Total cyst excision with Roux-en-Y hepaticojejunostomy for choledochal cyst management: a single-center experience

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Background
Choledochal cystis considered as a cystic dilatation of the biliary tract, which is a rare disease. It is more common in children, but its incidence and diagnosis are increasing in adults, representing 20% of the cases.
Aim
To evaluate morbidity and mortality rates following hepaticojejunostomy done for patients, either child or adult, with choledochal cyst.
Patients and methods
A total of 30 patients were evaluated, comprising 13 children and 17 adults. The study was done in the period from January 2013 till January 2020, in the Advanced Hepato-Pancreatico-Biliary Center, Zagazig University Hospital, Egypt.
Results
Abdominal pain was the commonest complaint in all patients. Jaundice was present in 80%. Total cyst resection with reconstruction of the biliary tract with a standard 60–60 cm Roux-en-Y hepaticojejunostomy was done in 28 (93.33%) patients. Left hepatectomy was done in two patients with Caroli's disease. There was no recurrence in our follow-up period.
Conclusion
Total cyst excision with Roux-en-Y hepaticojejunostomy is the standard treatment of choledochal cyst. It was associated with low incidence of recurrence and also decreased long-term postoperative complications and malignancy.

Keywords:
choledochal cyst, hepaticojejunostomy and Caroli's disease, Roux-en-Y

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Introduction

Choledochal cyst is defined as a cystic dilation of either extrahepatic biliary tree or intrahepatic bile ducts, or sometimes both of them [1]. The age at diagnosis is mostly in children younger than 10 years. The prevalence rate is 25% in the first year of life, whereas 80% in the first decade, and 20% in adulthood [2]. It has a higher prevalence in females than males (4 : 1), and it is increasingly diagnosed in adult patients [3,4].

Todani classified choledochal cyst according to its location into five types [5–7]. Caroli's disease was considered as type V, defined as a cystic dilation of intrahepatic part of the biliary system, which may be associated with cirrhosis and periportal fibrosis, in a lobe or bilobal [1].

Pancreatic or biliary tract symptoms are usually found in adult type, whereas typical triad of right upper quadrant pain, abdominal mass, and jaundice is found in children [8].

Regarding pathology, according to Balbbitt [6], there is an anomalous junction between the common bile and pancreatic ducts, which creates a long common channel before it enters the duodenal wall. This pathology was found in 80–90% of patients with choledochal cysts [9].

So at first, cyst excision with hepaticojejunostomy was accepted as the standard treatment for choledochal cyst, but it had serious postoperative complications, including hepatic stones, cholangitis, pancreatitis, and development of carcinoma in the bile duct remnant, which were found during long-term follow-up after cyst excision [10].

The decision shifted to total cyst excision with the extrahepatic bile duct up to near the level of the pancreaticobiliary junction, followed by Roux-en-Y hepaticojejunostomy, which is recommended to prevent these complications [11,12].

Diagnosis usually begins with ultrasound or contrast-enhanced computed tomography (CT)

and magnetic resonance cholangiopancreatography (MRCP) to determine the cyst type [13]. For infants or children, MRCP is the diagnostic modality of choice [14]. In adults, MRCP has an advantage of being noninvasive and its ability to characterize cyst anatomy in relation to the biliary tree and surrounding structures [15]. However, endoscopic retrograde cholangiopancreatography (ERCP) used in complicated cyst as a therapeutic procedure when needed [16].

The present study aims to demonstrate the surgical treatment and outcomes of the patients with choledochal cyst.

Patients and methods

This is a retrospective study done on 30 patients presented with choledochal cyst to the Advanced Hepato-Pancreatico-Biliary Center in Zagazig University in the period from January 2013 till January 2020. We take the ethical approval and patient consent before starting the research.

Demographic characteristics were investigated. Operative procedures, early morbidity, mortality, and later results from the surgical procedures were recorded. The types of choledochal cysts, based on radiologic and operative findings, in our study, were assigned according to the classification of Todani. Histopathological confirmation of the diagnosis was done in all patients.

