Presentation and Clinical Outcomes of Choledochal Cysts in Children and Adults
A Multi-institutional Analysis

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IMPORTANCE Choledochal cysts (CCs) are rare, with risk of infection and cancer.

OBJECTIVE To characterize the natural history, management, and long-term implications of CC disease.

DESIGN, SETTING, AND PARTICIPANTS A total of 394 patients who underwent resection of a CC between January 1, 1972, and April 11, 2014, were identified from an international multi-institutional database. Patients were followed up through September 27, 2014. Clinicopathologic characteristics, operative details, and outcome data were analyzed from May 1, 2014, to October 14, 2014.

INTERVENTION Resection of CC.

MAIN OUTCOMES AND MEASURES Management, morbidity, and overall survival.

RESULTS Among 394 patients, there were 135 children (34.3%) and 318 women (80.7%). Adults were more likely to present with abdominal pain (71.8% vs 40.7%; \( P < .001 \)) and children were more likely to have jaundice (31.9% vs 11.6%; \( P < .001 \)). Preoperative interventions were more commonly performed in adults (64.5% vs 31.1%; \( P < .001 \)), including endoscopic retrograde pancreatography (55.6% vs 27.4%; \( P < .001 \)), percutaneous transhepatic cholangiography (17.4% vs 5.9%; \( P < .001 \)), and endobiliary stenting (18.1% vs 4.4%; \( P < .001 \)). Type I CCs were more often seen in children vs adults (79.7% vs 64.9%; \( P = .003 \)); type IV CCs predominated in the adult population (23.9% vs 12.0%; \( P = .006 \)). Extrahepatic bile duct resection with hepaticoenterostomy was the most frequently performed procedure in both age groups (80.3%). Perioperative morbidity was higher in adults (35.1% vs 16.3%; \( P < .001 \)). On pathologic examination, 10 patients (2.5%) had cholangiocarcinoma. After a median follow-up of 28 months, 5-year overall survival was 95.5%. On follow-up, 13 patients (3.3%), presented with biliary cancer.

CONCLUSIONS AND RELEVANCE Presentation of CC varied between children and adults, and resection was associated with a degree of morbidity. Although concomitant cancer was uncommon, it occurred in 3.0% of the patients. Long-term surveillance is indicated given the possibility of future development of biliary cancer after CC resection.
first reported by Vater and Ezler\(^4\) in 1723, choledochal cysts (CCs) are rare cystic dilations of the biliary tract. Choledochal cysts are diagnosed in children and adults, although most (80%) develop in children younger than 10 years.\(^2\) Choledochal cysts are also 4 times more common in females than males. There is a higher incidence of CCs in Asian populations compared with other ethnicities,\(^3,4\) ranging from 1 in 100 000 Western individuals to 1 in 13 000 people within the Japanese population.\(^4,5\) The exact cause of CCs is unknown; however, in 1969, Babbitt\(^6\) first described the reflux of pancreatic enzymes into the biliary tree through an anomalous pancreaticobiliary duct union (APBDU), thereby leading to biliary dilation. Although rare in the general population, APBDU has been reported\(^7,8\) in more than 80% of children with CCs.

Typically, CCs present in a child as a right upper quadrant mass, abdominal pain, and jaundice.\(^9,10\) However, the expanded use of axial imaging has led to increased detection of CCs in asymptomatic adults. When CCs are discovered, appropriate management has important implications.\(^11\) Biliary cancer is reported\(^12\) in 5% to 10% of patients with CCs. Although malignant CCs are rare in pediatric populations, the incidence of cancer may increase with older age at presentation, and CC-associated cholangiocarcinoma carries a poor prognosis.\(^13,14\) Complete surgical resection of the CC is therefore recommended to reduce the long-term risk of cancer as well as pancreatitis and cholangitis.\(^15,16\)

Published reports\(^17,18\) on CCs are limited by small, single-institution experiences. Moreover, CC disease in non-Asian populations is rare, and little is known regarding presentation, management, and long-term outcomes in Western populations. Therefore, the objective of the present study was to define CC disease in a large Western cohort derived from an international multi-institutional database of 8 major hepatobiliary centers. We sought to evaluate and characterize the presentation, management, and outcomes of CCs in children and adults. In addition, we define and characterize the differences in CC disease among children vs adults. In doing this, we emphasize the management of this complex disease to gain a better understanding of the natural history of CC disease as well as its long-term implications.

