due to distal obstruction and thus acquired.¹⁰

Other authors speculate that all adult cysts are acquired due to distal obstruction, with longer, narrower stenosis leading to round lesions and shorter wider stenosis leading to fusiform lesions.^{10, 44} The distal obstruction may be due to sphincter of Oddi dysfunction or scarring and stone formation from an APBDJ.^{47, 48} The same theorists contend that type-IVA cysts result from combined distal as well as hilar and intrahepatic stenosis.¹⁰

Choledochal cysts are associated with many different developmental anomalies, which have given rise to some additional etiological theories. Such associations include colonic atresia, duodenal atresia, imperforate anus, pancreatic arteriovenous malformation, multiseptate gallbladder, OMENS plus syndrome, ventricular septal defect, aortic hypoplasia, pancreatic divisum, pancreatic aplasia, focal nodular hyperplasia, congenital absence of the portal vein, heterotopic pancreatic tissue and familial adenomatous polyposis.⁴⁷⁻⁶³

Embryologically, the pancreas forms when the ventral and dorsal pancreatic buds rotate, fuse and form connections with the biliary tree. Abnormal rotation and fusion may result in APBDJ and CC, pancreatic divisum and pancreatic aplasia.^{40, 50, 51, 56, 59, 64} Although the relation with enteric atresia is not clear, hypotheses include common developmental malformations or embryological cyst compression of either the gastrointestinal tract itself or its blood supply.^{52, 53, 58, 60} Familial adenomatous polyposis is associated with mutations in the adenomatous polyposis coli tumour suppression gene, which leads to interference with normal biliary cell-cell adherence, and therefore may lead to cystic dilation.⁴⁹ Reasons for the other associations remain unclear.

The above theories may explain the formation of type-I and type-IV cysts, but some authors contend that the aetiologies of the other types are quite distinct. As described previously, type-II cysts are true diverticula of the common bile duct, with histological evidence of little inflammation and carcinogenic potential. There also have been reports of "diverticular" cysts with no apparent communication with the biliary tree.⁶⁵ Therefore the question arises as to whether this is truly a cystic dilation caused by the above mechanisms or if it simply reflects a biliary duplication cyst. The etiology of choledochoceles is also not clear.

Wheeler⁶⁶ suggested that obstruction of the ampulla of Vater may result in localized dilation of the distal intramural bile duct. Others believe that increased pressure

owing to sphincter of Oddi dysfunction leads to such dilation. As mentioned previously, the inner lining of a choledochocoele can be biliary or duodenal epithelium, leading some authors to believe that these reflect either duodenal or biliary duplication cysts.^{67, 68}

Type-V CCs, or Caroli disease, is a disease entity quite separate from other CCs, with very different theories of etiology. Embryology of the intrahepatic biliary tree is as follows: a single layer of cells called a ductal plate forms around the portal branches, which then duplicates to form a double layer. Remodelling and selective resorption of the ductal plate commences in the 12th week and progresses to form the large bile ducts at the hilum to the small ductules in the periphery. Arrest of this remodelling results in Caroli disease. When such duct plate malformation occurs at the level of the large ducts, Caroli disease results. Malformation that continues to later stages of development such that the peripheral ductules are affected results in Caroli syndrome, with intrahepatic cysts reflecting large duct arrest and congenital hepatic fibrosis reflecting ductile arrest.^{7, 69} Caroli disease is associated with biliary atresia, which is also thought to be due to duct plate malformation. Caroli disease also is associated with both autosomal recessive and, less commonly, autosomal dominant polycystic kidney disease.⁷⁰⁻⁷² It is postulated that the genetic mutations responsible for the renal malformations also result in hepatic duct plate malformation.⁷¹⁻⁷³

Presentation

Clinical presentation can occur at any time, but 80% of patients present before the age of 10 years. The classic triad of symptoms, consisting of abdominal pain, jaundice and a palpable abdominal mass, occurs in less than 20% of patients, although almost two-thirds of patients present with 2 of the 3 symptoms.

Neonatal patients generally present with obstructive jaundice and abdominal masses, whereas adult patients present most commonly with pain, fever, nausea, vomiting and jaundice. Symptoms associated with CCs are usually due to the associated complications of ascending cholangitis and pancreatitis.

Complications associated with all types of CCs result from bile stasis, stone formation, recurrent superinfection and inflammation. Both dilated cysts and ductal stricture caused by chronic inflammation lead to proximal bile stasis, which in turn leads to stone and sludge formation and infected bile. Both of these factors lead to ascending cholangitis and further obstruction, resulting in the classic symptoms of episodic abdominal pain, fever and obstructive jaundice. Stone and protein plug formation in the distal common bile duct and pancreatic duct causes obstruction and resultant pancreatitis. Protein plug formation may be due to chronic inflammation and the formation of albumin-rich exudate or hyper secretion of mucin from dysplastic epithelium. Concurrent cholangitis in patients with type-IVA and type-V cysts is thought to be due to persistent bacterial colonization of the intrahepatic dilations and exacerbated by the presence of bile stasis,

sludge and stones. As the cysts are difficult to eradicate short of total excision and liver transplantation, these complications tend to be lifelong and may progress to liver abscess and life-threatening sepsis.

The obstruction and infections in all CCs, especially those with intrahepatic involvement, also lead to secondary biliary cirrhosis in 40%–50% of patients, such that patients can also present with signs and symptoms of portal hypertension such as upper gastrointestinal bleeds, splenomegaly and pancytopenia.

Portal hypertension⁹⁸ can also occur without cirrhosis, in which case the cyst can mechanically obstruct the portal vein.⁹⁹ Bile stasis can also lead to acalculous cholecystitis. About 1%–12% of patients with CCs present with spontaneous rupture and symptoms and signs of abdominal pain, sepsis and peritonitis.

The condition can be diagnosed when bilious paracentesis fluid is observed, bile stained ascites are found intraoperatively and there is peritoneal entry of contrast seen on Hydroxy iminodiacetic acid (HIDA) scan.¹⁰⁰ Ultrasounds may be misleading as the cyst may be decompressed from the rupture, and the biliary tree may thus appear normal.

The cause of spontaneous rupture has been hypothesized to be caused by mural fragility from chronic inflammation, increased ductal pressure due to distal obstruction or raised intrabdominal pressure.¹⁰⁰ The site of rupture is often at the junction of the cystic and common bile ducts, as this is a site of poor blood flow. Although patients with choledochoceles can also present with the above complications, they are often asymptomatic. Type-III cysts can also cause gastric outlet obstruction either by directly obstructing the duodenal lumen or by Intussusception.

Diagnosis

When patients present with the symptoms described, the first step toward making the correct diagnosis is imaging. The first imaging modality generally used for the biliary tree is ultrasonography, which, with the exception of type III and type-V cysts, will show a cystic mass in the right upper quadrant (usually at the porta hepatis) that is separate from the gallbladder. Diagnosis of a CC requires demonstration of continuity of the cyst with the biliary tree so that it can be differentiated from other intrabdominal cysts such as pancreatic pseudocysts, echinococcal cysts or biliary cystadenomas.¹⁰² Although most authors recommend other imaging modalities for this purpose. Akhan and colleagues¹⁰³ demonstrated continuity with the bile duct in 93% of their patients and recommended other imaging only when the diagnosis cannot be made based on an ultrasound. Sensitivity of ultrasonography in making the diagnosis is 71%–97%. Furthermore, given that ultrasonography is noninvasive and inexpensive, it is the modality of choice for follow-up surveillance. Reconstruction of 2-dimensional ultrasound images to form a 3-dimensional image has been advocated by some authors to view the cyst from different angles, allow full visualization of curved structures and estimate cystic

volume, all of which may be important for preoperative planning.¹⁰⁵ Unfortunately, all ultrasonography is limited by body habitus, bowel gas and overlying structures.¹⁰⁵ Furthermore, the size of the cyst may be underestimated by suboptimal probe pressure.¹⁰⁶ Endoscopic ultrasonography has been proven useful as it does not have any of these limitations and allows good visualization of the intrapancreatic portion of the common bile duct.¹⁰⁷

Another commonly used technique is a technetium-99 HIDA scan, which is recommended for viewing continuity with bile ducts. This type of scan will show an initial area of photopenia at the cyst, with subsequent filling and then delayed emptying into the bowel. The sensitivity of HIDA scans varies with type of cyst (100% for type-I and 67% for type-IV A cysts 54%) owing to the inadequacy of HIDA scans in visualizing the intrahepatic bile ducts. Neonatally, it is important to differentiate a CC from biliary atresia, both of which can present as an obstructive cyst in the porta hepatis. Biliary atresia requires urgent surgical correction via Kasai portoenterostomy within the first few weeks of life and carries a very poor prognosis of progression to cirrhosis, liver failure and death. Although it is difficult to distinguish a CC from biliary atresia on an ultrasound, a HIDA scan will show emptying of contrast into the bowel with CC, whereas retention of contrast owing to the distal obstruction indicates atresia. In addition, HIDA scans are useful for the diagnosis of cyst rupture, as this will show entry of contrast into the peritoneal cavity. Computed tomography (CT) scans are useful in showing continuity of the cyst with the biliary tree, its relation to surrounding structures and the presence of associated malignancy. It is superior to ultrasonography in imaging the intrahepatic bile ducts, distal bile duct and pancreatic head.¹⁰⁸ In patients with type-IV A cysts and Caroli disease, it is useful to delineate the intrahepatic dilations and the extent of disease such as diffuse hepatic involvement versus localized segmental involvement. This is important, as localized type-IV A cysts or Caroli disease can be treated with segmental lobectomy. Malignancy can be identified as a mass or a focal region of wall thickening on a CT scan.¹¹¹ Some authors recommend spiral CT to differentiate malignant cyst wall changes from reactive inflammation.¹¹³ Computed tomographic cholangiography (CTCP) has been used to delineate the full anatomy of the biliary tree to correctly plan surgery; this imaging modality is 93% sensitive for visualizing the biliary tree, 90% sensitive for diagnosing CCs and 93% sensitive for diagnosing lithiasis. Unfortunately, it was reported to be only 64% sensitive for imaging the pancreatic duct, as this depends on reflux of the contrast into the ducts. Virtual endoscopy based on CT images has been used to evaluate the biliary tree anatomy and identify defects successfully. Intravenous cholangiography and spiral CT can be combined to form a 3-dimensional image that very accurately delineates the postoperative anastomosis site. Of course, the drawbacks to using CT and CTCP are the risk of nephro- and hepatotoxicity with contrast and the exposure to ionizing radiation.

Endoscopic retrograde cholangiopancreatography (ERCP), percutaneous trans hepatic cholangiography (PTC) or intraoperative cholangiography is necessary for completely

delineating biliary anatomy preoperatively. Cholangiography is also useful for identifying an abnormal pancreaticobiliary duct junction or ductal filling defects, which may be stones or cancers. Although the use of cholangiography was previously ubiquitous in patients with CCs, it is slowly falling out of favour for a variety of reasons. For one, it is an invasive procedure with inherent risks of cholangitis and pancreatitis, which has been reported to be as high as 87.5% in patients with CCs. Given that many patients with cystic disease have long common channels, dysfunctional sphincter mechanisms and dilated ducts, this risk is greater in these patients than in the general population. Cholangiography also exposes the patient to ionizing radiation. Although ERCP has been reported to be the most sensitive imaging modality for CCs, this sensitivity does fall in certain situations. Recurrent inflammation and scarring may make cannulation of the ampulla difficult or impossible and may cause partial or complete obstruction at any point of the biliary tree, with no resultant biliary imaging. Full visualization of large cysts requires high dye load, and a compromise needs to be made between complete visualization and the risk of cholangitis or pancreatitis with increased amounts of dye. The use of a high volume of dye can also cause intense opacification, thus obscuring mucosal defects such as ulcers or malignancy, as well as dilate the cyst and overestimate its volume. Cholangiography is also not useful for postoperative imaging, as contrast is drained into the bowel without continuity to the hepatic duct. Additionally, although ERCP can be performed safely in paediatric patients, the procedure requires the administration anaesthesia. Finally, the sensitivity of ERCP and the quality of images is operator-dependent. Given the concerns regarding cholangiography,

Magnetic resonance cholangiopancreatography (MRCP) is now considered to be the gold standard.¹²⁰ Magnetic resonance imaging (MRI) and MRCP create images by differential signal intensity of stagnant pancreatic and bile secretions compared with surrounding structures. Unfortunately, intraductal air, blood, debris, stones or protein plugs, all of which are common in patients with CCs, can interfere with the signal and alter visualization¹⁰⁸. Nevertheless, sensitivity for diagnosis has been reported to be as high as 90%–100%. Although breath holding manoeuvres were previously necessary to negate the interference of motion artefact, new technology allows for quicker procedures and eliminates motion interference, such that breath holding is no longer necessary. This allows more convenient imaging for adults and obviates the need for anaesthesia in children. Magnetic resonance cholangiopancreatography is 84% sensitive for imaging of postoperative anastomosis. Unfortunately, sensitivity for assessing the pancreaticobiliary junction is as low as 46%–60%.¹²⁰ Magnetic resonance imaging is poor at imaging ducts or stones smaller than 5 mm and tortuous ducts.¹²⁰ Some authors suggest that the low sensitivity of MRCP in visualizing pancreaticobiliary junction is related to the small calibre of this junction, and they advocate the preimaging administration of secretin, which will increase pancreatic secretion and dilate the duct. Magnetic resonance cholangiopancreatography is 20% less

expensive than ERCP, although both modalities are twice as expensive as PTC.¹⁰⁸ Further advantages of MRCP over ERCP are that it avoids ionizing radiation; it is non-invasive and operator-independent; there are no complications of cholangitis and pancreatitis; and it can be coupled with MRI to image surrounding structures, lithiasis and malignancy. Endoscopic retrograde cholangiopancreatography allows for the performance of therapeutic procedures, but this is only necessary with type-III cysts.¹⁰⁸ Although all the information we have discussed so far pertains to the diagnosis of most CCs, type-III and type-V cysts deserve special consideration. Owing to their intramural nature, imaging abnormalities in choledochoceles are subtle, and the correct diagnosis is made preoperatively as little as 30% of the time. Generally, multiple imaging modalities are required to make the diagnosis. Upper gastrointestinal series (UGIS) may show a filling defect where the cyst bulges into the duodenal lumen. Endoscopy and ERCP will show smooth bulging of the papilla, and cannulation will opacify the dilated intramural common bile duct.¹⁰⁸ Magnetic resonance cholangiopancreatography and CTCP have been advocated by some authors for diagnosis, but these modalities do not offer the option of performing sphincterotomy for treatment of the choledochoceles. In contrast to other CCs, ultrasonography is not useful for the diagnosis of choledochoceles. The cysts are usually too small to visualize, and the normal diameter of the common bile duct makes connection to the biliary tree difficult to identify. Endoscopic ultrasonography, however, has been used with much success, as it achieves close proximity to the cyst and is not as hindered by surrounding bowel gas as traditional ultrasonography. Differential diagnosis for type-III cysts includes duodenal diverticuli and duplication cysts. Diverticuli fill up with contrast in an UGIS and fail to opacify with ERCP. Duplication cysts will have identical images to choledochoceles and are therefore very difficult to differentiate. Some authors claim that a muscular wall is present in duplication cysts and absent in choledochoceles. In patients with Caroli disease, ultrasounds and CT and MRI scans show multiple saccular dilations, which can be focal or diffuse and contain bile, sludge and stones. Computed tomography and MRI scans can also be used to diagnose associated cirrhosis, portal hypertension and varices, cholangitis, liver abscesses, malignancy and renal abnormalities.¹¹¹ Bloustein and colleagues described the “central dot sign,” which is a dilated duct surrounding a portal bundle, as pathognomic for Caroli disease. Initially found on ultrasounds, this sign can also be seen on MRI and CT scans.¹¹¹ Although the central dot sign does suggest Caroli disease, it is not pathognomic, as it is also seen in obstructive dilation. Also suggestive of Caroli disease is intraductal bridging, which involves echogenic septa traversing the duct. A beaded appearance of the intrahepatic bile ducts on HIDA scan can be diagnostic.

The differential diagnosis for Caroli disease includes 1) recurrent pyogenic cholangitis, 2) polycystic liver disease and 3) primary sclerosing cholangitis. Recurrent pyogenic cholangitis manifests as intra- and extrahepatic nonsaccular dilations with cast-like stones filling the entire lumen. Polycystic liver disease will have cysts that do not

communicate with the biliary tree. Primary sclerosing cholangitis manifests as mild, focal fusiform dilations with obvious distal obstruction and is associated with inflammatory bowel disease.

These differences can help differentiate Caroli disease from other conditions. Intrahepatic disease, such as type-IV A and type-V cysts. Once CCs have been diagnosed, careful treatment decisions need to be made. The third and final installment of this review series describes the management of biliary cystic.

