Surgical Management of Adult Choledochal Cysts

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Objectives: *Results of the surgical management of 17 choledochal cysts in adults at the Department of Surgery, Siriraj Hospital, Mahidol University, are presented.*
Material and Method: *All the patients who underwent diagnosis and were surgically managed during the period between October 1990 and January 1999 were analyzed retrospectively. Cysts were classified anatomically according to the descriptions of Todani et al. The authors assessed the clinical features, operative procedure and outcome of the patients.*
Results: *There were 15 females and 2 males, with ages ranging from 16-45 years. Only 2 patients (11.8%) had the clinical triad: jaundice, abdominal pain and mass. Clinical pancreatitis was presented in 3 patients (17.6%). There were 10 type I (58.8%), 6 type IVa (35.3%) and one type V (5.9%) according to Todani's classification. Cholangiocarcinoma was found in one patient (5.9%). Extrahepatic cyst excision with a Roux-en-Y hepatico-jejunostomy was performed on 16 patients with type I or IVa cysts (94.1%). There were no surgical deaths or complications. Ten survivors are well. The authors lost contact with 6 patients during follow-up (35.2%). The median follow up was 3.2 years. The patient with cholangiocarcinoma died 2 years after treatment.*
Conclusion: *This experience recommends total extra-hepatic cyst excision with Roux-en-Y hepaticojejunostomy as the treatment of choice for adult choledochal cyst type I and IV to eliminate the risk of recurrent cholangitis and malignancy.*

Keywords: *Adult choledochal cyst, Surgical management*

J Med Assoc Thai 2005; 88(7): 939-43
Full text. e-Journal: *http://www.medassocthai.org/journal*

Choledochal cyst, a congenital dilatation of the biliary tree, is a relatively rare abnormality. Their incidence is reported to be 1 in 13,000 births in Japan[1], about 150 times more than in the West where the incidence is 1 in 2,000,000 births[2], three to four times more frequently in females. In Thailand, the incidence of this condition has been reported in a small series which varied from being 1 in 2,400 to more than 1,000,000 births [3,4].

The condition is typically presented in infancy and childhood, but 20% of patients delay diagnosis until they are adults[5]. In adults, the choledochal cysts may have complex clinical features that influence the operative procedure and surgical outcome. The surgical result of these patients may not be as good as those patients who were treated during childhood because severe inflammation and malignant change of the lesion can be found. In Thailand, only 5 cases of Adult choledochal cysts have been reported[6]. During the past 10 years the authors have managed 17 patients who presented with adult choledochal cysts in Siriraj Hospital. The present report includes the largest series of surgical management of adult choledochal cysts in Thailand so far.

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Material and Method

Between October 1990 and January 1999, patients older than 16 years of age who underwent surgery for symptoms associated with choledochal cysts at the Department of Surgery, Faculty of

Medicine Siriraj Hospital, Bangkok 10700, Thailand, were included in the present study.

Gender, age, presenting symptoms, radiologic data, operative procedures, early morbidity, mortality and later results from the surgical procedures were recorded. The types of choledochal cysts, based on radiologic and operative findings, in the present study were assigned according to the classification of Todani et al in 1977 (Fig. 1)[7].

The medical records and radiographs of all patients were reviewed. Follow-up data were obtained via a review of outpatients' medical records and through telephone interviews, letters and follow-up visits. Histopathological confirmation of the diagnosis was obtained in all cases.

Results

Demographics

Of 17 patients, 15 (88.2%) were females and 2 (11.8%) were males. The median age on admission for the authors' care was 28 years, with a range from 16 to 45 years. The mean age for all 17 patients was 29.7 years. Ten patients (58.8%) had Type I choledochal cysts, six (35.3%) had Type IVa and one (5.9%) had Type V.

Clinical presentation

The mean duration of symptoms before initial surgical treatment was 2 years, with a range from 4 days to 16 years. The presenting symptoms and signs are given in Table 1. The most common presenting feature was jaundice (70.6%). Only two patients (11.8%) appeared with the classic triad of symptoms of abdominal pain, jaundice and abdominal mass.

Twelve patients had a coexistent hepato-biliary disease associated with the choledochal cyst (Table 2). Approximately 47% of the presented patients had biliarylithiasis. Only one patient presented with biliary carcinoma associated with cyst Type I.