**Methods**

**Patient Population**

Using an international, multi-institutional hepatobiliary database from 8 major centers in North America and Europe (The Johns Hopkins Hospital [n = 121], Medical College of Wisconsin [n = 57], University of Virginia [n = 10], Hospital Curry Cabral [n = 23], Ospedale San Raffaele [n = 8], Université Catholique de Louvain [n = 20], Stanford University School of Medicine [n = 89], and Emory University [n = 66]), 394 patients who received treatment for CC disease between 1972 and 2014 were identified. Patients of all ages with a diagnosis of CC were included in the analysis. Individuals 18 years or older were considered adults. Median follow-up was 28 months (children, 32.5 months; adults, 25.4 months), and patients were followed up through September 27, 2014. Data analysis was conducted from May 1, 2014, to October 14, 2014.

Data were collected after study protocol approval by the institutional review boards of each participating institution. The need for informed consent was waived owing to the observational nature of the study and minimal risk to participants.

**Data Collection**

Standard demographic variables, including age, sex, and race/ethnicity, as well as data on symptoms, medical history, and social history, were collected on review of medical records. Baseline laboratory values, including liver function test results and serum tumor marker levels, were also recorded. Diagnostic assessments were characterized, and details on all imaging, including computed tomography, ultrasonography, magnetic resonance imaging, magnetic resonance cholangiopancreatography, and hepatobiliary iminodiacetic acid scan, were obtained.

Data on operative treatment were collected. Resections were classified according to the following categories: internal drainage; cyst excision with extrahaepatic bile duct resection primary bile duct closure; cyst excision with extrahaepatic bile duct resection and hepaticoenterostomy; cyst excision with extrahaepatic bile duct resection, hepaticoenterostomy, and liver resection; Whipple procedure; liver transplant; and cholecystectomy. When performed, hepaticoenterostomy was further characterized as hepaticocholedochostomy, hepaticojejunostomy, or Roux-en-Y hepaticojejunostomy. Classification of CCs was performed using the Todani modification\(^17\) of the Alonso-Lej et al\(^18\) classification.

Postoperative complications were categorized until 30 days postoperatively and graded according to the Clavien-Dindo classification.\(^19\) Major complications corresponded to a Clavien-Dindo score of grade III or higher, which represents complications requiring surgical, endoscopic, or radiologic intervention; are life-threatening; or result in death. System-specific 30-day postoperative complications were assessed, including wound, hepatobiliary, and gastrointestinal tract complications. Perioperative mortality was calculated on the basis of the number of patients who died within 30 days after surgery. Long-term outcome data were obtained, including readmission rates, reoperation rates, need for additional biliary procedures, and overall survival at last follow-up.

**Statistical Analysis**

Categorical variables were reported as whole numbers and percentages. Continuous variables were presented as median with interquartile range or mean (SD). Baseline characteristics were compared according to the age group (children vs adults). Comparative analysis of categorical values was performed using χ\(^2\) or Fisher exact tests, and the Wilcoxon rank sum test was used for continuous variables. Overall survival was estimated using the Kaplan-Meier method calculated from the date of surgery to the date of the last follow-up or death, and differences in survival were examined using the log-rank test. A 2-tailed value of P < .05 was considered significant. All analyses were carried out with Stata, version 12.1 (StataCorp LP).
Results

