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EDITORIAL

Biliary cysts: Etiology, diagnosis and management

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Abstract

Biliary cysts (BC) are rare dilatations of different parts of a biliary tract. They account for approximately 1% of all benign biliary diseases. BC occur the most frequently in Asian and female populations. They are an important problem for pediatricians, gastroenterologists, radiologists and surgeons. Clinical presentation and management depend on the BC type. Cholangiocarcinoma is the most serious and dangerous BC complication. The other complications associated with BC involve cholelithiasis and hepatolithiasis, cholangitis, acute and chronic pancreatitis, portal hypertension, liver fibrosis and secondary liver cirrhosis and spontaneous cyst perforation. Different BC classifications have been described in the literature. Todani classification dividing BC into five types is the most useful in clinical practice. The early diagnosis and proper treatment are very important, because BC are associated with a risk of carcinogenesis. A malignancy risk increases with the age. Radiological investigations (ultrasonography, computed tomography, endoscopic retrograde cholangiopancreatography and magnetic resonance cholangiopancreatography) play an important role in BC diagnostics. Currently, prenatal diagnosis using ultrasonography is possible. It allows to differentiate biliary disorders in fetals and to perform the early surgical treatment that improves results. In most patients, total cyst excision with Roux-Y hepaticojejunostomy is the treatment of choice. Surgical treatment of BC is associated with high success rate and low morbidity and mortality. The early treatment is associated with a lower number of complications. Patients following BC surgery require permanent and careful postoperative observation using laboratory and imaging investigations because of possibility of biliary anastomosis stricture and biliary cancer in tissue remnant.

Key words: Biliary cysts; Biliary drainage; Hepaticojejunostomy

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INTRODUCTION

Biliary cysts (BC) are rare dilatations of different parts of a biliary tract. They account for approximately 1% of all benign biliary diseases. BC occur the most commonly in Asian populations and in females (with a female:male ratio of 3-4:1). Incidence of this pathology in Asian populations is 1:1000 and it is lower in western countries (1:100 000 to 1:150 000)[1-6]. They are usually a surgical problem of infancy or childhood; however in approximately 20% of cases they are recognized in adults[2]. The early diagnosis and proper treatment are very important, because BC are associated with a risk of carcinogenesis. According to the literature, biliary tract malignancy occurs in 2.5%-28% of patients with BC[3,4]. The risk of biliary cancer increases with the age. It is the lowest in the childhood in the first decade (< 1%)[6]. In the third decade the cancer

risk is > 10%[7,8]. According to the literature, cholangiocarcinoma is reported the most frequently at the age of 32 years in patients with BC (about 20 years earlier than in general population)[7]. There are reports of cholangiocarcinoma developing in adult many years after radical BC resection in infant[9]. Cholangiocarcinoma is the most serious and dangerous BC complication. The other complications associated with BC involve cholelithiasis and hepatolithiasis, cholangitis, acute and chronic pancreatitis, portal hypertension, liver fibrosis and secondary liver cirrhosis and spontaneous cyst perforation[8-12]. Nicholl *et al*[9] noted a higher number of complications in adults compared to children. Therefore, BC constitute an important problem for pediatricians, gastroenterologists, radiologists and surgeons. The role of pediatricians and gastroenterologists is a proper early diagnosis. Radiological investigations play an important role in BC diagnostics. Currently, the prenatal BC diagnosis using ultrasonography (USG) is possible[6,13,14].

Surgery is the treatment of BC. The goal of surgical treatment is to remove a cyst and to reconstruct the proper bile flow to the alimentary tract. In order to achieve this goal, different techniques are used. Roux-Y hepaticojejunostomy is the method of choice in most patients with BC[5-22].