Treatment

Choledochal cysts (CCs) are single or multiple dilations of the intrahepatic or extrahepatic biliary tree. If left untreated, they can cause morbidity and mortality from recurrent cholangitis, pancreatitis, sepsis, liver abscesses and cholangiocarcinoma. Comprehensive treatment involves medical management of complications, surgery and long-term follow-up. Management of CC depends on the type of cyst. Treatment of types I and IV A cysts has undergone much change in the past years. Although McWhorter first described cyst excision and hepaticojejunostomy in 1924, this surgery was initially abandoned because of multiple complications. Surgical strategies of cyst marsupialization and choledochoraphy failed because of significant mortality and morbidity. Subsequently, internal drainage of cysts via cystenterostomy became popular. Depending on anatomic proximity, cysts were incised and anastomosed to the duodenum or jejunum. Although this operation resulted in periprocedural relief of symptoms, multiple complications resulted. Reflux of the enteric contents into the cyst and biliary tree resulted in recurrent ascending cholangitis.

The site of anastomosis was also prone to stricture formation, resulting in obstruction, bile stasis, stone formation and recurrent cholangitis. Most importantly, surgeons found that leaving the cyst intact carried a significant risk of malignant transformation. The overall success rate of internal drainage procedures is 30%, the risk of postoperative malignancy is 30%, the mortality rate is 11%, and more than half who undergo this procedure require re-operation.¹⁴ Therefore, internal drainage is currently thought to be a dangerous and incomplete treatment of CC. Instead, surgeons favour complete cyst excision and hepaticoenterostomy. This separates the biliary tree from the pancreatic duct, thus ending the mixing of pancreatic and biliary secretions thought to be responsible for the pathogenesis of the disease; it also excises the damaged and presumably premalignant cyst tissue. If left in situ, the risk of cancer in the retained cyst is as high as 50% and occurs 15 years earlier than primary cancer. Therefore, the cyst should be excised completely from the hepatic hilum to the pancreatic duct. Recurrent inflammation, cholangitis and pancreatitis result in fibrosis of the ducts and adhesion to surrounding structures, making excision difficult. Intramural saline injection may separate the dissection planes and facilitate excision. If the cyst cannot be completely excised, the mucosa should either be stripped or destroyed by abrasion and iodine or alcohol application.

Any patient with a remnant cyst should receive regular surveillance via ultrasound.⁴⁶ The hepatico enterostomy can either be a hepaticoduodenostomy or a Roux-en-Y hepaticojejunostomy (RYHJ). The success rate of RYHJ has been shown to be as high as 92%.⁴⁶ This procedure has a reported complication rate of 7%, compared with a complication rate of 42% with hepaticoduodenostomy.¹⁵ Hepaticoduodenostomy carries with it the risk of bilious gastric reflux, gastritis and esophagitis, ulceration and malignant disease.

Furthermore, Todani and colleagues reverted from advocating hepaticoduodenostomy as the procedure of choice when they discovered a patient with hilar adenocarcinoma after excision. They hypothesize that the reflux of bile and active pancreatic enzymes from the duodenum can irritate the hilar epithelium and lead to malignant transformation. Many surgeons recommend end-to-end RYHJ to avoid the formation of a long blind pouch, which can result in bile stasis, reflux, cholangitis and stone formation.⁸ Authors also recommend creating a wide stoma at the hepatic hilum by extending the incisions up the lateral walls of the hepatic ducts to allow free drainage and avoid anastomotic stricture.¹⁰ The minimum diameter of the stoma has been suggested to be 3 cm. After cyst excision and hepaticoenterostomy, patients symptoms improve, intrahepatic duct dilations decompress and hepatic fibrosis and varices regress.¹¹¹

The complications of cystenterostomy and benefits of cyst excision and hepaticoenterostomy are both so substantial that surgeons now recommend revision of previous internal drainage procedures even for patients with no symptoms or complications. Early complications of cyst excision and hepaticoenterostomy include anastomotic leak, pancreatic leak with injury to the pancreatic duct, bowel obstruction due to intussusception, and bowel kinking due to manipulation or adhesions. Late complications include peptic ulcer disease, cholangitis, biliary calculi, pancreatitis, liver failure and cancer. Fibrosis and inflammation of cyst tissue at the time of surgery, such that the anastomosed margins are friable, result in poor healing, leakage and anastomotic stricture. Because fibrosis and inflammation increase with age, surgical complications become more common with older age at surgery, and surgery should be done as early as possible.

Cholangitis and calculi usually occur as a result of anastomotic stricture leading to bile stasis. Both cholangitis and pancreatitis can also result from duct stenosis or obstruction from debris, lithiasis and protein plugs. Pancreatic remnant cysts often cause these obstructive factors.¹¹² Many surgeons have had success and report a dramatic decrease in the complication rate with intraoperative cystendoscopy with identification and correction of stenosis and wash out of stones, debris and plugs. Cystendoscopy is also useful to identify the location of the pancreatic duct so that as much of the distal common bile duct can be excised without pancreatic injury. If intrahepatic duct stenoses are so high that they cannot be reached, a hepaticocutaneous jejunostomy for continued balloon dilation and stone extraction may be

warranted.¹¹⁰ Many authors also suggest the use of perioperative and long-term antibiotics to minimize the incidence of cholangitis.¹¹³ Postexcisional malignant disease, which has an incidence of 0.7%–6%, is thought to be due to remnant cyst tissue or subclinical malignant disease not detected before surgery. Therefore, some authors recommend intraoperative endoscopic ultrasonography and pathology of frozen sections to rule out dysplasia, hyperplasia and malignant disease.

All patients with CC also require life-long follow-up for cancer, usually via serial ultrasonography and monitoring of liver enzymes.⁷⁵ Surgery may be hindered by cirrhosis, portal hypertension and varices. Large pericystic varices, especially in the hepaticoduodenal ligament, increase the risk of postoperative bleeding. Therefore, if there is clinical or radiological evidence of cirrhosis or portal hypertension, esophagogastrosomy should be performed to identify and assess the extent of the varices. If large pericystic varices are a concern, portosystemic shunting can be performed for decompression before surgery.¹⁵ Gallstone ileus after hepatico- enterostomy has also been reported; this operation facilitates the passage of stones into the enteric tract.³³ Recently, many authors have reported success performing cyst excision and RYHJ via laparoscopy, with quicker recovery (mean hospital stay 5.5 days), less adhesions and improved cosmesis and ease of surgery because of magnification of the operative field.

Although initial laparoscopic surgeries took 9–10 hours, technological advances and operator experience have shortened this to a reasonable 4.5–5.5 hours.¹¹⁴ with the advent of robot-assisted surgery, which has improved manual dexterity; this operative time continues to become shorter. Laparoscopic treatment of CC is still evolving and promises many future benefits.

Various modifications to the surgical treatment described have been proposed. Shah and Shah¹¹⁵ proposed an appendiceal conduit. In this surgery, the cecum is mobilized to the splenic flexure, the appendix and its vascular pedicle are dissected, the caecal end is anastomosed to the hepatic duct, and the distal end is incised and then anastomosed to the jejunum as a tubular structure. Although the authors argue that this procedure is superior because of reduced risk of cholangitis from a high appendiceal lymphoid follicle content and physiologic duodenal bile drainage, this procedure has not gained popularity.¹¹⁶ Chang described a procedure in which a spur valve was placed in the ascending limb of the RYHJ in an attempt to prevent biliary reflux and cholangitis, but this resulted in a reoperation rate as high as 15% and complications of recurrent cholangitis and obstructive jaundice. Raffensperger¹¹⁶ and Zhang¹¹⁷ proposed the Chicago–Beijing procedure, which is still commonly used in China. This technique comprises cyst excision with a jejunal conduit between the hepatic stump and the duodenum, with an antireflux spur valve at the jejunoduodenal anastomosis.¹¹⁶ The benefits of this type of surgery are physiologic bile drainage and a valve that prevents reflux, whereas the disadvantages are a long, complicated procedure and anastomotic stricture formation. Surgeons performing this procedure report a

low reoperation rate of 0.8% in a large number of patients ($n = 481$). Other surgeons have abandoned this procedure, however, because of a high incidence of postoperative pain, which has been attributed to reflux biliary gastritis. Although all of these innovations appear great in theory, actual benefit has not yet been demonstrated, and cyst excision and RYHJ remain the procedures of choice.

Many patients are diagnosed with CC while acutely ill with active cholangitis, pancreatitis or rupture and bile peritonitis. In these conditions, the patient's physical state and intra-abdominal inflammation make the risks of surgery substantial. Furthermore, operating on acutely inflamed tissue results in poor healing, scarring and anastomotic stricture. Therefore, temporary measures should be performed, with definitive surgery performed when the patient's clinical condition allows.¹¹⁸ For active cholangitis and pancreatitis, the procedure of choice is external drainage via T-tube or percutaneous hepaticostomy. This is preferred over internal drainage because the only scarring is around the percutaneous tract, whereas the latter results in fibrosis of the duodenum, pericystic vascular structures and the hepaticoduodenal ligament, all of which make subsequent surgery very difficult.¹¹¹ Cyst rupture should include laparotomy and washout of the bile, external drainage and antibiotics for stabilization before definitive surgery.^{119, 120}

The risk of malignant disease with type II and III cysts is exceedingly low, and, thus, complete excision is not necessary. Simple excision of type II cysts is sufficient. Choledochoceles often just require endoscopic sphincterotomy to allow free duodenal drainage of bile and stones. Given the possibility that pancreatic and biliary secretion can mix within the choledochocoele and create a precancerous state, some authors recommend sphincterotomy even in asymptomatic patients.^{122, 123} Endoscopic excision via snare cautery is also possible for small cysts. Some surgeons believe that the common bile duct and pancreatic duct should be separated and reanastomosed to the duodenum to prevent pancreaticobiliary mixing. Large choledochoceles may cause biliary, duodenal or gastric outlet obstruction, in which case duodenotomy and cyst excision is warranted. For fruste CC are associated with significant risk of bile duct and gall bladder cancer, and treatment of this condition requires at least cholecystectomy. However, many authors believe that this is not enough to prevent malignant disease and advocate excision of the choledochus and hepaticenterostomy. Treatment of type IV A and V disease remains difficult. Type IV A is treated by cyst excision and a wide hilar hepaticenterostomy, but patients often continue to have symptoms because of intrahepatic disease. If the intrahepatic involvement is localized, a segmental hepatectomy may be performed. For diffuse disease, a percutaneous hepaticojejunostomy may allow for continuous stone extraction and dilation. Surgical or endoscopic unroofing of some intrahepatic cysts can also be performed for bile drainage.⁶⁵ Similarly, localized Caroli disease may be treated by hepatic lobectomy. Diffuse disease with recurrent or life-threatening cholangitis, liver failure, cirrhosis and portal hypertension or malignant disease requires orthotopic liver

transplantation. Some authors recommend early liver transplant if possible, because prognosis is very poor once malignant disease develops. Although prophylactic transplant is not warranted, aggressive surveillance for malignant disease in asymptomatic or minimally symptomatic patients is required. Complications of transplant include bleeding, sepsis, hepatic artery thrombosis and rejection. Recurrent lithiasis and cholangitis in both type IV A and V cysts can also be conservatively treated with prophylactic antibiotics for patients who are well, and intravenous and intraductal antibiotics can be used for ill patients; endoscopic or percutaneous lithotripsy and ursodeoxycholic acid can also be used. Ursodeoxycholic acid has proven effective in dissolving pre-existing stones and preventing the formation of new stones.

Malignant disease within the biliary tree mandates excision of the extrahepatic bile duct and adjacent liver, with regional lymph node excision. Unfortunately, less than 10% of cancers are resectable at diagnosis. Metastatic disease that affects the surrounding vasculature, organs or peritoneum may need percutaneous, endoscopic or surgical bile duct stent placement. A prophylactic or therapeutic gastroenterostomy to bypass the affected enteric tract and relieve obstruction may also be necessary. Distal malignant disease within the pancreatic head requires a Whipple procedure. Adjuvant chemotherapy or radiotherapy or both may increase survival, although prognosis after diagnosis of cancer is very poor.

Many previously asymptomatic women present during pregnancy for a number of reasons, including obstruction of the cyst by the gravid uterus, further stasis of pancreaticobiliary secretions because of biliary hypomotility, and cyst rupture because of increased intra-abdominal pressure during pregnancy and labour. Presenting symptoms are usually abdominal pain, fever and vomiting, usually due to cholangitis or pancreatitis. Diagnosis by ultrasonography may be difficult because of obscuration and alteration of normal anatomy by the gravid uterus. Given that computed tomography scans expose the foetus to ionizing radiation, magnetic resonance imaging (MRI) has been recommended as the imaging modality of choice.

Management of CC during pregnancy is difficult because of the surgical risk to both mother and foetus. Incidentally found CC should be followed with serial ultrasonography, and symptoms or rapid cyst enlargement should be treated conservatively. Patients with active cholangitis or pancreatitis should also receive conservative treatment of hospital admission and close observation, external drainage and antibiotics. Despite the label "conservative treatment," nonsurgical management should be aggressive because pancreatitis carries a maternal mortality rate of 20% and a foetal mortality rate of 38%. High intra-abdominal pressure during labour may cause cyst rupture, and many surgeons recommend elective caesarean section in the third trimester. Subsequently, definitive cyst excision and hepaticoenterostomy should be performed after delivery. Cyst rupture may mandate emergent surgery for bile evacuation and washout, but this should be

followed with external drainage, and definitive surgery should be performed during the postpartum period.

Ultrasound and MRI have been used to antenatally diagnose CC, even before the onset of signs or symptoms. Histopathology shows increased incidence and grade of liver fibrosis in pediatric patients with increasing age at surgery. Such fibrosis has been shown to regress after surgery. Furthermore, the longer surgery is delayed, the greater the potential for complications such as cyst rupture. Serial ultrasonography shows rapid cyst enlargement after birth at a rate of 2 mm per week, perhaps because of increased pancreatic and biliary secretion after the initiation of feedings. Additionally, the longer the biliary tree is exposed to the chronic inflammation associated with CC, the greater the risk of malignant transformation. Finally, the surgical complication rate is almost negligible in the neonatal period but increases with age at surgery. For all of these reasons, most pediatric surgeons advocate neonatal cyst excision for prenatally diagnosed CC, even before the onset of symptoms. While waiting for surgery, neonatal patients should receive serial ultrasonography and liver enzyme measurements; a rapidly enlarging cyst, cholangitis or worsening liver function should prompt expedient surgery.

Embryology of Biliary Ducts

Development of the Bile Duct

A hepatic diverticulum appears in the ventral wall of the primitive midgut early in the 4th week of intrauterine life in the development of the human embryo¹⁵². This small diverticulum is the anlage for the development of the liver, extrahepatic biliary ducts, gallbladder, and ventral pancreas. In the 4th week, two buds can be recognized in the hepatic diverticulum¹⁵³. The cranial bud becomes the liver and the extrahepatic biliary tree. The caudal bud develops into superior and inferior buds. From the superior bud, the gallbladder and cystic duct appear, and the right and left ventral pancreas develops from the inferior bud. By the 5th week, all elements of the biliary tree are recognizable¹⁵³. Meanwhile, bile canaliculi differentiate from hepatic cells. The terminal bile ducts grow out into the mesenchymal tissue of the septum transversum, which will produce the fibrous architecture of the liver, and the development of an intrahepatic duct system is completed by the 10th week¹⁵⁴. Marked elongation of the common duct occurs with plugging of the lumen by epithelial cells. Recanalization of the lumen of the common duct starts at the end of the 5th week and moves slowly distally. The lumen of the common duct extends into the cystic duct by the 7th week, but the gallbladder remains solid until the 12th week. By the 6th week, the common duct and ventral pancreatic bud rotate 180° clockwise around the duodenum. After completion of the rotation, the entrance of the common bile duct into the left posterior surface of the duodenum can be seen. Early in the 7th week, the bile and pancreatic ducts end in closed cavities of the duodenum. However, the cavity between the orifices of the dorsal pancreas and common bile duct is completely obliterated for the active epithelial proliferation of the developing common bile duct. Between the end of the 7th

and beginning of the 8th week, the bile duct develops two channels, and the ventral one continues into the lower segment¹⁵⁵. Still in the 8th week, in the distal segment of the duodenum, two parallel channels and vacuoles are still present¹⁵⁴.