Patient Demographics and Disease Presentation
Preoperative characteristics and disease presentation of the 394 patients are presented in eTable 1 in the Supplement. Most patients were adults (259 [65.7%] vs 135 [34.3%] children) and female (318 [80.7%]). The mean (SD) age at diagnosis for children and adults was 5.2 (4.9) and 45.0 (15.2) years, respectively. Race/ethnicity data were available on 389 patients; of these, most (342 [87.9%]) were non-Asian (white, 239 [61.4%]; black, 51 [13.1%]; Asian, 37 [9.1%]; and other, 52 [13.4%]). As expected, adults were more likely to present with at least 1 comorbidity compared with children (35.1% vs 1.5%; \(P < .001\)). Children were more likely to have congenital anomalies compared with adults (20 [14.8%] vs 10 [3.9%]; \(P < .001\)), including ventricular septal defect (3 patients [0.8%]), pancreatic divisum (6 [0.3%]), and biliary atresia (9 [0.3%]).

Abdominal pain was the most common presenting symptom (241 patients [61.2%]) followed by pancreatitis (74 [18.8%]) and jaundice (73 [18.5%]). Sixty-one patients (15.5%) were asymptomatic. None of the patients presented with the classic triad of abdominal pain, jaundice, and right upper quadrant mass. Adults were more likely to present with abdominal pain (71.8% vs 40.7%; \(P < .001\)) and children were more likely to have jaundice (31.9% vs 11.6%; \(P < .001\)). In addition, adults were more likely than children to have undergone previous biliary surgery, including cholecystectomy (32.4% vs 5.9%; \(P < .001\)), common bile duct resection (4.2% vs 0%; \(P = .01\)).

Diagnostic Evaluation and Disease Classification
Diagnostic assessment and interventions are described in eTable 2 in the Supplement. Imaging modalities included ultrasonography (61.9%), computed tomography (57.4%), magnetic resonance imaging (42.6%), magnetic resonance cholangiopancreatography (38.1%), and hepatobiliary isomiaid acid scan (6.6%). Children were more likely to have undergone ultrasonography (83% vs 51%; \(P < .001\)); adults were approximately 2-fold more likely to have undergone computed tomography (69.1% vs 34.8%; \(P < .001\)). Preoperative interventions were more common in adults (64.5% vs 31.1%; \(P < .001\)) and included endoscopic retrograde cholangiopancreatography (55.6% vs 27.4%; \(P < .001\)), percutaneous transhepatic cholangiography (17.4% vs 5.9%; \(P < .001\)), and endoscopic ultrasonography (0% vs 3.5%; \(P = .02\)). In addition, adults were more likely to have biliary stents preoperatively, including endobiliary stents (18.1% vs 4.4%; \(P < .001\)) and transhepatic stents (16.2% vs 2.2%; \(P < .001\)). Abnormal pancreaticobiliary ductal union was identified in 48 patients (12.2%), including 17 children and 31 adults. Forty-four patients (11.2%) received a diagnosis of a long common channel, with a mean length of 18.5 (6.7) mm. Thirty-five patients (8.8%) had both an APBDU and a long common channel (15 children and 20 adults).

Preoperative biopsies were rare in both groups (5.2% children, 8.5% adults). However, when biopsies were used, biliary brushings were most frequently performed (16 [55.2%]). Biliary clamshell (2 [6.9%]), percutaneous biopsy (6 [20.7%]), duodenal biopsy (2 [6.9%]), fine-needle aspiration (2 [6.9%]), and liver biopsy (2 [6.9%]) were also used. One patient underwent both a duodenal biopsy and biliary brushing. Of the 29 biopsy specimens, 65.5% showed benign biliary epithelium or were nondiagnostic. Five other biopsy specimens (17.2%) showed atypia or inflammatory changes. Finally, 4 biopsy specimens (1 biliary brushing, 1 percutaneous biopsy, and 2 fine-needle aspirations) demonstrated cancer.