ETIOLOGY AND PATHOGENESIS OF BC

Many theories explain the etiology and pathogenesis of BC[2]. In 1936, Yotsuyanagi supposed that BC arouse from inequality in the cellular proliferation of the biliary tract during the early fetal life[23]. Babbitts's theory of the "common channel" (1969) is the most widely accepted in the literature. According to this theory, the common cannel is formed by abnormal pancreaticobiliary junction (APBJ) of the pancreatic and bile ducts outside the ampulla of Vater. This condition leads to pancreaticobiliary reflux and mixing of the pancreatic and biliary juices and activations of pancreatic enzymes, because the maximal pressure within the pancreatic duct is two to three timed higher than within a biliary tract. Activated pancreatic enzymes cause inflammation and deterioration of the bile duct wall that leads to biliary dilatation[24]. A number of studies analyzed the levels of amylase within the bile duct. The higher amylase concentrations have been observed in patients with BC compared to the control groups. Also, association between the amylase level, earlier presentation and dysplasia grade in patients with BC has been reported in the literature[25,26]. Therefore, there is a theory, that the pancreaticobiliary reflux leads to inflammation and dysplasia in patients with BC[2,26-29]. APBJ occurs only in 50%-80% patients with BC. Another counterargument to this hypothesis is a fact, that immature neonatal acini do not produce sufficient pancreatic enzymes to explain antenatal BC[2,30,31]. The counterargument supporting Babbitt's theory is arbitral definition of the long common channel that depends on imaging modality and angles. Therefore, a defined length of a long common channel has been described from 10 mm to 45 mm in the literature. Therefore, APBJ can occur in more patients than radiological investigations show[2]. According to Okada *et al*[32], long common channel as any pancreaticobiliary junction is defined, if it lies outside of the duodenal wall and thus could result in pancreaticobiliary reflux and mixing. APBJ has been divided into three types by Komi *et al*[33]: (1) A right-angled union without an accessory pancreatic duct; (2) An acute-angled union without an accessory pancreatic duct; and (3) A right- or acute-angled union with an accessory pancreatic duct.

Table 1 Types of an anomalous pancreaticobiliary junctions (Komi classification)

Type name	Type description
Type Ⅰ	A narrowed common bile duct joins the pancreatic duct at a right angle
Ⅰ A	A slender common channel
Ⅰ B	A short or long ectatic common channel
Type Ⅱ	A pancreatic duct joins the common bile duct at an acute angle
Ⅱ A	A slender common channel
Ⅱ B	A short or long ectatic common channel
Type Ⅲ	It is complicated by a patent accessory pancreatic
Ⅲ A	A classic pancreas divisum
Ⅲ B	A pancreas divisum without a pancreatic duct
Ⅲ C	A patent accessory pancreatic duct with an intricate network
Ⅲ C1	A tiny communicating duct between the main and accessory ducts
Ⅲ C2	A communicating duct with the same caliber as the main and accessory ducts
Ⅲ C3	A total or partial dilatation of the ductal system

These types are subdivided into subtypes according to the shape of the common channel. The Komi's classification is presented in Table 1. The type of BC and clinical presentation can depend on the type of an angle of the anomalous ductal junction. A right-angled union is associated with cystic dilatation of the common bile duct. In patients with this type of union a palpable mass or jaundice as the main sign of disease are observed. An acute-angled union is associated with fusiform dilatation of the common bile duct. Patients with such dilatation usually complain on abdominal pain, and an erroneous diagnosis of acute pancreatitis is occasionally made. Type Ⅰ B, Ⅱ B, and Ⅲ junctions are associated with a dilated common channel and an accessory pancreatic duct. They are frequently complicated by relapsing pancreatitis leading to chronic pancreatitis[34]. Therefore, it's important to describe the type of APBJ. The type of APBJ is based on a cholangiopancreatographical picture[33-35]. The other theories regarding BC pathogenesis are less popular. According to Singham *et al*[2], biliary dilation can be a result of embryologic overproliferation of epithelial cells within solid bile ducts during the fetal life. According to Davenport and Basu, dilations are due to a lower number of neurons and ganglions in patients with BC that have been recognized pathologically. They suggested that round cysts were congenital with distal obstruction due to agan-

