Surgical Management of Adult Choledochal Cysts

Thawatchai Akaraviputh, MD*,
Wiroon Boonnuch, MD*, Prasit Watanapa, MD, PhD, FRCS, FACS*,
Narong Lert-akayamanee, MD, FRCS*, Darin Lohsiriwat, MD*

* Department of Surgery, Faculty of Medicine, Siriraj Hospital, Mahidol University

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


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.

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 hepatobiliary disease associated with the choledochal cyst (Table 2). Approximately 47% of the presented patients had biliary lithiasis. 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.
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 chole-
dochal cysts in adults frequently have complex clinical
and pathological characteristics that influence surgical
management(8,9). In adults, cystic disease of the intra-
hepatic ducts and coexisting hepatobiliary conditions
(e.g. biliary lithiasis, pancreatitis and malignant disease)
present problems that require an individualized therapeu-
tic 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)
occurring in 60-70% of patients. A palpable
mass is uncommon (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 cholangio-
graphy 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 chole-
dochal 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). Cystoentero-
scopy is no longer recommended. Not only problems
associated with retention of the cyst, including biliary
stasis, recurrent cholangitis, stone formation, pan-
creatitis 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 1. Frequency of symptoms and signs associated
with choledochal cysts in 17 adults

<table>
<thead>
<tr>
<th>Symptoms and signs</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jaundice</td>
<td>70.6</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>58.8</td>
</tr>
<tr>
<td>Fever</td>
<td>52.9</td>
</tr>
<tr>
<td>Abdominal mass</td>
<td>41.2</td>
</tr>
<tr>
<td>Classical triad</td>
<td>10.8</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Pathologic finding</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biliary lithiasis</td>
<td>8 (47.0%)</td>
</tr>
<tr>
<td>Acute pancreatitis</td>
<td>3 (17.6%)</td>
</tr>
<tr>
<td>Cholangiocarcinoma</td>
<td>1 (5.9%)</td>
</tr>
</tbody>
</table>
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.

References
22. Lai HS, Duh YC, Chen WJ, Chen CC, Hung WT, Lee PH, et al. Manifestations and surgical treat-
การผ่าตัดรักษาผู้ป่วยโรคถุงน้ำของท่อน้ำดีร่วมในผู้ใหญ่

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

วัตถุประสงค์: คณะผู้วิจัยได้ศึกษาถึงผลการผ่าตัดรักษาผู้ป่วยโรคถุงน้ำของท่อน้ำดีร่วมในผู้ใหญ่ ภาควิชาศัลยศาสตร์โรงพยาบาลศิริราช

วัสดุและวิธีการ: การศึกษาแบบย้อนหลังตั้งแต่เดือนตุลาคม พ.ศ. 2533 ถึงเดือนมกราคม 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 ผู้ป่วยมารถอาการของโรคตับอ่อนอักเสบ 3 ราย (17.6%) จากการแบ่งชนิดตาม Todani พบว่าเป็นชนิดที่ 1, 10 ราย (58.8%), ชนิดที่ 4, 6 ราย (35.3%), และชนิดที่ 5, 1 ราย (5.9%). พบผู้ป่วยมะเร็งถุงน้ำของท่อน้ำดีร่วม 1 ราย (5.9%) ผู้ป่วย 16 ราย (94.1%) ได้รับการผ่าตัดรักษาโดยการตัดเอาถุงน้ำของท่อน้ำดีร่วมออกและนำลำไส้เล็กต่อกับท่อน้ำดีที่ขั้วตับ (hepaticojejunostomy) ไม่มีผู้ป่วยรายใดเสียชีวิตจากการผ่าตัด ไม่มีภาวะแทรกซ้อนหลังการผ่าตัด ผู้ป่วย 10 รายมีอายุมากกว่า 35 ปี (35.2%) ขาดการติดต่อไป การติดตามผู้ป่วยโดยเฉลี่ยในระยะเวลา 3.2 ปี ผู้ป่วยที่เป็นมะเร็งถุงน้ำของท่อน้ำดีมีชีวิตอยู่ได้ 50-80 ปี ภายหลังการผ่าตัด

สรุป: การผ่าตัดถุงน้ำดีของท่อน้ำดีร่วมในการรักษาผู้ป่วยจะทำให้เกิดผลดีต่อหอตันน้ำดีที่ขั้วตับเป็นการรักษาที่เหมาะสมที่สุดสำหรับโรคนี้ในผู้ใหญ่เนื่องจากมักเกิดในเด็กและอาการเป็นมะเร็งถุงน้ำของท่อน้ำดีร่วม