

# COMPLICATIONS AFTER EXCISION OF CHOLEDOCHAL CYSTS

Trung Bui Hai MD  
Hung Le Hoang MD  
Tri Tran Thanh MD  
General surgery Department

# CHOLEDOCHAL CYSTS

- Congenital dilatations of the extra and/or intrahepatic bile ducts
- Most common site: choledochus
- Significantly more common in Asia
- Female dominance ( 3-4/1)

# CHOLEDOCHAL CYSTS

**TABLE 44-1 The Mean Common Bile Duct Diameter and the Range According to a Patient's Age**

<b>Age (Years)</b>	<b>Range (mm)</b>	<b>Mean (mm)</b>
≤4	2-4	2.6
4-6	2-4	3.2
6-8	2-6	3.8
8-10	2-6	3.9
10-12	3-6	4.0
12-14	3-7	4.9

*Adapted from Witcombe JB, Cremin BJ. The width of the common bile duct in childhood. Pediatr Radiol 1978;7:147-9.*

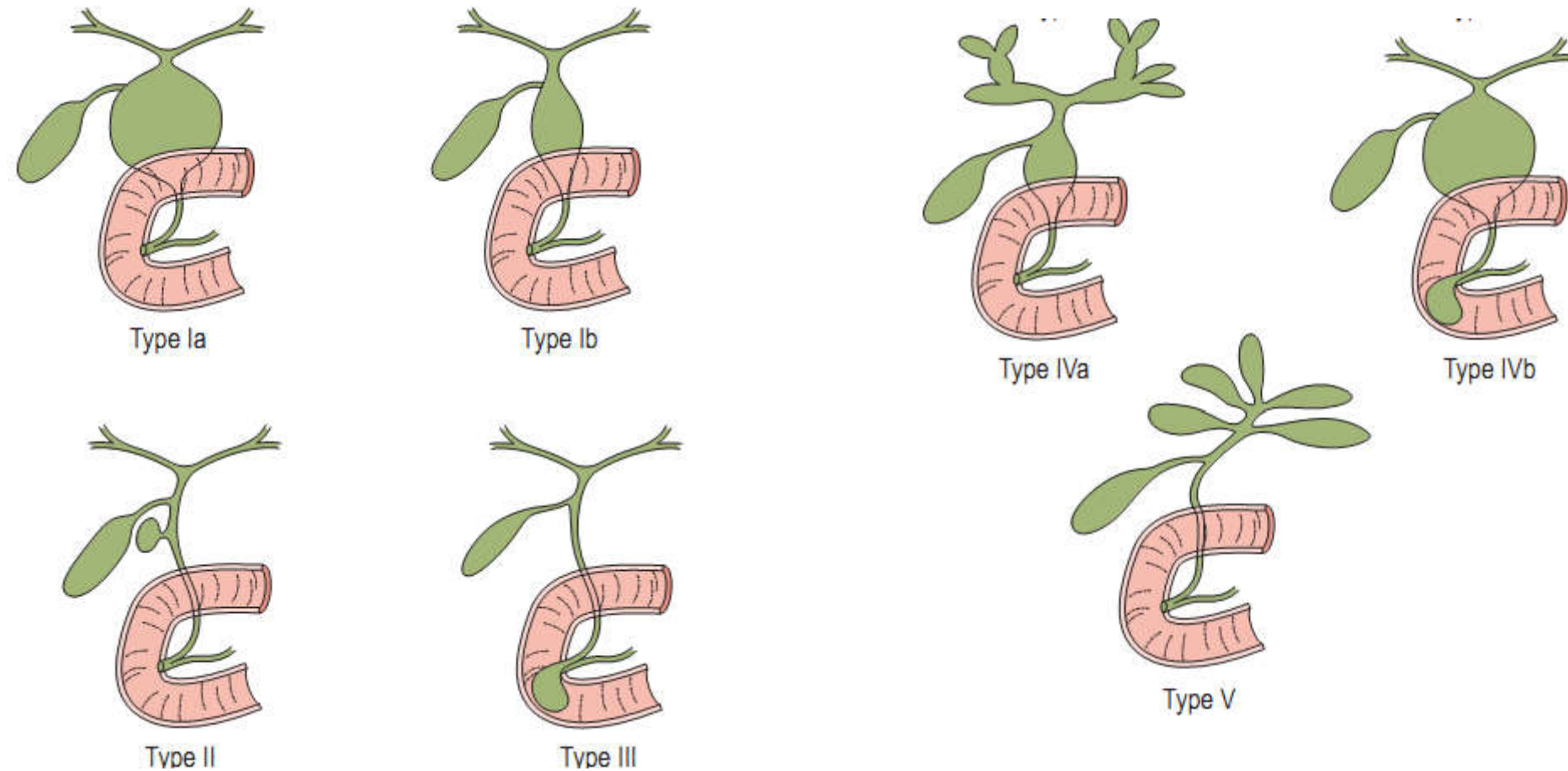
# CHOLEDOCHAL CYSTS

- Etiology:

- Pancreaticobiliary maljunction (PBM) → reflux of pancreatic fluid into the bile duct
- Distal obstruction at the level duodenum



# CLASSIFICATION

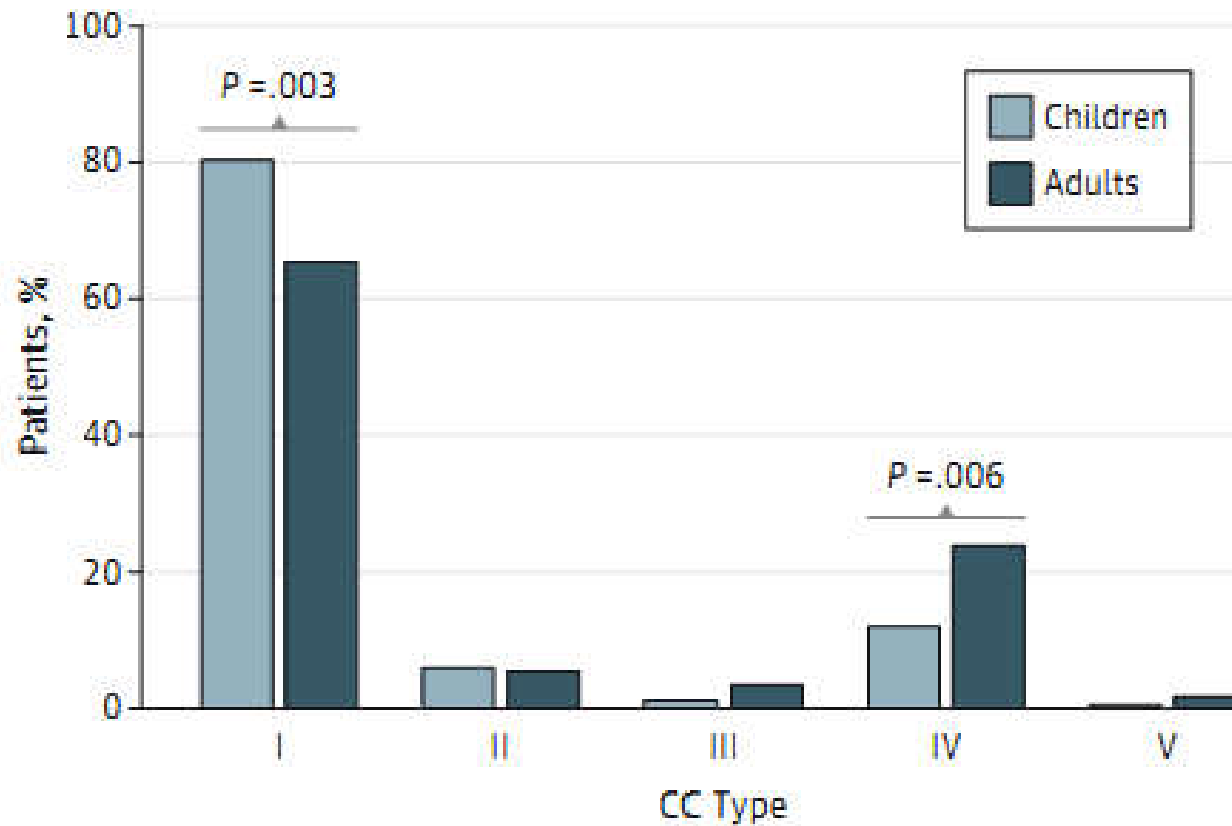


**FIGURE 44-1** ■ These diagrams depict the five classifications of choledochal cyst according to Todani. (From Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cyst: Classification, operative procedure, and review of 37 cases including cancer arising from choledochal cyst. *Am J Surg* 1977;134:263–9.)

