Long-term outcomes after hepaticojejunostomy for choledochal cyst: a 10- to 27-year follow-up

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Abstract

Introduction: Choledochal cyst (CC) is closely associated with anomalous arrangement of the pancreaticobiliary duct, which is considered a high-risk factor for biliary tract malignancy. Early diagnosis and early treatment for CC could lead to a good prognosis. This study investigated late complications and long-term outcomes after surgery for CC.

Patients and Methods: Fifty-six patients with CC and over 10 years of postoperative follow-up were analyzed retrospectively. All patients had undergone total resection of the extrahepatic bile duct and hepaticojejunostomy.

Results: Six patients showed liver dysfunction manifested in the first 10 years after surgery, but all returned to normal thereafter. Dilatation of intrahepatic bile ducts persisted in 6 postoperatively, and in 3, this was still apparent more than 10 years after. Recurrent abdominal pain was encountered in 3, 1 had pancreas divisum with a pancreatic stone, and 1 had adhesive small bowel obstruction. Two patients developed biliary tract malignancy. A 14-year-old girl died of recurrent common bile duct cancer 2 years after the initial resection of CC with adenocarcinoma. A 26-year-old man with repeated cholangitis owing to multiple intrahepatic bile stones developed cholangiocarcinoma 26 years after the initial resection of CC. Event-free survival rate and overall survival rate were 89% (50/56) and 96% (54/56), respectively.

Conclusions: Choledochal cyst generally has an excellent prognosis with early total resection and reconstruction. Long-term surveillance for the development of malignancy is still essential, especially if there is ongoing dilatation of the intrahepatic bile duct or biliary stones.

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Key words:
Choledochal cyst; Late complications; Long-term outcome

Choledochal cyst (CC) is a congenital dilatation of the biliary tree, which is closely associated with an anomalous arrangement of the pancreaticobiliary duct (AAPBD). In 1969, Babbitt [1] reported an AAPBD in 3 children with CCs, in which he proposed that was was the cause of the dilatation. The combination is widely held to be a risk factor for biliary tract malignancy [2], and several investigators have reported cholangiocarcinoma developing in patients younger than 20 years [3-9] and others have shown carcinoma arising from the intrahepatic bile duct or residual common bile duct following resection of their CC [10,11]. Nonetheless, most patients with CC have a good prognosis when diagnosed and treated early. In this review, we investigate late complications and long-
term outcomes of a series of patients who are now older than 10 years after their definitive surgery.

1. Patients and methods

Between 1981 and 2008, 86 children with CC were treated in our institution. Of these 86 patients, data from 56 patients (≥10-year follow-up) were retrospectively reviewed with respect to development of late complications. These were defined as a complication occurring at 3 months or more postoperatively. We also subdivided complications into those occurring at less than 10 years and those occurring more than 10 years.

2. Results

There were 56 (37 female) subjects with a mean age of 21 (range, 11-33) years. Mean age at original operation was 4.2 years (2 months to 13 years). Mean follow-up was 17 (10-27) years. All patients had undergone total resection of the extrahepatic bile duct and hepaticojejunostomy (Roux-en-Y). Using Todani’s classification [12], there were 25 type I (cystic or fusiform dilatation of the extrahepatic bile duct alone) and 14 as type IV-A (cystic or fusiform dilatation of both intrahepatic and extrahepatic bile duct). Of the remaining 17, 10 showed nondilatation of the common bile duct (so-called forme fruste choledochal malformation) and 7 were not able to be classified. Forty-six of 56 patients demonstrated AAPBD on endoscopic retrograde cholangiopancreatography.

2.1. Late complications within 10 years postoperatively

Late complications within 10 years postoperatively are shown in Table 1. Six of 56 patients showed biochemical liver dysfunction persisting for several months to several years. None of the patients were jaundiced postoperatively.

Six patients had persistent dilatation of intrahepatic bile duct after surgery. All 6 patients had shown dilatation of the intrahepatic bile duct preoperatively (ie, Todani IV-A type). Three patients developed recurrent abdominal pain. However, there were no abnormal findings on periodical ultrasound or computed tomographic scan.

<table>
<thead>
<tr>
<th>Late complications (&lt;10 years postoperatively)</th>
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<tbody>
<tr>
<td>Late complications</td>
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<tr>
<td>Biochemical liver dysfunction</td>
</tr>
<tr>
<td>Persistent dilatation of intrahepatic bile duct</td>
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<tr>
<td>Recurrent abdominal pain</td>
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<td>Recurrence of common bile duct adenocarcinoma</td>
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2.2. Late complications over 10 years postoperatively

Illustrated in Table 2 is the biochemical data from 6 patients, who had shown liver dysfunction previously now normalized.