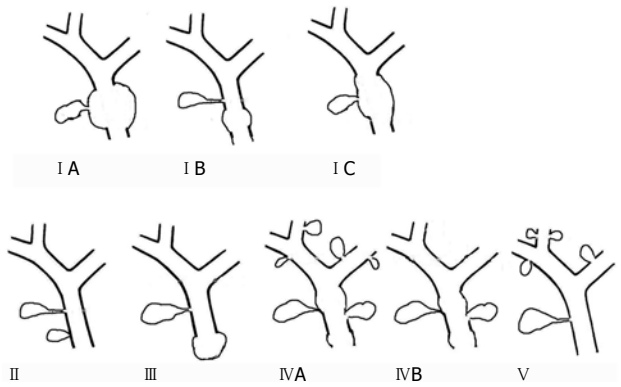

Figure 1 Types of biliary cysts according to Todani classification. Ⅰ: Solitary extrahepatic cyst (40%-85%): Ⅰ A: Common type; Ⅰ B: Segmental extrahepatic duct dilatation; Ⅰ C: Diffuse extrahepatic dilatation; Ⅱ: Extrahepatic diverticulum (2%-3%); Ⅲ: Choledochocele (1.4%-5.6%); ⅣA: Multiple extra- and intrahepatic cysts; ⅣB: Multiple extrahepatic cysts (18%-20%); Ⅴ: Multiple intrahepatic cysts (Caroli's disease) (rare).

glionosis and proximal dilation similarly to Hirschprung's disease. In this theory, chronic inflammation and clinical manifestation are caused by bile stasis within dilated bile ducts[2,31,36].

CLASSIFICATIONS OF BC

The first classification system for BC was proposed by Alonso-LEJ *et al*[37] in 1959. This classification involves three BC types: congenital cystic dilatation (the most common), congenital diverticulum of the common bile duct, and congenital choledochocele[37]. In 1977, Todani *et al*[38,39]modified BC classifications distinguishing five types (Figure 1). The most frequent (more than 90%) is type Ⅰ cyst. The type Ⅰ A cyst involves dilatation of the common bile duct, with marked dilatation of part or all of the extrahepatic bile ducts. The type Ⅰ B cyst involves segmental dilatation of the common bile duct, usually of its most distal part. The type Ⅰ C cyst involves fusiform dilatation of the common bile duct, along with diffuse, cylindric dilatation of the common hepatic duct and common bile duct. In 2011, Michaelides *et al*[40] reported and proposed a new BC variant and called it the Ⅰ D type. In this new variant, apart from the dilatation of the common hepatic and the common bile duct, dilatation of the central portion of the cystic duct was also observed, giving a bicornal configuration to the cyst. The type Ⅱ cyst is a diverticulum of the common bile duct usually arising laterally but may arise in the pancreatic portion. The type Ⅲ cyst (which is called a choledochocele) is a cystic dilatation of the intraduodenal portion of the common bile duct. According to some authors, choledochocele is different from other types of BC. In 2010, Ziegler *et al*[41], compared choledochocele with other types of BC (types Ⅰ, Ⅱ, Ⅳ and Ⅴ according to Todani classification). Based on study including 146 patients with BC (46 patients with choledochoceles), they concluded that classifications of choledochal cysts should not include choledochoceles. They reported that patients with choledochoceles differed from patients with other choledochal cysts with respect to age, gender, presentation, pancreatic ductal anatomy, and their management. Patients with cholechoceles were the most frequently male, older, complained with acute pancreatitis rather than jaundice or cholangitis, and were managed with endoscopic therapy. A pancreas divisum and a low risk of malignancy compared to other BC were observed with patients with choledochoceles. The type Ⅳ cyst involves dilatation of the intrahepatic and extrahepatic bile ducts. In the type ⅣA cyst, both the intrahepatic and extrahepatic bile ducts are dilated. The type ⅣB cyst involves dilatation of multiple segments of the extrahepatic bile ducts. The type Ⅴ cyst (Caroli's disease) involves dilatation of one or several segments of the intrahepatic bile ducts. First, it was described by Todd in 1818, but precise definition of this disorder was constructed by Jaques Caroli in 1958[41-45]. There are two descriptions (Caroli's disease and Caroli's syndrome) in the literature. Caroli's syndrome is the more serious condition compared to Caroli's disease. In Caroli's disease, cholangitis due to bile stasis within dilated intrahepatic bile ducts, is observed. In Caroli's syndrome, recurrent bouts of cholangitis due to bile stasis, hepatolithiasis, gallbladder stones lead to liver fibrosis with portal hypertension and liver failure. Caroli's syndrome can be associated with autosomal recessive polycystic kidney disease caused by a mutation of the *PKHD1* gene[46,47].