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Figure 1. Type of Choledochal Cysts (CCs) in Adult and Pediatric Populations in 394 Patients

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Type I CCs were more common in children; type IV CCs were more common in adults.

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# CLINICAL FEATURES

TABLE 1. CLINICAL MANIFESTATIONS (N = 400)

<i>Manifestation</i>	<i>Number</i>	<i>%</i>
Abdominal pain	352	88.0
Fever	113	28.3
Vomiting	194	45.8
Icterus	99	24.8
Discolored stool	50	12.5
Abdominal tumor	30	7.5
Classic triad	9	2.2

# DIAGNOSIS

- Abdominal ultrasound: first imaging modality(sens 71-97%), antenatal diagnosis.
- Technetium-99m HIDA scan
- CT scan, CT cholangiography
- ERCP, PTC, intraoperative cholangiogram: risk of pancreatitis, cholangitis
- MRCP: now considered the gold standard



# SURGICAL MANAGEMENT

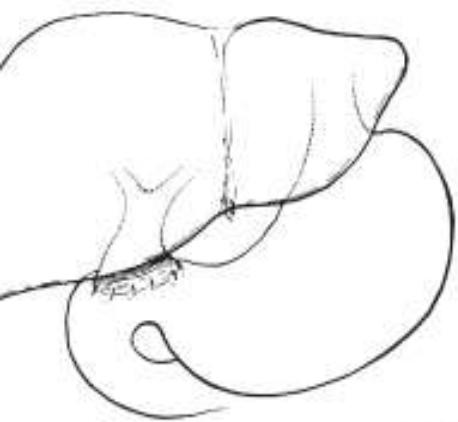
- Laproscopic surgery
- Open surgery

Table 3 Surgical modalities used for the treatment of choledochal cyst

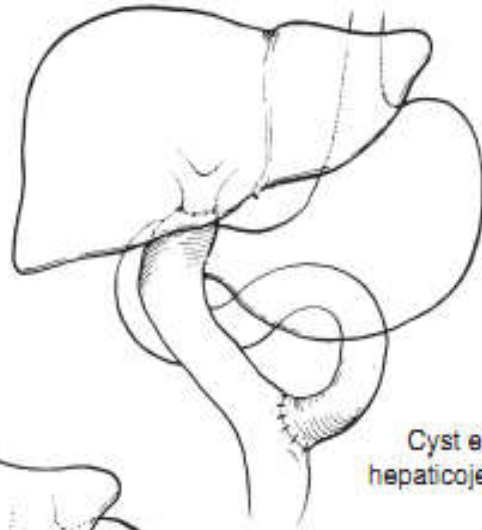
Type of surgery	Number of patients (n, %)
Roux-en-Y hepaticojejunostomy <sup>a</sup>	26 (74.28)
Hepaticoduodenostomy	5 (14.28)
Cystoduodenostomy	2 (5.71)
External T-tube drainage	3 (8.57)

<sup>a</sup>The most common operation performed for choledochal cyst.

