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Reoperation After Cyst Excision with Hepaticojejunostomy for Choledochal Cysts: Our Experience in 18 Cases

Authors' Contribution:
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Background: Complete cyst excision with biliary reconstruction is the treatment of choice for choledochal cyst (CC). The aim of this article is to review our experience in patients who underwent reoperation between January 1995 and December 2014.


Material/Methods: The records of 18 patients (female/male, 15/3) were retrospectively analyzed including age, sex, cyst type, initial procedure, lab and imaging findings, indications for reoperation, intraoperative findings, and results of reoperation. The median follow-up period was 6 years.

Results: The rate of reoperation in this study was about 6.8%. Eighteen patients (7 type Ia, 2Ic, 9 IV-A) developed severe postoperative complications and required surgical intervention. The median time interval from primary surgery to reoperation was 19.5 months (range, 3 days to 8 years). Two early complications required surgery due to anastomotic bile leakage and intussusception. Sixteen late complications occurred, including 3 intrahepatic bile duct stenosis with calculi, 5 anastomotic strictures with/without stones, 4 intrapancreatic cyst remnants, 3 adhesive bowel obstructions, and 1 internal hernia. For patients with persistent dilatation of the intrahepatic bile duct or anastomotic stricture, removal of stones and revision of hepaticojejunostomy were performed, with additional hepatic ductoplasty when necessary. Radical excision of the dilated cystic remnant in the head of pancreas was performed in 4 patients, with 1 needing additional pancreaticojejunostomy procedure. No deaths occurred. Sixteen patients recovered uneventfully after reoperation, and 2 wound infections developed.

Conclusions: A wide hepaticojejunostomy with/without ductoplasty is essential to prevent cholangitis, anastomotic stricture, and calculi formation. Complete cyst excision, including the intrapancreatic portion, should be meticulously pursued.

MeSH Keywords: **Choledochal Cyst • Postoperative Complications • Reoperation**

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Background

Choledochal cyst (CC) is a congenital dilatation of the extrahepatic and/or intrahepatic bile duct, and is commonly associated with pancreaticobiliary maljunction. Cyst excision combined with hepaticoenterostomy (hepaticoduodenostomy or hepaticojejunostomy) has become the standard procedure for children with choledochal cyst [1–3]. The majority (over 85–90%) have satisfactory surgical outcomes. However, 6–10% of patients need reoperative surgery due to severe postoperative complications, including bile leakage, anastomotic stricture, intrahepatic cholelithiasis, recurrent pancreatitis, adhesive bowel obstruction, and malignancy [2,4,5]. The aim of this study was to review our experience with 18 patients who underwent reoperation over the past 20 years.

Material and Methods

Between January 1995 and December 2014, 275 children with CC were diagnosed and surgically treated in 2 tertiary hospitals (Children's Hospital of Shanghai Jiao Tong University and Children's Hospital of Fudan University). Cysts were categorized according to Todani's classification: type Ia (n=176), type Ic (n=64), type III (n=1), type IV-A (n=33), and type V (n=1). The study was approved by the Institutional Review Board and the Ethics Board of Children's Hospital of Shanghai Jiao Tong University and Children's Hospital of Fudan University. Written informed consents were obtained from the parents. Regular follow-up was carried out by phone or interview in our clinics by an experienced surgeon (Y. Wu). In this study, procedures performed after primary surgery, such as percutaneous peritoneal drainage (n=2) and gastroscopy (n=1), were excluded. The medical records of 18 patients who underwent reoperation were retrospectively analyzed, including sex, cyst type, initial procedure, age at time of primary surgery and reoperation, clinical presentations, lab and imaging findings, indications for reoperation, intraoperative findings, and type and results of reoperation. Data were recorded as numeric or nominative variables. SPSS 17.0 software package (SPSS Inc, Chicago, IL) was used for statistical analysis. Differences in frequency distribution were compared using the χ^2 test or Fisher's exact test. Other results are expressed as mean \pm standard deviation (SD). A P-value of less than 0.05 was considered significant ($\alpha=0.05$, two-tailed).