Most patients presented with laboratory values within the reference ranges, including tumor markers CA19-9 (median, 13.75 ng/dL) and carcinoembryonic antigen (median, 1.5 ng/dL [conversion to micrograms per liter is 1:1]) as well as total bilirubin (median, 0.65 mg/dL [to convert to micromoles per liter, multiply by 17.104]), aspartate aminotransferase (median, 37 U/L [to convert to micromoles per liter, multiply by 1.136]), and prothrombin time (median, 12.3 seconds). Twelve patients were evaluated for biliary amylase levels (median biliary amylase, 38 150 U/L), of which one-third had an APBDU and 2 patients had a long common channel. Aspiration of cyst fluid was performed in 15 patients (3.8%). Median cyst CA19-9 and carcinoembryonic antigen levels were 93 108.5 ng/dL and 1.35 ng/dL, respectively.

The CC subtypes according to the Todani modification of the Alonso-Lej et al classification within this cohort are described in Figure 1. Most patients presented with a type I CC (70.1%). There were similar numbers of types II, III, and V CCs within each population (all \(P > .05\)). Type I CCs were more commonly seen in children compared with adults (79.7% vs 64.9%; \(P = .003\)); type IV CCs predominated in the adult population (23.9% vs 12.0%; \(P = .006\)).

Operative Therapies
Data for CC resection, perioperative outcomes, and long-term outcomes were available for 390 patients (98.9% of the cohort) (eTable 3 in the Supplement). Most patients (313 [80.3%]) underwent cyst excision with extrahepatic biliary resection and hepaticoenterostomy. Few patients (20 [5.1%]) underwent cyst excision with extrahepatic biliary resection with primary bile duct closure. Adults were more likely to undergo liver resection or pancreaticoduodenectomy (\(P < .05\)).

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure1.png}
\caption{Type of Choledochal Cysts (CCs) in Adult and Pediatric Populations in 394 Patients}
\end{figure}
indicated, Roux-en-Y hepaticojejunostomy was the most commonly used technique of bilioenteric reconstruction in both cohorts (323 of 344 patients with data available [93.9%]). Nine patients (2.3%) underwent noncurative resections.

The mean (SD) CC size was 3.6 (2.9) cm. Intraoperative frozen section was performed in 131 patients (33.2%), and cancer was identified in 7 (5.3%) of these analyses. Cancer was confirmed on the final pathology laboratory report in all 7 of these patients. Five other patients (3.8%) also had cancer identified on the final pathology report. Of these 5 patients, 3 had negative frozen section analyses. Operative time, length of stay, and intensive care unit stay were equivalent between the 2 age groups. However, adults were more likely to have higher mean (SD) blood loss (328.6 [344.2] mL vs 81.6 [127.9] mL; \( P < .001 \)) and received more packed red blood cell transfusions (2 U vs 1 U; \( P = .04 \)).

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### Postoperative Outcomes, Complications, and Hospital Readmission

Thirty-day postoperative complications are summarized in Table 1. The incidence of major morbidity, defined as Clavien-Dindo classification of 3 or higher, was similar in both groups (adults, 56.1%; children, 54.2%; \( P = .86 \)). Overall perioperative morbidity was higher in adults (35.1% vs 16.3%; \( P < .001 \)). However, adults were more likely to have wound, hepatobiliary, or gastrointestinal complications (\( P < .05 \)). Specifically, seromas (3.1% vs 0%; \( P = .04 \)), wound infections (9.7% vs 1.5%; \( P < .001 \)), and perihepatic abscesses (7.7% vs 0%; \( P < .001 \)) were more common in adults. However, children had more anastomotic leaks (3.0% vs 0%; \( P = .01 \)) and gastrointestinal tract perforations (3.0% vs 0%; \( P = .01 \)). There was no significant difference in hemorrhagic/thrombotic, renal/urologic, cardiovascular, and pulmonary complications between the 2 groups. Overall 30-day mortality was 5.6% (adults, 6.9%; children, 3.0%; \( P = .10 \)).