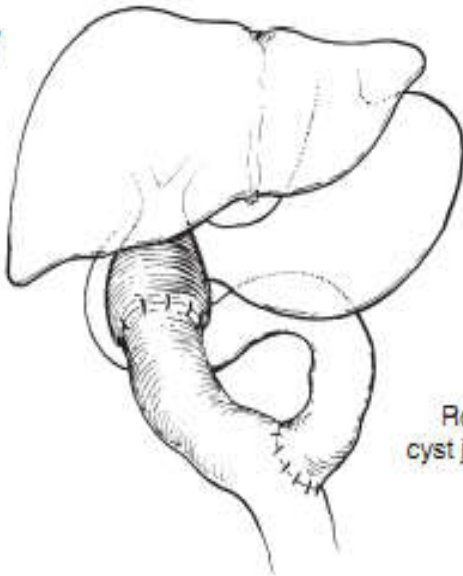
# SURGICAL MANAGEMENT



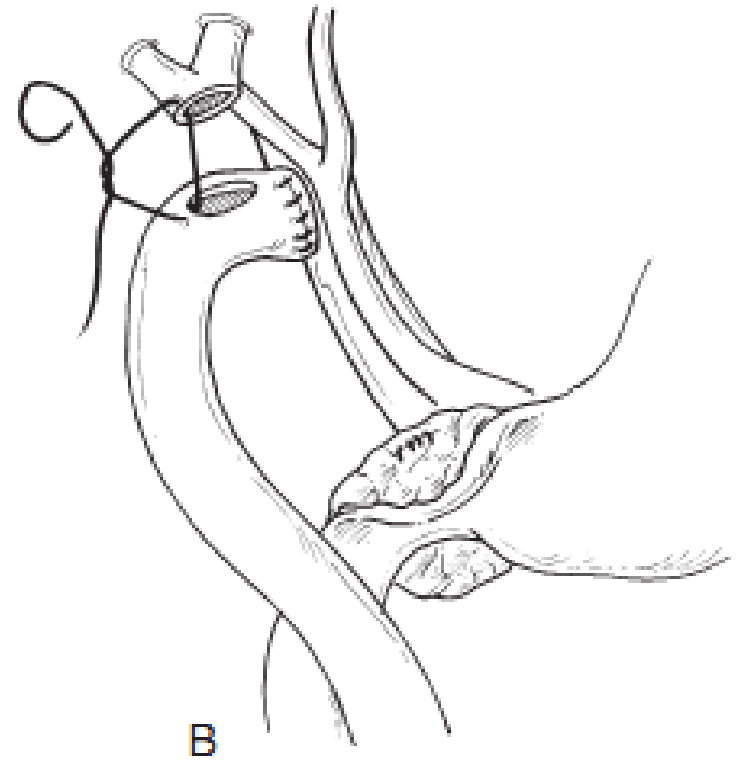
Cyst duodenostomy



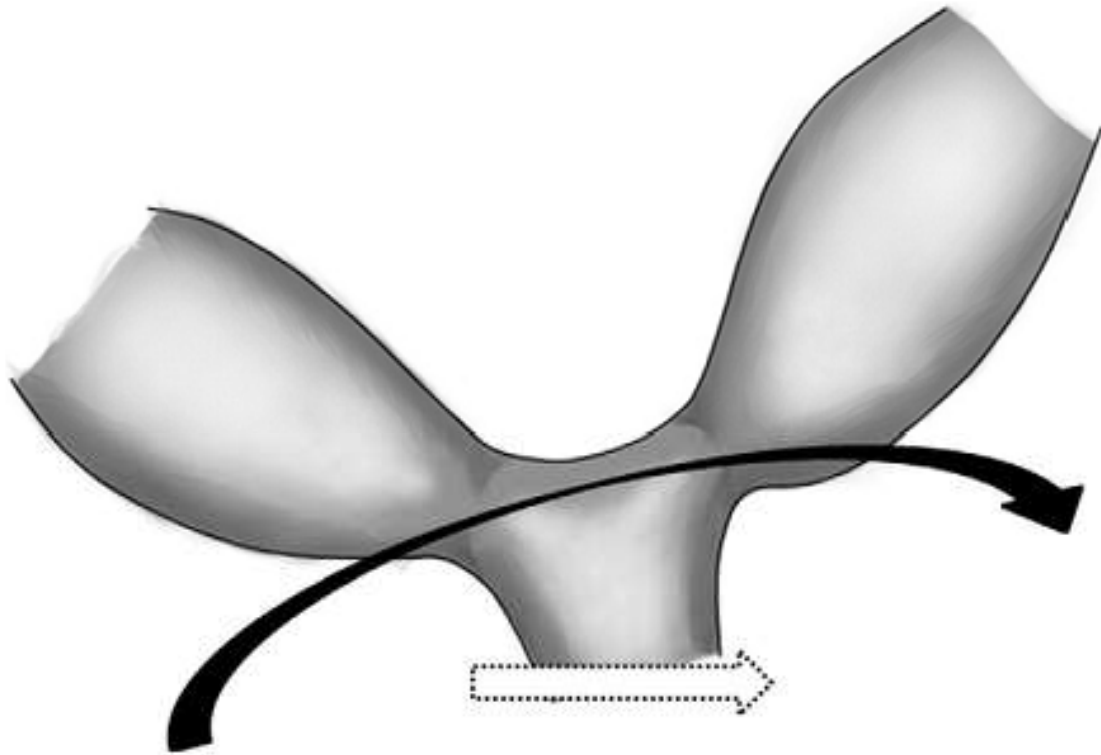
Cyst excision,  
hepaticojejunostomy



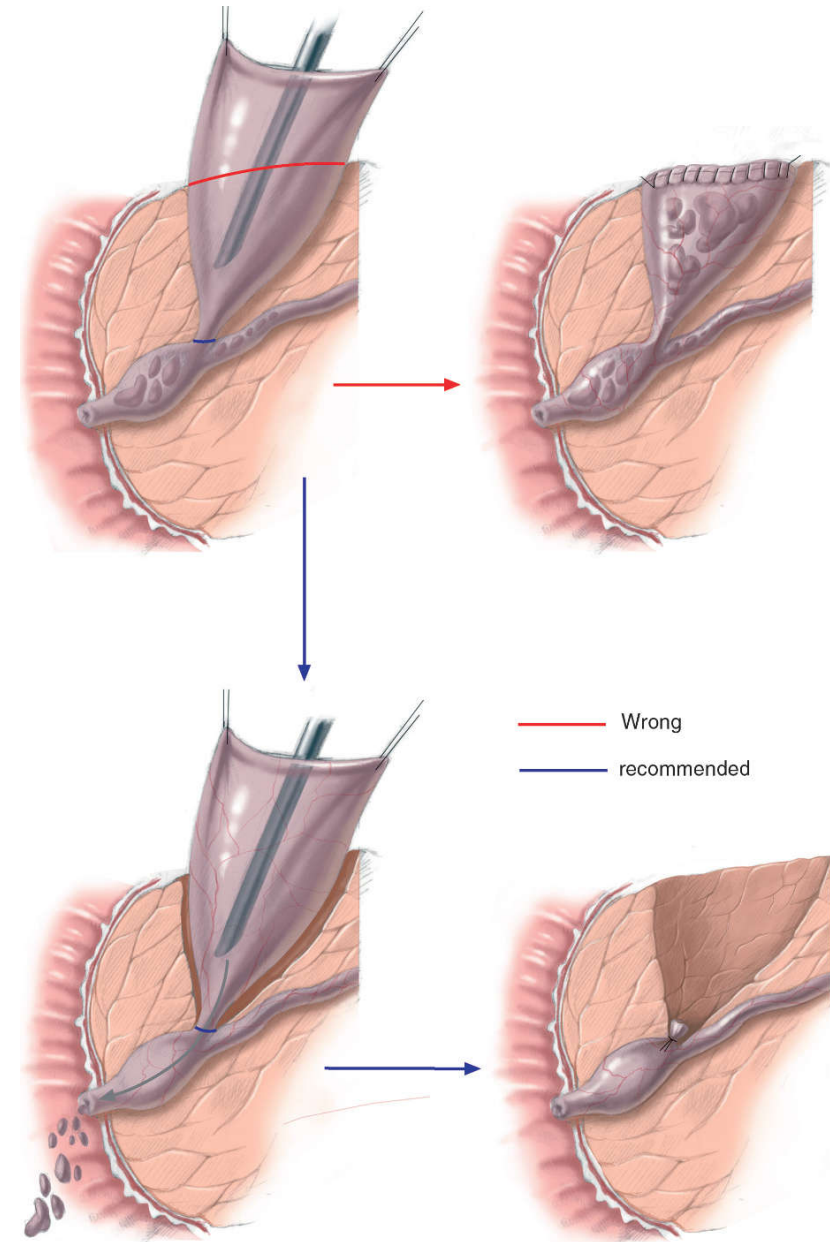
Roux-en-Y  
cyst jejunostomy

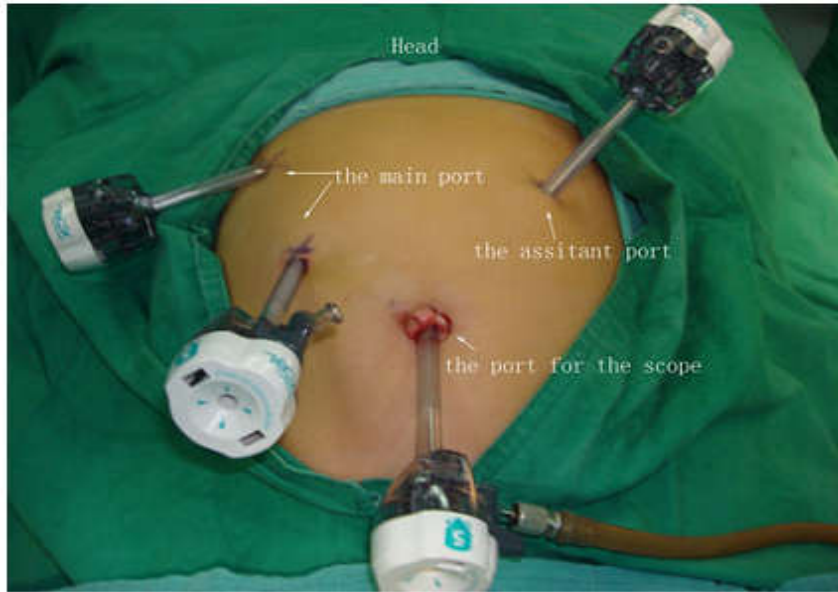


B

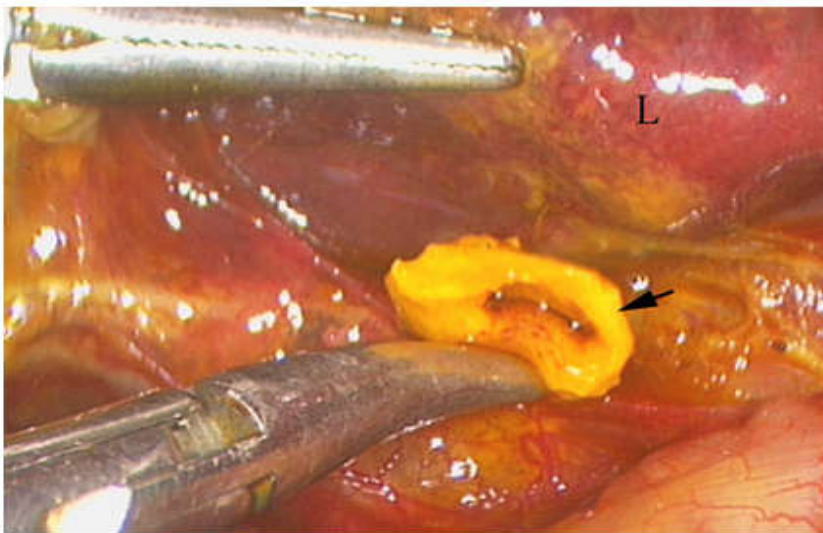


**Fig. 1** Anastomotic line for hepaticojejunostomy below the hepatic hilum (white arrow) and wide hilar hepaticojejunostomy (black arrow).