Results

Initial operation was performed by open or laparoscopic cyst excision and end-to-side hepaticojejunostomy using a 30- to 40-cm Roux limb without additional anti-reflux procedure. Enteric drainage (Roux-en-Y cystojejunostomy) was abandoned after

January 1995. Twelve children (8 type Ia, 4 type Ic) were lost to follow-up. Of the remaining 263 patients, 43 children developed early (less than 30 days, n=18) or late (greater than 30 days, n=25) postoperative complications. Eighteen (6.8%, 15 females) children required surgical intervention (Table 1). They underwent definitive primary surgery (16 open, 2 laparoscopic) at the median age of 6 years (range, 11 months to 11 years). Diagnostic workup before reoperation was carried out, including thorough history-taking, meticulous clinical examination, lab and radiological studies (abdominal X-ray film, ultrasonography, computed tomography (CT), and magnetic resonance cholangiopancreatography (MRCP). The median time interval from primary surgery to reoperation was 19.5 months (range, 3 days to 8 years). Abdominal pain, fever, jaundice, and vomiting were seen in the great majority of cases. There was no significant difference in reoperation rate between open and laparoscopic procedure (7.5%, 16/212; 3.9%, 2/51; P=0.539).

Two early postoperative complications required surgery due to anastomotic bile leakage (1 case) and intussusception (1 case) (Table 2). A 38-month-old girl presented with fever and abdominal pain 6 days after the primary procedure. A CT scan showed perihepatic fluid collection and eventration of the diaphragm on the right side (Figure 1). Ultrasonography-assisted paracentesis demonstrated bilious ascites. The patient recovered after leakage repair and peritoneal drainage. In the other patient, an 11-month-old girl, vomiting developed 3 days after primary surgery and intussusception was diagnosed by ultrasonography and CT scan (Figure 2). Laparotomy revealed ileo-ileal intussusception and manual reduction was performed. Other early complications were managed conservatively with good results, including wound infection (n=4), wound dehiscence (n=1), gastrointestinal bleeding (n=2), chylous ascites (n=3), acute pancreatitis with pancreatic juice leakage (n=1), anastomotic leakage (n=2), and intra-abdominal infection or abscess formation (n=3).

Twenty-five late complications occurred and 16 of these patients underwent relaparotomy (Table 3), including intrahepatic bile duct stenosis with stones (n=3), anastomotic stricture with or without stone formation (n=5), intrapancreatic cyst remnant (n=4), adhesive bowel obstruction (n=3), and internal hernia formation (n=1). No malignant change was observed. Postoperative cholangitis occurred significantly more often in type IV-A than in type I patients (27.2%, 9/33; 1.3%, 3/228; P<0.001).

Stenosis of the intrahepatic bile duct and calculus formation developed in 3 type IV-A patients (Figure 3). The time interval between the initial surgery and the first identification of intrahepatic stones ranged from 21 months to 7 years. Intrahepatic stenosis occurred near the orifice of the left hepatic duct in all 3 children. One patient had multiple stenoses.

Table 1. Summary of reoperation in 18 patients with choledochal cyst.

No.	Sex	Age	Type	Initial operation	Complications	Reoperation	Outcome
1	F	8y	IV-A	Open	IHBD stenosis with stones	Wide anastomosis	Good
2	M	9y	IVa	Open	Anastomotic stricture	Redo HJ	Good
3	F	9y	Ic	Open	Residual IPCC	Radical excision	Good
4	M	6y	IV-A	Open	Anastomotic stricture	Redo HJ	Good
5	F	11y	IV-A	Open	Residual IPCC	Radical excision	Wound infection
6	F	6y	Ia	Open	Adhesive bowel obstruction	Adhesiolysis	Good
7	F	11y	Ia	Open	Residual IPCC	Radical excision	Good
8	M	9y	Ia	Open	Adhesive bowel obstruction	Adhesiolysis	Good
9	F	8y	Ia	Open	Anastomotic stricture	Redo HJ	Good
10	F	6y	IV-A	Open	IHBD stenosis with stones	Wide anastomosis	Good
11	F	10y	Ia	Laparoscopic	Residual IPCC	Radical excision with PJ	Good
12	F	4y	Ic	Open	Anastomotic stricture	Redo HJ	Good
13	F	7y	IV-A	Open	Adhesive bowel obstruction	Adhesiolysis	Good
14	F	5y	Ia	Laparoscopic	Internal herniation,	Bowel resection	Good
15	F	9y	IV-A	Open	Anastomotic stricture	Redo HJ	Good
16	F	11m	Ia	Open	Intussusception	Manual reduction	Good
17	F	13y	IV-A	Open	IHBD stenosis with stones	Wide anastomosis	Good
18	F	3y	IV-A	Open	Bile leakage	Repair & drainage	Wound infection

y – year; m – month; F – female; M – male; IHBD – intrahepatic bile duct; IPCC – intrapancreatic choledochal cyst; HJ – hepaticojejunostomy; PJ – pancreaticojejunostomy.