Operative procedures

A total extrahepatic cyst excision with Roux-en-Y hepaticojejunostomy was performed for type I or IVa cysts in 15 of the 17 patients (88.2%) as definitive treatment. One patient with biliary carcinoma (5.9%) underwent Whipple's operation because of invasion

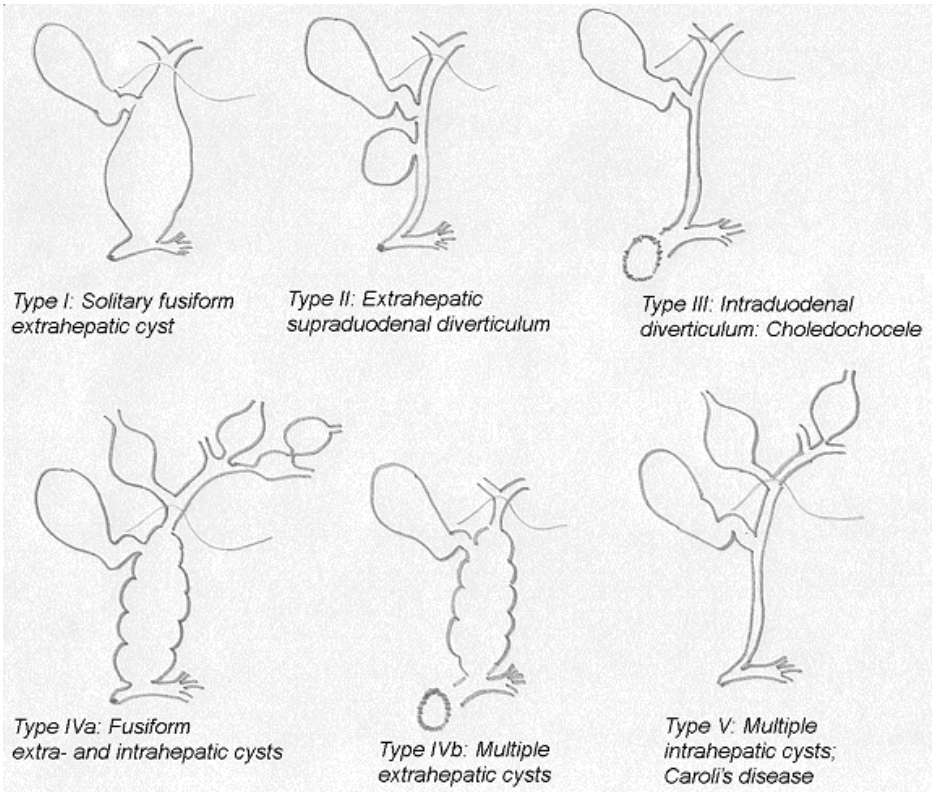

Fig. 1 Todani's classification of choledochal cysts[7]

Table 1. Frequency of symptoms and signs associated with choledochal cysts in 17 adults

Symptoms and signs	Frequency (%)
Jaundice	70.6
Abdominal pain	58.8
Fever	52.9
Abdominal mass	41.2
Classical triad	10.8

Table 2. Hepatobiliary disease associated with choledochal cysts in 17 adults

Pathologic finding	No. of patients (%)
Biliarylithiasis	8 (47.0%)
Acute pancreatitis	3 (17.6%)
Cholangiocarcinoma	1 (5.9%)

of the pancreas by cancer that had developed in the cyst. An opened cholecystectomy with exploratory common bile duct for stone removal was performed in one case (5.9%), Type V.

Postoperative complications

There was no operative death in the present series. Only two patients developed early post-operative complications including fever, bile or pancreatic juice leakage and wound infection; all recovered with conservative treatment.

Follow-up

The median (range) follow-up was 3.2 years (1 month to 10 years). One patient with cholangio-carcinoma who underwent Whipple's operation died 2 years after the operation. The 10 patients who are alive and available for follow-up have remained symptom free after the treatment.

Discussion

The present findings confirm that choledochal cysts in adults frequently have complex clinical and pathological characteristics that influence surgical management[8,9]. In adults, cystic disease of the intrahepatic ducts and coexisting hepatobiliary conditions (eg. biliarylithiasis, pancreatitis and malignant disease) present problems that require an individualized therapeutic approach.

Most choledochal cysts occur in women. Adults with initial manifestation of choledochal cysts usually have nonspecific symptoms. In the present study, the most common clinical presentation of adults with choledochal cyst was cholangitis, including jaundice and abdominal pain, similar to previous reports[10-13] occuring in 60-70% of patients. A palpable mass is uncommom [14]. The presented cases fit this pattern. These symptoms are indistinguishable from those biliary calculus diseases. Abdominal Utrasound and CT scan usually delineate the cyst. At the present time, documentation of the extent of the cyst by endoscopic retrograde cholangio-pancreatography (ERCP) preoperatively or by intraoperative cholangiography is essential in planning the surgical approach. Although not observed in the present study, other pancreatic and biliary anomalies could be reported to be present with an anomalous pancreatico-biliary union [15] by ERCP.

The association of pancreatitis with choledochal cyst is well-recognized[16]. The presence of acute pancreatitis may suggest other pancreatic ductal anomalies or that the etiologic factors were not all eradicated with a total excision of the choledochal cyst[12,17]. In the present study, the authors found three patients, one Type I and two Type IVa, with clinical symptoms of acute pancreatitis. After the operation, the patients had no evidence of recurrent pancreatitis. At the present time, a satisfactory explanation of this phenomenon is not clear.