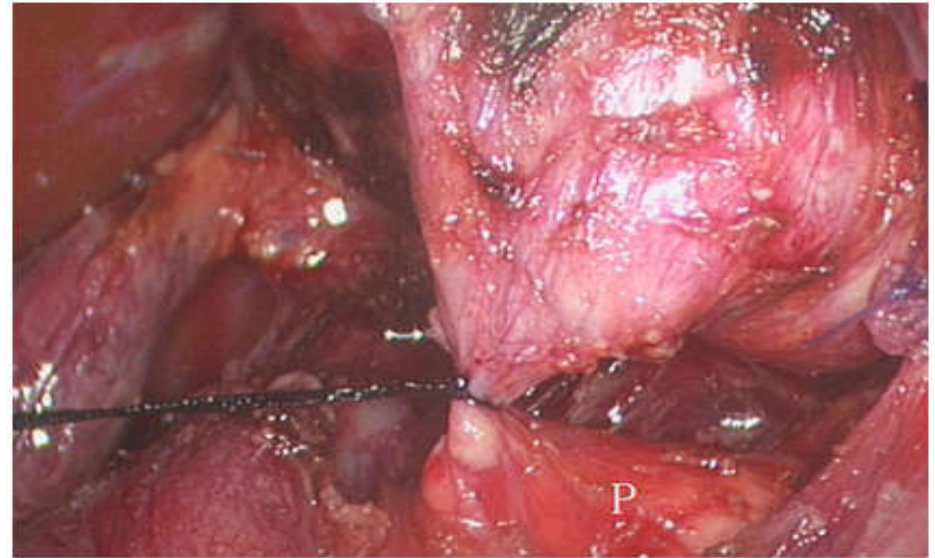




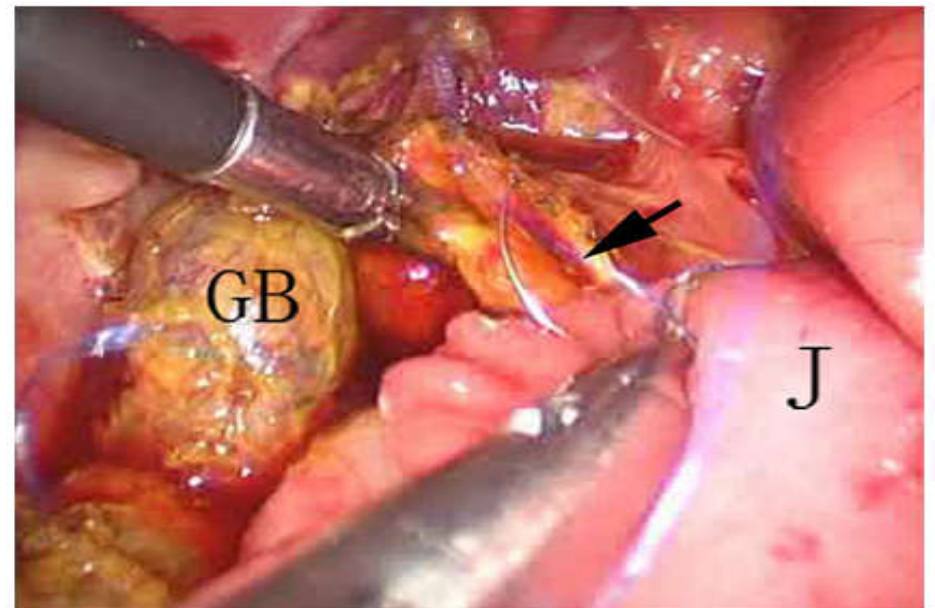
**Fig. 1** Locations of laparoscopic ports



**Fig. 3** Proximal part of the cyst dissected up to the normalized hepatic ducts (*arrow*) and then removed at this level



**Fig. 2** Distal choledochus ligated with a 4-0 silk suture. *P* pancreas



**Fig. 4** Hepaticojejunostomy performed by the laparoscopic approach. *GB* gallbladder, *Arrow* the proximal hepatic duct, *J* jejunum

# Early and Late Results of Excision of Choledochal Cysts

By Htut Saing, Htoo Han, K.L. Chan, W. Lam, F.L. Chan, W. Cheng, and P.K.H. Tam  
*Hong Kong*

**Background/Purpose:** Reports on the late results of choledochal cyst excision with hepaticojejunostomy in children are relatively few.

**Methods:** Of the 84 patients who had choledochal cyst who were under our care, 79 have had definitive surgery, three are awaiting surgery, one is being observed with Caroli's disease, and the parents of one child have refused surgery. Eight patients treated decades ago had internal drainage procedures. Since 1972, 41 patients have had cyst excision with hepaticojejunostomy using a 40-cm Roux loop without an antireflux procedure. Early complications in those who underwent cyst excision with hepaticojejunostomy included anastomotic leak in three patients who required reoperation, cholangitis in two, and fluid collection in the gallbladder bed that required no intervention in one.

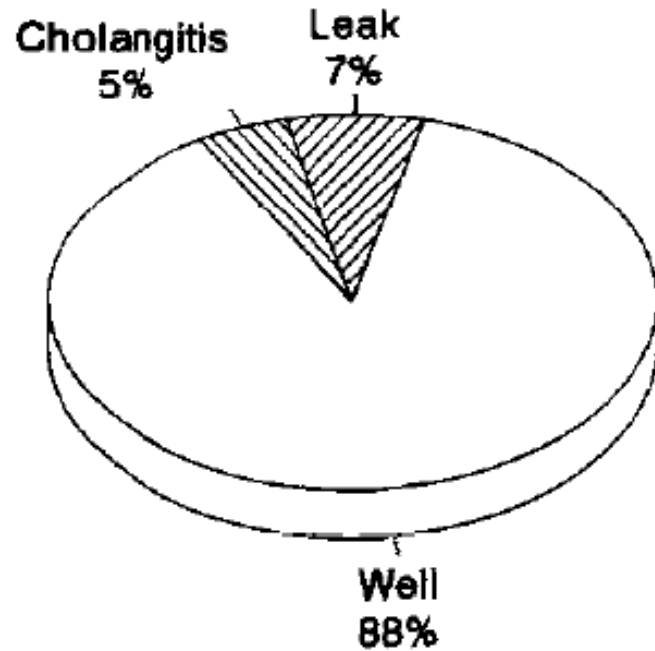
**Results:** During a follow-up period ranging from 4 months to 15 years (mean, 8.5 years), anastomotic stricture, cholangitis,

and intrahepatic stone formation developed in two children after being well for 8 years and over 11 years. These children required additional surgical procedures to overcome these problems. Asymptomatic intrahepatic stones 2 years after cyst excision with hepaticojejunostomy developed in a third child. There was no mortality in the entire group that underwent cyst excision and they are all enjoying a good quality of life.

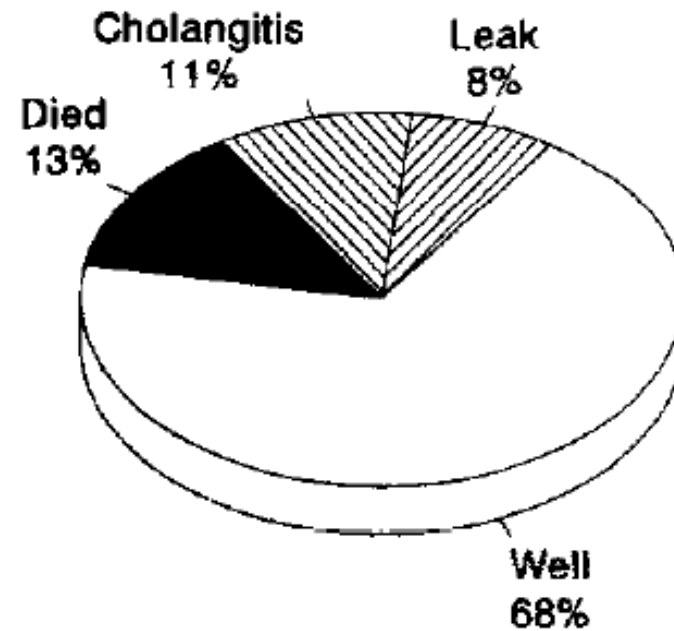
**Conclusions:** Careful, long-term follow-up is important in children who have choledochal cyst excision with hepaticojejunostomy.

*J Pediatr Surg* 32:1563-1566. Copyright © 1997 by W.B. Saunders Company.