BC can be associated with the biliary atresia. Three biliary atresia (BA) types have been distinguished in the literature: 1 BA (atresia of the common bile duct), 2 BA (atresia of the common hepatic duct), and 3 BA (atresia of the porta hepatis)[18]. Muise *et al*[18], concluded that BA associated with BC formed a distinct subtype of BA, characterized by a preponderance of type 1 BA, a relatively good clinical outcome after surgery, and an absence of associated congenital anomalies. In 2004, Visser *et al*[48] proposed modified classification of BC and distinguished the following BC types: choledochal cyst, choledochal diverticulum, choledochocele, and Caroli's disease. All BC classifications are presented in Table 2.

CLINICAL PRESENTATION OF BC

The classic triad of clinical symptoms was described by Alonso-LEJ *et al*[37] and it involves right hypochondriac pain, palpable abdominal mass and jaundice. All clinical signs are observed only in 20%-30% patients[49]. Two of three clinical symptoms occur in 2/3 patients. The most frequently (in 80% of patients) clinical presentation appears before the age of 10 years[44]. Clinical presentation is different and it depends on the patient's age. According to the age, patients are divided into two groups: neonatal patients (under 12 mo) and adults (above 13 mo). Prenatally, BC are diagnosed incidentally during prenatal ultrasonography visualized as an intra-abdominal cystic mass[6,13,14]. In neonatal patients, obstructive jaundice

Table 2 Classifications of biliary cysts

Ref.	Classification
Alonso *et al*[37]	Congenital cystic dilatation
	Congenital diverticulum of the common bile duct
	Congenital choledochocele
Todani *et al*[38,39]	Ⅰ A Common type
	Ⅰ B Segmental extrahepatic duct dilatation
	Ⅰ C Diffuse extrahepatic duct dilatation
	Ⅱ Diverticulum type in the whole extrahepatic duct
	Ⅲ Choledochocele
	ⅣA Multiple cysts of the extra- and intra-hepatic ducts
	ⅣB Multiple cysts of the extrahepatic ducts
	Ⅴ Caroli's disease (intrahepatic bile duct cysts)
Vesser *et al*[48]	Choledochal cyst
	Choledochal diverticulum
	Caroli's disease

and abdominal mass are present the most frequently. Pain, fever and nausea are the most common for adult patients[44]. According to Lopez *et al*[21], pain is the commonest clinical sign in adults. Recurrent cholangitis and pancreatitis are complications of BC due to pancreaticobiliary reflux[49,50]. The other complications observed commonly in adults are the following: cholangiocarcinoma, cholecystolithiasis and cholecystitis, and liver cirrhosis[5]. Cholecystolithiasis and cholecystitis occur due to bile stasis in patients with BC. The secondary liver cirrhosis leads to different complications associated with portal hypertension such as upper gastrointestinal bleeding, splenomegaly and pancytopenia[49]. Portal hypertension can also occur without liver cirrhosis due to mechanical obstruction of the portal vein by the cyst[48]. An spontaneous BC rupture occurs in about 1%-12% of patients. It is presented as abdominal pain, peritonitis signs and sepsis[49,51]. Patients with choledochocele are usually asymptomatic. The Ⅲ BC type can be also presented by gastric outlet obstruction symptoms caused by direct obstruction of the duodenal lumen or intussusceptions[44]. It should be highlighted that BC are premalignant disorder. The overall reported risk of cancer is 10%-30% and it increases with age (from 0% in patients aged < 10 years to 75% in patients aged 70-80 years). Nicholl *et al*[9] reported direct correlation between the patient age and cancer risk: 0 year to 30 years (0%), 31 years to 50 years (19%), and 51 years to 70 years (50%). The histopathological types of cancer are the following: *adenocarcinoma* (73%-84%), *anaplastic carcinoma* (10%), *undifferentiated cancer* (5%-7%), *squamous cell carcinoma* (5%), and others (1.5%). The locations of cancer are the following: the extrahepatic bile duct (50%-62%), gallbladder (38%-46%), intrahepatic bile ducts (2.5%), the liver (0.7%), and the pancreas (0.7%)[49]. Todani *et al*[52] reported that 68% of cancers were associated with type-Ⅰ, 5% type-Ⅱ, 1.6% type-Ⅲ, 21% type-Ⅳ and 6% type-Ⅴ BC. Cholangiocarcinoma occurs as a result of chronic inflammation, cell regeneration and DNA breaks, leading to dysplasia[49,52]. The following molecular lesions have been reported in BC during carcinogenesis: microsatellite instability, k-ras mutations, expression of COX-2 and bcl-2, and increased telomerase activity that occur early and involvement of cyclin D1, beta-catenin, DPC-4/Smad4 and p53 that occur later[7].