Development of the Sphincter of Oddi

By the 7th week, further elongation of the hepatopancreatic duct pushes the junction of bile and pancreatic ducts out to the level at which the intestinal muscle is forming. In the 8th week, the site of the junction is initially retracted through the slit of the duodenal wall, and soon comes to lie within the submucosa of the intestine¹⁵⁵. The muscle of the sphincter of Oddi develops from a concentric ring of mesenchyme surrounding the periampullary portion of the bile and pancreatic ducts. At about the 10th week, 4 weeks after the intestinal muscle has formed, the muscle of the sphincter of Oddi undergoes differentiation. The sphincter choledochus inferior appears first around the bile duct, which is carried away from the intestinal muscle, setting it up as an independent agent. After that, the sphincter choledochus superior is formed. By the 12th week, the bile and pancreatic ducts enter the duodenum obliquely, as a result of which the fibres appearing on the mucosal side of the bile duct seem to extend between the two margins of the transverse slit in the circular muscle layer of the duodenum. At this time, the liver begins to secrete bile that flows via the extrahepatic biliary tree into the duodenum. In the 16th week, the muscularis propria extends from just outside the fenestra to the upper end of the ampulla. By the 28th week, the musculus proprius is differentiated almost to the distal end of the ampulla.

Anatomy of Hepatobiliary System

Liver Anatomy

To understand the anatomy and physiology of the biliary tract and the production of bile, it is necessary to briefly outline the anatomy of the liver. The liver is divided macroscopically into the right and left lobe by the falciform ligament anteriorly (Fig. 1.1). Inferiorly, this corresponds to the round ligament and umbilical fissure.

The right lobe is further divided by the gallbladder fossa into the right hemiliver to the right of the gallbladder and the quadrate lobe to the left. The fourth lobe (caudate) is posterior and surrounds the inferior vena cava. Hence, anatomically the liver is divided into two main lobes and two accessory lobes. With improved understanding of liver function, the concept of functional anatomy has developed.

This was initiated by Cantlie in 1898 and was enhanced by McIndoe in 1929, Ton That Tung in 1939, and Couinaud in 1957. In December 1998, the Scientific Committee of the International Hepato-Pancreato-Biliary Association created a terminology committee to deal with confusion in the nomenclature of hepatic 3 anatomy and liver resections. This committee formulated a new terminology termed The Brisbane 2000 Terminology of Liver Anatomy and Resections. This is now internationally accepted. It is

anatomically and surgically correct, consistent, self-explanatory, linguistically correct, precise and concise.

The liver was divided into three functional livers: the right, the left and the caudate. The separation between the right and left hemi liver is at Cantlie's line, which is an oblique plane extending from the centre of the gallbladder bed to the left border of the inferior vena cava. In this plane runs the middle hepatic vein, which is an important radiological landmark.

The right hemi liver is divided further into two sections by the right portal scissura (anterior and posterior sections), within which runs the right hepatic vein. Each section is then divided on the basis of their blood supply and bile drainage into two segments. The anterior section is divided into segment 5 (inferior) and segment 8 (superior) and the posterior section into segment 6 (inferior) and segment 7 (superior) (Tables 1.1, 1.2 and 1.3).

The left hem liver is divided into three segments. Segment 4 (quadrate lobe) is known as the left medial section, which lies to the right of the falciform ligament and its right margin forms the right margin of the left hemi liver. Segment 3, which lies in the anterior part, and segment 2, which lies in the posterior part of the left hemi liver, form the left lateral section. The left lateral section lies on the left of the falciform ligament. Between segment 2 and segment 3 runs the left hepatic vein. The caudate hemi liver (segment 1) is considered separately because of its separate blood supply and venous and bile drainage².

Blood Supply and Venous Drainage

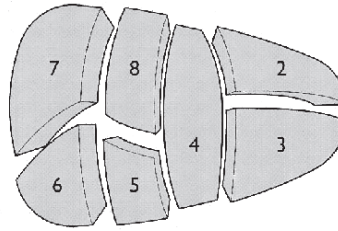
The arterial supply to the liver in early gestation life is from three main sources: the left hepatic artery from the left gastric artery; the middle hepatic artery (common hepatic artery) from the celiac trunk; and the right hepatic artery from the superior mesenteric artery. With further development, the blood supply assumes the adult pattern, with atrophy of both the right and left hepatic arteries and the common hepatic artery (middle hepatic) supplying the whole liver (Fig.1.2)³. This adult pattern occurs in around 67% of individuals⁴. The common hepatic artery gives the right and left hepatic arteries, which supply the right and left hemi livers, respectively. In 90% of cases, segment 4 is supplied by a named branch (middle hepatic) from either the right or left hepatic artery (45% each).

The other variations that occur are: The common hepatic supplying the right liver and the left hepatic arising from the left gastric in 8%. The common hepatic supplying the left liver and the right hepatic arising from the superior mesenteric artery in 11%.

- Persistence of all three arteries in 3%.

Table 1.1: First Order Division

Schematic
diagram of the segments
for reference in the table
(for purposes of clarity Sg1 is not shown)



Anatomical term

Couinaud segments
referred to

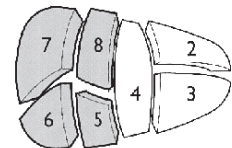
Term for surgical resection

Diagram
(pertinent area is in
heavy black outline)

Right hemiliver
OR
Right liver

Sg5-8(+/-Sg1)

Right hepatectomy
OR
Right hemihepatectomy
(stipulate +/- segment 1)



Left hemiliver
OR
Left liver

Sg2-4 (+/-Sg1)

Left hepatectomy
OR
Left hemihepatectomy
(stipulate +/- segment 1)

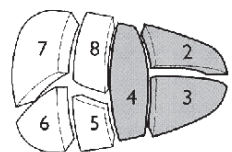
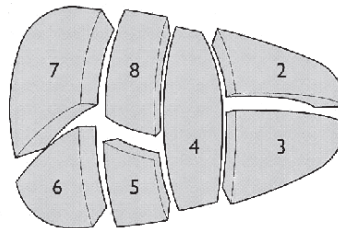


Table 1.2: Second Order Division

Schematic
diagram of the segments
for reference in the table
(for purposes of clarity Sg1 is not shown)



Anatomical term

Couinaud segments
referred to

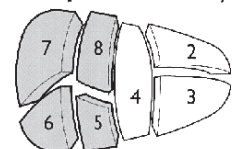
Term for surgical resection

Diagram
(pertinent area is in
heavy black outline)

Right hemiliver
OR
Right liver

Sg5-8(+/-Sg1)

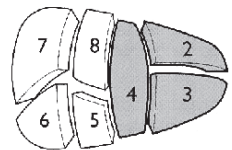
Right hepatectomy
OR
Right hemihepatectomy
(stipulate +/- segment 1)



Left hemiliver
OR
Left liver

Sg2-4 (+/-Sg1)

Left hepatectomy
OR
Left hemihepatectomy
(stipulate +/- segment 1)

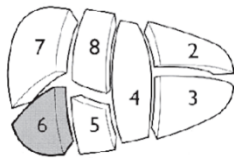
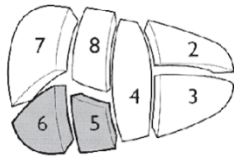


- Atrophy of the common hepatic artery in 12%, with the liver supplied by the:
- Right hepatic in 9%
- Left hepatic in 1%
- Both right and left in 2%.

The left hepatic arising from the left gastric is usually easy to identify in the gastrohepatic ligament. When this artery is present, care should be taken not to damage it when performing a gastrectomy.

The right hepatic artery arising from the superior mesenteric artery, on the other hand, is more variable. It ascends behind the pancreas in relation to the portal vein, and in the portal pedicle it assumes a posterior location, usually slightly to the left of the portal vein.

Table 1.3: Third Order Division

Anatomical term	Couinaud segments referred to	Term for surgical resection	Diagram
Segments 1–9	Any one of Sg1 to Sg9	Segmentectomy	 (Segmentectomy 6)
2 contiguous segments	Any two of Sg1 to Sg9 in continuity	Bisegmentectomy	 (Bisegmentectomy 5,6)

The venous drainage of the liver is into the inferior vena cava through the right, middle and left hepatic veins. The union of superior, middle and inferior branches usually forms the right vein, where the superior is the largest branch.

The right hepatic vein trunk joins at the right margin of the vena cava at a point separate and slightly above the trunk that is formed by the middle and left vein. The middle hepatic vein forms from two veins arising from segment 4 and segment 5. The middle hepatic vein joins the left hepatic vein to form a common trunk before draining into the vena cava in 90% of people. The left hepatic vein is more variable and is usually formed by the union of the branches from segment 2, segment 3 and segment 4.

Intrahepatic Bile Ducts

There are more than 2 km of bile ductules and ducts in the adult human liver. These structures are far from being inert channels, and are capable of significantly modifying biliary flow and composition in response to hormonal secretion. Bile secretion starts at the level of the bile canaliculus, the smallest branch of the biliary tree. They form a meshwork between hepatocytes with many anastomotic interconnections. Bile then enters the small terminal bile ductules (canals of Hering), which provide a conduit through which bile may traverse to enter the larger perilobular or interlobular bile ducts. The interlobular bile ducts form a richly anastomosing network that closely surrounds the branches of the portal vein. These ducts increase in calibre and possess smooth muscle fibres within their wall as they reach the hilum of the liver.

Furthermore, as they become larger, the epithelium becomes increasingly thicker and contains many elastic fibres. These ducts anastomose to form the segmental branches (from segment 1 to segment 8). In 80 to 85% of individuals, these segmental branches anastomose to form the anterior (segment 5 and segment 8) and posterior sectorial bile ducts (segment 6 and segment 7) in the right hemi liver. With the union of these two sectorial ducts, in 57% of individuals the right hepatic duct is formed. The right hepatic duct is usually short—approximately 9 mm in length. In the left hemi liver the segmental branches 2 and

3 anastomose to form the left hepatic duct in the region of the umbilical fissure. The anastomosis of segment 4 to the left hepatic duct usually occurs as a single trunk to the right of the umbilical fissure in 67% of individuals¹³³. The left hepatic duct is generally longer and more surgically accessible than the right hepatic duct. The caudate lobe (segment 1) is drained by both right and left hepatic ducts. Its arterial supply is also from both right and left portal vein and hepatic artery, with small venous branches draining directly to the inferior vena cava. The anatomy of this third hemi liver is revealed in certain pathologic conditions, such as Budd–Chiari syndrome where the outflow of the three hepatic veins is obstructed, leading to diversion of blood to the caudate lobe resulting in hypertrophy¹³⁴.

Extrahepatic Bile Ducts

The joining of the right and left hepatic ducts forms the common hepatic duct. The accessory biliary apparatus, composed of the gallbladder and cystic duct, joins the common hepatic duct to form the common bile duct that drains bile into the duodenum. This comprises the extra hepatic biliary system. The confluence takes place at the right of the hilum of the liver, anterior to the portal venous bifurcation and overlying the origin of the right branch of the portal vein (Fig. 1.5). The biliary confluence is separated from the posterior aspect of segment 4 of the left liver by the hilar plate, which is the fusion of connective tissue enclosing the biliary and vascular structures with Glisson's capsule.

Gallbladder and Cystic Duct

The gallbladder is a reservoir of bile in the shape of a piriform sac partly contained in a fossa on the inferior surface of the right hepatic lobe. It extends from the right extremity of the porta hepatis to the inferior border of the liver. It is 7 to 10 cm long and 3 to 4 cm broad at its widest part, and can hold from 30 to 50 ml. The gallbladder is divided into a fundus, body, infundibulum and neck.

The fundus extends about 1 cm beyond the free edge of the liver. The body is the largest segment. The infundibulum is the transitional area between the body and the neck.

Hartmann's pouch is a bulge on the inferior surface of the infundibulum. Gallstones may become impacted here and can cause obstruction of the cystic duct. The neck is the tapered segment of the infundibulum that is narrow and joins the cystic duct.

The cystic duct is 3 to 4 cm long and passes posteriorly inferior and to the left from the neck of the gallbladder to join the common hepatic duct to form the common bile duct (CBD). The mucosa of the cystic duct is arranged with spiral folds known as the valves of Heister.

The arterial supply to the gallbladder is from the cystic artery. Because the cystic artery is an end artery, the gallbladder is more susceptible to ischemic injury and necrosis as a result of inflammation or interruption of the artery. The cystic artery can originate from the right hepatic, left hepatic or the common hepatic artery, and it can be anterior or posterior to the common hepatic duct.

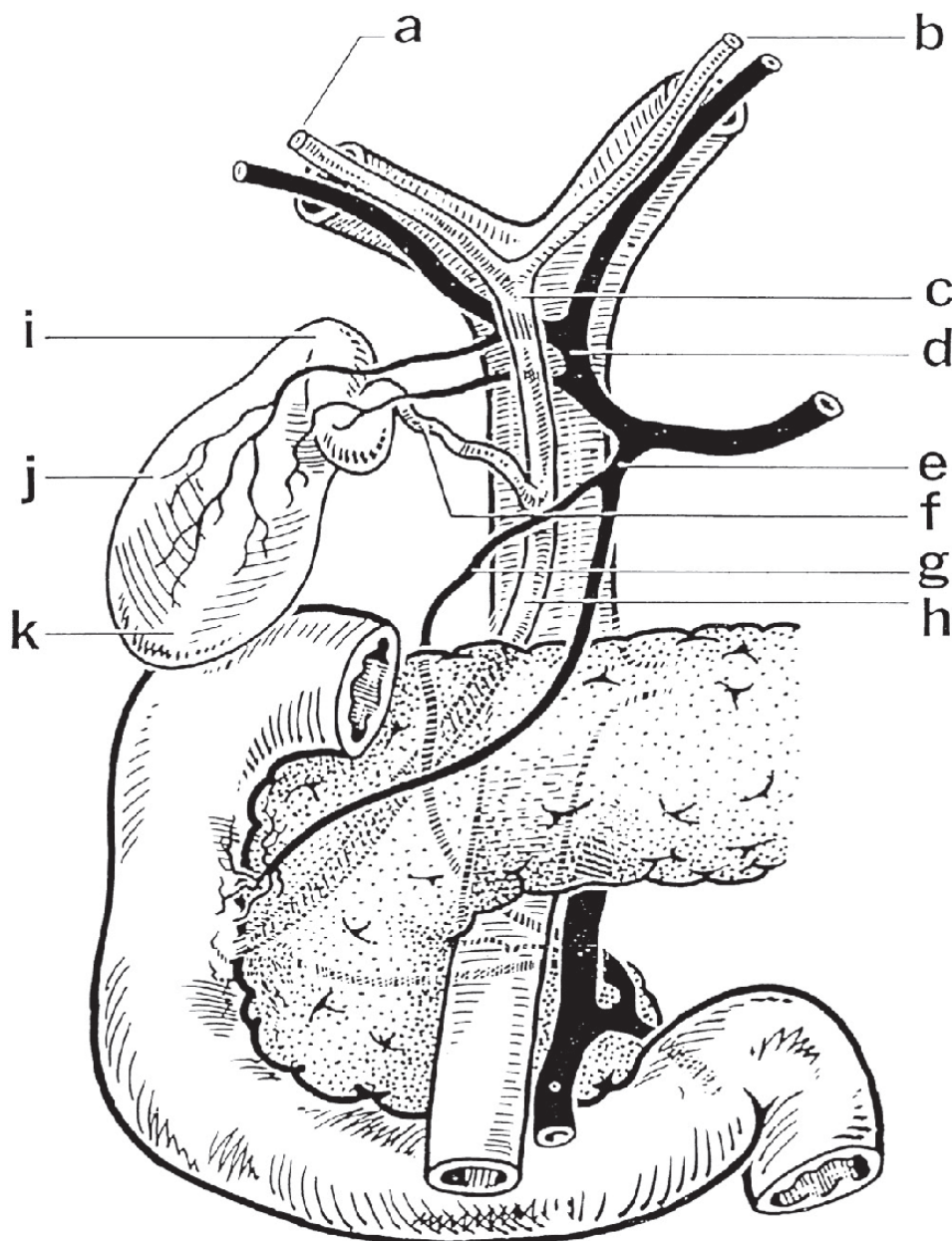


Figure 1.1 The anatomy of the extrahepatic biliary system: (a) right hepatic duct, (b) left hepatic duct, (c) common hepatic duct, (d) hepatic artery, (e) gastroduodenal artery, (f) cystic duct, (g) retroduodenal artery, (h) common bile duct, (i) neck of the gallbladder, (j) body of the gallbladder, (k) fundus of the gallbladder. (Reprinted from Blumgart LH, ed. Surgery of the liver and biliary tract, 3rd ed., p. 14. © 2000)

The venous drainage is through the cystic vein, which drains into the portal vein. There are also some small veins that drain directly into the liver to the hepatic veins. The lymphatic drainage of the gallbladder proceeds mainly by four routes, which form two pathways that drain in the

thoracic duct. 1 Superior and external, drains the fundus (around 6% of cases). 2 Superior and medial, drains the medial aspect of the gallbladder (around 10% of cases). 3 Inferior and external, drains the body of the gallbladder

(present in 82% of cases). 4 Inferior and medial, from the body of the gallbladder (constant).

All four routes drain to both pathways, except the inferior and external which drain only to the inferior pathway. This is important in cases of gallbladder cancer, which can spread to the liver; because of its extensive lymph drainage to both pathways, cure by radical surgery is difficult. The gallbladder is innervated by the vagus nerve through its hepatic branch from the anterior vagal trunk. The gallbladder is also innervated by the sympathetic nervous system through the celiac plexus. Fibres in the right phrenic nerve may also be distributed to the gallbladder through the hepatic plexus.