Diagnosis is usually suspected with ultrasound or CT and confirmed with MRCP. ERCP is done in cases with adult type complicated with repeated attacks of cholangitis (Fig. 1).

Operative details

Type I (Fig. 2):

(1) A right subcostal incision is made in the adult type, whereas in children, transverse supra-umbilical incision is done.
(2) The cyst is usually easily identified and easily freed of attachments. The dissection is approached from a combination of cephalic and caudal positions.
(3) The gallbladder was removed and intraoperative cholangiography was done from the cystic duct to detect the cyst type and to determine the cyst extension.
(4) We should reach the distal-most portion of the cystic dilatation and encircle it as it enters the pancreas.
(5) The duct is then transected as distal as possible provided no injury to pancreatic duct.
(6) Proximally, the cyst was usually transected just below the hepatic bifurcation.
(7) After totally excision of the cyst, a standard 60–60 cm Roux-en-Y loop is used for an end-to-side hepaticojejunostomy with biliary stent (epidural stent) at both right and left hepatic duct to be removed after 3 months.
(8) The removed cyst was sent for histopathology to rule out malignancy and safety margins.

Type IV:

(1) Midline extension of the wound was done (J-shaped incision) to facilitate left lateral hepatic resection.
(2) Extrahepatic biliary resection, as with type I cysts, was done.
(3) The entire cystic portion of the extrahepatic biliary tree should be resected when possible.
(4) Reconstruction was done at the hepatic duct bifurcation with separate right and left duct hepaticojejunostomy with two biliary stents.

Type V (Caroli's disease):

(1) In our patients with Caroli's disease, the biliary dilatation was confined to the left hepatic lobe (two cases).
(2) Left hepatectomy was done through J-shaped incision.
(3) The common bile duct (CBD) was not removed.
(4) Biliary stent was inserted inside CBD extended to right hepatic duct to be removed after 3 months.

All our patients were followed up in our clinic at 1 week, 1 month, and 3 months after discharge and at 6 monthly intervals thereafter. The range of follow-up period was from 6 months to 7 years, with a mean of 2±3.6 years. All patients during the follow-up period underwent liver function test, CA 19-9, and ultrasound. MPCP was done only in complicated patients with jaundice and hepaticojejunostomy stricture.

