Choledochal Cysts: Age of Presentation, Symptoms, and Late Complications Related to Todani’s Classification

Amsterdam, The Netherlands

Purpose: The aim of this study was to compare presentation, complications, diagnosis, and treatment of choledochal cysts in pediatric and adult patients.

Methods: Forty-two patients were analyzed after subdivision into 3 groups: group A, less than 2 years (n = 10); group B, 2 to 16 years (n = 11); group C, greater than 16 years (n = 21).

Results: The cysts were classified as extrahepatic (n = 33), intrahepatic (n = 5), and combined (n = 4). Seventy-six percent of patients presented with abdominal pain, (20 of 21 group C), and 57% with jaundice, (10 of 10 group A). Cholangiocarcinoma occurred in 6 patients, 4 of whom had previously undergone internal drainage procedures. Excision of the extrahepatic cyst was performed in 27 of 37 patients. Five patients, of whom, 4 had cholangiocarcinoma, were beyond curative treatment at the time of diagnosis. Six patients had died at the closure of this study, 5 of them had carcinoma.

Conclusions: 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 extrahepatic cysts. Patients who had undergone internal drainage in the past still should undergo resection of the cyst.

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INDEX WORDS: Choledochal cyst, cholangiocarcinoma.

Choledochal Cyst is a rare congenital dilatation of the bile ducts. The estimated incidence in Western countries varies between 1 in 100,000 and 1 in 150,000. The incidence is higher in Asia and occurs more in women, with a male to female ratio of 1:3 to 4.

The most widely used subdivision of choledochal cysts is Todani’s classification (Fig 1), which is a modification of the Alonso-Lej classification. Type I cysts are the most frequently encountered. The intrahepatic part of type IVa and type V cysts occur diffusely or in a part of the liver. Not shown in this figure is type IVb, featuring multiple extrahepatic dilatations, which is a very uncommon condition.

Choledochal cysts belong to the fibropolycystic disorders. Type V (Caroli’s disease) and probably the intrahepatic part of type IVa cysts are thought to be ductal plate malformations (DPM). The precise etiology of extrahepatic cysts is unclear. Type I cysts are associated with an abnormal arrangement of the pancreatobiliary ducts (APBD), also known as “common channel,” which is seen in up to 92% of the patients with choledochal cysts. A long common channel (>2 cm) can be the cause of a variety of pathologic conditions, such as pancreatitis, stenosis of the papilla of Vater, and choledochal cysts (Fig 2). Although a common channel may occur without a choledochal cyst, an APBD is believed to enhance reflux of pancreatic juice into the bile duct, leading to exposure of the common bile duct wall to pancreatic enzymes and to higher pressures in the choledochal duct finally resulting in cyst formation.

If choledochal cysts are not resected, a high incidence (20% to 30%) of cholangiocarcinoma has been reported, mainly after the second decade of life, which formed the basis of resection as state of the art surgical treatment. This policy is further supported by a study that found increasing rate of premalignant changes in resected cysts with advancing age.

The aim of this study was to evaluate the difference in presentation, complications, diagnosis, and treatment of choledochal cysts in children and adults in 2 academic centers.

MATERIALS AND METHODS

Patients

Between 1972 and 2000, 42 patients with choledochal cysts were treated in the Academic Medical Center (n = 36) or the Academic Hospital of the Vrije Universiteit Medical Center (n = 6), Amsterdam,
the Netherlands. The median age was 16 years (range, 0 to 72 years); the male to female ratio was 1:3.

**Methods**

The study was designed as a case-cohort report. Data were collected using patients’ files, operative reports, and office notes. The following data were collected: presenting symptoms, complications of the disease, diagnostic strategy, and treatment of choledochal cysts. Patients were subdivided into 3 age groups: group A, patients below 2 years of age; group B, patients from 2 to 16 years; and group C, patients older than 16 years. Statistical significance of the results was evaluated using the $\chi^2$ test, with a $P < 0.05$ as the level of significance. Significance is mentioned in text and tables when relevant. In general, the $P$ value was calculated from the value of a parameter in a group or category compared with the expected value based on the total of the parameter in all patients.