**INDEX WORDS:** Choledochal cyst, cyst excision, hepaticojejunostomy.

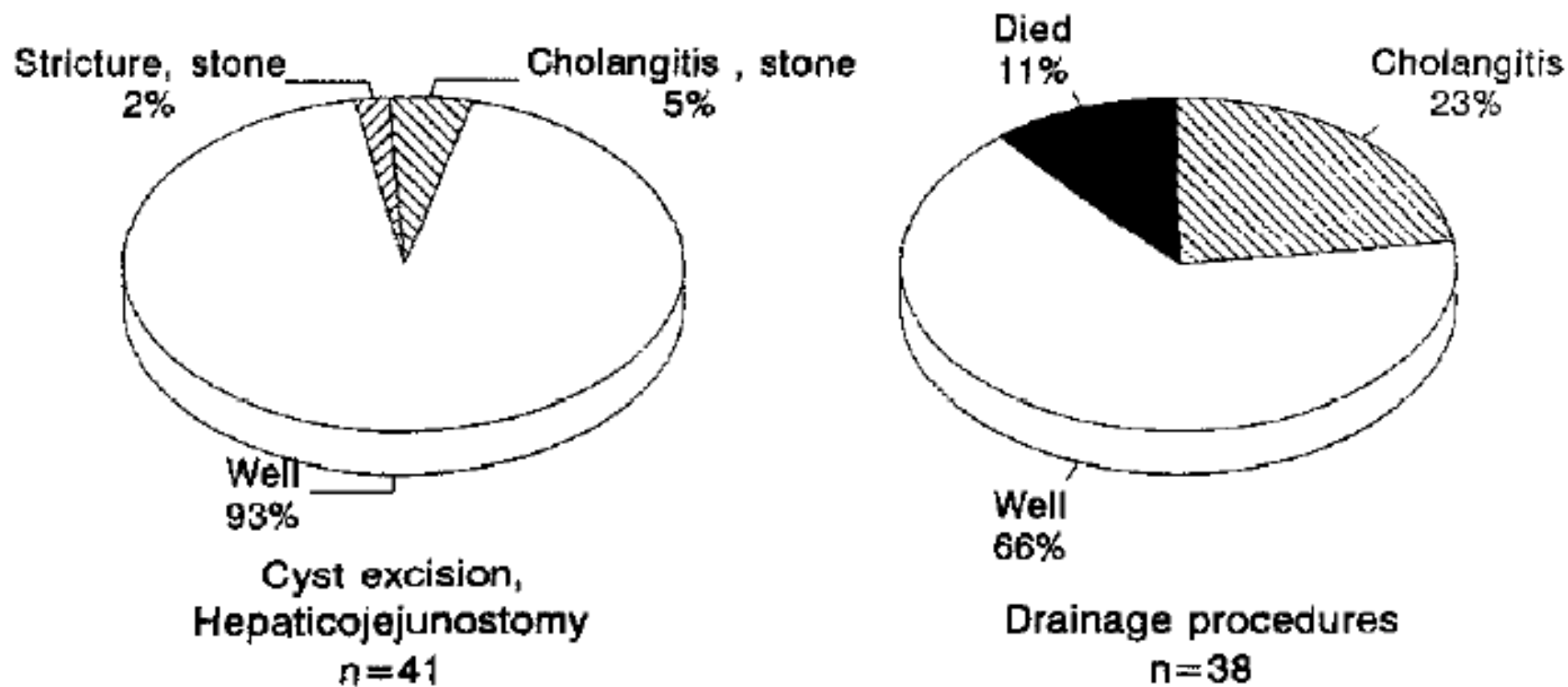


Cyst excision,  
Hepaticojejunostomy  
n=41



Drainage procedures  
n=38

**Fig 2. Internal drainage procedures are associated with a high incidence of early morbidity and mortality, whereas cyst excision with hepaticojejunostomy is associated with acceptable morbidity and zero mortality.**



**Fig 3. Late results of surgical treatment show that cyst excision is a far better procedure with minimal late complications and no mortality.**

**Table 2. Late Complications After Cyst Excision, Management, and Outcome**

Sex/Age (yr) at 1st Op	Recurrent Cholangitis After Initial Op (yr)	Anastomotic and Intrahepatic Duct Strictures	Intrahepatic Stones Detected After Op (yr)	Age at Intervention for Stones (yr)	Surgical Procedures	Remarks
F/1	11	+	13.5	14.5	Revision of anastomotic stricture Hepaticocutaneous jejunostomy Endoscopic stone clearance	Well for 4 years since last procedure A course of drug therapy for liver flukes
F/0.6	8	+	10	11	PTBD followed by balloon dilatation of anastomotic stricture Revision of anastomotic stricture Hepaticocutaneous jejunostomy Endoscopic stone clearance	Well for 1 year since last procedure
M/4	Asymptomatic	+	6	Awaiting intervention	No procedure performed yet	Completely asymptomatic 1 year since stones were detected

Abbreviations: Op, operation; PTBD, percutaneous transhepatic biliary drainage.



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Original Investigation

# Presentation and Clinical Outcomes of Choledochal Cysts in Children and Adults

## A Multi-institutional Analysis

Kevin C. Soares, MD; Yuhree Kim, MD, MPH; Gaya Spolverato, MD; Shishir Maithel, MD; Todd W. Bauer, MD;  
Hugo Marques, MD; Mafalda Sobral, MD; Maria Knoblich, MD; Thuy Tran, MD; Luca Aldrighetti, MD;  
Nicolas Jabbour, MD; George A. Poultsides, MD; T. Clark Gamblin, MD; Timothy M. Pawlik, MD, MPH, PhD

**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.

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Published online April 29, 2015.

## Morbidity and Mortality Rates After Choledochal Cyst Surgery

Complication	No. (%)			P Value
	Overall (N = 394)	Children (n = 135)	Adults (n = 259)	
Death	42 (10.7)	4 (3.0)	38 (14.7)	<.001
Cholangitis	8 (2.0)	0	8 (3.1)	.001
Cholecystitis	27 (6.9)	2 (1.5)	25 (9.7)	<.001
Cholelithiasis	10 (2.5)	2 (1.5)	8 (3.1)	.3
Cholelithiasis	67 (17.0)	14 (10.4)	53 (20.5)	.001
Cholangitis	17 (4.3)	3 (2.2)	14 (5.4)	.1
Common bile duct obstruction	8 (2.0)	1 (0.7)	7 (2.7)	.1
Common bile duct stricture	4 (1.0)	1 (0.7)	3 (1.2)	.7
Common bile duct stones	2 (0.5)	2 (1.5)	0	.0
Common bile duct hepatic abscess	20 (5.1)	0	20 (7.7)	<.001
Common bile duct leak	22 (5.6)	5 (3.7)	17 (6.6)	.2
Common bile duct stomotic leak	4 (1.0)	4 (3.0)	0	.0
Common bile duct stricture	3 (0.8)	0	3 (1.2)	.2

gastrointestinal tract	42 (10.7)	4 (3.0)	38 (14.7)	<.001
bleeding	3 (0.8)	1 (0.7)	2 (0.8)	.5
level obstruction	15 (3.8)	5 (3.7)	10 (3.9)	.9
abscess	9 (2.3)	2 (1.5)	7 (2.7)	.4
ascites	7 (1.8)	0	7 (2.7)	.0
pancreatitis	10 (2.5)	1 (0.7)	9 (3.5)	.3
perforation	4 (1.0)	4 (3.0)	0	.0
paralytic ileus	1 (0.3)	0	1 (.4)	.4
hemorrhagic/thrombotic	20 (5.1)	4 (3.0)	16 (6.2)	.3
hepatic/urologic	13 (3.3)	3 (2.2)	10 (3.9)	.4
neurovascular	5 (1.3)	0	5 (1.9)	.3
respiratory	10 (2.5)	3 (2.2)	7 (2.7)	.8
total	7 (1.8)	5 (3.7)	2 (0.8)	.0
total	113 (28.7)	22 (16.3)	91 (35.1)	<.001
non-Dindo <sup>19</sup> classification $\geq 3^a$	92 (55.8)	13 (54.2)	78 (56.1)	.8
mortality	22 (5.6)	4 (3.0)	18 (6.9)	.3

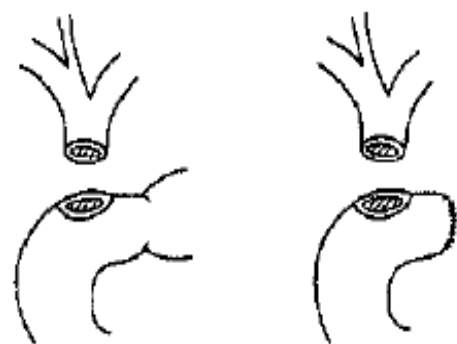
*Journal of Pediatric Surgery, Vo130, No 3 (March), 1995: pp 478-48*

# **Biliary Complications After Excisional Procedure for Choledochal Cyst**

By T. Todani, Y. Watanabe, N. Uru|shihara, T. Noda, and Y. Morotomi  
*Kagawa, Japan*

During the last 25 years, from 1969 to 1994, the authors treated 97 choledochal cysts by surgical excision. Biliary reconstruction consisted of 67 hepaticoduodenostomies and 30 hepaticojejunostomies. The common hepatic duct was the site of anastomosis in 9 of the duodenostomies and 13 of the jejunostomies and of the bifurcation of the hepatic ducts in 58 duodenostomies and 17 jejunostomies. Reoperation was required in 10 cases because of recurrent cholangitis with intrahepatic gallstones.

BELOW HILUM



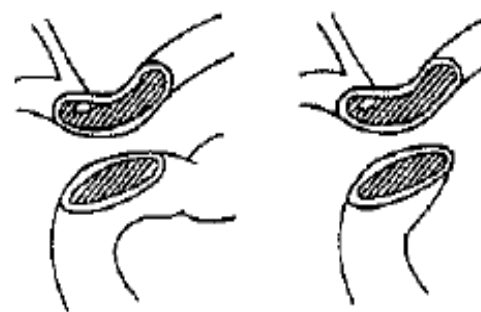
hepaticoduodenostomy

9 (4)

hepaticojejunostomy  
with blind pouch

13 (5)

