BILE DUCT CYSTS

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Aim of the study was analysis of methods of surgical treatment of patients with bile duct cysts.

Material and methods. Retrospective analysis of data from 30 patients who underwent surgical treatment for bile duct cysts in Clinic of General, Transplantation and Liver Surgery, Warsaw Medical University, between 1 October 2001 and 31 December 2009.

Results. Bile duct cysts are more common in females; female to male ration is 4:1. Most of the treated patients had bile duct cysts belonging to type I according to Todani classification – 13 patients (43.3%). Six patients (20%) had type IV cysts, 8 patients (26.7%) had type V cysts according to Todani classification. Three patients (10%) with isolated intrahepatic bile duct cysts were not classified to any group according to Todani classification. The most common type of surgical treatment was complete resection of bile ducts cysts with choledocho-intestinal Roux-en-Y anastomosis that was performed in 17 patients (56.7%). The other patients generally underwent various types of resections of the liver and bile ducts or only of the liver. Five patients (16.7%) required liver transplantation. Nine patients (30%) developed complications. One patient (3.3%) who underwent liver transplantation and retransplantation, died from progressive multiorgan failure and renal failure.

Conclusions. First line treatment of patients with bile duct cysts involves their resection, sometimes with requirement of resection of liver parenchyma. Most of these patients underwent reconstruction of bile duct through choledocho-intestinal Roux-en-Y anastomosis. Some patients undergo liver transplantation. Surgical treatment of patients with bile duct cysts is demanding from the technical point of view and should be undertaken in centers that specialize in hepatobiliary surgery.

Key words: bile duct cysts, bile duct resection, liver resection

Bile duct cysts are rare, hereditary anomalies, manifesting as simple or complex dilatations of the biliary tree, of its extrahepatic, intrahepatic segment or both (1-4). Female to male ratio is from 3:1 to 5:1 (1, 3, 5-9). Only approximately 20% of cysts are diagnosed in adults; they are more common in children (1). Incidence of bile duct cysts is variable and ranges from 1:100 000 to 1:150 000 in Western countries versus 1:1000 in Asian countries (Japan, China) (10).

The term „cyst” originates from the Greek „kystis” (pocket, bladder) (8). „Choledochal cyst”, a term used in English literature to indicate a bile duct cyst, is not completely correct since dilation can occur both in intrahepatic and extrahepatic bile ducts while the term “choledoch” refers only to the common bile duct. Therefore the following terms should be used: “bile ducts cysts” or “biliary cysts” (2, 8). The most typical signs and symptoms include: abdominal pain, nausea and vomiting, jaundice, fever, cholangitis, pancreatitis, rarely an abnormal epigastric mass. The most common out of the above mentioned signs and symptoms is abdominal pain (1-4, 6-9). Triad of signs and symptoms that includes: abdominal pain, jaundice and palpable epigastric mass, is found occasionally (1-4, 6-9). In adults correct diagnosis is often challenging and delayed due to
nonspecific and periodic nature of symptoms (1, 11). Bile duct cysts are associated with increased risk of bile duct cancer, ranging from 6 to 21% (1, 3, 4, 6, 8, 9).

The most common theory of pathogenesis of bile duct cyst is Babbit’s “long common channel theory” (12). Abnormally long common channel (longer than 15 mm) results from anomaly of pancreatobiliary junction. It leads to regurgitation of pancreatic juice to the common bile duct, resulting in formation of inflammatory lesions and epithelial damage. Walls of bile ducts become thin and more prone to develop dilatation. The second most common pathogenetic theory is oligogangionosis introduced by Kusonoki (13). Abnormal autonomic innervation of the distal part of the common bile duct, related to inadequate number of ganglionic cells results in a functional stricture and dilatation of the proximal part of the biliary tree. Stricture of the distal part of the common bile duct is another possible etiological factor of bile duct cysts (14).

First classification of extrahepatic bile duct cysts was introduced by Alonso-Lej (15) in 1959 (fig. 1, 2, 3).

In 1977 this classification was expanded by Todani to include type IV and V and became the most popular classification system (16). This classification categorizes bile duct cysts into 5 main types. The most common type I is a single dilatation of extrahepatic biliary tree. It is further subdivided into three subtypes according to the cyst shape: vesicular – Ia (fig. 4), focal – Ib (fig. 5) and fusiform – Ic (fig. 6).