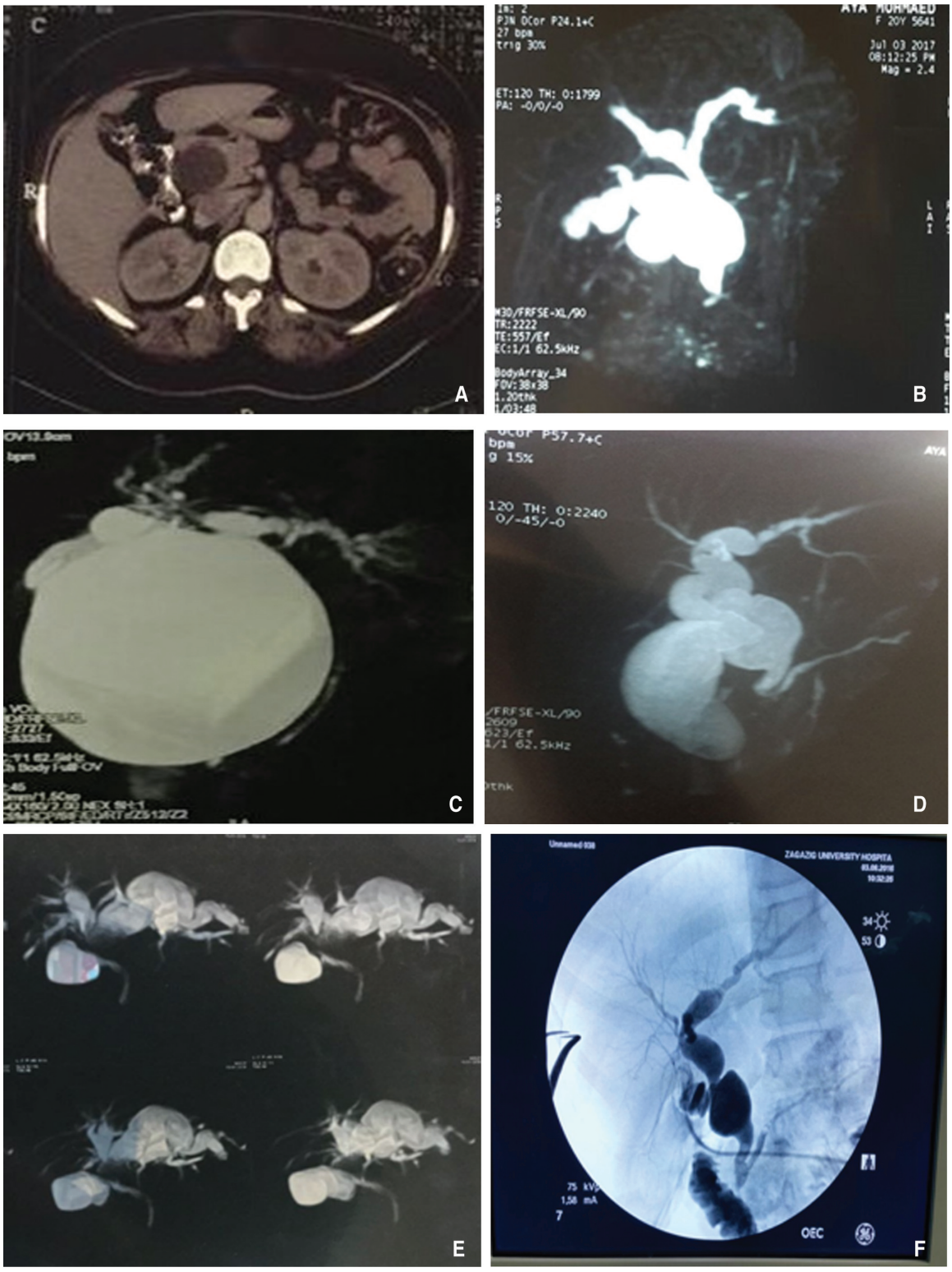
Statistical analyses

Data are presented as number (%) or mean±SD and were statistically analyzed using SPSS software, version 19.0 (SPSS Inc., Chicago, Illinois, USA).

Results

A total of 30 female patients with choledochal cyst were included in this study. Our study included 13