AT HILUM



hepaticoduodenostomy

58

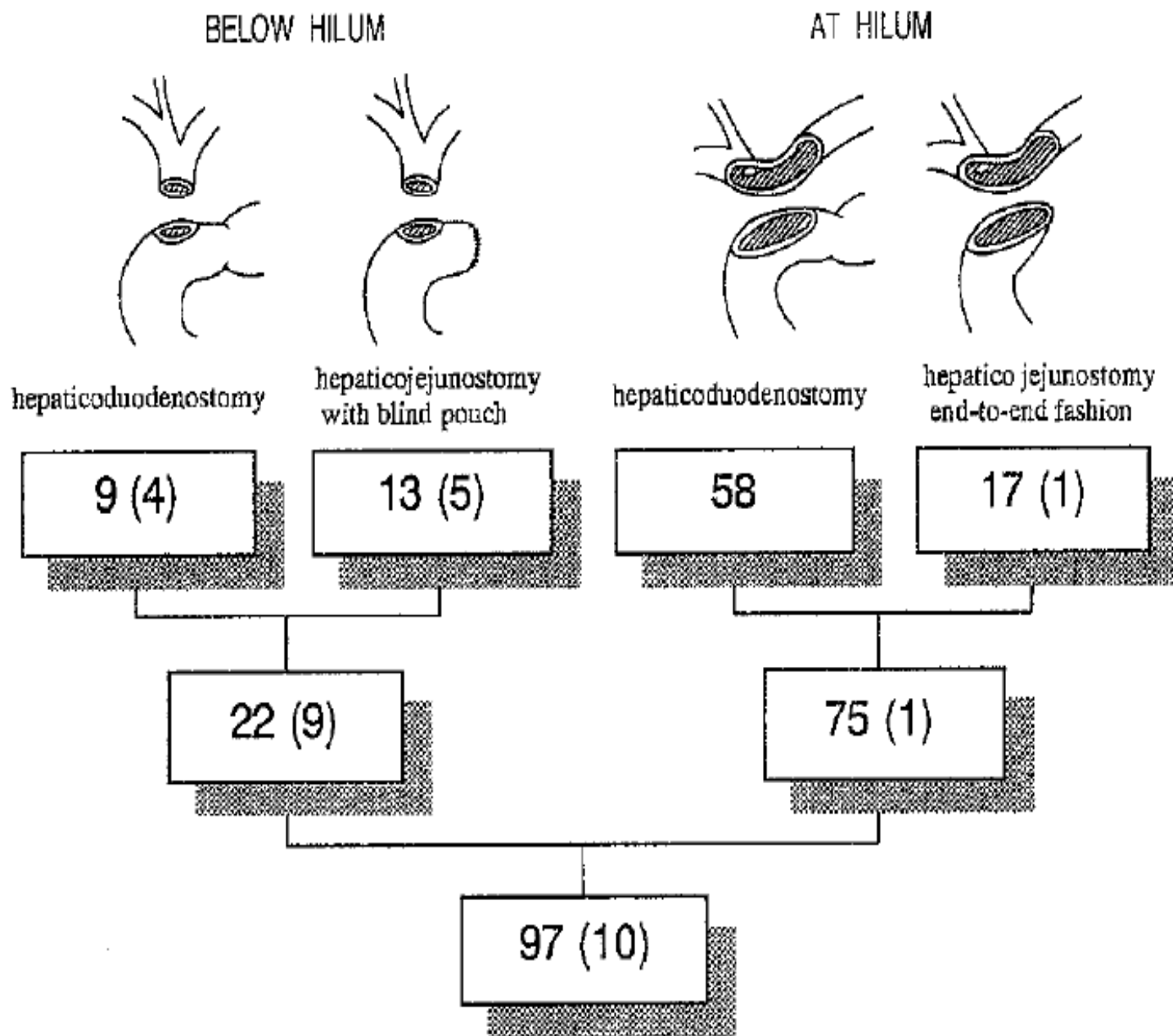
hepatico jejunostomy  
end-to-end fashion

17 (1)

22 (9)

75 (1)

97 (10)



**Table 3. Case Summary of Reoperation**

Type	First Operation	Age (yr)	Redo-Operation	Age (yr)	Follow-Up (yr)
<i>Anastomotic stricture</i>					
1. I	Hepaticojejunostomy*	10	Hepaticojejunostomy	15	4
2. I	Hepaticojejunostomy	9	Hepaticojejunostomy and right hepatectomy	30	3
3. I	Hepaticojejunostomy*	16	Hepaticojejunostomy*	23	7
			Hepaticojejunostomy	26	2
4. IV	Hepaticoduodenostomy	11	Hepaticojejunostomy	18	17
5. IV	Hepaticojejunostomy*	2	Hepaticoduodenostomy	6	7
6. IV	Hepaticoduodenostomy	5	Lateral segmentectomy with hepaticojejunostomy	11	6
7. IV	Hepaticojejunostomy	13	Hepaticojejunostomy	28	5
8. IV	Hepaticoduodenostomy	18	Hepaticojejunostomy*	33	4
9. IV	Hepaticoduodenostomy*	15	Hepaticojejunostomy	24	3
<i>Primary ductal stricture</i>					
10. IV	Hepaticojejunostomy	17	Lateral segmentectomy	32	3

\*Performed elsewhere.

Journal of pediatric Surgery, Vol31, No 10 (October), 1996: pp 1417-1421

# **Hepaticoenterostomy After Excision of Choledochal Cyst in Children: A 30-Year Experience With 180 Cases**

By Takeshi Miyano, Atsuyuki Yamataka, Yoshifumi Kato, Osamu Segawa, Geoffrey Lane, Shigeru Takamizawa,  
Sumio Kohno, and Toshio Fujiwara  
*Tokyo, Shizuoka, and Tochigi, Japan*



**Table 1. Types of Hepaticoenterostomy Anastomoses Performed in 174 Patients**

Type of Anastomosis	No. of Patients
Conventional hepaticojejunostomy	169*
Conventional hepaticoduodenostomy	2
Intrahepatic cystojejunostomy	2
Hepaticojejunostomy at the hepatic hilum	1

\*Three of the 169 patients required another operation because of IHBD stones: two had a conventional hepaticojejunostomy, and one had a hepaticojejunostomy at the hepatic hilum.

**Table 2. Cases of Postoperative IHBD Stone Formation**

Case No.	Sex/Age (yr)	Type of Choledochal Cyst (IHBD dilatation)	Operation	Stoma Diameter (mm)	Symptom	Cause	Subsequent Surgery		
							Procedure	Age	Result
1	F/12	Cystic (+)	Cyst excision; H-J (end-to-side anastomosis)	> 20	Cholangitis, jaundice	Anastomotic stricture	Revision of H-J	26	Excellent
2	F/7	Cystic (+)	Cyst excision; H-J (end-to-side anastomosis)	8	Cholangitis, jaundice	Anastomotic stricture	Revision of H-J	18	Excellent
3	M/6 16	Cystic (-)	C-J Cyst excision; H-J (end-to-side anastomosis)	> 20	Cholangitis Cholangitis	Anastomotic stricture	H-J at the hepatic hilum	21	Excellent
4	F/6	Cystic (+)	Cyst excision; H-J (end-to-side anastomosis)	10	Epigastric pain	Blind pouch	PTCS	21	Excellent

Abbreviations: H-J, Roux-en-Y hepaticojejunostomy; C-J, cystojejunostomy performed at another hospital; PTCS, percutaneous transhepatic cholangioscopy.<sup>12</sup>

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

Shigeru Ono\*, Shigehisa Fumino, Shinichi Shimadera, Naomi Iwai

*Department of Pediatric Surgery, Graduate School of Medical Science, Kyoto Prefectural University of Medicine, Kyoto 602-8566, Japan*

Received 23 October 2009; accepted 27 October 2009

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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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**Table 1** Late complications (<10 years postoperatively)

Complications	n
Biochemical liver dysfunction	6
Persistent dilatation of intrahepatic bile duct	6
Recurrent abdominal pain	3
Presence of common bile duct adenocarcinoma	1

**Table 2** Late complications (>10 years postoperatively)

## Late complications

Biochemical liver dysfunction

Persistent dilatation of intrahepatic bile duct<sup>a</sup>

Recurrent abdominal pain

Adhesive small bowel obstruction

Pancreas divisum with pancreatic stones

Repeated cholangitis<sup>a</sup>Intrahepatic lithiasis and cholangiocarcinoma<sup>a</sup><sup>a</sup> Same patient.

# Long-term outcomes after excision of choledochal cysts at a single institution: Operative procedures and late complications

Yoshihiro Urushihara\*, Koji Fukumoto, Hiroaki Fukuzawa, Maki Mitsunaga, Masaharu Watanabe, Takeshi Aoba, Masaya Yamoto, Hiromu Miyake

*Department of Pediatric Surgery, Shizuoka Children's Hospital, 860 Urushiyama, Aoi-ku, Shizuoka 420-8660, Japan*

Received 22 August 2012; accepted 1 September 2012

## Abstract

**Purpose:** The purpose of this study was to evaluate long-term outcomes for a minimum of 3 years after cyst excision in children with choledochal cysts, focusing on the relationship between operative procedures and outcomes.

**Methods:** Between 1977 and 2008, 138 children underwent cyst excision. Follow-up results were obtained from 120 patients. Their mean age was 20.9 years (range 4–49). The mean interval between surgery and conducting the analysis was 16.6 years (range 3–34). These patients were divided into two groups based on their operative procedures: group A (1977–2000) comprising 76 patients who underwent cyst excision with hepaticojejunostomy below the hilum, and group B (2001–2008) comprising 44 patients who underwent excision of the extrahepatic bile duct from the confluence of the hepatic duct to near the level of the pancreatobiliary junction with wide hilar hepaticojejunostomy. When hepatic strictures were seen near the hilum, duct plasty was made. We evaluated the long-term outcomes in the two groups.