A total of 28.7% of the patients were readmitted to the hospital secondary to their initial operative resection. Sixty-four patients (58.2% of all readmissions) were readmitted less than 6 months after their operation (Table 2). Both groups had a similar incidence of multiple readmissions (9.6% children and 9.7% adults; \( P = .99 \)). Adults were more likely to undergo postoperative biliary procedures (5.2% vs 22.4%; \( P < .001 \)). In total, 28 patients (7.1%) underwent more than 1 biliary procedure, with adults being more likely to undergo multiple proce-
dures after resection than children (9.3% vs 3.0%; P = .02). Fifty patients (33 adults and 17 children) underwent reoperation related to their initial resection. Forty-eight percent of these reoperations occurred within 6 months of the initial procedure; 28.0% occurred more than 2 years after the resection.

Disease recurrence was noted in 29 patients; 82.8% of the recurrences were symptomatic. Patients with recurrence most commonly presented with cholangitis (8 [29.6%]), abdominal pain (6 [22.2%]), and pancreatitis (4 [14.8%]). Adults with recurrence were more likely to develop a biliary cancer after CC resection (12 [63.2%] vs 1 [10.0%; P = .01]; children were more likely to have recurrent CCs (9 [90.0%] vs 7 [36.8%]; P = .01). At follow-up, 13 patients (44.8%) had cancer; median overall survival was 3.5 years. These patients initially had presented with type I (n = 6), type IV (n = 6), and type V (n = 1) CCs.

### Prognosis

Overall, patients in both cohorts did well, with a 95.5% five-year survival (adults, 94.6%; children, 97.2%; P = .09) (Figure 2A). As expected, patients with a malignant CC at the time of resection had a worse outcome than patients without cancer (P < .001) (Figure 2B). Twelve patients had malignant disease at the time of their CC resection (Table 3). Ten of these patients were adults (n = 10), and 1 of these 10 adults had an APBDU. The adults ranged in age from 24 to 81 years; the children were aged 3 and 4 years. Both children had embryonal rhabdomyosarcomas. All patients with malignant CC underwent cyst excision with Roux-en-Y hepaticojejunostomy. Four adults underwent an additional liver resection (1 right hepatectomy, 1 extended right hepatectomy, and 2 left hepatectomies). Cancer was confined to the bile duct in 9 patients, the pancreas in 1 patient, and the gallbladder in 1 patient. The lo-
cation of 1 malignant CC was unspecified. Among patients who presented with cancer, the CC type included type I (n = 6), type IV (n = 5), and type III (n = 1). At last follow-up, 5 of the 10 adults with cancer at the time of resection had died and both children with cancer were alive with no evidence of disease.

Discussion

Choledochal cysts are diagnosed in patients of all ages and harbor a risk of becoming malignant as well as producing serious complications. To our knowledge, available data on these rare lesions are limited to small single-center series. In the present study, we report on the experience of 8 major hepatobiliary centers from the Western hemisphere in the treatment of both pediatric and adult CC disease. More than 80% of the patients were non-Asian, making it, to our knowledge, the largest series of Western CC disease reported in the literature. Our study focused on the natural history of CC disease and its implications for management. The report is important because we examined a large multi-institutional cohort of patients and identified characteristic presentations of children and adults. Moreover, we identified population-specific complications of CC disease as well as defined the long-term implications and complications of CCs.