**RESULTS**

**Clinical Presentation**

Table 1 shows the presenting symptoms with subdivision into the age groups. Abdominal pain is the most frequent symptom (32 of 42 patients [76%]) at presentation, with a significantly higher incidence in the adult group (20 of 21 patients; $P < 0.05$). Jaundice is the main presenting symptom in children and was seen in all 10 patients below 2 years of age, resulting in a significantly higher incidence ($P < 0.05$) in this age group. Overall, we found cholangitis (cholestasis in combination with fever) in more than one third of the patients (15 of 42 patients (36%)). Pancreatitis was present in only 7 of 42 patients (17%). Although not significant, it was most frequently seen in the age group between 2 and 16 years (4 of 11 patients, 36%).

Table 2 shows the presenting symptoms compared with the type of cyst. Abdominal pain is the main symptom in most types. Jaundice is mainly seen in type I (17 of 30 patients; $P > 0.05$) and IV (4 of 4 patients; $P < 0.05$) cysts, both (partly) extrahepatic. Exclusively intrahepatic cysts (type V) present primarily with both cholangitis (4 of 5 patients; $P < 0.05$) and gall stones (4 of 5 patients; $P < 0.05$).

The classic triad, which consists of abdominal pain, jaundice, and a palpable mass, was seen in only 2 patients.

**Diagnostic Procedures**

The following studies were performed: ultrasound scan (39 of 42 patients [93%]), ERCP (29 of 42 patients
[69%], Fig 2), PTC (3 of 42 patients [7%]), MRCP (1 of 42 patients [2%], Fig 3), abdominal computed tomography scan (10 of 42 patients [24%]), abdominal x-ray (3 of 42 patients [7%]), laparoscopy (2 of 42 patients [5%]), gastroduodenoscopy (2 of 42 patients [5%]), and biopsy (2 of 42 patients [5%] one biopsy of the duodenal papilla at ERCP and one percutaneous of the liver, both positive for [metastatic] cancer). Laboratory studies, like serum amylase, were not systematically performed. ERCP was used less frequently in younger than in older patients: group A, 5 of 10 patients (50%); group B, 5 of 11 patients (45%); group C, 20 of 21 (95%).

**Diagnosis**

Table 3 shows the types of cysts among the study group. The majority of the cysts were extrahepatic, mostly type I cysts: 30 of 42 patients (71%). There were no type IVb cysts. Type V cysts (5 of 42 patients [12%]), were seen only in the adult group. Three of 5 of type V cysts were diffusely intrahepatic, and 2 of 5 were confined to the left lobe of the liver.

Extrahepatic cholangiocarcinoma was encountered in 6 of 42 patients, all group C (5 with type I and I with type IVa cysts) and one in combination with liver metastases. In only 29 of 42 of the patients an ERCP or PTC was available on which the presence of a common channel could be assessed. A common channel was seen in 10 of 29 of these patients. In 1 of these 10 patients a cholangiocarcinoma presented in conjunction with a common channel. All other patients with carcinoma did not have a common channel, although in 4 patients this was difficult to judge because of earlier internal drainage procedures.

**Interval Between First Presentation and (Attempted) Resection**

In 14 of 42 patients (33%) the interval between first presentation and (attempted) resection was more than one year (range, 2 to 33 years; mean, 14 years). In 7 of 14 patients, this was because of a late diagnosis, and in 5 of 14 patients, initially an internal drainage procedure had been performed. In 2 of 14 patients (both group B) there was both a late diagnosis and an earlier internal drainage procedure. A long interval was most frequently seen in group C (9 of 21, 43%).

Four of the patients with carcinoma belonged to the 14 patients with an interval longer than one year. In one patient, the choledochal cyst was first thought to be a pancreatic pseudocyst, which was treated by marsupialisation; 3 patients were treated initially with a cyst-enterostomy.

**Treatment**

Most patients underwent a resection, as shown in Table 4. This consisted of resection of the extrahepatic cyst (type I, II and the extrahepatic part of type IVa) with reconstruction of a biliary digestive anastomosis by a Roux-Y loop in the majority of the patients.