DIAGNOSTICS OF BC

Laboratory investigations

Laboratory investigations may demonstrate mildly abnormal liver function and cholestasis tests (serum bilirubin, alkaline phosphatase, γ-glutamyltranspeptidase, alanine and aspartate aminotransferases) or amylase values, but these findings are not specific[34,49-56].

Radiological investigations

Imaging investigations are the most useful in BC diagnosis. The following radiological examinations are performed in order to visualize biliary dilatation: ultrasonography (USG) of the abdominal cavity, computed tomography (CT) of the abdominal cavity, and cholangiography[34,49-56].

USG of abdominal cavity

USG is the initial and easy to perform examination. It allows imaging of intrahepatic and extrahepatic bile ducts with measurement of the diameter of common bile duct or common hepatic duct and BC. It demonstrates (with the exception of type-Ⅲ and type-Ⅴ cysts) a cystic mass in the right upper quadrant (usually at the porta hepatis) separately from the gallbladder[49].

Use of USG in prenatal diagnosis

USG is very useful in prenatal diagnosis of BC. USG shows BC as a round intra-abdominal cystic mass located in the upper abdominal quadrant. Color Doppler USG shows no significant flow within the mass. Differential diagnosis of the cystic mass in prenatal USG should involve simple hepatic cysts, biliary atresia, ovarian, omental or mesenteric cysts, duodenal or gall bladder duplications, adrenal cysts, renal cysts, dilated loop of bowel, hydronephrotic renal pelvis and situs inversus. Proper early differential diagnosis between BC and another biliary disorder such as BA is very important, because BA needs an immediate surgery[6,13,14]. When the differential diagnosis of a cystic mass in the right upper quadrant is difficult to make by conventional USG alone, ultrasound-guided BC aspiration may serve as an alternative to the prenatal diagnosis of BC in the fetus[57,58]. Tanaka *et al*[14] compared imaging (USG and CT) and laboratory investigations in BC and BA in order to differentiate these two pathologies. They concluded that patients with biliary disorder smaller than 21 mm, direct bilirubin level higher than 2.5 mg/dL, and total bile acid level higher than 111 μmol/L in the neonatal period were more likely to have BA than BC and needed a surgery as soon as possible before irreversible liver cirrhosis. Okada *et al*[17] described the differential diagnosis between BC and BA by immunohistological examination using liver biopsy specimens.

They indicated that CD56-positive biliary duct cells were present in prenatally diagnosed type-1 cystic BA.

Cholangiography

Proper diagnosis requires demonstration of continuity of the cyst with the biliary tract, because it allows to differentiate BC from other intrabdominal cysts such as pancreatic pseudocysts, echinococcal cysts or biliary cystadenomas[49]. In order to show it, cholangiography is performed. There are the following methods of cholangiography: endoscopic retrograde cholangiopancreatography (ERCP), magnetic resonance-cholangiopancreatography (MRCP), percutaneous transhepatic cholangiography (PTC) and intraoperative cholangiography. These investigations allow to demonstrate the anatomy of biliary tree and define the BC type[49].

ERCP

ERCP is invasive investigation, but it has got therapeutic possibility. It accurately reveals the presence of any associated intraductal pathology or an APBJ. In the a type Ⅲ BC, it allows to perform a therapeutic papillotomy simultaneously[49].

MRCP

MRCP is a non-invasive procedure compared to ERCP. It is a favoured alternative to ERCP, but it has a lower accuracy in the detection of APBDJ and lacks the therapeutic possibility in case of the type Ⅲ BC. MRCP, as a non-invasive procedure, is investigation of choice in preoperative imaging of biliary tree[5,49,54-56]. A gadoxetic acid-enhanced MRCP in order to diagnose the biliopancreatic bile reflux and pancreatico-biliary reflux of pancreatic secretions in patients with anomalous union of the pancreatico-biliary duct has been described in the literature. Gadoxetic acid-enhanced MRCP can visualize the physiology of bile excretion, in contrast to conventional T2-weighted MRC which can visualize fluid filled space by heavily T2-weighted and fat-suppressed images. It is associated with specific properties of the gadoxetic acid which is uptaken by hepatocytes. Gadoxetic acid is excreted into the bile ducts that allows visualization of the bile ducts on hepatobiliary phase T1-weighted images[59].