Type II (fig. 7) is a saccular diverticulum of the common bile duct. Type III (fig. 8) is a paraduodenal dilatation of intramural part of the common bile duct, also referred to by its Latin term „choledochoecele“.

Type IVa (fig. 9), the second most common, indicates multiple dilatations of intrahepatic and extrahepatic bile ducts. In type IVb (fig. 10), the dilatations are limited to extrahepatic biliary tree.

Type V (fig. 11) indicated multiple dilations of intrahepatic bile ducts. This type of changes is also referred to as Caroli’s disease (17).

Transabdominal ultrasound imaging is an useful modality in a preliminary diagnostics of bile duct cysts (fig. 12) (1, 2, 4, 7, 18).
Furthermore computed tomography imaging (fig. 13) can be used to establish the extent of the disease process (1, 18).

Computed tomography is also useful in the postoperative period to visualize choledochointestinal junction and diagnose its strictures (11). Endoscopic retrograde cholangiopancreatography (ERCP) (fig. 14) and percutaneous transhepatic cholangiography (PTC) play a critical role in the diagnostics of bile duct cysts because their accurately establish anatomy of intrahepatic and extrahepatic biliary tree (4, 6, 7, 9). Both modalities also detect anomalies of choledochopancreatic junction (2, 6, 7).

Magnetic resonance cholangiopancreatography (MRCP) (fig. 15) precisely demonstrates anatomy of the biliary tree and is currently considered as a "gold standard" in the imaging diagnostics of the bile duct cysts (11, 18). Unless definite diagnosis can be established using US and CT imaging or when more precise establishment of the bile duct anatomy is required, CRCP, MRCP or PTC should be used (4).
MATERIAL AND METHODS

Thirty patients with bile duct cysts underwent surgical treatment in Clinic of General, Transplantation and Liver Surgery, Medical University of Warsaw, between 2001 and 2009. The study group included 24 women and 6 men, with female to male ratio 4:1.

RESULTS

An average patient age was 37.3 years (20–70 years). The cysts were classified according to Todani classification. Type I cysts were found in 13 patients (43.3%), type IV in 6 patients (20%) including 4 cysts (13.3%) of type IVb and 2 cysts (6.6%) of type IVa, and 8 patients (26.7%) were diagnosed with type V according to Todani classification. Three patients (10%) were diagnosed with isolated dilatation of intrahepatic bile duct that did not meet any type according to Todani classification.

We did not find any patient with type II or III bile duct cyst according to Todani classification. None of the patients was diagnosed with malignant changes.

All patients with type I cyst according to Todani classification underwent complete resection of extrahepatic bile ducts with choledocho-intestinal Roux-en-Y anastomosis (fig. 16a, 16b, 16c, 16d, 16e).

One patient, a 66-year old woman, who had undergone side-to-side cystoduodenostomy (outside our hospital) required reoperation due to recurrent infections of bile ducts. The primary anastomosis was removed, the duodenum was sutured and then extrahepatic bile duct was resected from the site of junction of the right and left hepatic duct to head of the pancreas, with subsequent choledochojejunostomy Roux-en-Y.

Two patient with type IVa underwent the following management. A 47-year old female patient with cysts of the extrahepatic bile ducts and left hepatic duct underwent only resection of extrahepatic bile ducts with choledocho-intestinal Roux-en-Y anastomosis. Left hemihepatectomy was impossible due to anatomical variant of intrahepatic bile ducts involving presence of 4 right hepatic ducts that had independent connections with the left hepatic duct, completely involved by cystic lesions. This was the only case in which not all lesions were resected. The patient is still being monitored and up til now she has not required surgical reintervention. A 20-year old female patient with multiple cysts of intrahepatic and extrahepatic bile ducts underwent piggy-back liver transplantation. Unfortunately, she developed liver failure and early thrombosis of the hepatic artery after the surgical procedure. She underwent classic retransplantation in an urgent setting. The patient died from multiorgan failure and renal failure. This is the only case of postoperative death in the presented material.

Treatment of 4 patients with type IVb cysts involved complete resection of extrahepatic bile ducts...
Fig. 16b. Status post resection of extrahepatic bile duct cyst. The hepatic duct was prepared for Roux-en-Y anastomosis.