Table 2. Early complications after definitive surgery.

Complications	Number
Early complications	18 (2)
Wound infection	4
Wound dehiscence	1
Gastrointestinal bleeding	2
Bile leakage	3 (1)
Acute pancreatitis with pancreatic leakage	1
Chylous ascites	3
Intra-abdominal infection/abscess	3
Intussusception	1 (1)

The parentheses demonstrate the number of patients requiring reoperation.



Figure 1. CT coronal scan shows suprahepatic fluid and eventration of the right diaphragm (patient No. 18).

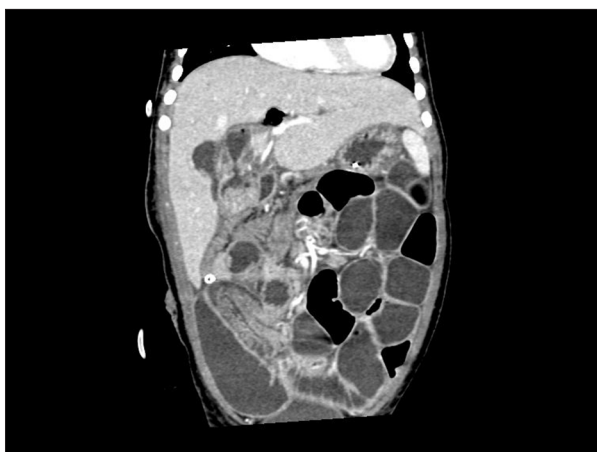


Figure 2. CT coronal scan shows intussusception (patient No. 16).

Table 3. Late complications after definitive surgery.

Complications	Number
Late complications	25 (16)
Intrahepatic calculus	3 (3)
Anastomotic stricture	5 (5)
Intrapancreatic residual cyst	4 (4)
Internal herniation	1 (1)
Adhesive bowel obstruction	8 (3)
Cholangitis*	2
Persistent dilatation of intrahepatic bile duct	2

The parentheses demonstrate the number of patients requiring reoperation. * These two patients had symptoms of cholangitis without other findings.

No anastomotic strictures were found at the hepaticojejunostomy site. Organisms cultured from the bile were reported: *Escherichia coli* (2 cases) and negative culture (1 case). The stenotic bile duct was incised from the hepatic hilum to the dilated segment and irrigated via a catheter. Then a wide oblique anastomosis was performed.

Anastomotic stricture was identified in 5 patients (3 type IV-A, 1 type Ia, and 1 type Ic) (Figure 4). Symptoms of cholangitis, including abdominal pain, high fever, and jaundice, were observed. Stones were found proximal to the site of the hepaticojejunostomy in 3 children. The mean diameter of the anastomosis was 4.8 ± 0.8 mm. The stricture including the proximal hepatic duct and jejunal wall was excised en bloc, stones were removed, and intrahepatic bile ducts were lavaged with normal saline. Then a redo hepaticojejunostomy was performed.



Figure 3. Intraoperative cholangiography of patient No. 17 demonstrates left hepatic bile duct dilatation with multiple calculi formation (arrow).

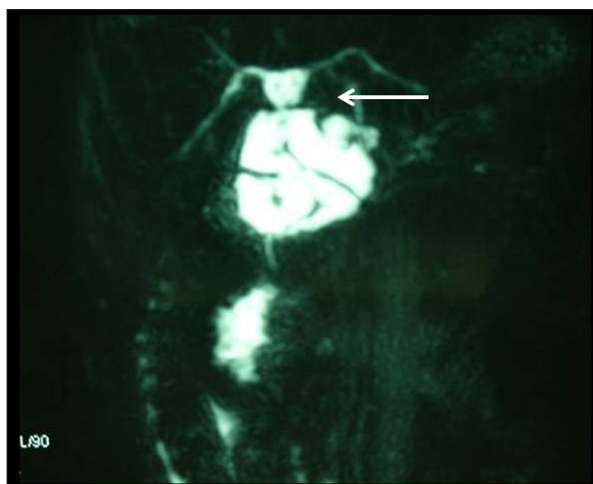


Figure 4. Postoperative MRCP shows anastomotic stricture (arrow, patient No. 2).