Several series of adults with CC in Asian populations have suggested an increased incidence of adult CC disease.\textsuperscript{3,11,13,14,16,20,21} The present analysis also demonstrated an adult predominance. Although this finding could be secondary to referral bias within participating institutions, these findings suggest that adult CC disease may be increasing in incidence and therefore need to be considered in adults with abdominal pain and an enlarged ductal system. It is unclear whether a CC has a slow progression, remains undetected for some time, or develops late in life. In the present study, we noted that pediatric and adult CC disease presentation and management varied, although several important similarities were apparent. Specifically, we confirmed a female predominance in CC disease with an approximately 1:4 male to female ratio in both children and adults.\textsuperscript{15,13,14,16,21,22} In addition, abdominal pain was the most common presenting symptom in both

![Figure 2. Select Survival Analyses](image-url)

**A** Age survival analysis

![Time, mo](time-axis) ![Proportion of Patients Alive](proportion-axis)

Log-rank: P = .09

**B** CCA survival analysis

![Time, mo](time-axis) ![Proportion of Patients Alive](proportion-axis)

Log-rank: P < .001

Table 3. Concurrent Choledochal Cyst and Biliary Cancer

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Cancer, No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>APBDU</td>
<td></td>
</tr>
<tr>
<td>Present</td>
<td>0</td>
</tr>
<tr>
<td>Absent</td>
<td>2 (100.0)</td>
</tr>
<tr>
<td>Operation</td>
<td></td>
</tr>
<tr>
<td>Cyst excision</td>
<td></td>
</tr>
<tr>
<td>Extrahepatic biliary resection, bile duct closure</td>
<td>0</td>
</tr>
<tr>
<td>Extrahepatic biliary resection, HE</td>
<td>2 (100.0)</td>
</tr>
<tr>
<td>Extrahepatic biliary resection, HE, liver resection</td>
<td>0</td>
</tr>
<tr>
<td>Location of cancer (n = 11)</td>
<td></td>
</tr>
<tr>
<td>Bile duct</td>
<td>2 (100.0)</td>
</tr>
<tr>
<td>Gallbladder</td>
<td>0</td>
</tr>
<tr>
<td>Pancreas</td>
<td>0</td>
</tr>
<tr>
<td>Type of cancer (n = 11)</td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td>0</td>
</tr>
<tr>
<td>Anaplastic carcinoma</td>
<td>0</td>
</tr>
<tr>
<td>Embryonal rhabdomyosarcoma</td>
<td>2 (100.0)</td>
</tr>
<tr>
<td>Undifferentiated carcinoma</td>
<td>0</td>
</tr>
<tr>
<td>Palliative procedure</td>
<td>0</td>
</tr>
<tr>
<td>Survival, median, mo</td>
<td>NA</td>
</tr>
</tbody>
</table>

Abbreviations: APBDU, anomalous pancreaticobiliary duct union; HE, hepaticoenterostomy; NA, not available.
groups. However, children were more likely to develop jaun-
dice and biliary perforation.14-16 In contrast, adults more com-
monly had symptomatic cholelithiasis and cholangitis, al-
though the latter finding could be secondary to their increased
likelihood of a preoperative biliary intervention. Nearly one-
third of the adults in our series had undergone previous bili-
ary surgery and/or preoperative biliary stenting, but previous
biliary interventions were rare in the pediatric population.

In examining our findings in light of previous data on Asian
populations, we noted several similarities between CC dis-
eease in Eastern and Western populations. Shah and collea-
gues16 performed a single-center review of 92 patients (19 children,
73 adults) in India. Similar to our findings, Shah et al16 re-
ported a significantly higher rate of previous biliary surgery
in adults, and abdominal pain was the most common present-
ing symptom. Moreover, these authors16 and others3,11,13 noted
a higher incidence of type I CC in children and type IV CC in
adults, finding consistent with our results. Many reports on
Eastern populations have described the classic triad of jaun-
dice, abdominal mass, and pain in nearly one-third of af-
fected children.3,14 In our cohort, this classic triad was not seen.
It is possible that the lack of the classic triad in Western popu-
lations may be secondary to socioeconomic factors and acces-
sibility to health care, such as the ubiquitous use of cross-
sectional imaging as well as Western patients being more likely
to have access to and seek medical care at the onset of symp-
toms. Regardless of these differences, the overall presenta-
tion of CC in all age groups seems similar between Eastern and
Western societies.