PTC

Preoperative PTC allows to define the proximal extent of biliary dilatation and to use this information in preoperative resection planning[49]. According to Lipsett *et al*[53], the preoperative placement of a percutaneous ring catheter after cholangiography is useful for the surgeon during the operative procedure and can be used as a stent for the biliary reconstruction.

A technetium-99 HIDA investigation

A technetium-99 HIDA investigation is recommended to visualize the continuity BC with bile ducts. This investigation shows an initial area of photopenia within BC, with subsequent filling and then delayed emptying into the bowel[49].

CT of abdominal cavity

CT is useful in showing continuity of the cyst with the biliary tree, its relation to surrounding structures and the presence and staging of associated malignancy. It is better than USG in visualization of the intrahepatic bile ducts, distal bile duct and pancreatic head. In patients with type-ⅣA cysts and Caroli's disease, it is useful to describe the intrahepatic dilations and the extent of disease (diffuse hepatic or localized segmental involvement). It is important for a surgeon before operation, because localized type-ⅣA BC or Caroli's disease can be treated with segmental lobectomy[49]. Computed tomography cholangiography (CTC) after infusion of meglumine iodoxamate with subsequent 3-dimensional rendering has been described in the literature. Fumino *et al*[60] compared CTC cholangiography with MRCP in visualizing the pancreaticobiliary maljunction in 53 children. They noted superior visualization of the intrahepatic duct and the pancreatic system by MRCP compared with CTC. But the great advantage of CTC was its ability to produce high-quality images without respiratory artifacts in young infants in whom performing a good-quality MRCP is very difficult.

MANAGEMENT IN BC

BC require surgical intervention in order to avoid complications associated with pancreaticobiliary reflux. Management depends on the BC type. Currently, complete cyst excision with cholecystectomy followed by biliary reconstruction using a Roux-en-Y hepatico-jejunostomy is the treatment of choice[53,61,62].

Internal drainage (cystenterostomy) of BC

Currently, this method of treatment has been abandoned due to a high risk of morbidity and malignancy up to 50% following internal biliary drainage. This surgical procedure involved incision of BC and anastomosis it to the duodenum or jejunum depending to its location. Despite relief of clinical symptoms in operated patients, it was associated with a high postoperative mortality rate due to biliary reflux. Reflux of the enteric juice into the retained cyst and biliary tract leaded to recurrent ascending cholangitis. The another complication of this procedure was biliary anastomosis stricture. The most important complication of this treatment method was malignant transformation within the wall of retained BC[62-64]. Therefore, internal drainage is currently not considered to perform and all patients previously operated in this way should be reoperated in order to totally remove the cyst[62,63,65,66].

Total excision of BC with hepaticoenterostomy

Currently, total BC excision is recommended in order to avoid above mentioned complications. Total BC excision

with hepaticoenterostomy separates the biliary tree from the pancreatic duct that ends mixing pancreatic juice with bile. In situations where the intensity of fibrosis precludes safe periductal or pericystic dissection, Lilly's technique is useful. In this technique, the most densely adherent portion of the cyst wall is retained on the hepatoduodenal ligament, removing only the less adherent portion. The mucosal lining of the retained cyst wall should be ablated by diathermy or be stripped or destroyed by abrasion and iodine or alcohol application, because 57% of the bile duct cancer in BC arises from the posterior wall of the cyst[54,63-70].

There are two possible methods of hepaticoenterostomy: hepaticoduodenostomy and Roux-Y hepaticojejunostomy. According to the literature, the success rate of Roux-Y hepaticojejunostomy is 92%, with complication rate of 7%, compared with a complication rate of 42% following hepaticoduodenostomy. Some authors have compared these two reconstructions[63]. Shimotakahara *et al*[71] did not recommend hepaticoduodenostomy for reconstruction after BC excision due to a higher number of complications (33.3%) such as bilious gastritis due to duodenogastric bile reflux and adhesive bowel obstruction and cholangitis. Contrary to this report, Mukhopadhyay *et al*[72] recommended hepaticoduodenostomy as a simple and quick procedure with preservation of normal anatomy and physiology and minimum complications.