The Duct of Luschka

The duct of Luschka is a small bile duct, running in the bed of the gallbladder, outside the wall. It is present in 50% of individuals. This duct is surgically significant because it may be injured during cholecystectomy and may result in bile fistula unless ligated. Recent reports demonstrated a 1.5 to 2.0% incidence of bile leak from the duct of Luschka after laparoscopic cholecystectomy. Ligation has no consequences as it is an end duct that drains an isolated segment.

Common Bile Duct

The common bile duct forms by the junction of the cystic duct with the common hepatic duct. Its course is divided into supraduodenal, retroduodenal, pancreatic and intraduodenal (joins the main pancreatic duct to form the sphincter of Oddi, which will be discussed separately).

The supraduodenal segment usually lies in the free border of the hepatoduodenal ligament. It runs to the right of the hepatic artery and anterior to the portal vein. The retroduodenal segment descends posterior to the first part of the duodenum and slightly obliquely from right to left. The pancreatic segment is related to the head of the pancreas; it can run entirely retropancreatic or travel through its parenchyma. The diameter of the common bile duct is often used as an indication of biliary pathology. Its "normal" size varies depending on the modality used to measure it, and a range of 4 to 13 mm has been reported.

The most common modality to examine the common bile duct diameter is ultrasound, and a diameter up to 6 mm is considered normal. Some consider the equivalent in contrast radiology to be 10 mm; this depends on the magnification.

Sphincter of Oddi

The common bile duct enters the duodenum approximately 8 cm from the pylorus in the second part of the duodenum. The site entry is marked by a papilla (major papilla). Its

position can be variable; in approximately 13% of individuals it can be located at the junction of the second and third part of the duodenum, or even more distally¹³⁵. A transverse fold of mucosa usually covers the papilla. The papilla is identified as a small nipple or pea-like structure in the lumen the duodenum.

The main pancreatic duct of Wirsung joins the common bile duct and forms a common channel in approximately 85% of individuals. In 15%, they open either separately or as a V junction with the duodenal mucosa. In 4% of individuals, the body and tail of the pancreas drain via the duct of Santorini (pancreas divisum) to the minor papilla. In this instance, only the ventral aspect of the pancreas drains through the duct of Wirsung. The minor papilla is located proximal and slightly anterior to the major papilla.

The human sphincter of Oddi is generally a continuous smooth muscle structure that is subdivided into several parts that largely reflect the arrangements found in other animal species (Fig. 1.3).

1. Sphincter choledochus consists of circular muscle that surrounds the common bile duct.
2. Pancreatic sphincter surrounds the intraduodenal portion of the pancreatic duct before its juncture with the ampulla.
3. Fasciculi longitudinales are composed of longitudinal muscle fibers between the pancreatic and bile ducts.
4. Sphincter ampullae are composed of longitudinal muscle fibers that surround the papilla.

Blood Supply

The blood supply to the common bile duct is also divided into three segments (Fig. 1.4). The supraduodenal segment of the duct essentially has an axial blood supply. The blood supply originates from the retroduodenal artery, right hepatic artery, cystic artery, gastroduodenal artery and the retroportal artery. On average there are eight small arteries with the main two running along the side of the common bile duct at 3 and 9 o'clock. Sixty percent of the arterial blood supply occurs from the duodenal end of the duct, and 38% is from the hepatic end. Only 2% of the arterial supply is nonaxial, arising directly from the main hepatic trunk.

The second segment is the retropancreatic part of the duct, which is supplied by the retroduodenal artery. It provides blood to the multiple small vessels running around the duct to form a mural plexus.

The third segment is the hilar duct, which receives its blood supply from the surrounding blood vessels, forming a rich network. The veins draining the bile duct correspond to the described arteries. They drain into veins at 3 and 9 o'clock on the side of the common bile duct.

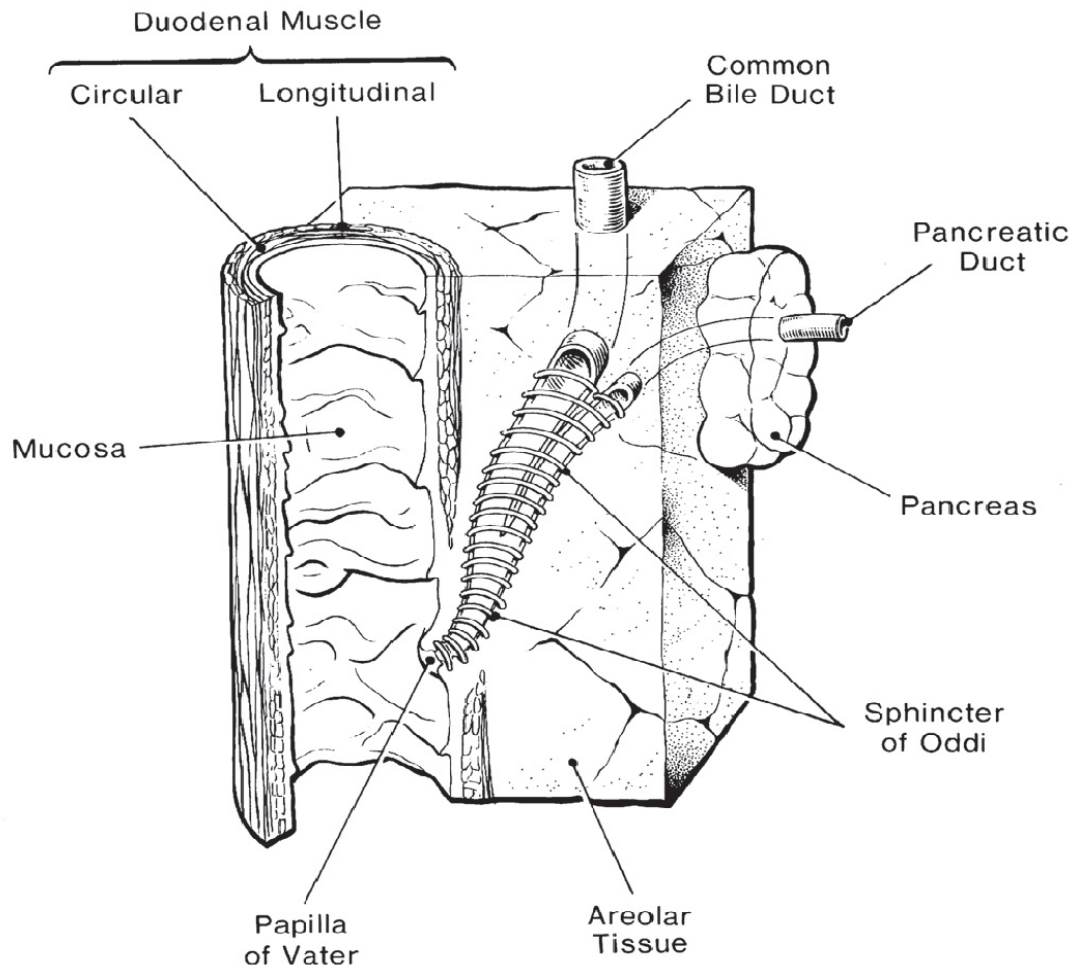


Figure 1.2The choledochoduodenal junction. The sphincter muscle is predominantly circular in orientation, and extends beyond the wall of the duodenum. There is a small extension along the pancreatic duct

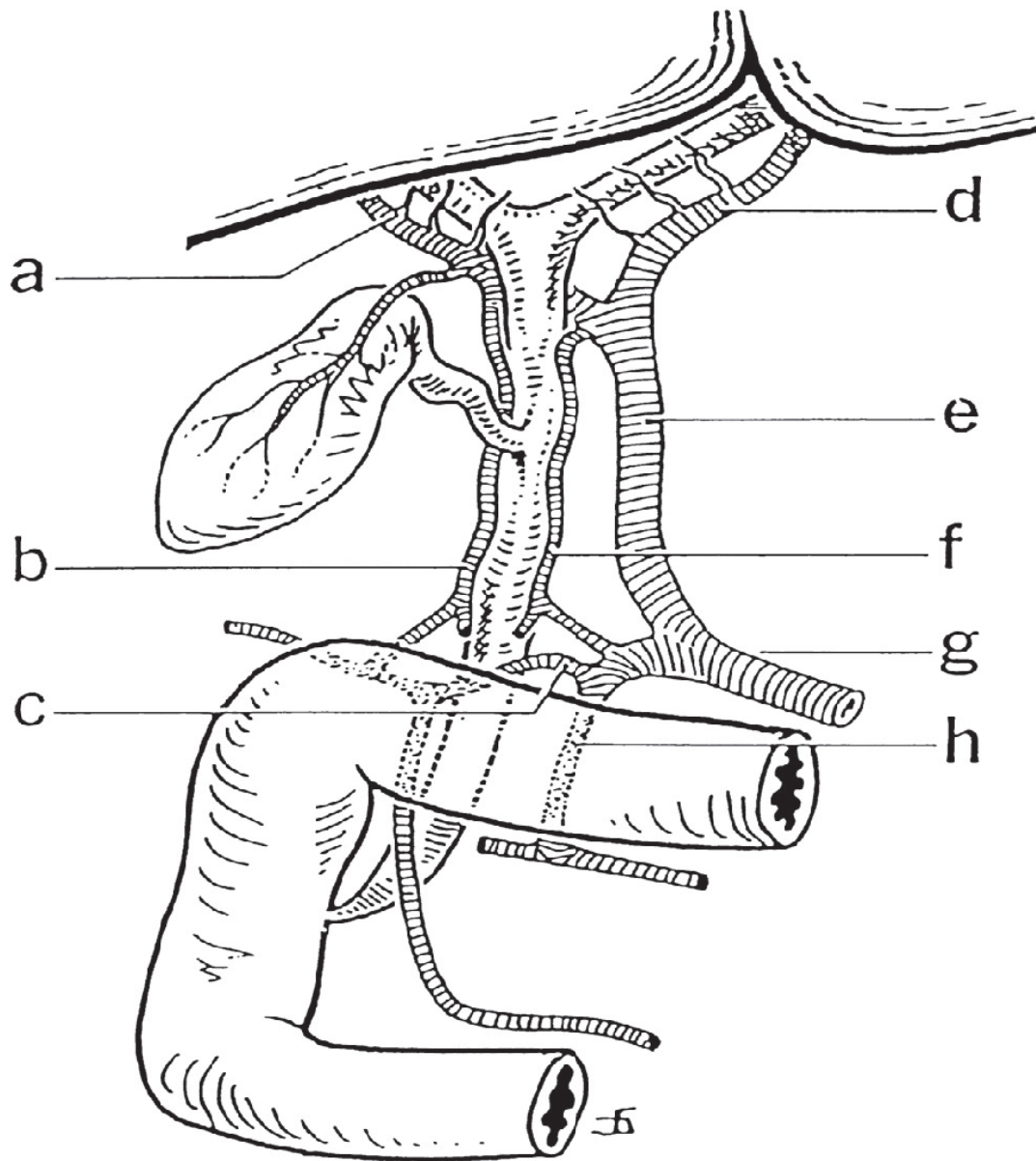


Figure 1.3 Blood supply to the extrahepatic bile ducts: (a) right hepatic artery, (b) 9 o'clock artery, (c) retroduodenal artery, (d) left hepatic artery, (e) hepatic artery, (f) 3 o'clock artery, (g) common hepatic artery, (h) gastroduodenal artery. (Reprinted from Blumgart LH, ed. Surgery of the liver and biliary tract, 3rd ed., p. 21. © 2000, with permission from Elsevier.)

4. Materials and Methods

All patients >12years who were operated for choledochal cyst their age; gender; presenting clinical symptoms; physical, laboratory and histological findings; diagnostic methods; and operative procedures were recorded using electronic data and case record sheets in our KEM Hospital, Mumbai. Detailed analysis was made of the clinical presentation, radiological and anatomical anomalies, management, complications and outcomes.

Investigation of the patients was dictated by their presentation, but generally involved assessment of the biliary tree by endoscopic retrograde cholangiopancreatography (ERCP) (Figure 1.4) and computed tomography (CT) scanning (Figure 1.5), and MRCP (Figure 1.6).

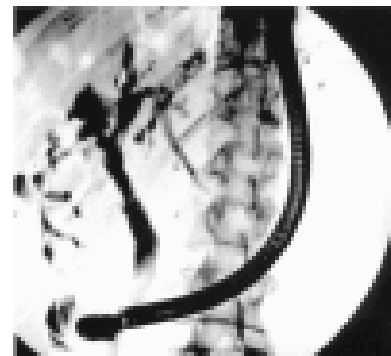


Figure (1.4) ERCP demonstrating extrahepatic and intrahepatic biliary duct dilatation in a type IV choledochal cyst

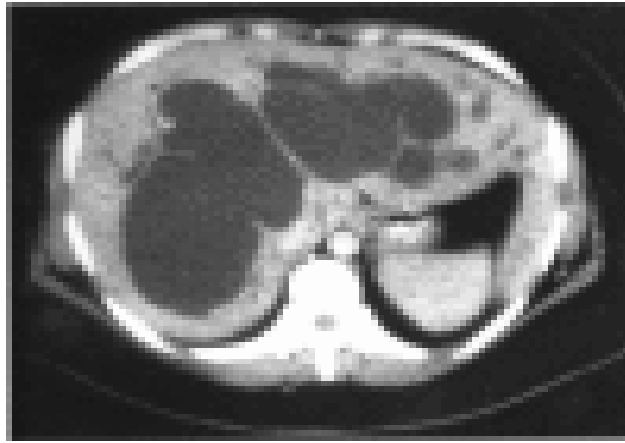


Figure (1.5) CT scan demonstrating intrahepatic biliary duct dilatation in a type IV choledochal cyst

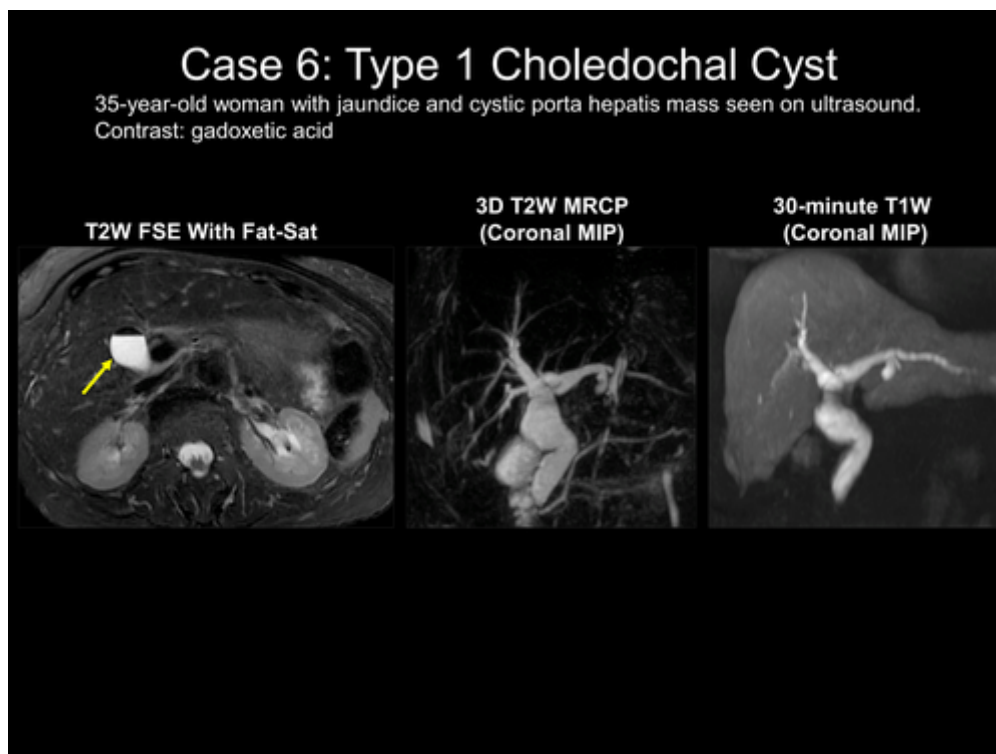


Figure (1.6) MRCP showing type I choledochal cyst

Patients with choledochal cysts in situ underwent total cyst excision with reconstruction of the biliary tract by means of a Roux-en-Y hepaticojejunostomy. Patients with Caroli's disease were excluded from our series.

Modified Todani's classification is used for the determination of the cyst type (Janakie Singham, MD Eric M. Yoshida, MD Charles H. Scudamore, MD, From the Departments of Medicine and Surgery, the University of British Columbia, Vancouver, BC, Accepted for publication Feb. 22, 2008). Ultrasonography (USG), scintigraphy, computerized tomography (CT), percutaneous transhepatic cholangiography (PTC), oral and magnetic resonance (MR) cholangiography were used as diagnostic tools.