Fig. 16a. Intraoperative image of extrahepatic bile duct cysts. A catheter was inserted to the cystic duct to perform intraoperative cholangiography.

Fig. 16c. Choledochojunostomy.

Fig. 16d. Jejunojunostomy ending the surgical procedure. Roux-en-Y loop has the length of 70 cm and is located retrocolic.

Fig. 16e. Resected extrahepatic bile duct cyst. The image demonstrates “deflated” cyst connected via cystic duct with a gall-bladder.

Bile duct and creation of choledocho-intestinal Roux-en-Y anastomosis.

Types of surgical procedures in the treatment of patients with type V cysts were variable. The treatment of choice of 2 patients with lesions limited to bile ducts of the left hepatic lobe was left hemihepatectomy. In one case, in which cysts were limited to intrahepatic portion of the biliary tree of the right lobe and segment IV, extended right hemihepatectomy, resection of extrahepatic bile ducts and anastomosis of the left hepatic duct with Roux-Y loop was performed. One patient, a 38-year-old female with cysts of intrahepatic bile ducts of the left lobe, with a history of choledochojunostomy, underwent left lateral bisegmentectomy with resection of extrahepatic bile ducts, and then choledocho-intestinal Roux-en-Y anastomosis with right hepatic duct. Piggyback liver transplantation was performed in the remaining 4 patients. Secondary biliary cirrhosis was an indication for liver transplantation in a 32-year-old woman after resection of extrahepatic bile ducts with choledocho-intestinal Roux-en-Y anastomosis. In the remaining 3 patients (2 men, 1 woman), cysts...
occupied a significant portion of both liver lobes.

Surgical management of isolated dilatation of intrahepatic bile ducts was also variable. One patient underwent simple cyst resection (of segments I and IV along with tissues in the area of the liver hilum), in another two patients with a cyst of the intrahepatic segment of the hepatic ducts, left lateral bisegmentectomy was performed (fig. 17a, 17b, 17c, 17d, 17e, 17f, 17g, 17h)

Complications occurred in 9 patients (30%) and included: infection of a postoperative wound found in 4 patients (13.3%), temporary external biliary fistula in 3 patients (10%), acute pancreatitis in 1 patient (3.3%) and a fluid collection in a cavity after resection of the liver with a cyst also in 1 patient (3.3%). No case required repeated surgical treatment. Mortality was 3.3%.

Six liver transplantation procedures (including 1 retransplantation) were performed between 2004 and 2008 due to bile duct cyst. 466 liver transplantations were performed during this time in Department of General, Transplant and Liver Surgery, Medical University of Warsaw. Therefore liver transplantations due to bile duct cysts accounted only for 1.3% of all liver transplantations.

**DISCUSSION**

Liver cysts are more common in females, with female to male ration of 4:1.

Irrespective of the fact that the aim of surgical treatment is to completely resect the
cysts, type of management depends on type of lesions and particular cyst type (1). Anatomical cyst variants may require more extensive surgical procedure (7).

The most common method of management of our patients with extrahepatic bile duct cysts was complete resection of extrahepatic bile ducts with choledochojejunostomy Roux-en-Y, performed in 17 of 30 patients (56.7%) – 13 with type I according to Todani classification and 4 with a type IVb cyst.

Other authors suggest identical management for the patients with type I, II and IVb (1, 3, 9). This type of surgical treatment results in resection of mucosa characterized by the highest risk of malignant transformation (8). As our experience suggests, too, the resection should involve a segment of the bile duct from the site of junction of the right and left hepatic duct to head of the pancreas (7, 8). Unless adequately extensive distal resection can be performed without of damage of the pancreatic parenchyma, the patient may require pancreatoduodenectomy (7). Such treatment was not required in our group of patients.

Cystoenterostomy is currently not recommended due to high rate of patients that developed cholangiocarcinoma in the remaining cyst (1, 3, 6, 8, 9). We do not use this type of treatment at our site. Patients who previously had undergone only internal drainage (cystoenterostomy) should undergo excision of the underlying cyst (1, 3, 6, 8). Malignant transformation in patients who underwent cystoenterostomy could be secondary to reflux of in-
testinal bacteria and enzymes, through choledochojjunostomy (5).