Some technical aspects of Roux-Y hepaticojejunostomy should be discussed. According to the literature, the anastomosis should be wide in order to prevent the postoperative stricture and Roux-Y jejunal loop should be 40-50 cm long[66]. Conventionally, an adult's standard of a 40-cm loop is adopted in Roux-Y hepatojejunostomy) also in children, irrespective of patient size. Diao *et al*[73] compared two patients groups according to Roux-loop length: (1) conventional group (n = 108) where a standard 35-40 cm Roux-loop length was used regardless of the child's size, and (2) short Roux-loop group (n = 110) in which the Roux-loop length was based on the distance between hepatic hilum and umbilicus. The mean Roux-loop length in patients in the second group was significantly shorter than in the conventional group. There was no significant difference between both groups in age, operative blood loss, operative time, postoperative hospital stay, and duration of drainage. In the conventional group, 2 of (1.8%) 108 patients developed Roux-loop obstruction was observed in 1.8% patients in 1st group and did not appear in 2nd group. Mild reflux was reported in 2 patients in 1st group and 1 patient of 2nd group. Authors concluded that shorter Roux-Y loop according to patient's size can be safe and effective alternative for biliary reconstruction in children with BC.

Another technical aspect of BC surgery is management with the distal bile duct stump, after cyst excision, which is usually ligated. Diao *et al*[74] compared two patients groups in order to investigate the feasibility of selectively leaving distal stump unligated. In this study, patients were divided into two groups: (1) non-ligation group (n = 207), where the distal stump was stenotic and was left unligated, and (2) ligation group (n = 63), where the distal stump was not stenotic and was ligated. The pancreatic juice leakage rates were compared. Authors concluded that not ligating distal stump is a feasible approach in patients with the stenotic distal common bile duct. In authors' opinion, this management simplifies the operative procedure and may minimize pancreatic duct injury.

Different treatment depending on BC type

Total cyst excision with Roux-Y hepatojejunostomy has been recommended by most authors in the treatment of I and Ⅳ types of BC[62]. The risk of malignancy in type Ⅱ and Ⅲ BC is low. Therefore, complete cyst excision is not necessary. Simple excision of type Ⅱ BC is sufficient. Choledochoceles often just require endoscopic sphincterotomy in order to allow free duodenal drainage of bile and stones. Large choledochoceles should be treated surgically and excised *via* duodenostomy because of biliary, duodenal or gastric outlet obstruction. In some cases of small cysts, endoscopic excision is possible. Type ⅣA BC is treated by surgical cyst excision and a wide hilar hepaticoenterostomy, but clinical symptoms are frequently observed in patients following operation that is caused by the intrahepatic involvement of disease. If the intrahepatic involvement is localized, a segmental hepatectomy can be performed. In case of diffuse disease, a percutaneous hepaticojejunostomy, surgical or endoscopic unroofing of some intrahepatic cysts can be made. Unilobar Caroli's disease can be treated by hepatic lobectomy. Diffuse Caroli's disease with recurrent cholangitis, liver failure and cirrhosis and portal hypertension or malignant disease requires orthotopic liver transplantation[61]. There are a lot reports regarding laparoscopic excision of BC with extracorporeal biliary reconstruction. Total intracorporeal biliary reconstruction has been also described in the literature. This minimally invasive treatment method is safe and effective, and it is associated with earlier postoperative feeding and discharge from hospital[75,76].

The timing of BC surgery

Early surgical treatment in order to prevent further complications is recommended. It has been observed that early diagnosis and surgical treatment was associated with a lower morbidity compared with later diagnosis and treatment. Therefore, prenatal diagnosis is such important[77,78]. Foo *et al*[77] evaluated the clinical outcomes of patients with prenatally diagnosed choledochal cysts compared with those diagnosed after birth and the optimal timing of definitive treatment. The mean age at operation for the prenatally diagnosed group was 4.4 mo. For the postnatal diagnosed group, the mean age at operation was 5.7 years. Based on analysis of 45 patients, they concluded that prenatal diagnosis of BC resulted in earlier

definitive surgery. In patients operated in older age, more adverse complications were observed[77]. Tsai *et al*[79] compared results of surgical treatment according to patients age at the time of surgery. They compared three patients groups that had been operated: < 1 year old (group Ⅰ, n = 26), 1-16 years old (group Ⅱ, n = 48), and > 16 years old (group Ⅲ, n = 33). Group I suffered significantly fewer surgical complications and less severe liver fibrosis compared with groups Ⅱ and Ⅲ. Therefore, authors recommended early surgical treatment. Germani *et al*[80] recommended the surgical treatment after 3 mo of life to increase the success according to the size of the anatomic structure. They recommended the early treatment before 3 mo of life, only in case of severe symptoms.