- (1) STUDY DESIGN: Observational retrospective study
- (2) SAMPLE SIZE: 30 (Thirty).
- (3) INCLUSION CRITERIA: Patients age >12 years.

- Patients presented with pain in upper abdomen, fever and jaundice
- Patients present with obstructive jaundice
- Patients present with cholangitis or choledocholithiasis or pancreatitis.
- Sometimes patients present with only pain in abdomen and diagnosed as choledochal cyst radiologically.

(4) EXCLUSION CRITERIA:

- Patients of age <12 years.
- Patients presented with pain, fever and jaundice but not radiologically diagnosed.
- Patients presented obstructive jaundice but not diagnosed by radiology or ERCP.
- Patients present with cholangitis, choledocholithiasis or cholangitis or pancreatitis or carcinoma of gall bladder but not diagnosed by radiology or ERCP.
- Type v choledochal cyst.

(5)STUDY PROCEDURE: Data are collected from case records of the patients admitted and operated for choledochal cyst in KEM hospital, Mumbai, India, General Surgery Department over 5years.

5. Results and Data Analysis

We have done an audit of 30 cases of choledochal cyst operated in our tertiary health care hospital. Following observations were made.

Demography

Age Distribution: Table 1.4

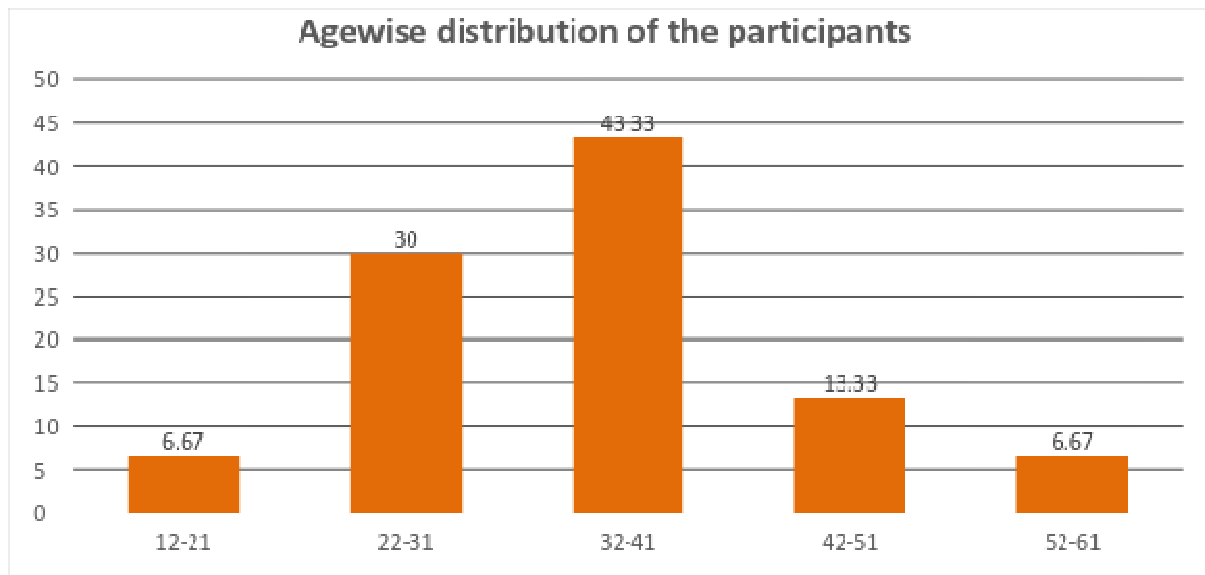
Percentage	frequency	Age group
6.67	2	12-21
30	9	22-31
43.33	13	32-41
13.33	4	42-51
6.67	2	52-61
100	30	Total

Patient's age group range from 12yrs to 61yrs

Peak incidence was between 32 to 41 years amounting for 43.33% of total cases.

Average age of incidence was 35.9.

Figure 1.7:



2) Gender Distribution

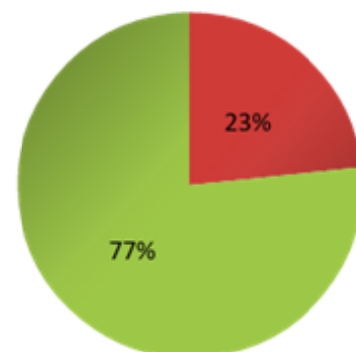
Table 1.5

Female	Male
23	7

Figure 1.8

SEXWISE DISTRIBUTION

SEX MALE FEMALE



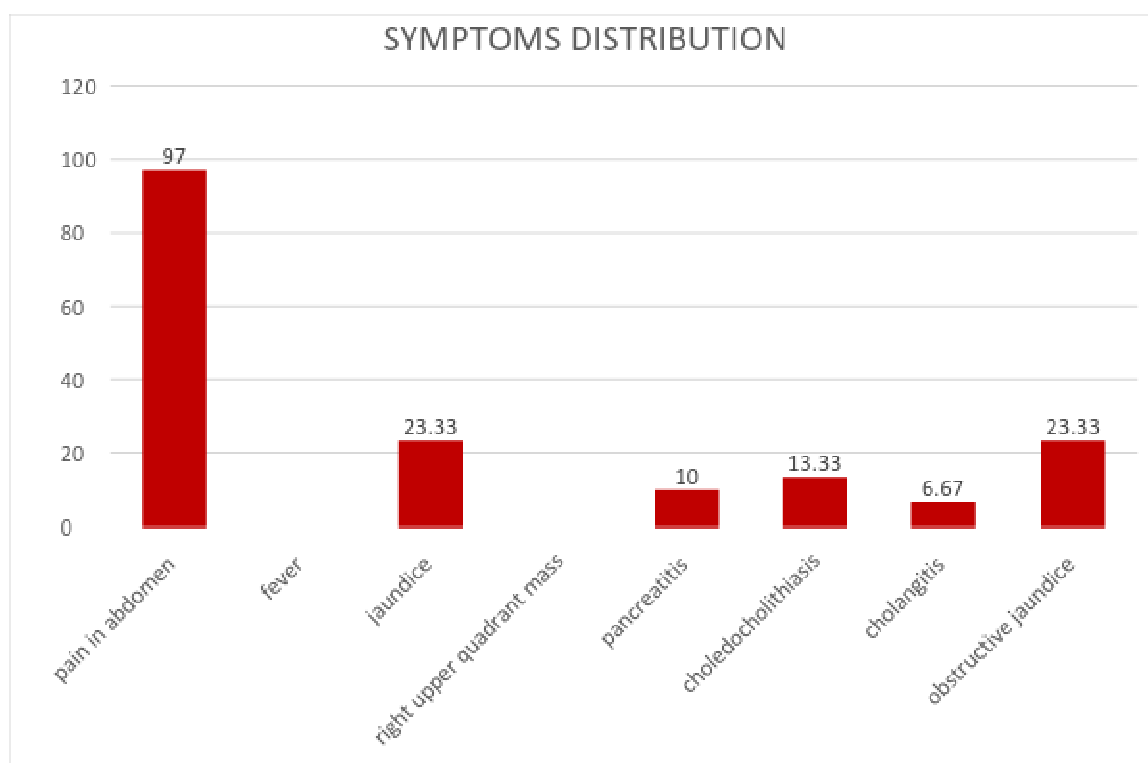
In our study females are 77% & males are 23% affected.

Symptomatology

Table 1.6

Percentage	No of Patients	Clinical Features
97%	29	pain in upper abdomen
0	0	fever
23.3	7	jaundice
0	0	right upper quadrant mass
10	3	pancreatitis
13.3	4	choledocholithiasis
6.6	2	cholangitis
23.3	7	obstructive jaundice

Figure 1.9:



Symptoms of the disease varied from pain to cholangitis and pancreatitis compelling the patients for medical line of investigation.

Most common presenting symptom is pain in abdomen which constitutes 97% of total symptoms. Fever and right upper quadrant mass are least common.

Investigations

Table 1.7

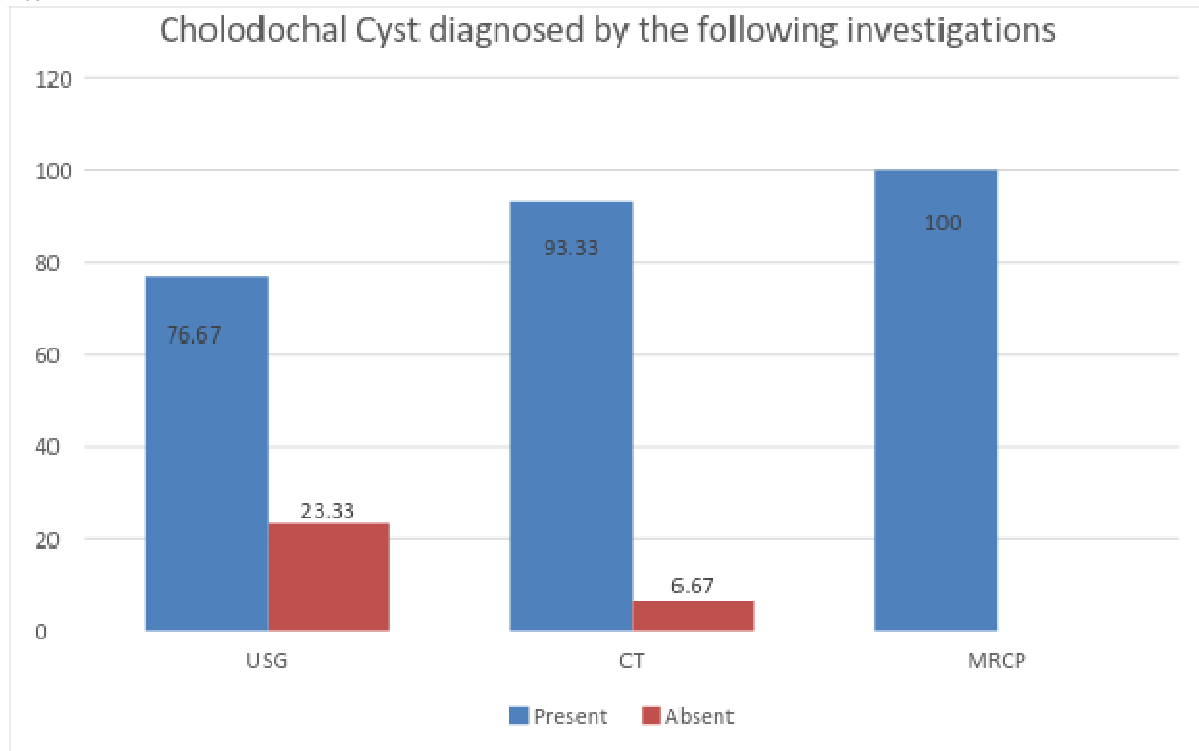
USG diagnosis	choledochal Cyst
23	Present
7	Absent
30	Total
CT SCAN diagnosis	
28	Present
2	Absent
30	Total

MRCP diagnosis	
30	Present
0	Absent
30	Total

30	Total
----	-------

	Choledochal cyst	
Absent (%)	Present (%)	Investigations
23.33	76.67	USG
6.67	93.33	CT
0	100	MRCP

Figure 2.0



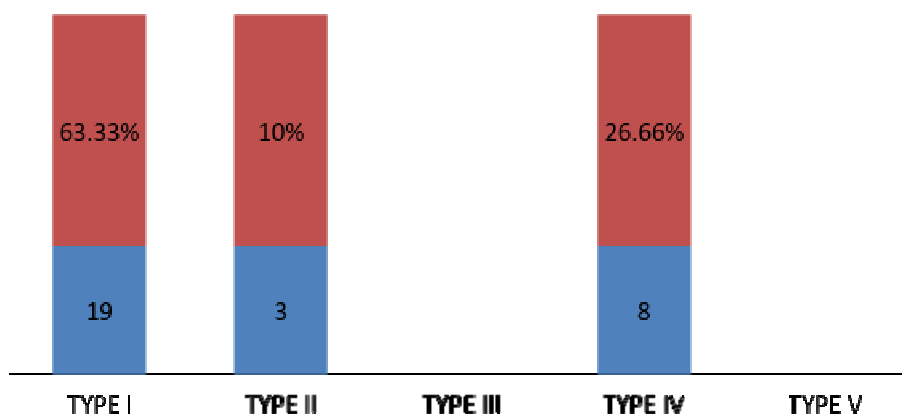
Distribution of Choledochal Cysts

Table 1.8:

Percentage (%)	No of Patients	Type of Cysts
63.33	19	I
10	3	II
0	0	III
26.66	8	IV
0	0	V

Figure 2.1

distribution of choledochal cyst



Out of 30 patients 63.33% patients are type I choledochal cyst followed by type iv and least are type III and type V.

Treatment**Table 1.9**

Mortality	Re Explorati on	Cure d	No of Patient s	Types of Surgery
0	0	22	22	open roux en y HJ
0	0	1	1	Open roux en y HJ+PJ
0	0	5	5	lap assisted roux en y HJ
0	0	1	1	choledochoduodenosto my
1	0	0	1	whipples procedure
1	0	29	30	TOTAL

Out of 30 patients in 22 patients open cc exision with roux en y hepaticojejunostomy done. In 5 patients lap assisted cyst exision done. One mortality due to malignancy.

6. Discussion

Choledochal cysts are a congenital anomaly, and they show dilatation of the intra- or extrahepatic biliary tree. These cysts are uncommon in Western countries, but are not rare in Asian countries. Choledochal cysts are classified into five groups based on location or shape of the cysts. Types I and IV-A cysts are the most common types, which are associated with ano-malous pancreaticobiliary junction (APBJ), but other cysts are not associated with APBJ. Types I and IV-A cysts appear to belong to a different category from other cysts embryologically. Type I and IV-A cysts accompany anomalies of the pancreas. Type I and IV-A cysts might occur when left ventral anlage persists, and with disturbed recanalization of the common bile duct. Endoscopic retrograde cholangiopancreatography is the gold standard for detecting APBJ, but it is an invasive procedure.

Magnetic resonance cholangiopancreatography (MRCP) is a non-invasive imaging tool for detecting pancreatic and biliary trees. MRCP is the first-choice modality for diagnosing choledochal cysts and APBJ in pediatric patients. Cystoenterostomy has been performed because of high complication and mortality rates. Complete excision of the cysts with Roux-en-Y hepatojejunostomy is a standard procedure for choledochal cysts to prevent postoperative complications, including development of cancer.

Choledochal cysts are resected more often in childhood. Presenting symptoms are age dependent with jaundice prevailing in children and abdominal pain in adults. In view of the high risk of cholangiocarcinoma, early resection and not internal drainage is the appropriate treatment of type I and II and the extrahepatic part of type IV biliary cysts. Patients who only had internal drainage in the past still should undergo resection of the cyst remnant.

Nearly 25% of choledochal cysts are detected in the first year of life and 60% in the first decade, but 20% are diagnosed after the age of 20 years were reported.^{88, 124} In our series average age was between 32 to 41 years. Out of

30 patients its 13 which constitute 43.33% of total cases. This is possibly because below 12 years of age group patients goes to paediatric surgery and some patients also go to gastroenterology surgery department in our hospital.

Female: Male Ratio

Out of 30 patients operated in our study 23 were females and 7 were males. Females constitute 77% and males constitute 23% in our study. Female: male ratio being (3.28). This is comparable to series reported by LAMIGAN.P.D (1975), YAMAGUCHI.M (1980), and HOWARDS.E.R (1991)^{2, 125, 126}. In their reported study female: male ratio was 3: 1 to 4: 1.

Clinical Presentation

According to TODANI et al⁶ Classification of choledochal cysts with relative incidences. Type I: Dilatation of hepatic and common bile duct (40% to 85%); II, Type II: Diverticulum of the common bile duct (2% to 3%); Type III: Intraduodenal common bile duct dilatation (1.4% to 5.6%); Type IVa: Intra- and extrahepatic bile duct dilatation (18% to 20%); Type V: Intrahepatic bile duct dilatation (rare). In our study the incidence are 63.33% type I CC, 26.66% are type IV CC, 10% are type II CC which can be comparable to series reported in literature. Type III and type v not detected because we have only taken operated cases of CC. Hepatectomy or liver transplantation in type v cc done in surgical gastroenterology department. Upper abdominal pain is most common presenting symptoms in our study (29 out of 30(97%)), which is significantly higher in adults age group. We found jaundice in 7 patients (23.33%). No patients presented with fever or right upper quadrant mass which is most commonly found in children. Though we found 2 patients with cholangitis(6.6%) but none has fever out of 30 patients. Although not significant, 3 patients (10%) had pancreatitis and 4(13.33%) had choledocholithiasis. Classical triad like pain, fever and jaundice we did not find in any patients.