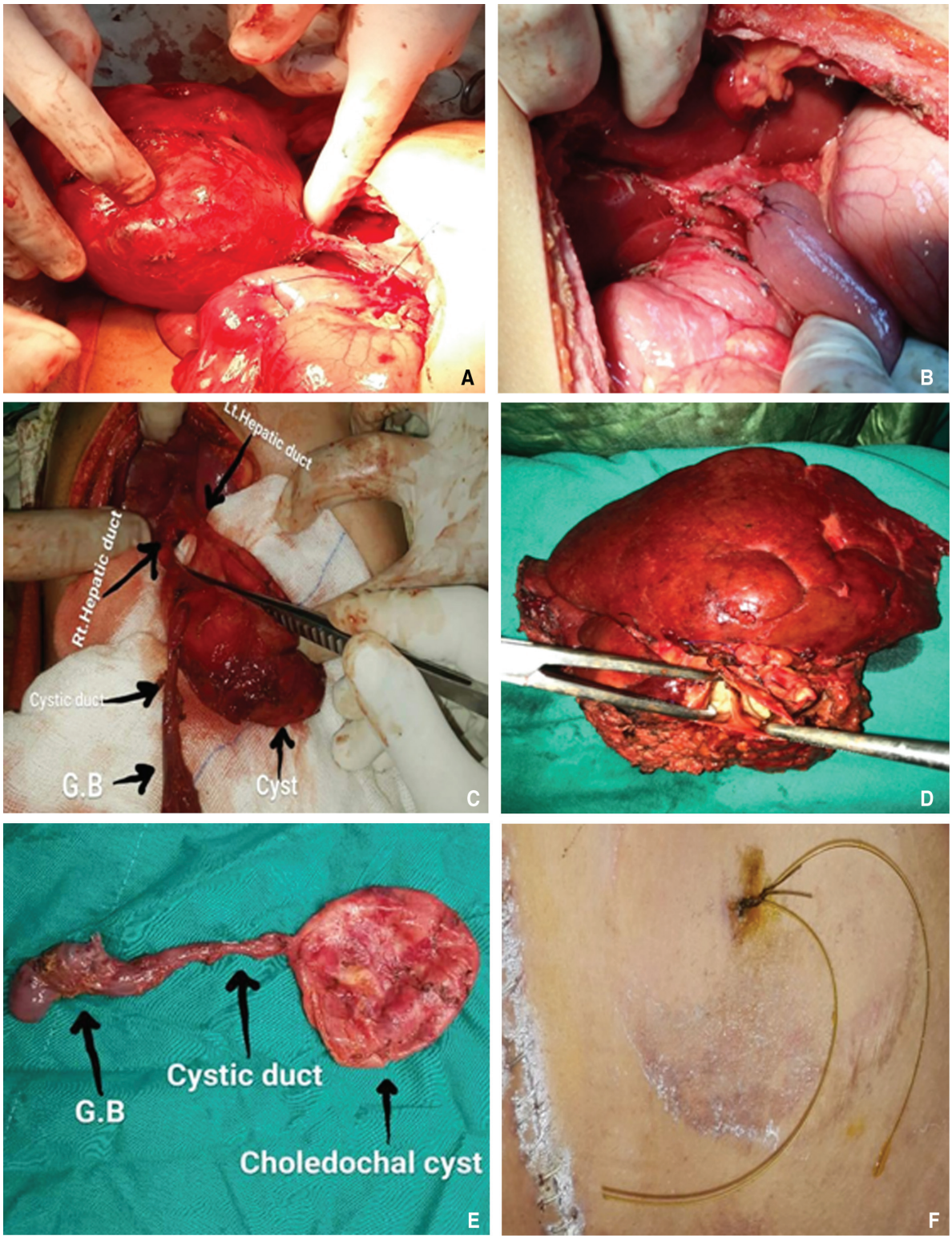
Figure 1



Types of choledochal cyst. (a) CT scan: type I choledochal cyst. (b) MRCP: type I choledochal cyst. (c) MRCP: type IVB choledochal cyst. (d) MRCP: type IVA choledochal cyst. (e) MRCP: type V choledochal cyst (Caroli's disease). (f) Intraoperative cholangiogram: type IVA extending to left lateral duct. CT, computed tomography; MRCP, magnetic resonance cholangiopancreatography.

(43.33%) children and 17 (56.67%) adults, with mean age of 18±8.6 years old (5–48 years). Symptoms, associated hepatobiliary disease in adult patients, and type of the cysts are demonstrated in Table 1. Seven patients had biliary stones that needed preoperative ERCP and stenting. Two adult patients with type IVA cyst had previously undergone laparoscopic cholecystectomy.

Figure 2



Intraoperative details of surgical management of choledochal cyst. (a) Huge type I choledochal cyst. (b) Roux-en-Y hepaticojejunostomy. (c) Type IVB choledochal cyst. (d) Left hepatectomy in type V choledochal cyst. (e) Excised choledochal cyst with gallbladder. (f) Epidural stent as biliary stent inside the hepaticojejunostomy.

A total extrahepatic total cyst excision with a standard 60–60 cm Roux-en-Y hepaticojejunostomy was performed in type I, type IVA, and type IVB cyst in 28 (93.33%) patients. In the two patients with Caroli's disease, there was no extrahepatic biliary dilatation so left hepatectomy was done alone without hepaticojejunostomy. Only one 5-year-old child had three separate anastomosis between (segment IV duct, right anterior sector duct, and right posterior sector duct) with the jejunal lobe.