Postoperative pancreatitis with or without stone formation was detected in 4 patients (2 type IV-A, 1 type Ia, and 1 type Ic) after a median follow-up period of 9.5 months (range, 4 months to 50 months). Elevated serum amylase levels (726 ± 250 U/L) were detected in all 4 children. Ultrasonographic examination and CT scan of the abdomen revealed dilated residual cysts in the head of pancreas (Figure 5). Radical excision of the dilated cystic remnant was completed in 4 children (with 1 needing an additional pancreaticojejunostomy procedure due to significant pancreatic duct dilatation). It is important to keep the pancreatic parenchyma and duct intact during choledochal remnant dissection.

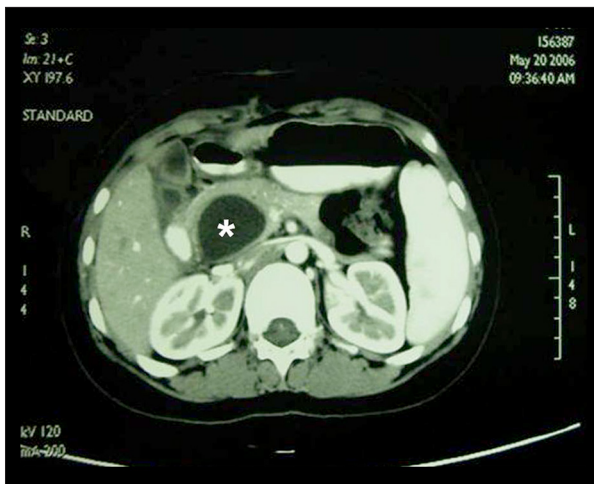


Figure 5. CT axial scan shows intrapancreatic residual cyst (asterisk, patient No. 7).

One girl developed acute bowel obstruction due to internal herniation through the mesenteric defect 2 months after primary surgery. The patient underwent relaparotomy and necrotic bowel resection. Of the 8 children with adhesive intestinal obstructions, 3 underwent surgical interventions and 5 were treated medically.

The median follow-up period after reoperation was 6 years (range, 1 year to 13 years). No mortality was documented. Wound infection developed in 2 patients and was managed by changing the dressing, and the other 16 patients recovered uneventfully.

Discussion

Complete cyst excision with biliary reconstruction is the treatment of choice for choledochal cyst. Understanding the pathophysiology and anatomy of CC is crucial for successful management. Despite recent advances in surgical techniques and peri-operative management, early and late complications are not rare in children, while postoperative complications are more common in adults [5,6]. Reoperation is required in less than 10% of pediatric patients and remains a great challenge to pediatric surgeons. The rate of reoperation in this study was about 6.8%, which is similar to other reports from Japan and UK [2,4,5].

Our experience demonstrates that most early complications can be treated conservatively. Anastomotic bile leakage is thought to be caused by surgical inexperience and severe inflammation. In some cases, the diagnosis of bile leakage is difficult and delayed because the symptoms are nonspecific. Imaging findings of ultrasonography and CT cannot be differentiated from other causes of postoperative intra-abdominal fluid collection. Some

patients might succumb to the subsequent damage, such as septicemia and septic shock. Chavhan et al. [7] reported gadobenate-enhanced MR cholangiography can be used to diagnose and accurately localize the site of bile leakage noninvasively. Minor leakage can be treated effectively by adequate drainage. Reoperation is considered only after the failure of conservative treatment. The leakage can be repaired by a circumferential buried suture around the anastomotic site, peritoneal lavage, and effective drainage. Postoperative intussusception may develop after a variety of abdominal operations [8]. The majority (87.2%) of them were small bowel intussusceptions (jejuno-jejunal, jejuno-ileal, or ileo-ileal). The median onset time of intussusception was 6 days after the operation. Relaparotomy and manual reduction was always carried out. Therefore, intussusception, although very rare, should be considered, in addition to adhesive ileus and bowel twist.