In this study, all of the choledochal cysts were diagnosed in age group more than 12 years. The type of symptoms depends largely on the age at presentation. Abdominal pain has been reported to be the most frequent symptom at presentation and is the main symptom in adults found in study by LIPSETT PA et al⁸⁸ and by S.R BANERJESUDASON, DHAWAN et al¹³¹ (57 adult choledochal cyst study) which was also found in our series. Jaundice is reportedly the main presenting symptom in infants¹²⁷ but in our study only 23.33% patients presented with jaundice. Among them 3 patients associated with choledocholithiasis which is a known complication of cc. It has been suggested that age-related difference in presentation is determined by whether there is reflux of activated pancreatic juice.³⁵ In literature it is found that patients with choledochal cysts presenting with abdominal pain were older than 1 year and that in these patients there is a relation with elevated serum amylase and signs of chronic inflammation in histologic sections of the resected cyst. Because we have not studied paediatric patients and serum amylase was not assessed in our series

we could not confirm this notion. Further, according to literature pancreatitis more often in children from 2 to 16 years than in the other age groups. However, this is not statistically significant. In our study also it is not statistically significant. The finding of jaundice as the main presenting symptom of extrahepatic cysts and cholangitis and gallstones of intrahepatic cysts is similar to those of earlier reports.^{128, 129, 130} This may be explained by the localization of the lesion. Extra hepatic cysts may give complete obstruction of the biliary tree leading to jaundice, whereas intrahepatic cysts will lead to partial obstruction giving late and localized complications. The classic triad of abdominal pain, jaundice, and abdominal DHAWAN et al¹³⁰ mass has proved to be rare in study by SAMUEL M, SPITZ.⁸⁸ STRINGER M. This could not be confirmed in our study because we have taken the age group more than 12 years but this triad mainly found under 2 years age group still it is rare. It may thus be less classic then is usually thought.

Choledocholithiasis and cholangitis were managed with ERCP stenting, and papillotomy. In our study choledocholithiasis was 13.33% and cholangitis was 6.67%. S.R BANERJEE et al¹³¹ reported 22.8% in his 57 adult cc study. In other associated pathology gall stone commonly found. In our study 9 out of 30 patients (30%). J.S DE VRIES et al reported 4 patients out of 42 patients (9.5%).¹³² S.R BANERJEE et al¹³¹ reported in his study 57.1% incidence of gall stone.

The etiology of CCs is still unclear, although many theories have been put forth. Babbitt's²¹ theory of cysts caused by an abnormal pancreaticobiliary duct junction (APBDJ) such that the pancreatic duct and the common bile duct meet outside the ampulla of Vater, thus forming a long common channel, has gained much popularity. This theory postulates that the long common channel allows mixing of the pancreatic and biliary juices, which then activates pancreatic enzymes. These active enzymes cause inflammation and deterioration of the biliary duct wall, leading to dilation.²¹ Furthermore, greater pressures in the pancreatic duct can further dilate weak-walled cysts.²² Many studies have measured the amylase level in CC bile, which is always higher in patients than in controls.^{23, 24}

Furthermore, higher levels of amylase were significantly associated with younger age of symptom onset and higher grade of dysplasia. This lends credence to the theory that pancreaticobiliary reflux not only exists in these patients, but also leads to inflammation and dysplasia.²⁴⁻²⁸ The authors also postulated that high levels of reflux (and thus amylase) results in earlier symptoms, whereas low levels result in chronic, insidious disease that presents with complications later in life. Although amylase may be a marker for pancreatic reflux, it is more likely that the other enzymes actually cause epithelial breakdown.

Therefore further studies have been conducted to assess trypsinogen and phospholipase A2 levels in CC bile, which were also found to be elevated.^{24, 29-35} Interestingly, 61% of the trypsinogen in the bile duct and 65% of the trypsinogen in the gallbladder was activated to trypsin,

which can only be accomplished by the presence of enterokinase.²⁴ Although normal epithelium does not produce enterokinase outside of the duodenum, it is secreted by dysplastic biliary epithelium, including the epithelium of patients with APBDJ.^{29, 31} Therefore it is theorized that enterokinase from diseased biliary epithelium activates trypsinogen to trypsin, which in addition to its digestive and irritating effects activates phospholipase A2. Activated phospholipase A2 hydrolyzes epithelial lecithin to lysolecithin, resulting in further inflammation and bile wall breakdown.^{34, 35}

It is well accepted that a CC is a premalignant state, with cancer not only occurring more often in these patients but also 10–15 years earlier.⁷⁴ The overall risk of cancer has been reported to be 10%–15%, and increases with age.^{35, 75} The risk rises from 2.3% in patients aged 20–30 years to 75% in patients aged 70–80 years, and histopathology shows increasing dysplasia with increasing age.^{76, 77}

Distribution of the types of cancer found in patients with CC are as follows: adenocarcinoma 73%–84%, anaplastic carcinoma 10%, undifferentiated cancer 5%–7%, squamous cell carcinoma 5% and other carcinoma 1.5%.⁷⁸⁻⁸⁰ The site of cancer is the extra hepatic bile duct in 50%–62% of patients, gallbladder in 38%–46%, intrahepatic bile ducts in 2.5%, and the liver and pancreas in 0.7% each.⁷⁷

A review by Todani and colleagues⁷⁹ found that 68% of cancers were associated with type-I, 5% type-II, 1.6% type-III, 21% type-IV and 6% type-V CCs. Abnormal pancreaticobiliary duct junction has a 16%–55% risk of malignancy with or without bile duct dilation, and cancer has been reported in 12%–39% of form fruste patients.^{77, 81, 82} Cancer usually occurs within the cyst in CC and in the gallbladder in form fruste CC, leading some authors to postulate that malignancy occurs at the site of bile stasis, irritation and inflammation (within the dilated cyst in CC and within the gallbladder when no cyst exists).^{77, 82-84} Caroli disease is associated with a cancer risk of 7%–15%.^{64, 69, 85}

The incidence of malignancy with choledochocoele is usually reported as 2.5%, but 1 study reported a 27% incidence.⁸⁶ Although not typically associated with APBDJ, some authors claim that the choledochocoele itself may be a site of pancreatic and biliary juice mixing, as the pancreatic duct and the CBD may both open into the cyst, thus creating the same inflammatory and precancerous milieu as with an APBDJ.^{86, 87} Cancer occurs as a result of chronic inflammation, cell regeneration and DNA breaks, leading to dysplasia. The inflammation can be from either recurrent cholangitis or pancreaticobiliary reflux.^{75, 76, 88-90} Chronic inflammation also destroys the protective mucin-producing epithelial cells.⁷⁵

Furthermore, chronic post obstructive infection by gram-negative bacteria such as *Escherichia coli* metabolizes bile acids into carcinogens.⁷⁵ K-ras mutations and over expression of p53, which have been demonstrated in many other malignancies, are also present in malignant, precancerous dysplastic and chronically inflamed bile ducts in CC and APBDJ. This suggests that pancreatic

reflux causes K-ras mutation, cellular atypia, p53 overexpression and carcinogenesis.^{25, 91} Although most CC-associated malignancy presents with abdominal pain, weight loss and obstructive jaundice, many can be asymptomatic, and therefore vigilant surveillance is necessary.^{74, 75, 84, 90}

The overall finding of cholangiocarcinoma is comparable with most recent series in this table. In our study one patient out of 30 patients had been detected as cholangiocarcinoma(3.3%).It is seen in lower side in our study and it is possible because patients in our hospital (<12years)are considered in paediatric surgery and some adults to gastroenterology surgery.

No of Patients with Malignancy (%)	No of Patients	Study, Year
8(10)	80	Jan et al, ²⁰ 2000
6(13)	48	Bismuth and Krissat, ⁷⁵ 1999
5(12)	42	Lenriot et al, ²² 1998
2(14)	14	Hewitt et al ¹²⁴ 1995
6(26)	27	Stain et al, ²³ 1995
3(10)	42	Lipsett et al ⁸⁸ 1995
4(9)	46	Chijiwa and Koga, ²⁵ 1993
2(15)	13	Robertson and Raine ²⁶ 1988
8(10)	82	Todani et al, ⁶ 1987
6(14)	42	current study, 2000
50(11)	437	Total

Diagnosis

Ultrasonography provides adequate information about the intra- and extra-hepatic biliary tree and is an extremely useful initial investigation, but it does have limitations, including the fact that its effectiveness is dependent on operator experience, that cysts on US images may be misinterpreted as the gallbladder or other structures, and that US scanning suffers decreased sensitivity in the presence of overlying bowel gas, pancreatitis, cholangitis, or other inflammatory processes. Differentiating a choledochal cyst from a hepatic cyst, hepatic abscess, acute fluid collection, or pancreatic pseudocyst may be difficult. Diagnosis of a CC requires demonstration of continuity of the cyst with the biliary tree so that it can be differentiated from other intrabdominal cysts such as pancreatic pseudocysts, Ecchinococcal cysts or biliary cystadenomas⁷.Although most authors recommend other imaging modalities for this purpose, AKHAN AND COLLEAGUES et al¹⁰⁴ demonstrated continuity with the bile duct in 93% of their patients and recommended other imaging only when the diagnosis cannot be made based on an ultrasound. Sensitivity of ultrasonography in making the diagnosis is 71%–97% reported by HUANG SP et al¹⁰¹. Furthermore, given that ultrasonography is non-invasive and inexpensive, it is the modality of choice for follow-up surveillance. Reconstruction of 2-dimensional ultrasound images to form a 3-dimensional image has been advocated by some authors to view the cyst from different angles,

allow full visualization of curved structures and estimate cystic volume, all of which may be important for preoperative planning¹⁰³.In our study diagnostic sensitivity in US was 76.76%, which is comparable.

Computed tomography (CT) scans are useful in showing continuity of the cyst with the biliary tree, its relation to surrounding structures and the presence of associated malignancy. It is superior to ultrasonography in imaging the intrahepatic bile ducts, distal bile duct and pancreatic head¹⁰⁵.In patients with type-IVA cysts and Caroli disease; it is useful to delineate the intrahepatic dilations and the extent of disease such as diffuse hepatic involvement versus localized segmental involvement. This is important, as localized type-IV A cysts or Caroli disease can be treated with segmental lobectomy. Malignancy can be identified as a mass or a focal region of wall thickening on a CT scan¹⁰⁵. Some authors recommend spiral CT to differentiate malignant cyst wall changes from reactive inflammation¹⁰⁵.Computed tomographic cholangiography (CTCP) has been used to delineate the full anatomy of the biliary tree to correctly plan surgery; this imaging modality is 93% sensitive for visualizing the biliary tree, 90% sensitive for diagnosing CCs and 93% sensitive for diagnosing lithiasis. Unfortunately, it was reported to be only 64% sensitive for imaging the pancreatic duct, as this depends on reflux of the contrast into the ducts¹⁰⁵. Virtual endoscopy based on CT images has been used to evaluate the biliary tree anatomy and identify defects successfully¹³⁹.

Intravenous cholangiography and spiral CT can be combined to form a 3-dimensional image that very accurately delineates the postoperative anastomosis site¹⁴⁰.Of course, the drawbacks to using CT and CTCP is the risk of nephro- and hepatotoxicity with contrast and the exposure to ionizing radiation. In our study 28 patients diagnosed in CT scan with sensitivity 93.33%, which is comparable to series reported in literature.

Although ERCP has been reported to be the most sensitive imaging modality for CCs, this sensitivity does fall in certain situations. Recurrent inflammation and scarring may make cannulation of the ampulla difficult or impossible and may cause partial or complete obstruction at any point of the biliary tree, with no resultant biliary imaging.¹⁰⁴ Full visualization of large cysts requires high dye load, and a compromise needs to be made between complete visualization and the risk of cholangitis or pancreatitis with increased amounts of dye. The use of a high volume of dye can also cause intense opacification, thus obscuring mucosal defects such as ulcers or malignancy, as well as dilate the cyst and overestimate its volume. Cholangiography is also not useful for postoperative imaging, as contrast is drained into the bowel without continuity to the hepatic duct¹³⁹.

ERCP defines the anatomy of the biliary tract accurately and reveals the presence of any associated intraductal pathology or an APBDJ. In the rare instance of a type III cyst, ERCP facilitates a therapeutic papillotomy simultaneously. MRCP, being a non-invasive procedure, is emerging as a favoured alternative to ERCP, but it has a

lower accuracy in the detection of APBDJ and lacks the above therapeutic option in case of a type III cyst. PARK DH, KIM MH, LEE SK, LEE SS, CHOI JS, LEE YS, SEO DW, WON HJ, KIM MY et al¹³⁶. The overall detection rate of MRCP for choledochal cysts was 96% (69/72). The sensitivity, the specificity, the positive predictive value, and the negative predictive value of MRCP for classifying choledochal cysts according to Todani's classification were 81%, 90%, 86%, and 86% in type I, respectively; 73%, 100%, 100, and 95% in type III, respectively; 83%, 90%, 80%, and 91% in type IVa, respectively; 100%, 100%, 100%, and 100% in type IVb, respectively; and 100%, 100%, 100%, and 100% in type V, respectively. The sensitivity, the specificity, and the accuracy of MRCP for detecting ductal anomalies were 83%, 90%, and 86%, respectively. The detection rate of MRCP for concurrent cholangiocarcinoma and choledocholithiasis was 87% (13/15) and 100% (8/8), respectively. In our study 30 /30 patients diagnosed by MRCP (100% sensitivity).so ultrasonography is the initial diagnostic modality. MRCP is mandatory in all cases of cc. So it is comparable to other series in literature.

Treatment

The treatment of choice for choledochal cysts is removal of the cysts by surgery. Internal drainage by cystojejunostomy or cystoduodenostomy has been performed because of low mortality rates and little technical difficulty². However, cystoenterostomy without total excision has not been performed, because of complications after surgery, including recurrent cholangitis, intrahepatic calcification, and carcinoma arising from cysts¹¹¹. A high incidence of complications after internal drainage has been reported. CHIJIIWA AND KOGA et al¹⁴¹ reported complications of cystoenterostomy, including cholangitis in 88% of patients, choledocholithiasis in 25% of patients, and hepatolithiasis in 33% of patients. Seventy percent of these patients required reoperation¹⁴¹. BESNER et al¹⁴² reported that patients with choledochal cysts who were treated with either cystoduodenostomy or cystjejunostomy still had symptoms in 65% of them, and a second operation was required in 40% of patients¹⁴². ATKINSON et al¹⁴³ reported that more than 80% of patients who had cystoenterostomy performed had recurrent pancreatitis and cholangitis related to residual cysts. Other complications, including stone formation, pancreatitis, portal hypertension, and hepatic abscess after cystoenterostomy or external drainage have been reported. Reoperation was necessary because of intrahepatic stone formation in 58.8% of patients who had external drainage or cystoenterostomy performed. In our study 5 patients had undergone laparoscopic cyst excision with roux en y hepaticojejunostomy and one patients due to chonic pancreatitis pancreaticojejunostomy done. one patients cyst excision with choledochodudenostomy done. In maximum number (22) of patients open Roux en y hepaticojejunostomy done. One patients detected malignancy so whipples procedure done. No post-operative complication found. One mortality due to malignancy..

Cholangiojejunostomy is a common surgical procedure for the treatment of biliary tract diseases. Among all surgical cholangiojejunostomy procedures, Roux-en-Y cholangiojejunostomy (RCJS) is considered to have a lower incidence of postoperative cholangitis and recurrent stones.¹ With the development of laparoscopy, laparoscopic RCJS (LRCJS) has become a new choice for the treatment of choledochal cyst. In most report literature, an assisted mini-incision was needed for surgery. Because of the expertise and complicated technique needed in total LRCJS (TLRCJS), reports of large case series pertaining to this surgical procedure are rare.

Indications for LRCJS were treatment of bile duct injury, congenital choledochal cyst resection, obstructive jaundice caused by advanced pancreatic head or periampullary carcinoma, and biliary tract reconstruction after cholangiocarcinoma resection. In treating injury of the bile duct, the optimal management was dependent on the timing of recognition of injury, the extent of bile duct injury, the patient's condition, and the availability of experienced hepatobiliary surgeons. Since 2004, it has been shown that the excision of cystic dilation of the common bile duct (CBD) combined with Roux-en-Y hepaticojejunostomy done laparoscopically has been effective and safe for the treatment of choledochal cyst.¹⁴⁹ However, most of the methods reported were a combination of intracorporeal and extracorporeal procedures. The reports on TLRCJS were rare.¹⁴⁹ In 2011, Urushihara et al. reported total laparoscopic cyst excision in the CBD, but the number of cases was only 8 and they were children.¹⁵⁰ To complete TLRCJS, the surgeon was required to master surgical skills, such as laparoscopic excision of lesions, portal hepatic basin-like bile tract plasty, and cholangiojejunostomy.