**Results:** Late complications were seen in a total of 18 patients (15.0%). In group A, 16 patients (21.1%) had late complications, which included cholangitis and/or hepatic stones in 9, stones in residual intrapancreatic cysts in 4, intestinal obstruction in 2, and pancreatitis in 1. Of these 16 patients, 12 patients (15.8%), including 7 with hepatic stones (6 IV-A and 1 Ic cysts), 4 with remnant intrapancreatic cysts, and 3 with intestinal obstruction underwent surgical intervention. In group B, none of the patients developed cholangitis, pancreatitis, or stone formation. However, 2 patients (4.5%) developed intestinal obstruction that required surgery.

**Conclusions:** Although a longer follow-up period is necessary, late complications were more frequent in group A than in group B patients and with type IV-A cysts. We believe that excision of the extrahepatic bile duct with wide hilar hepaticojejunostomy is essential for the prevention of postoperative complications.

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**Table 3** Intrahepatic stones and surgical management.

Age	Sex	Type of cyst	Initial operation (age)	Causes	Surgical management	Outcome (age)
25 y	F	IV-A	Cyst ex & H-J (12 y)	Anastomotic stricture	Revision H-J	Disease free (40 y)
18 y	F	Ic	Cyst ex & H-J (6 y)	Anastomotic stricture	Revision H-J	Disease free (39 y)
21 y	F	IV-A	Cyst ex & H-J (6 y)	Left hepatic duct stricture	PTCS	Disease free (37 y)
16 y	M	IV-A	Cyst ex & H-J (1 y)	Anastomotic stricture	Left hepatectomy & repeated PTCS	Recurrent hepatic stones & cholangitis (31 y)
20 y	F	IV-A	Cyst ex & H-J (10 d)	Left hepatic duct stricture	PTCS	Recurrent cholangitis (20 y)
17 y	M	IV-A	Cyst ex & H-J (16 y)	Anastomotic stricture	Revision H-J	Disease free (31 y)
16 y	F	IV-A	Cyst ex & H-J (8 y)	Anastomotic stricture	Revision H-J	Disease free (19 y)

Cyst ex indicates cyst excision; H-J, hepaticojejunostomy; PTCS, percutaneous transhepatic cholangioscopy.

**Table 4** Stones of the residual intrapancreatic cyst and surgical management.

Age	Sex	Type of cyst	Initial operation (age)	Causes	Surgical management	Outcome (age)
13 y	M	IV-A	Cyst ex & H-J (7 y)	Incomplete mucosectomy	Excision of residual cyst	Disease free (40)
30 y	F	IV-A	Cyst ex & H-J (2 y)	Incomplete resection	Excision of residual cyst	Disease free (33)
12 y	F	la	Cyst ex & H-J (7 y)	Incomplete resection	Excision of residual cyst	Disease free (34)
17 y	M	IV-A	Cyst ex & H-J (16 y)	Incomplete resection	Excision of residual cyst	Disease free (31)

Cyst ex indicates cyst excision; H-J, hepaticojejunostomy.

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# **Complications After Cyst Excision With Hepaticoenterostomy for Choledochal Cysts and Their Surgical Management in Children Versus Adults**

By Atsuyuki Yamataka, Kiyohiko Ohshiro, Yasuhiro Okada, Yataro Hosoda, Toshio Fujiwara, Sumio Kohno,  
Masakatsu Sunagawa, Shunji Futagawa, Noburu Sakakibara, and Takeshi Miyano  
*Tokyo, Shizuoka, and Tochigi, Japan*



**Table 2. Post-CEHE Complications in Children Versus Adults**

Incidence in 200 Children	Post-CEHE Complications	Incidence in 40 Adults
3	Ascending cholangitis	9
3	Intrahepatic bile duct stones	5
3*	Intrapancreatic terminal cho- ledochus calculi	1
1	Pancreatic duct calculus	1
1*	Stones in the blind pouch of the end-to-side Roux-en-Y hepaticojejunostomy	0
9†	Bowel obstruction	3‡
0	Cholangiocarcinoma	2
0	Liver dysfunction	1
5	Pancreatitis	5
25 (18)	Total	27 (17)

NOTE. The numbers in parentheses indicate the no. of patients who had post-CEHE complications (18 children and 17 adults had 25 and 27 post-CEHE complications, respectively).

\*One patient with intrapancreatic terminal choledochus calculi also had a stone in the blind pouch of the end-to-side hepaticojejunostomy.

†Adhesions in six and intussusception in three.

‡Adhesions in all three.

**Table 4. Number of Patients With Post-CEHE Stone Formation:  
Children Versus Adults**

	145 Children Aged $\leq 5$ yr*	55 Children $\geq 6$ yr*	40 Adults $\geq 16$ yr*
No. of patients with post-CEHE stones	0 (0%)†	7‡ (12.7%)	7 (17.5%)

NOTE. The percentages in parentheses indicate the incidence of post-CEHE stone formation.

\*Age at cyst excision combined with hepaticoenterostomy.

† $P < .0001$  for children aged  $\leq 5$  yr versus children aged  $> 5$  yr or adults.

‡One child formed two stones (one in the residual intrapancreatic terminal choledochus, the other in the blind pouch of the end-to-side Roux-en-Y hepaticojejunostomy).

## **Malignant change in the biliary tract after excision of choledochal cyst**

T. ISHIBASHI, K. KASAHARA, Y. YASUDA, H. NAGAI, S. MAKINO and K. KANAZAWA

*Department of Surgery, Jichi Medical School, Minamikawachi, Tochigi, Japan 329-04*

*Correspondence to: Dr T. Ishibashi*

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**Background** Choledochal cyst is a rare congenital condition with a high risk of malignant change if untreated. The risk of malignancy after surgical excision of choledochal cyst is not known.

**Methods** Forty-eight patients with choledochal cysts managed over a 21-year period were reviewed, to determine the risk of malignant change after cyst excision. Thirty-nine of 48 patients had no carcinoma at first admission; their mean(s.d.) age was 20(18) years. Thirty-seven of 39 patients underwent cyst excision and cholecystectomy followed by hepaticoenterostomy.

**Results** Cyst excision was incomplete in 28 of the 37 patients because dilated portions of the biliary ducts remained proximally and/or distally. In these 37 patients, no carcinoma has developed in the remnant proximal hepatic duct or the terminal bile duct after mean(s.d.) follow-up of 9.1(6.4) years. In the remaining nine patients, biliary carcinoma was diagnosed at the first visit. Six patients died from recurrence with a mean(s.d.) survival time of 13(11) months, while three patients were alive and free from recurrence 2 months, 1 year and 7 years after operation.

**Conclusion** Malignant change has not been observed after total or subtotal excision of choledochal cysts in this series.

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**Table 1** Choledochal cyst with biliary carcinoma

Patient no.	Age (years)	Sex	Cyst type	Site of carcinoma	Previous operation	Operation	Outcome
1	63	F	I	Cyst wall	Cholecystectomy Cystojejunostomy (10 years before)	Partial resection of cyst	Died at 11 months
2	28	M	I	Cyst wall	Cystoduodenostomy (27 years before)	Left hepaticostomy	Died at 6 months
3	48	F	IV-A	Cyst wall	Cholecystectomy (15 months before)	Cyst excision and hepaticojejunostomy	Died at 22 months
4	34	F	IV-A	Cyst wall	None	Pancreatoduodenectomy	Died at 32 months
5	66	F	I	Gallbladder	None	Cyst excision with hepaticojejunostomy; cholecystectomy with gallbladder bed resection	Died at 5 months
6	33	M	IV-A	Cyst wall	None	Right extended hepatectomy; cyst excision with hepaticojejunostomy	Alive at 86 months
7	60	F	I	Gallbladder	None	Exploratory laparotomy	Died at 2 months
8	59	M	I	Cyst wall	None	Cyst excision and cholecystectomy with hepaticojejunostomy	Alive at 12 months
9	47	M	V	Intrahepatic duct	None	Right extended hepatectomy	Alive at 2 months