The breakdown of CC disease between adult and pediat-
ratic patients in our series is consistent with that of previous
publications.11-13 Type I CCs were predominant in the pediat-
ric population; adults had significantly more type IV CCs. Al-
though surgery was generally well tolerated, nearly one-
third of the patients required readmission after operative
resection, with most readmission occurring within the first 6
postoperative months. Despite acceptable short-term morbid-
ity and mortality, patients required continued observation and
interventions as a consequence of CC resection. Specifically,
although uncommon, a small subset of patients (3.3%) devel-
oped biliary cancer after surgical excision. More than 40% of
patients with recurrence developed CC-associated cancer, and
these patients had poor long-term survival. Similar to previ-
ous reports,14,23,24 development of biliary cancer after CC re-
section was predominantly seen in patients with type I and IV
cysts. Although the risk of biliary cancer is reduced with op-
erative resection of CCs,15 the risk of malignancy does not re-
turn to baseline population levels.12 Whether this increased
malignancy risk is related to incomplete excision or inher-
ently greater biliary cancer susceptibility remains unclear.
However, data from the present study serve to emphasize that close
surveillance and a higher degree of clinical suspicion is war-
ranted in patients with CCs, even after surgical excision.

This study had several limitations. The retrospective de-
velopment increased the likelihood for information bias and selec-
tion bias. It is unlikely, however, that this bias was systematic
and therefore would be unlikely to affect the main findings.
The multi-institutional design of our study provides a wide-
ranging report on the Western experience with CCs. The long
time span of the study may also be a possible limitation. The
effect of advances in technology, improved detection, and sur-
gical advances, as well as changes in the CC system, could not
be assessed.

Conclusions

Biliary cancer was rare in pediatric CC disease but increased
with the patient’s age at presentation. Operative resection of CCs
is mandatory. Although concomitant cancer was uncommon, it oc-
curred in 3.0% of the patients. Resection of CCs with adherence
to strict oncologic principles is warranted. The data strongly
suggest that long-term surveillance is indicated given the possibil-
ity of future development of biliary cancer after CC resection.

REFERENCES

Lingering Questions on Choledochal Cysts

David Linehan, MD; Eva Galka, MD

Soares and colleagues\(^1\) are to be commended for assembling a large, multicenter analysis of children and adults with resected choledochal cysts (CCs). Although the study provides useful information, it has several limitations including the retrospective design, the long time span reviewed, and the fact that most patients analyzed had symptomatic disease. As is the case in most surgical series, the risk of associated cancer may be overstated since the true denominator of patients with asymptomatic disease is unknown. Furthermore, although surgical intervention in symptomatic patients is straightforward, treatment recommendations for the asymptomatic patient are more controversial. The series presented by Soares et al\(^1\) offers little guidance for this more vexing clinical problem.

Guidelines for treatment of asymptomatic patients are important, especially with the significant surgical morbidity and mortality found in the series. The major morbidity rate was greater than 50%; nearly one-third of the patients required re-admission, often undergoing multiple biliary procedures, and the 30-day mortality rate was 5.6%. This is a high price to pay for a prophylactic operation in an asymptomatic patient, especially if the risk of subsequent cancer is not known. Hence, if one is considering surgery in an asymptomatic patient, risk-benefit analysis is paramount. Fortunately, most single-center series and high-volume centers show\(^2-4\) lower morbidity and mortality rates than those reported by Soares et al.\(^1\)

The most striking finding of this study is that recurrent CCs will be malignant in 3.0% of patients who have undergone resection. We have not routinely performed postoperative surveillance imaging in the absence of symptoms or abnormal liver function test results, but Soares et al\(^1\) suggest that perhaps we should. Are some patients at higher risk? It would certainly be helpful to identify a subgroup in which this extensive surveillance is warranted. Unfortunately, this retrospective study offers us little guidance, but we look forward to future work in this endeavor.

REFERENCES