In 2010, it has been reported total laparoscopic resection of a type I choledochal cyst in an adult and Roux-en-Y hepaticoenterostomy, with good effects.¹⁵¹

Although robotic-assisted results and outcomes abound for many procedures, only limited information has been published on robotic-assisted choledochal cyst excision. We found only 4 cases in the literatures.¹⁴⁸ Interestingly, the Roux limb could be created entirely intracorporeally by the robot or extra corporeally through a small incision, which could decrease the robotic time and total operative times. In our case, we did an extracorporeal jejuno-jejunoanastomosis, and therefore our operative time was significantly shorter than the others report in literature. The minor leakage of hepatico-jejunoanastomosis found may be caused by unsecured suturing technique from the early experiences in robotic surgery.

Disadvantages include the size of the robotic hardware in relation to patient body; the loss of haptic feedback; and the overall cost of the hardware, drapes, and maintenance of the robotic system.

Finally, the robotic approach to the complex hepatobiliary surgery is feasible and safe in selected patients. Three-dimensional visualization, articulating instruments, and fine-motion filtering are the principle advantages. Robotic

surgery may increase the variety of procedures, which can be accomplished with a minimally invasive approach and may also enable more general surgeons to perform these complex procedures. Surgeons need to become familiar with these improvements as the technology continues to progress¹⁵¹.

7. Conclusion

1. Overall age incidence in cc is in mid age groups (32yr to 41yr) in adults who required surgical cure in our study population.
2. Choledochal cyst predominantly occurs in females, in our study and series reported in literature.
3. Though ultrasonography is the initial mortality of investigation, MRCP is most sensitive and specific evaluating mortality. ERCP is an invasive procedure. So only reserved for the patients presented with cholangitis, choledocholithiasis or malignancy.
2. Classical triad of symptoms may not found in adult age groups. Pain abdomen is most common presenting symptom in adult age group.
3. Patients diagnosed as having CCs as adults, regardless of symptoms, should undergo complete excision of the extra hepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy. The outcome of that approach was excellent. It may be laparoscopic or robotic and open surgical excision of cyst. Laparoscopic and robotic surgeries are in evolving stage. It mainly needs thorough expertise and resources. Its risk benefit ratio need further study to evaluate.

Bigliography

- [1] Vater A. Dissertation in auguralis medica. poes diss. qua. Scirrhis viscerum dissert. c. s. ezlerus. Edinburgh: University Library; 1723; 70: 19
- [2] Howard ER. Choledochal cysts. In: Howard ER, ed. *Surgery of liver disease in children*. Oxford: Butterworth-Heinemann; 1991.p.78-90
- [3] Gigot J, Nagorney D, Farnell M, et al. Bile duct cysts: a changing spectrum of disease. *J Hepatobiliary Pancreat Surg* 1996; 3: 405-11
- [4] O'Neill JA Jr. Choledochal cyst. *Curr Probl Surg* 1992; 29: 361-410
- [5] Alonso-Lej F, Rever WB, Pessango DJ. Congenital choledochal cyst, with a report of 2, and analysis of 94 cases. *Int Abstr Surg* 1959; 108: 1-30
- [6] Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. *Am J Surg* 1977; 134: 263-9
- [7] Levy AD, Rohrman CA. Biliary cystic disease. *Curr Probl Diagn Radiol* 2003; 32: 233-63
- [8] Yamaoka K, Tazawa J, Koizumi K, et al. Choledochocoele with obstructive jaundice: a case report and a review of the Japanese literature. *J Gastroenterol* 1994; 29: 661-4
- [9] Sarris GE, Tsang D. Choledochocoele: case report, literature review and a proposed classification. *Surgery* 1989; 105: 408-14

- [10] Todani T, Watanabe Y, Toki A, et al. Classification of congenital biliary cystic disease: special reference to type Ic and IVA cysts with primary ductal stricture. *J Hepatobiliary Pancreat* 2003; 10: 340-4
- [11] Keane F, Hadzic N, Wilkinson ML, et al. Neonatal presentation of Caroli's disease. *Arch Dis Child Fetal Neonatal Ed* 1996; 77: F145-6
- [12] Levy AD, Rohrman CA, Murukata LA, et al. Caroli's disease: radiologic spectrum with pathologic correlation. *AJR Am J Roentgenol* 2002; 179: 1053-7
- [13] Lilly JR, Stellin GP, Karrer FM. Forme fruste choledochal cyst. *J Pediatr Surg* 1985; 20: 449-51
- [14] Sarin YK, Sengar M, Puri AS. Forme fruste choledochal cyst. *Indian Pediatr* 2005; 42: 1153-5
- [15] Shimotakahara A, Yamataka A, Kobayashi H, et al. Forme fruste choledochal cyst. Long-term follow-up with special reference to surgical technique. *J Pediatr Surg* 2003; 38: 1833-6. Can J Surg, Vol. 52, No. 5, October 2009 **439**
- [16] Thomas S, Sen S, Zachariah N, et al. Choledochal cyst sans cyst — experience with six “forme fruste” cases. *Pediatr Surg Int* 2002; 18: 247-51
- [17] Kaneyama K, Yamataka A, Kobayashi H, et al. Mixed type I and II choledochal cyst: A new clinical subtype? *Pediatr Surg Int* 2005; 21: 911-3
- [18] Loke TKL, Lam SH, Chan CS. Choledochal cyst. An unusual type of cystic dilatation of the cystic duct. *AJR Am J Roentgenol* 1999; 173: 619-20
- [19] Visser BC, Suh I, Wy LW, et al. Congenital choledochal cysts in adults. *Arch Surg* 2004; 139: 855-62
- [20] Wiseman K, Buczkowski AK, Chung SW, et al. Epidemiology, presentation, diagnosis, and outcomes of choledochal cysts in adults in an urban environment. *Am J Surg* 2005; 189: 527-31
- [21] Babbitt DP. Congenital choledochal cyst: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. *Ann Radiol (Paris)* 1969; 12: 231-40
- [22] Han SJ, Hwang EH, Chung KS, et al. Acquired choledochal cyst from anomalous pancreatobiliary duct union. *J Pediatr Surg* 1997; 32: 1735-8
- [23] Sugiyama M, Haradome H, Takahara T, et al. Biliopancreatic reflux via anomalouspancreaticobiliary junction. *Surgery* 2004; 135: 457-9.
- [24] Todani T, Narusue M, Watanabe Y, et al. Management of congenital choledochal cyst with intrahepatic involvement. *Ann Surg* 1978; 187: 272-80.
- [25] Jeong IH, Jung YS, Kim H, et al. Amylase level in extrahepatic bile duct in adult patients with choledochal cyst plus anomalous pancreatobiliary ductal union. *World J Gastroenterol* 2005; 11: 1965-70.
- [26] Zhan JH, Hu XL, Dai GJ, et al. Expressin of p53 and inducible nitric oxide synthase in congenital choledochal cysts. *Hepatobiliary Pancreat Dis Int* 2004; 3: 120-3.
- [27] Iwai N, Yanagihara J, Tokiwa K, et al. Congenital choledochal dilatation with emphasis on pathophysiology of the biliary tract. *Ann Surg* 1992; 215: 27-30.

- [28] Xiao X, Li H, Wang O, et al. Abnormal pancreatic isoamylases in the serum of children with choledochal cysts. *Eur J Pediatr Surg* 2003; 13: 26-30.
- [29] Ochiai K, Kaneko K, Kitagawa M, et al. Activated pancreatic enzyme and pancreatic stone protein (PSP/reg) in bile of patients with pancreaticobiliary maljunction/choledochal cysts. *Dig Dis Sci* 2004; 49: 1953-6.
- [30] Narita H, Hashimoto T, Suzuki T, et al. Clinical and experimental studies on the activation mechanism of pancreatic enzymes refluxing into the biliary tract with an anomalous pancreatobiliary ductal junction. *Nippon Shonika Gakkai Zasshi* 1990; 26: 609-15.
- [31] Shimada K, Yanagisawa J, Nakayama F. Increased lysophosphatidylcholine and pancreatic enzyme content in bile of patients with anomalous pancreatobiliary ductal junction. *Hepatology* 1991; 13: 438-44.
- [32] Yagi T, Miyoshi Y, Komi N. An activation mechanism of pancreatic proenzymes refluxing into the biliary tract in APBD patients. *Shikoku Igakkai Zasshi* 1993; 49: 106-13.
- [33] Yamashiro Y, Sato M, Shimizu T, et al. How great is the incidence of truly congenital common bile duct dilatation? *J Pediatr Surg* 1993; 28: 622-5.
- [34] Mizuno M, Kato T, Koyama K. An analysis of mutagens in the contents of the biliary tract in pancreaticobiliary maljunction. *Surg Today* 1996; 26: 597-602.
- [35] Okada A, Hasegawa T, Oguchi Y, et al. Recent advances in pathophysiology and surgical treatment of congenital dilatation of the bile duct. *J Hepatobiliary Pancreat Surg* 2002; 9: 342-51.
- [36] Tajiri T, Tate G, Inagaki T, et al. Mucinous cystadenoma of the pancreas 17 years after excision of gallbladder because of a choledochal cyst. *J Gastroenterol* 2004; 39: 181-7.
- [37] Oguchi Y, Okada A, Nakamura T, et al. Histopathologic studies of congenital dilatation of the bile duct as related to an anomalous junction of the pancreatobiliary ductal system: clinical and experimental studies. *Surgery* 1988; 103: 168-73.
- [38] Matos C, Nicaise N, Deviere J, et al. Choledochal cysts: comparison of findings at MR cholangiopancreatography and endoscopic retrograde cholangiopancreatography in eight patients. *Radiology* 1998; 209: 443-8.
- [39] Imazu M, Iwai N, Tokiwa K, et al. Factors of biliary carcinogenesis in choledochal cysts. *Eur J Pediatr Surg* 2001; 11: 24-7.
- [40] Hosoki T, Hasuiki Y, Michita T, et al. Visualization of pancreaticobiliary reflux in anomalous pancreatobiliary junction by secretin stimulated dynamic magnetic resonance cholangiopancreatography. *Acta Radiol* 2004; 45: 375-82.
- [41] Davenport M, Stringer MD, Howard ER. Biliary amylase and congenital choledochal dilatation. *J Pediatr Surg* 1995; 130: 474-7.
- [42] Cheng SP, Yang TL, Jeng KS, et al. Choledochal cyst in adults: aetiological considerations to intrahepatic involvement. *ANZ J Surg* 2004; 74: 964-7.
- [43] Yotsuyanagi S. Contribution to aetiology and pathology of idiopathic cystic dilatation of the common bile duct with report of three cases. *Gann* 1936; 30: 601-752.
- [44] Davenport M, Basu R. Under pressure: choledochal malformation manometry. *J Pediatr Surg* 2005; 40: 331-5.
- [45] Ohkawa H, Sawaguchi S, Yamazaki Y, et al. Experimental analysis of the ill effect of anomalous pancreatobiliary ductal union. *J Pediatr Surg* 1982; 17: 7-13.
- [46] Tao KS, Lu YG, Wang T, et al. Procedure for congenital choledochal cysts and curative effect analysis in adults. *Hepatobil Pancr Dis Int* 2002; 1: 442-5.
- [47] Elton E, Hanson BL, Biber BP, et al. Dilated common channel syndrome: endoscopic diagnosis, treatment, and relationship to choledochocoele formation. *Gastrointest Endosc* 1998; 47: 471-8.
- [48] Craig AG, Chen LD, Saccone GT, et al. Sphincter of Oddi dysfunction associated with choledochal cyst. *J Gastroenterol Hepatol* 2001; 16: 230-4.
- [49] Behrns KE, Shaheen NJ, Grimm IS. Type I choledochal cyst in association with familial adenomatous polyposis. *Am J Gastroenterol* 1998; 93: 1337-8.
- [50] Kinjo T, Aoki H, Sunagawa H, et al. Congenital absence of the portal vein associated with focal nodular hyperplasia of the liver and congenital choledochal cyst: a case report. *J Pediatr Surg* 2001; 36: 622-5.
- [51] Suzuki K, Uchida T, Nakayama H. Heterotopic pancreatic tissue associated with intra- and extrahepatic choledochal cysts. *Pathol Int* 1999; 49: 759-62.
- [52] Shih HS, Ko SF, Chaung JH. Is there an association between duodenal atresia and choledochal cyst? *J Pediatr Gastroenterol Nutr* 2005; 40: 378-81.
- [53] Arbell D, Orkin B, Naveh Y, et al. Duodenojejunal atresia with absent dorsal mesentery, choledochal cyst, and malrotation in a premature newborn — a case report. *J Pediatr Surg* 2006; 41: E11-3.
- [54] Kapoor S, Mukherjee SB, Paul R, et al. OMENS-plus syndrome. *Indian J Pediatr* 2005; 72: 707-8.
- [55] Rayamajhi A, Singh R, Prasad R, et al. An unusual case of Type IV A choledochal cyst with subaortic ventricular septal defect. *Pediatr Int* 2006; 48: 187-9.
- [56] Li L, Yamataka A, Segawa O, et al. Coexistence of pancreas divisum and septate common channel in a child with choledochal cyst. *J Pediatr Gastroenterol Nutr* 2001; 32: 602-4.
- [57] Dalvi AN, Pramesh CS, Prasanna GS, et al. Incomplete pancreatic divisum with anomalous choledochopancreatic duct junction with choledochal cyst. *Arch Surg* 1999; 134: 1150-2.
- [58] Komuro H, Takahashi MI, Matoba K, et al. Rare association of severe hypoplasia of the abdominal aorta with imperforate anus, colonic atresia, and choledochal cyst. *Pediatr Surg Int* 2006; 22: 289-92.
- [59] Oyachi N, Ohhama N, Take H, et al. Aplasia of the dorsal pancreas and choledochal cyst. *Pediatr Surg Int* 2006; 22: 557-9.