More important is the question of the development of cancer. Cancer may occur in the cyst of a patient who is not diagnosed until adulthood, or in a cyst that was initially treated by cyst enterostomy. The reported incidence is 2.5% to 15.6% of cases[18]. When a cancer has developed, treatment of cancer related to the choledochal cyst points to extensive excision of the biliary system because the malignancy is not only limited to the cyst but also can occur in other areas of the biliary tract as well, such as the gallbladder, pancreas and liver[19,20].

Total extrahepatic cyst excision with Roux-en-Y hepaticojejunostomy is now the preferred option for treatment of the choledochal cyst[21]. Cystoenterostomy is no longer recommended. Not only problems associated with retention of the cyst, including biliary stasis, recurrent cholangitis, stone formation, pancreatitis and carcinoma of bile duct[10], but also the cyst excision was successfully performed without an increased risk of death or severe complications[10,11,13], as demonstrated in the present study. In addition, early surgery is suggested for young patients with a choledochal cyst detected by routine screening.

Table 3. Surgical management of Choledochal cysts

Type I	Total cyst excision with Roux en Y hepaticojejunostomy
Type II	Simple cyst excision
Type III	Endoscopic sphincterotomy
Type IVa	Extrahepatic cyst excision with Roux en Y hepaticojejunostomy
Type IVb	Total cyst excision with Roux en Y hepaticojejunostomy
Type V (Caroli's disease)	Medical management of symptoms (? Liver transplantation)

Surgery should be performed after the diagnosis is made[22,23], which varies according to the type of cyst (Table 3.).

In summary, surgical experience of 17 patients with choledochal cysts in adults was reviewed. Total extrahepatic cyst excision with Roux-en-Y hepaticojejunostomy was the treatment of choice. Recurrence of symptoms was minimally found in the long-term follow-up. In addition, most of them responded to conservative treatment.

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การผ่าตัดรักษาผู้ป่วยโรคถุงน้ำของท่อน้ำดีร่วมในผู้ใหญ่

ธวัชชัย อัครวิพุธ, วิรุณ บุญนุช, ประสิทธิ์ วัฒนาภา, ณรงค์ เลิศอรรฆยมณี, ดรินทร์ โล่ห์สิริวัฒน์

วัตถุประสงค์: *คณะผู้วิจัยได้ศึกษาถึงผลการผ่าตัดรักษาผู้ป่วยโรคถุงน้ำของท่อน้ำดีร่วมในผู้ใหญ่ ภาควิชาศัลยศาสตร์ โรงพยาบาลศิริราช*

วัสดุและวิธีการ: *การศึกษาแบบย้อนหลังตั้งแต่เดือนตุลาคม พ.ศ. 2533 ถึง เดือนมกราคม พ.ศ. 2542*

ผลการศึกษา: *พบว่ามีจำนวนผู้ป่วยทั้งหมด 17 ราย เป็นผู้หญิง 15 ราย ชาย 2 ราย อายุระหว่าง 16-45 ปี มีผู้ป่วยเพียง 2 ราย (11.8%) ที่มีอาการครบ 3 อย่างคือ ตาเหลืองตัวเหลือง ปวดท้องและคลำก้อนที่ท้องได้ ผู้ป่วยมาด้วยอาการของโรคตับอ่อนอักเสบ 3 ราย (17.6%) จากการแบ่งชนิดตาม Todani พบว่าเป็นชนิดที่ 1, 10 ราย (58.8%), ชนิดที่ 4 เอ, 6 ราย (35.3%), และชนิดที่ 5, 1 ราย (5.9%). พบผู้ป่วยมะเร็งถุงน้ำของท่อน้ำดีร่วม 1 ราย (5.9%) ผู้ป่วย 16 ราย (94.1%) ได้รับการผ่าตัดรักษาโดยการตัดเอาถุงน้ำของท่อน้ำดีร่วมออกร่วมกับการนำลำไส้เล็กต่อเข้ากับท่อน้ำดีที่ขั้วตับ (hepaticojejunostomy) ไม่มีผู้ป่วยรายใดเสียชีวิตจากการผ่าตัด ไม่มีภาวะแทรกซ้อนหลังการผ่าตัด ผู้ป่วย 10 รายมีสุขภาพดี, 6 ราย (35.2%) ขาดการติดต่อไป การติดตามผู้ป่วยโดยเฉลี่ยเป็นระยะเวลา 3.2 ปี ผู้ป่วยที่เป็นมะเร็งถุงน้ำของท่อน้ำดีมีชีวิตอยู่ได้นาน 2 ปี ภายหลังการผ่าตัดรักษา*

สรุป: *การผ่าตัดถุงน้ำดีของท่อน้ำดีร่วมออกร่วมกับการนำลำไส้เล็กต่อกับท่อน้ำดีที่ขั้วตับเป็นการรักษาที่เหมาะสมที่สุดสำหรับโรคนี้ในผู้ใหญ่เพื่อป้องกันภาวะทางเดินน้ำดีอักเสบและการกลายเป็นมะเร็งถุงน้ำของท่อน้ำดีร่วม*