Based on our experience, unrecognized primary bile duct stenosis, residual debris, and anastomotic stricture are the main causes of postoperative cholangitis. In the present study, 4 of the 12 patients with cholangitis responded to antibiotics treatment, and biochemical liver dysfunction normalized. Intrahepatic bile duct dilatations of type IV-A always accompany a downstream ductal stenosis, frequently around the hepatic hilum. Interestingly, Tsuchida et al. [9] found that 40% of patients developed postoperative intrahepatic cholelithiasis among patients with dilated intrahepatic ducts and downstream stenosis, while only 2% occurred in patients without any dilatation of the intrahepatic bile ducts. Therefore, it is clinically meaningful to detect the intrahepatic bile ductal stricture. However, it can be very difficult to distinguish between type I and type IV-A preoperatively in some cases [10,11]. Excision of the cyst, ductoplasty, and wide hepaticoenterostomy at the hilum may be imperative to prevent bile stasis, recurrent cholangitis, stone formation, and biliary carcinoma. It is also important to perform endoscopic examination and wash out debris or stones from the dilated intrahepatic bile duct during cyst excision [1,2,12]. Liver resection has been performed in adult patients with type IV-A CC, but it is too aggressive for infants and children. Urushihara et al. [13] reported hepaticojejunostomy at hilum combined with intrahepatic cystojejunostomy (double anastomoses) was an attractive alternative procedure for more upstream intrahepatic ductal cyst. In terms of anastomotic stricture, improvement of surgical skills, preservation of blood supply, no or mild inflammation of cyst wall, and construction of a wide (larger than 1 cm) and tension-free stoma are key factors to reduce anastomosis-related complications [10,14,15]. There should be no delay in surgical or endoscopic intervention once the biliary obstruction develops postoperatively, but a great deal of planning and a thoughtful workup are required. Revision of hepaticojejunostomy with ductoplasty, when necessary, is usually performed in children. Kim et al. [15] reported that percutaneous transhepatic

cholangioscopy with stone removal and balloon dilatation was useful in 7 adult patients with anastomotic stricture. Recently, Ono et al. [16] successfully treated pediatric patients with intrahepatic bile duct stones using double-balloon enteroscopy (via the limb of the Roux-en-Y reconstruction). However, our team has no personal experience with this endoscopic technique.

Pancreatitis occurs in patients with CC, both preoperatively and postoperatively. Fujishiro et al. [17] reported that the incidence of pancreatitis after cyst excision was about 4.2%. In this study, pancreatitis was observed in 5 (1.9%) children after primary surgery. Complete resection of the distal portion of the cyst, removal of debris and protein plugs in the long common channel and pancreatic duct, and correction of anomalous arrangement of the pancreaticobiliary duct junction are essential to minimize pancreatic complications after the operation. Overly aggressive resection might cause damage to the pancreatic duct. Recently, Koga et al. [18] reported that intraoperative ureteroscopy during laparoscopic excision of CC was successfully used to more accurately dissect the intrapancreatic cyst. However, a fine-caliber endoscope is not available in our institutes, so we instead use a flexible catheter to irrigate the common channel. Intraoperative cholangiography can be performed to confirm the clearance. Intrapancreatic cyst remnant is a significant risk factor for pancreatic carcinoma [19–21]. Thus, radical excision of the residual cyst is strongly advocated. In some unusual cases with pancreatic duct stenosis and upstream dilatation, pancreaticojejunostomy (Puestow procedure) is required.

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Adhesions after intra-abdominal surgery are common and are a major cause of small bowel obstruction. Patients should be closely observed for any sign of deterioration after initial management. In the absence of clinical improvement, observation longer than 48 h carries a greater risk of bowel necrosis and bowel resection [22]. However, the optimal time of surgery has not been clearly defined. Furthermore, it is necessary to pay particular attention to the length and placement of the Roux loop during biliary reconstruction [23,24].

Although there was no subsequent biliary malignancy documented in this series, long-term surveillance remains mandatory for patients with CC. The main limitations of this study are its retrospective design and small number of patients. Precise and thorough surgical treatment is necessary to minimize early and late postoperative complications [25,26].

Conclusions

Detection and correction of the intrahepatic bile duct stenosis, and a wide hepaticojejunostomy (with ductoplasty when necessary) are essential to prevent cholangitis, anastomotic stricture, and calculus formation. Moreover, complete cyst excision, including the intrapancreatic portion, should be strongly recommended.

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