- [60] Al-Wafi A, Morris-Stiff G, Lari A. Colonic atresia secondary to a choledochal cyst. *Pediatr Surg Int* 1998; 13: 422-3.
- [61] Mizuno K, Itoh K, Monoe T, et al. Pancreaticobiliary arteriovenous malformation with common bile duct dilation in a patient with hemobilia. *J Clin Gastroenterol* 2001; 33: 61-3.
- [62] Turkvatan A, Erden A, Celik M, et al. Ectopic hypoplastic and multiseptate gallbladder with coexisting choledochal cyst: evaluation with sonography and magnetic resonance cholangiopancreatography. *J Clin Ultrasound* 2006; 34: 88-91.
- [63] Ramnarine IR, Mulpur AK, McMahon MJ, et al. Pleuro-biliary fistula from a ruptured choledochal cyst. *Eur J Cardiothorac Surg* 2001; 19: 216-8.
- [64] Tanaka T. Pathogenesis of choledochal cyst. *Am J Gastroenterol* 1995; 90: 685.
- [65] Jindal RM, Harris N, McDaniel HM, et al. Presentation of choledochal cysts without intrabiliary communication on endoscopic retrograde cholangiopancreatography. *Liver Transpl Surg* 1996; 2: 468-71.
- [66] Wheeler W. An unusual case of obstruction of the common bile duct (choledochoceles?). *Br J Surg* 1940; 27: 446-8.
- [67] Kagiya S, Okazaki K, Yamamoto Y. Anatomic variants of choledochoceles and manometric measurements of pressure in the cecum and the orifice zone. *Am J Gastroenterol* 1987; 82: 641-9.
- [68] Venu RP, Geenen JE, Hogan WJ, et al. Role of endoscopic retrograde cholangiopancreatography in the diagnosis and treatment of choledochoceles. *Gastroenterology* 1984; 87: 1144-9.
- [69] Fulcher AS, Turner MA, Sanyal AJ. Case 38: Caroli disease and renal tubular ectasia. *Radiology* 2001; 220: 720-2.
- [70] Mousson C, Rabec M, Cercueil JP, et al. Caroli's disease and autosomal dominant polycystic kidney disease: a rare association? *Nephrol Dial Transplant* 1997; 12: 1481-3.
- [71] Ninan VT, Nampoori MRN, Johnny KV, et al. Caroli's disease of the liver in a renal transplant patient. *Nephrol Dial Transplant* 2002; 17: 1113-5.
- [72] Sgro M, Rosetti S, Barozzino T, et al. Caroli's disease: prenatal diagnosis, postnatal outcome and genetic analysis. *Ultrasound Obstet Gynecol* 2004; 23: 73-6.
- [73] Aguilera V, Rayón M, Pérez-Aguilar F, et al. Caroli's syndrome and imaging: report of a case. *Rev Esp Enferm Dig* 2004; 96: 74-6.
- [74] Tsuchiya R, Harada N, Ito T, et al. Malignant tumors in choledochal cysts. *Ann Surg* 1977; 186: 22-8.
- [75] Bismuth H, Krissat J. Choledochal cystic malignancies. *Ann Oncol* 1999; 10 Suppl 4: 94-8.
- [76] Benjamin IS. Biliary cystic disease: the risk of cancer. *J Hepatobiliary Pancreat Surg* 2003; 10: 335-9.
- [77] Todani T, Watanabe Y, Fujii M, et al. Carcinoma arising from the bile duct in choledochal cyst and anomalous arrangement of the pancreatobiliary ductal union. *Biliary Tract Pancreas* 1985; 6: 525-35.
- [78] Todani T, Watanabe Y, Tokai A, et al. Carcinoma related to choledochal cysts with internal drainage operations. *Surg Gynecol Obstet* 1987; 164: 61-4.
- [79] Todani T, Tabuchi K, Watanabe Y, et al. Carcinoma arising in the wall of congenital bile duct cysts. *Cancer* 1979; 44: 1134-41.
- [80] Fieber SS, Nance FC. Choledochal cyst and neoplasm: a comprehensive review of 106 cases and presentation of two original cases. *Am Surg* 1997; 63: 982-7.
- [81] Okamura K, Hayakawa H, Kuze M. Triple carcinomas of the biliary tract associated with congenital choledochal dilatation and pancreaticobiliary maljunction. *J Gastroenterol* 2000; 35: 465-71.
- [82] Miyano G, Yamataka A, Shimotakahara A. Cholecystectomy alone is inadequate for treatment of form fruste choledochal cyst: evidence from a rare but important case. *Pediatr Surg Int* 2005; 21: 61-3.
- [83] Sugiyama M, Haradome H, Takahara T, et al. Anomalous pancreaticobiliary junction shown on multidetector CT. *AJR Am J Roentgenol* 2003; 180: 173-5.
- [84] Franko J, Nussbaum ML, Morris JB. Choledochal cyst cholangiocarcinoma arising from adenoma: case report and a review of the literature. *Curr Surg* 2006; 63: 281-4.
- [85] Totkas S, Hohenberger P. Cholangiocellular carcinoma associated with segmental Caroli's disease. *Eur J Surg Oncol* 2000; 26: 520-1.
- [86] Ohtsuka T, Inoue K, Ohuchida J. Carcinoma arising in choledochoceles. *Endoscopy* 2001; 33: 614-9.
- [87] Horaguchi J, Fujita N, Kobayashi G. Clinical study of choledochoceles: Is it a risk factor for biliary malignancies? *J Gastroenterol* 2005; 40: 396-401.
- [88] Lipsett PA, Pitt HA. Surgical treatment of choledochal cysts. *J Hepatobiliary Pancreat Surg* 2003; 10: 352-9.
- [89] Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. *J Hepatobiliary Pancreat Surg* 1999; 6: 207-12.
- [90] Yoshida H, Itai Y, Minami M. Biliary malignancies occurring in choledochal cysts. *Radiology* 1989; 173: 389-92.
- [91] Iwasaki Y, Shimoda M, Furihata T, et al. Biliary papillomatosis arising in a congenital choledochal cyst: report of a case. *Surg Today* 2002; 32: 1019-102.
- [92] O'Neill JA Jr, Templeton JM, Schnauffer L et al. Recent experience with choledochal cyst. *Ann Surg* 1987; 205: 553-40.
- [93] Areja Ibars E, Artiges Sánchez de Rojas E, Villalba Ferrer F, et al. Pseudopancreatitis and choledochal cyst. *Rev Esp Enferm Digest* 2004; 96: 277-8. Available: http://scielo.isciii.es/scielo.php?pid=S1130-01082004000400007&script=sci_arttext (accessed 2009 Aug. 27).
- [94] Nakano K, Mizuta A, Oohashi S, et al. Protein stone formation in an intrapancreatic remnant cyst after resection of a choledochal cyst. *Pancreas* 2003; 26: 405-7.
- [95] Nakano K, Mizuta A, Oohashi S, et al. Protein stone formation in an intrapancreatic remnant cyst after

- resection of a choledochal cyst. *Pancreas* 2003; 26: 405-7
- [96] Gupta AK, Gupta A, Bhardwaj VK, et al. Caroli's disease. *Indian J Pediatr* 2006; 73: 233-5.
- [97] Martin LW, Rowe GA. Portal hypertension secondary to choledochal cyst. *Ann Surg* 1979; 190: 638-9.
- [98] Moss RL, Musemeche CA. Successful management of ruptured choledochal cyst by primary cyst excision and biliary reconstruction. *J Pediatr Surg* 1997; 32: 1490-1.
- [99] Ramos A, Castell J, Pinto I. Intestinal intussusception as a presenting feature of choledochoceles. *Gastrointest Radiol* 1990; 15: 211.
- [100] Levy AD, Rohman CA. Biliary cystic disease. *Curr Probl Diagn Radiol* 2003; 32: 233-653. Akhan O, Demirkazik FB, Ozmen MN, et al. Choledochal cysts: ultrasonographic findings and with other imaging modalities. *Abdom Imaging* 1994; 19: 243-7.
- [101] Huang SP, Wang HP, Chen JH, et al. EUS and peroral cholangioscopy in choledochoceles with choledocholithiasis. *Gastrointest Endosc* 1999; 50: 568-71.
- [102] Spottswood SE, Jolles PR, Haynes JH, et al. Choledochal cyst with biliary atresia scintigraphy and correlative imaging. *Clin Nucl Med* 2001; 26: 555-6.
- [103] Sood A, Senthilnathan MS, Deswal S. Spontaneous rupture of a choledochal cyst and the role of hepatobiliary scintigraphy. *Clin Nucl Med* 2004; 29: 392-3.
- [104] Arshanskiy Y, Vyas PK, Type IV. Choledochal cyst presenting with obstructive jaundice: role of MR cholangiopancreatography in preoperative evaluation. *AJR Am J Roentgenol* 1998; 171: 457-8.
- [105] Lam WM, Lam TP, Saing H. MR. Cholangiography and CT cholangiography of pediatric patients with choledochal cysts. *AJR Am J Roentgenol* 1999; 173: 401-5.
- [106] Han JK, Choi BI. Carcinoma in a choledochal cyst. *Abdom Imaging* 1996; 21: 179-81.
- [107] Kim MJ, Han SJ, Yoon CS, et al. Cholangiopancreatography to reveal anomalous pancreaticobiliary ductal union in infants and children with choledochal cysts. *AJR Am J Roentgenol* 2002; 179: 209-145
- [108] Tao KS, Lu YG, Wang T, et al. Procedure for congenital choledochal cysts and curative effect analysis in adults. *Hepatobiliary Pancreat Dis Int* 2002; 1: 442-5.
- [109] Saing H, Han H, Chen KL. Early and late results of excision of choledochal cyst. *J Pediatr Surg* 1997; 32: 1563-6.
- [110] Todani T, Watanabe Y, Toki A, et al. Reoperation for congenital choledochal cyst. *Ann Surg* 1988; 207: 142-7.
- [111] Chaudhary A, Dhar P, Sachdev A. Reoperative surgery for choledochal cysts. *Br J Surg* 1997; 84: 781-4.
- [112] Koshinaga T, Hoshino M, Inoue M, et al. Pancreatitis complicated with dilated choledochal remnant after congenital choledochal cyst excision. *Pediatr Surg Int* 2005; 21: 936-8
- [113] Ure BM, Schier F, Schmidt AI. Laparoscopic resection of congenital choledochal cyst, choledochojejunostomy, and extraabdominal Roux-en-Y anastomosis. *Surg Endosc* 2005; 19: 1055-7
- [114] Chowbey PK, Katrak MP, Sharma A, et al. Complete laparoscopic management of choledochal cyst: report of two cases. *J Laparoendosc Adv Surg Tech A* 2002; 12: 217-21
- [115] Shah AA, Shah AV. Appendix as a biliary conduit for choledochal cyst in children. *Eur J Pediatr Surg* 2005; 15: 128-31. 46. Chang CC. Antireflux spur valve in cholerostomy. *Jpn Soc J Pediatr Surg* 1982; 18: 73-5
- [116] Raffensperger JG. *Swenson's pediatric surgery*. 5th ed. Norwalk (CT): Appleton & Lange; 1995.
- [117] Zhang JZ, Wang YX, Wei LQ, et al. The spur valve jejunal interposition in choledochus cystectomy. *Chin Med J (Engl)* 1987; 100: 535-40
- [118] Woon CYL, Tan YM, Oei CL. Adults choledochal cysts: an audit of surgical management. *ANZ J Surg* 2006; 76: 981-6.
- [119] Lee CC, Levine DA, Tunik MG, et al. A case report: type I choledochal cyst induced pancreatitis in a 15-month-old child. *Pediatr Emerg Care* 2000; 16: 265-7
- [120] Gulati MS, Srivastava DN, Paul SB, et al. Pre-operative management of congenital choledochal cyst with ultrasound-guided percutaneous choledochalcystostomy. *Australas Radiol* 1999; 43: 514-6
- [121] Kiresi DA, Karabacakoglu A, Dilsiz A, et al. Spontaneous rupture of choledochal cyst presenting in childhood. *Turk J Pediatr* 2005; 47: 283-6.
- [122] Horaguchi J, Fujita N, Kobayashi G. Clinical study of choledochoceles: Is it a risk factor for biliary malignancies? *J Gastroenterol* 2005; 40: 396-401.
- [123] Masetti R, Antinori A, Coppola R. Choledochoceles: changing trends
- [124] Hewitt PM, Krige JE, Bornman PC, Terblanche J. Choledochal cysts in adults. *Br J Surg*. 1995; 82: 382-5. [PubMed] diagnosis and management. *Surg Today* 1996; 26: 281-5.
- [125] Flanagan, P.D. (1975) Biliary cysts. *Annals of Surgery*, **182**, 635-643. doi: 10.1097/0000658-197511000-00017
- [126] Yamaguchi, M. (1980) Congenital choledochal cyst. Analysis of 1433 patients in the Japanese literature.
- [127] Samuel M, Spitz L: Choledochal cyst: Varied clinical presentation and long-term results of surgery. *Eur J Pediatr Surg* 6: 78-81, 1995
- [128] Lu S: Biliary cysts and strictures, in N Kaplowitz (eds): *Liver and Biliary Diseases*, Baltimore, MD, Williams and Wilkins, 1996,
- [129] Taylor A, Palmer K: Caroli's disease. *Eur J Gastroenterol Hepatol* 10: 105-108, 1998
- [130] Stringer M, Dhawan A, Davenport M, et al: Choledochal cysts: lessons from 20 year experience. *Arch Dis Child* 73: 528
- [131] Management of adult choledochal cysts – a 15-year experience Professor S. R. Banerjee Jesudason, Mark Ranjan Jesudason, Rajiv Paul Mukha, Frederick L. Vyas, Sanjay Govil, and John C. Muthusami
- [132] Choledochal Cysts: Age of Presentation, Symptoms, and Late Complications Related to

- Todani's Classification By J.S. de Vries, S. de Vries, D.C. Aronson, D.K. Bosman, E.A.J. Rauws, A. Bosma, H.A. Heij, D.J. Gouma, and T.M. van Gulik Amsterdam, The Netherlands
- [133] Healey Jr JE, Schroy PC. Anatomy of the biliary ducts within the human liver. Analysis of the prevailing patterns of branching and the major variation of the biliary ducts. *Arch Surg* 1953; 66: 599.
- [134] Bismuth H. Surgical anatomy and anatomical surgery of the liver. *World J Surg* 1982; 6: 3–9.
- [135] Lindner HH, Penz VA, Ruggeri RA, et al. A clinical and anatomical study of anomalous termination of the common bile duct into the duodenum. *Ann Surg* 1976; 198: 626
- [136] Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts? Park DH, Kim MH, Lee SK, Lee SS, Choi JS, Lee YS, Seo DW, Won HJ, Kim MY
- [137] Haliloglu M, Akata D, Gurel S, et al. Choledochal cysts in children: evaluation with three-dimensional sonography. *J Clin Ultrasound*. 2003; 31: 478–80. [PubMed]
- [138] Han JK, Choi BI. Carcinoma in a choledochal cyst. *Abdom Imaging*. 1996; 21: 179–81. [PubMed]
- [139] Hamada Y, Tanano A, Takada K, et al. Magnetic resonance cholangiopancreatography on postoperative work-up in children with choledochal cysts. *Pediatr Surg Int*. 2004; 20: 43–6. [PubMed]
- [140] Wiedmeyer DA, Stewart E, Dodds WJ, et al. Choledochal cyst: findings on cholangiopancreatography with emphasis on ectasia of the common channel. *AJR Am J Roentgenol*. 1989; 153: 969–72. [PubMed]
- [141] Chijiwa, K. and Koga, A. (1993) Surgical management and long-term follow-up of patients with choledochal cysts. *American Journal of Surgery*, 165, 238-242. doi: 10.1016/S0002-9610(05)80518-5
- [142] Besner, G.E., Paddock, H., Nguyen, L., et al. (2008) Surgical treatment. *Pediatric Choledochal Cyst Surgery Treatment & Management*. <http://emedicine.medscape.com/article/934267-treatment>
- [143] Atkinson, H.D., Fischer, C.P., de Jong, C.H., et al. (2003) Choledochal cysts in adults and their complications. *HPB (Oxford)*, 5, 105-110. doi: 10.1080/13651820310001144
- [144] Choledochal cysts Part 1 of 3: Classification and pathogenesis Janakie Singham, MD, Eric M. Yoshida, MD Charles H. Scudamore, MD From the Departments of Medicine and Surgery, the University of British Columbia, Vancouver, BC Accepted for publication Feb. 22, 2008, Correspondence to: Dr. C.H. Scudamore, Department of Surgery, University of British Columbia, Gordon and Leslie Diamond Health Care Centre, 2775 Laurel St., floor 5, Vancouver BC V5Z 1M9
- [145] Choledochal Cyst – Presentation and Treatment in an Adult Ferat Sallahu, Antigona Hasani, [...], and Hysni Jashari
- [146] JSLS : Journal of the Society of Laparoendoscopic Surgeons Society of Laparoendoscopic Surgeons Total Laparoscopic Roux-en-Y Cholangiojejunostomy for the Treatment of Biliary Disease Dexing Chen, Prof Dr Med, Andong Zhu, Prof Dr Med, and Zhibo Zhang
- [147] JSLS : Journal of the Society of Laparoendoscopic Surgeons Society of Laparoendoscopic Surgeons Total Laparoscopic Roux-en-Y Cholangiojejunostomy for the Treatment of Biliary Disease Dexing Chen, Prof Dr Med, Andong Zhu, Prof Dr Med, and Zhibo Zhang
- [148] World Journal of Surgical Oncology, Robot-assisted complete excision of choledochal cyst type I, hepaticojejunostomy and extracorporeal Roux-en-Y anastomosis: a case report and review literature Thawatchai Akaraviputh, Aththaphorn Trakarnsanga, and Nutnicha Suksamanapun
- [149] Jia J, Li L, Liu G, Huang LM. Total cyst excision with ultrasonic scalpel under the laparoscope for choledochal cyst. *Zhonghua Wai Ke Za Zhi*. 2004; 42(17): 1056–1059.[PubMed]
- [150] Urushihara N, Fukuzawa H, Fukumoto K, et al. Totally laparoscopic management of choledochal cyst: Roux-en-Y Jejunostomy and wide hepaticojejunostomy with hilar ductoplasty. *J Laparoendosc Adv Surg Tech A*. 2011; 21(4): 361–366. [PubMed]
- [151] Tian Y, Wu SD, Zhu AD, Chen DX. Management of Type I choledochal cyst in adult: totally laparoscopic resection and Roux-en-Y hepaticoenterostomy. *J Gastrointest Surg*. 2010; 14(9): 1381–1388. [PubMed]
- [152] Meehan JJ, Elliott S, Sandler A. The robotic approach to complex hepatobiliary anomalies in children: preliminary report. *J Pediatr Surg*. 2007; 42: 2110–2114. doi: 10.1016/j.jpedsurg.2007.08.040. [PubMed] [Cross Ref]
- [153] 1 Bremer JL: Description of a 4 mm human embryo. *Am J Anat* 1906; 5: 459–480.
- [154] Kanagasuntheram R: Some observations on the development of the human duodenum. *J Anat* 1960; 94: 231–240.
- [155] Irving IM, Rickham PP: Duodenal atresia and stenosis: annular pancreas; in Rickham PP, Lister J, Irving IM (eds): *Neonatal Surgery*, ed 2. London, Butterworths, 1978, pp 355–370.
- [156] Schwegler RA, Boyden EA: The development of the pars intestinalis of the common bile duct in the human fetus, with special reference to the origin of the ampulla of Vater and the sphincter of Oddi. I. The involution of the ampulla. *Anat Rec* 1936–1937; 67: 441

Annexure

1. APBDJ: Abnormal pancreatobiliary duct junction
2. CC: choledochal cyst
3. ERCP: endoscopic retrograde cholangiopancreatography
4. MRCP: magnetic resonance cholangiopancreatography
5. US: ultrasonography
6. TLRCJS: total laparoscopic Roux-en-Y cholangiojejunostomy