Simple cyst resection may be adequate in type II (1, 4, 6). In cholangiocarcinoma, authors recommend complete resection of extrahepatic bile ducts in combination with resection of head of the pancreas and reconstruction by choledochojjunostomy Roux-en-Y anastomosis (1). Treatment options of type III cysts include additionally endoscopic sphincterotomy or surgical transduodenal papillotomy (3, 4, 6, 9). Patients with type III cysts may undergo medical treatment provided they undergo close monitoring for complications such as cholelithiasis, bile duct strictures, pancreatitis and cancer (7). Patients from this group should undergo pancreateoduodenectomy if cholangiocarcinoma does not necessarily result in malignant transformation (4).

Management of type IVa is the subject of controversy (1, 7, 9). If the disease involves an almost whole liver, the patient may require heptatectomy and liver transplantation (7). We subjected one of our female patients to such management. Liver resection along with removal of extrahepatic bile ducts is possible in type Iva, provided that the lesions involve extrahepatic bile ducts in one part of the liver, so that the resection can be performed (2, 3, 6, 7). Resection involving liver parenchyma is also required when malignant lesions are present (1). However, one must remember that complete resection of type IVa cysts is not universally possible due to extensive involvement of intrahepatic biliary tree or involvement of head of the pancreas (1, 2, 7). We adopted such treatment strategy to another our patient. According to some authors, many type IVa bile duct cysts may be successfully treated with resection of intrahepatic lesions with choledochojjunostomy Roux-en-Y anastomosis (3, 7). This is a controversial treatment option since one can never be sure whether resection of only extrahepatic lesions is sufficient to prevent malignant transformation. However, literature data indicate that cholangiocarcinoma does not necessarily result in patients with type IVa cysts in whom cysts have not been completely resected, who underwent primary treatment with choledochointestinal anastomosis (1).

In type V (Caroli disease), partial liver resection is a possible treatment option only in some patients (1, 3). Should complications reappear, orthotopic liver transplantation is indicated (1). It is also required in the event of involvement of intrahepatic bile ducts to the degree that makes liver resection impossible. Such situation was present in our 4 patients. Various types of surgical treatment are useful in type V cysts, as it was the case with another our 4 patients. Selection of adequate technique depends on the number and location of cystic lesions in the intrahepatic biliary tree.

Diagnosis and treatment of bile duct cysts is of utmost importance due to a significant risk of cholangiocarcinoma formation (4). Therefore, complete cyst resection with restoration of bile outflow tract is the treatment of choice (4). It is up to a surgeon to decide whether the risk related to incomplete cyst resection in the intrahepatic and extrahepatic biliary tree justifies against the risk of possible complications related to more extensive surgical procedure (7). It is impossible to predict whether cholangiocarcinoma develops after resection of cystic lesions, and time between the surgical treatment and its diagnosis may range from 10 to 20 years (7). Liver resection or heptatectomy and the organ transplantation in type IVa and V cysts are very extensive and rarely performed procedures, however they are indicated if removal of the whole intrahepatic biliary tree is required (7). Liver transplantation is also the only method eliminating the risk of cholangiocarcinoma in patients with type IVa or V cysts (4). Complete cyst resection is the best management, however choice of treatment depends on patient’s age, comorbidities, anatomical conditions and surgeon’s assessment (7).

**SUMMARY**

Bile duct cysts are a significant problem in the surgical practice, never mind their challenging diagnostics. Treatment of patients with bile duct cysts is not free from complications and could be life threatening. Adequate resection can be impossible due to extensive involvement of intrahepatic biliary ducts by the disease process or due to atypical anatomic variants of the intrahepatic biliary tree. In such cases postoperative patient monitoring is indicated. Liver transplantation is the only radical method of treatment in some patients.

Surgical treatment of patients with bile duct cysts should be undertaken only in centers that specialize and have experience in hepatobiliary surgery, that use a complete
panel of diagnostic methods, are equipped with specialist devices and perform all range of procedures in the liver surgery, including liver transplantation. All patients who undergo surgical treatment for bile duct cysts should be closely monitored due to the risk of cholangiocarcinoma or serious liver complications, such as recurrent cholangitis.

REFERENCES


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