Dilatation of the intrahepatic bile duct persisted in 3 patients. One patient developed multiple intrahepatic bile duct stones and underwent repeated lithotripsy with a percutaneous transhepatic cholangioscope. Thereafter, he developed cholangiocarcinoma some 26 years after his initial surgery (this has been reported previously in greater detail [11]).

Repeated abdominal pain persisted in 2 patients; one was diagnosed as having adhesive obstruction of the small intestine in which surgical intervention was later required, and the other was diagnosed as having pancreas divisum with pancreatic stone, in which papilloplasty was later performed with good effect.

2.3. Biliary tract malignancy

Two of our series developed biliary tract malignancy. A 12-year-old girl had adenocarcinoma in an existing CC evident at initial surgery. This was resected, and she received chemotherapy postoperatively. However, she later died of recurrence at 2 years after the initial surgery. A 26-year-old man with repeated cholangitis and febrile episode owing to multiple intrahepatic bile stones developed cholangiocarcinoma 26 years after the initial total resection of CC. He underwent a left lobectomy with reenconstruction by right hepaticojejunostomy. However, he died of carcinomatous peritonitis with recurrence of cholangiocarcinoma at 1 year. None of the patients in our series developed pancreatic carcinoma.

All other patients are alive, with a symptom-free survival and overall survival rates of 89% (50/56) and 96% (54/56), respectively.

Table 2 Late complications (>10 years postoperatively)

<table>
<thead>
<tr>
<th>Late complications</th>
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<tbody>
<tr>
<td>Biochemical liver dysfunction</td>
<td>0</td>
</tr>
<tr>
<td>Persistent dilatation of intrahepatic bile duct</td>
<td>3</td>
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<tr>
<td>Recurrent abdominal pain</td>
<td>2</td>
</tr>
<tr>
<td>Adhesive small bowel obstruction</td>
<td>1</td>
</tr>
<tr>
<td>Pancreas divisum with pancreatic stones</td>
<td>1</td>
</tr>
<tr>
<td>Repeated cholangitis</td>
<td>1</td>
</tr>
<tr>
<td>Intrahepatic lithiasis and cholangiocarcinoma</td>
<td>1</td>
</tr>
</tbody>
</table>

* Same patient.

A 14-year-old girl developed recurrence of common bile duct cancer 2 years after the initial resection of CC with adenocarcinoma (this has been reported in greater detail previously [3]).
3. Discussion

Choledochal cyst is characterized as a congenital dilatation of the extrahepatic and/or intrahepatic bile duct and is usually accompanied with an AAPBD. Early diagnosis and surgical treatment provide a good prognosis with few complications in most. Currently, total resection of the extrahepatic bile duct and hepaticojejunostomy is the accepted form of treatment. Previous complications such as cholangitis, pancreatitis, intrahepatic lithiasis, biliary tract malignancy, and pancreatic carcinoma have been recognized [12]. In our series, most dilatation of the intrahepatic bile duct resolved after 10 years. As Todani et al [13] emphasized, a wide anastomosis aims to prevent persistent cholestasis; however, congenital stenosis of the intrahepatic bile duct is occasionally present, which might require special attention and surgical treatment at the initial operation [14]. Postoperative stenosis and repeated cholangitis of the intrahepatic bile duct lead to bile stasis and intrahepatic lithiasis [13-15] and may be a high-risk factor for carcinogenesis. Certainly, this seemed to be the mechanism in our case of carcinoma 26 years after the initial surgery.

There have been previous reports of cholangiocarcinoma developing in children with CC with AAPBD [3-9], and biliary epithelial hyperplasia has been observed in some [2]. This hyperplastic change seems to be triggered by chronic inflammation caused by the reflux of pancreatic juice leading perhaps to metaplasia [16]. Early surgical biliary diversion tract seems reasonable even for this with minimal biliary dilatation [2,17,18].

Choledochal cyst generally shows an excellent long-term prognosis with early resectional surgery. Nevertheless, biliary tract malignancy may still occur, although in our case, there were obvious ongoing problems. Long-term surveillance remains essential in patients, especially if there was dilatation of the intrahepatic bile duct or biliary stones.

References