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**Table 2. Presentation and Cyst Type**

<table>
<thead>
<tr>
<th>Cyst Type</th>
<th>I (n = 30)</th>
<th>II (n = 2)</th>
<th>III (n = 1)</th>
<th>IV (n = 4)</th>
<th>V (n = 5)</th>
<th>Total (n = 42)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal pain</td>
<td>21</td>
<td>1</td>
<td>1</td>
<td>4</td>
<td>5</td>
<td>32</td>
</tr>
<tr>
<td>Jaundice</td>
<td>17</td>
<td>1</td>
<td>0</td>
<td>4</td>
<td>2</td>
<td>24</td>
</tr>
<tr>
<td>Vomiting</td>
<td>17</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>19</td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>11</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>14</td>
</tr>
<tr>
<td>Weight loss</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>10</td>
</tr>
<tr>
<td>Palpable mass</td>
<td>7</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Portal hypertension</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Seisps</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Cholangitis</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>2</td>
<td>4*</td>
<td>15</td>
</tr>
<tr>
<td>Pancreatitis</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>Gallstones</td>
<td>4*</td>
<td>0</td>
<td>0</td>
<td>2</td>
<td>4*</td>
<td>10</td>
</tr>
</tbody>
</table>

*P < .05.

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**Table 3. Diagnosis**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Total (n = 42)</th>
<th>Group A (n = 10)</th>
<th>Group B (n = 11)</th>
<th>Group C (n = 21)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>30</td>
<td>9</td>
<td>9</td>
<td>12</td>
</tr>
<tr>
<td>Type II</td>
<td>2</td>
<td>0</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Type III</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Type IVa</td>
<td>4</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Type V</td>
<td>5</td>
<td>0</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Common channel</td>
<td>10</td>
<td>3</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Carcinoma</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>6</td>
</tr>
</tbody>
</table>
patients with a type I cyst initially underwent a cystoduodenostomy, all before 1985. One of these had an excision of the cyst remnant, which occurred uneventfully. In 2 patients (type IVa and type V) a partial liver resection was performed. The other 4 patients with type V cysts were treated conservatively. One patient with a type III cyst was treated by sphincterotomy.

Of 6 patients with carcinoma, 2 patients underwent resection. In one patient, after earlier cystenterostomy, a Whipple operation was performed, but surgical margins contained tumor cells. The other patient underwent a local resection that proved to be radical. In 2 patients (type I and type IV cyst), both after earlier cystenterostomy, the tumor was found to be unresectable at exploration. In the remaining 2 patients the carcinoma was judged unresectable at diagnosis, one of these patients had carcinoma after earlier cystenterostomy.

One infant with a type I cyst died before any treatment could be instituted. Information about the exact cause of death could not be retrieved from available file.

**Procedure-Related Complications**

Procedure-related complications were noted in 7 of 42 (17%) of the patients. Complications consisted of wound infection (2 patients) and wound haematoma (one patient). Two patients had a subhepatic abscess, one patient in combination with cholangitis, the other in combination with a bile leak. In one patient a bowel perforation occurred when a failing abdominal drain was replaced. The last one of these 7 patients had an incisional hernia for which he had to be reoperated.

**Follow-Up**

Overall, 6 patients have died. Five of 6 of these patients had a cholangiocarcinoma. One infant, as stated earlier, died before treatment could be initiated. The remaining patient with carcinoma was still alive with no evidence of disease 8 years after radical excision.

**DISCUSSION**

In this study, half of the choledochal cysts were diagnosed in children ≤ 16 years, which is in accordance with the literature. 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, which was also found in our series. Jaundice is reportedly the main presenting symptom in infants, as in the present series.

It has been suggested that age-related difference in presentation is determined by whether there is reflux of activated pancreatic juice. It was 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 serum amylase was not assessed in our series we could not confirm this notion. Further, we found pancreatitis more often in children from 2 to 16 years than in the other age groups. However, this 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. This may be explained by the localization of the lesion. Extrahepatic 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 mass has proved to be rare. This was confirmed in our series. It may thus be less classic than is usually thought.