RESULTS OF THE SURGICAL TREATMENT OF BC

Short-term results and early complications

Surgical treatment of BC is successful in more than 90%. It is associated with a low postoperative morbidity (2.5%-27%) and mortality (0%-6%) rate according to the surgical center and patient's age. The following early complications have been reported in the literature: biliary-enteric anastomosis dehiscence with biliary fistula, intra-abdominal bile collection (biloma) or abscess and acute peritonitis, pancreatic leakage and pancreatic fistula due to injury to the pancreatic duct, acute pancreatitis, acute cholangitis, ileus caused by bowel obstruction due to intussusception or bowel kinking due to manipulation or adhesions, wound infection and wound dehiscence, gastrointestinal bleeding, hepatic failure, and multiple system organ failure[49,54,64,65,67,68,78,81-87].

Long-term results

Late postoperative complications occur in up to 25% patients and they are the following: biliary-enteric anastomosis stricture, peptic ulcer disease, cholangitis, biliary and intrahepatic stones, pancreatitis, liver failure and biliary cancer. Authors recommend to perform a wide bilio-enteric anastomosis in order to prevent its stricture[49,54,64,67,68,77,80-88]. According to the literature, 0%-6% incidence of malignancy occurs following surgery. It is caused by remnant cyst tissue or subclinical malignant disease which had not been detected before surgery. Therefore, some authors recommend intraoperative endoscopic ultrasonography and histopathological investigation of frozen sections to rule out dysplasia, hyperplasia and malignant disease. All patients with BC require long-term follow-up for bile duct cancer, using ultrasonography and laboratory investigations including liver parameters and cancer markers [carcinoembryonic antigen (CEA), CA 19-9, CA-125][49,87-89]. CA 19-9 is the mosts significant, because it is elevated in up to 85% of patients with cholangiocarcinoma. CEA is raised in about 30% of patients and CA-125 in 40%-50% of patients with cholangiocarcinoma. These markers are not specific for bile duct cancer, because their levels are also raised in other neoplasms and inflammatory diseases[90]. Lee *et al*[91] analyzed factors predicting cholangiocarcinoma in patients operated for BC. They pointed the following factors: age > 40 years, the absence of a gallstone, elevated CEA or cancer antigen 19-9 serum level, and the presence of anomalous pancreaticobiliary ductal union in univariate analysis, and an elevated cancer antigen 19-9 level in multivariate analysis. Therefore, laboratory investigations of CEA and CA 19-9 in patients following surgery for BC are useful.

Cytology of bile and bile ducts specimens taken during ERCP ora PTC by brush or needle biopsy play an additional role in cholangiocarcinoma diagnosis. Negative cytology from brushing does not exclude malignancy. Combined brush and biopsy cytology specimens increase sensitivity to 40%-70%[90].

Frequency of follow-up in patients following BC resection has not been clearly established in the literature. In my opinion, laboratory investigations including liver parameters and cancer markers (CA 19-9 and CEA) and ultrasonography should be performed as a screening every 6-12 mo.

In cases of the intrahepatic lithiasis and Caroli's disease, the use of ursodeoxycholic acid (UDCA) in the treatment has been described in the literature. Ros *et al*[92] UDCA observed that therapy with UDCA caused clinical remission, return to normal liver function, and dissolution of intrahepatic stones on USG in all patients. Therefore, litholytic therapy was indicated by authors for intrahepatic stones in Caroli's syndrome. Successful treatment of patients with the hepatilithiasis and Caroli's disease was also described by Guma *et al*[93].

In conclusion, clinical presentation and management of BC depend on the cyst type according to Todani classification. The early and proper treatment allows to avoid serious complications (including bile duct cancer) in patients with BC. The early treatment is also associated with a lower number of complications. In most cases, total cyst excision with Roux-Y hepaticojejunostomy is the treatment of choice. Surgical treatment of BC is associated with low morbidity and mortality. Patients following surgery for BC require permanent and careful postoperative observation because of possibility of biliary anastomosis stricture and biliary cancer in tissue remnant.

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