Table 3 Reported cases of carcinoma developing after an excisional procedure

Reference	Year	Age when cancer detected (years)	Sex	Interval between cyst excision and detection of carcinoma	Cyst type	Site of carcinoma	Remarks
<b>Carcinoma in proximal duct</b>							
Thistlethwaite <i>et al.</i> <sup>25</sup>	1967	25	M	46 months	I	Anastomotic site	Anaplastic adenocarcinoma
Gallagher <i>et al.</i> <sup>26</sup>	1972	58	F	7 years	IV-A	Intrahepatic duct	Remnant intrahepatic involvement
Terauchi <sup>27</sup>	1978	25	M	15 years	IV-A	Intrahepatic duct	Remnant intrahepatic involvement or anastomotic stricture
Chaudhuri <i>et al.</i> <sup>28</sup>	1982	38	F	17 years	I (or IV-A)	Intrahepatic duct	Multiple calculi with anastomotic stricture; squamous cell carcinoma
Kaneta <i>et al.</i> <sup>29</sup>	1984	54	F	7 years	I	Intrahepatic duct	Multiple calculi with anastomotic stricture
Deziel <i>et al.</i> <sup>8</sup>	1986	—	—	—	IV-A	Intrahepatic duct	
Deziel <i>et al.</i> <sup>8</sup>	1986	—	—	—	IV-A	Remnant cyst wall	Insufficient excision
Joseph <sup>9</sup>	1990	29	M	3 years	I	Intrahepatic duct	Extensive cholangiocarcinoma
<b>Carcinoma in distal end</b>							
Miyoshi <i>et al.</i> <sup>30</sup>	1984	29	—	9 years	I	Distal remnant cyst wall	Insufficient excision
Kure <i>et al.</i> <sup>31</sup>	1984	35	M	4 years	IV-A	Head of pancreas	
Yoshikawa <i>et al.</i> <sup>32</sup>	1986	27	F	12 years	IV-A	Distal remnant cyst wall	Insufficient excision

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

J.S. de Vries, S. de Vries, D.C. Aronson, D.K. Bosman, E.A.J. Rauws, A. Bosma, H.A. Heij, D.J. Gouma, and T.M. van Gulik  
*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 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 for 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.

**Table 5. Incidence of Malignancy in Choledochal Cysts Reported in Literature**

Study, Year	No. of Patients	No. of Patients With Malignancies (%)	Malignancies After Internal Drainage (% of All Malignancies)	Age at Presentation of Malignancy
Lin et al, <sup>20</sup> 2000	80	8 (10)	3 (38)	50 (32-81)
Smuth and Krissat, <sup>21</sup> 1999	48	6 (13)	2 (33)	39 (17-57)
Enriot et al, <sup>22</sup> 1998	42	5 (12)	3 (60)	39 (29-51)
Switt et al, <sup>13</sup> 1995	14	2 (14)	0 (0)	46 (30-62)
Gain et al, <sup>23</sup> 1995	27	6 (26)	1 (17)	48 (34-60)
Possett et al, <sup>24</sup> 1994	42	3 (10)	0 (0)	Adults
Mijiiwa and Koga, <sup>25</sup> 1993	46	4 (9)	1 (25)	61 (42-71)
Bertson and Raine, <sup>26</sup> 1988	13	2 (15)	1 (50)	41 (41-41)
Adani et al, <sup>27</sup> 1987	82	8 (10)	3 (38)	?
Current study 2000	42	6 (14)	4 (67)	36 (20-62)
Total	437	50 (11)	18 (36)	

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# Intrahepatic cholangiocarcinoma developed after cyst excision for choledochal cyst

HIRO WATANABE, AKIRA TOKI, and TAKUJI TODANI

Department of Pediatric Surgery, Kagawa Medical University, 1750-1 Miki, Kita, Kagawa 761-0793, Japan

**Abstract:** Oncogenesis after cyst excision for choledochal cyst and suitable surgical procedures for this operation are discussed. The clinical data of 23 patients with cancer of the biliary tree after excision of choledochal cyst reported in the English-language and Japanese literature were reviewed, and data for 1353 Japanese patients with choledochal cyst and/or pancreaticobiliary malunion were analyzed. In the 23 patients reported in the literature, age at cyst excision ranged from 1 to 55 years (average,  $23.0 \pm 13.7$  years), and cancers were detected at age 18–60 years (average,  $32.1 \pm 12.2$  years), with intervals between cyst excision and cancer detection of 1–19 years (average,  $9.0 \pm 5.5$  years). Sites of cancer development were: intrahepatic, six; anastomotic, eight; hepatic side residual cyst, three; and the intrapancreatic duct, six. In the Japanese patients with choledochal cyst and/or pancreaticobiliary malunion, the incidence of cancer associated with primary choledochal cyst and/or pancreaticobiliary malunion was 16.2% (219/1353). The incidence of cancer development after cyst excision in this population, of whom 1291/1353 underwent surgery, was assumed to be 0.7%. Nearly half of the 23 patients in the literature had undergone inadequate cyst excision. Oncogenesis of cancers after cyst excision is possibly different from that of choledochal cysts.



**Table 1.** Bile duct carcinomas that developed after cyst excision

Patient number	Sex	Type	Age at internal drainage (years)	Age at cyst excision (years)	Procedures of the operation	Complications after cyst excision	Age at cancer detection (years)	Interval between cyst excision and cancer detection (years)	Year reported
Carcinomas developed in the intrahepatic bile duct									
1	F	?	—	19	Ex, HD jejunal interposition		20	1	1987 <sup>1</sup>
2	M	I	—	26	Ex, HJ		29	3	1990 <sup>2</sup>
3	F	I	—	47	Ex, HJ	Cholangitis, gallstone	54	7	1984 <sup>3</sup>
4	F	IV-A	36	51	Partial Ex, CJ	None (cholangitis before excision)	58	7	1972 <sup>4</sup>
5	M	?	11	32	Ex, HJ	Cholangitis	42	10	1994 <sup>5</sup>
6	F	IV-A	—	21	Ex, HJ	Cholangitis, gallstone	38	17	1982 <sup>6</sup>
Carcinomas developed in the hepatic duct at or near anastomosis									
7	M	I	—	21	Ex, HJ with Brown	None	25	4	1967 <sup>7</sup>
8	M	?	—	18	Ex, HJ		23	5	1990 <sup>8</sup>
9	F	I	—	55	Ex, HJ	None	60	5	1996 <sup>9</sup>
10	F	IV-A	—	40	Ex, HJ	None	45	5	1997 <sup>10</sup>
11	F	IV-A	—	27	Ex, HJ	Gallstone	33	6	1987 <sup>11</sup>
12	M	IV-A	—	10		Cholangitis, anastomotic stricture	25	15	1978 <sup>12</sup>
13	M	IV-A	—	20	Ex, HJ	Cholangitis	35	15	1988 <sup>13</sup>
14	F	IV-A	—	1	Ex, HD	None	19	18	1998 <sup>14</sup>
Carcinomas developed in the remnant choledochal cyst (hepatic side)									
15	F	?	—	16	Ex		18	2	1998 <sup>15</sup>
16	F	?	—	15	Ex		24	9	1998 <sup>15</sup>
17	F	?	—	16	Ex		35	19	1998 <sup>15</sup>
Carcinomas developed in the intrapancreatic remnant bile duct or choledochal cyst									
18	F	?	—	24	Ex, HJ		28	4	1982 <sup>16</sup>
19	F	I	—	17	Partial Ex		23	6	1985 <sup>17</sup>
20		I	—	20	Partial Ex	Cholangitis	29	9	1984 <sup>18</sup>
21	F	IV-A	—	11	Partial Ex, CJ		23	12	1988 <sup>19</sup>
22	F	IV-A	—	14	Ex, HJ	None	27	12	1986 <sup>20</sup>
23	M	?	—	8	Ex, HJ	None	25	17	1996 <sup>21</sup>

Ex, Cyst excision; HJ, hepaticojejunostomy; CJ, choledochojejunostomy; HD, hepaticoduodenostomy

2. Age distribution of patients with choledochal cyst and/or pancreaticobiliary malunion and cancer (registered in Japan, 1997) and patients with cancer developed after cyst excision (present review of literature)

Age at operation (years)	Number of patients with choledochal cyst and/or pancreaticobiliary malunion	Number of patients with bile duct cancer	Incidence of cancer (%)	Number of patients with cancer after cyst excision
< 10	461	0	0.0	
10-19	88	0	0.0	2
20-29	130	3	2.3	12
30-39	106	6	5.7	4
40-49	135	34	25.2	2
50-59	156	62	39.7	2
60-69	123	53	43.1	1
70-79	63	31	49.2	
80-89	8	6	75.0	
≥ 90	21	5	23.8	
Total of operated patients	1291	200	15.5	
Not operated or treatment unknown	62	19	30.7	
	1353	219	16.2	23

# CONCLUSION

Excellent prognosis with early total resection and reconstruction

Long-term surveillance for the development of malignancy still essential

**THANK YOU FOR YOUR ATTENTION**