In most patients, ultrasound scan is the primary imaging technique for detection of choledochal cysts and usually suffices to establish the diagnosis. A form of cholangiography is mandatory to define the precise anatomy, as was performed in most of our adult patients. Although there is no higher risk of complications,
invasive cholangiography was less frequently performed in children in this study, because neonatal ERCP was not available in the earlier years of this study. More recently, MRCP has become available and, as a noninvasive method, is a promising alternative. CT may be of help in patients with intrahepatic cysts and patients suspected of malignancy. Plain abdominal films, laparoscopy, and gastroduodenoscopy are not used as standard diagnostic tools for choledochal cysts and in this study were mainly performed during workup of the patient when the diagnosis was still unclear. Preoperative biopsies were performed when cancer was suspected, and the lesion was accessible.

Like in most series, the majority of the patients had a type I cyst. Because the necessary information was not available, a further subdivision of type I cysts into cystic and fusiform cysts, as used by other investigators, was not possible in this series. Interestingly, we found more type V cysts than in older studies, which may be caused by more sophisticated imaging techniques, or by section bias in a tertiary referral center. Although all type V cysts were seen in adult patients, there was no statistical significance regarding the incidence of any of the types of cysts in the different age groups.

Cholangiocarcinoma in choledochal cysts has been reported in up to 26% of the patients. However, the overall finding of 14% (non intrahepatic) cholangiocarcinoma is comparable with most recent series (Table 5).

<table>
<thead>
<tr>
<th>Study, Year</th>
<th>No. of Patients</th>
<th>No. of Patients With Malignancies (%)</th>
<th>Malignancies After Internal Drainage (% of All Malignancies)</th>
<th>Age at Presentation of Malignancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jan et al,20 2000</td>
<td>80</td>
<td>8 (10)</td>
<td>3 (38)</td>
<td>50 (32-81)</td>
</tr>
<tr>
<td>Bismuth and Krissat,21 1999</td>
<td>48</td>
<td>6 (13)</td>
<td>2 (33)</td>
<td>39 (17-57)</td>
</tr>
<tr>
<td>Lenriot et al,22 1998</td>
<td>42</td>
<td>5 (12)</td>
<td>3 (60)</td>
<td>39 (29-51)</td>
</tr>
<tr>
<td>Hewitt et al,13 1995</td>
<td>14</td>
<td>2 (14)</td>
<td>0 (0)</td>
<td>46 (30-62)</td>
</tr>
<tr>
<td>Stain et al,23 1995</td>
<td>27</td>
<td>6 (26)</td>
<td>1 (17)</td>
<td>48 (34-60)</td>
</tr>
<tr>
<td>Lipsett et al,24 1994</td>
<td>42</td>
<td>3 (10)</td>
<td>0 (0)</td>
<td>Adults</td>
</tr>
<tr>
<td>Chijiiwa and Koga,25 1993</td>
<td>46</td>
<td>4 (9)</td>
<td>1 (25)</td>
<td>61 (42-71)</td>
</tr>
<tr>
<td>Robertson and Raine,26 1988</td>
<td>13</td>
<td>2 (15)</td>
<td>1 (50)</td>
<td>41 (41-41)</td>
</tr>
<tr>
<td>Todani et al,27 1987</td>
<td>82</td>
<td>8 (10)</td>
<td>3 (38)</td>
<td>?</td>
</tr>
<tr>
<td>Current study 2000</td>
<td>42</td>
<td>6 (14)</td>
<td>4 (67)</td>
<td>36 (20-62)</td>
</tr>
<tr>
<td>Total</td>
<td>437</td>
<td>50 (11)</td>
<td>18 (36)</td>
<td></td>
</tr>
</tbody>
</table>

The treatment of our patients was in accordance with this policy, except for 3 patients with extrabacterial cysts who were treated in the earlier years of this series when drainage was still the established treatment. Currently, excision of extrabacterial cysts after internal drainage is recommended, even in the absence of symptoms. Excision of the cyst remnant has been advised in our 3 patients who previously had cyst-enterostomy. One of them has undergone reoperation already.

Partial liver resection for type V (intrabacterial) cysts was limited to symptomatic patients with unilateral liver involvement, because the risk of cholangiocarcinoma is considered lower in this condition. However, liver transplantation has been suggested for prevention of malignancy in extensive intrabacterial cysts.

**CONCLUSION**

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 extrabacterial part of type IV biliary cysts. Patients who only had internal drainage in the past still should undergo resection of the cyst remnant.

**REFERENCES**

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