Table 1 Demographic data of all patients with biliary cystic dilatation

Demographic data	Children (*N*=13) [*n* (%)]	Adult (*N*=17)	Total (*N*=30)
Age	4–5 years: 6 (46.15)	20–48 years	5–48 years
	6–8 years: 2 (15.38)	25±6.3 years	18±8.6
	12–17 years: 5 (38.47)		
Symptoms			
Abdominal pain	13	17	30
Classical triad	9 (69.23)	2 (11.76)	11 (36.67)
Jaundice	11 (84.61)	14 (82.35)	25 (83.33)
Recurrent cholangitis	2 (15.38)	12 (70.59)	14 (46.67)
Associated hepatobiliary disease in adult patients			
Gallbladder stones		4 (23.53)	2 Caroli's disease and 2 type I
CBD stones		7 (41.18)	3 type I, 2 type IVA, and 2 Caroli's disease
Intrahepatic stones		2 (11.76)	Caroli's disease
Liver abscess		1 (5.88)	Type IVA
Type of the cyst			
Type I	8 (61.54)	11 (64.7)	19 (63.33)
Type IVA	2 (15.39)	2 (11.67)	4 (13.33)
Type IVB	3 (23.07)	2 (11.67)	5 (16.67)
Type V		2 (11.67)	2 (6.67)

CBD, common bile duct.

Table 2 Operative details of all patients with choledochal cysts

	Children (*N*=13)	Adult (*N*=17)	Total (*N*=30)
Type of operation			
Roux-en-Y HJ	11	13	28
Liver resection	2	2	6
Left lateral with HJ		2	4
Formal left without HJ			2
Number of anastomosis [*n*/*N* (%)]			
Type I cyst	8/13 (61.54)	11/17 (82.35)	19/30 (63.33)
Single anastomosis	6/13 (46.15)	8/17 (64.71)	14/30 (46.67)
Two in one	1/13 (7.69)	2/17 (11.76)	3/30 (10)
Two anastomoses	1/13 (7.69)	1/17 (5.88)	2/30 (6.67)
Type IVA	2/13 (15.38)	2/17 (11.76)	4/30 (13.33)
Two anastomoses	1/13 (7.69)	2/17 (11.76)	3/30 (10)
Three anastomoses	1/13 (7.69)		1/30 (3.33)
Type IVB	3/13 (23.07)	2/17 (11.76)	5/30 (16.67)
Two anastomoses	3/13 (23.07)	2/17 (11.76)	5/30 (16.67)

HJ, hepaticojejunostomy.

Type of operation and number of anastomosis are demonstrated in Table 2.

The operation time was high in type IVA and type V, about 190±42 min, whereas was 140±22 min in type I cyst. Blood and plasma transfusion were needed only in the six patients with liver resection.

The total hospital stay was 5±2.3 days. All the preschool patients needed ICU stay, whereas three adult patients needed ICU stay (one patient with type IVA cyst who had liver abscess and the two patients with Caroli's disease). The average ICU stay was 2±1.7 days. There was no postoperative mortality in our study. Postoperative complications occurred in six (20%) patients, including biliary leakage, wound infection, chest infection, and pleural effusion. All had been recovered with conservative treatment.

There was no recurrence in our patients. Only one 28-year-old female patient with type IVB cyst underwent two anastomosis. She developed biliary stricture after 16 months, which was managed by one session of percutaneous balloon dilatation. Another 6-year-old child experienced slipped biliary stent in the hepatic ducts; the patient passed the stent spontaneously in the stool after 8 months from the operation.

Discussion

A choledochal cyst, which is believed to be congenital in origin, may occur at any age. Overall, 20% of cases

occur in patients older than 20 years, but the majority affects children younger than 10 years [17]. In our study, all 30 cases were female patients, where 13 (43.33%) cases were children and 17 (56.67%) were adult, with mean age of 18±8.6 years old (5–48 years). Regarding etiology, there is a theory concerning the reflux of pancreatic enzymes into the common bile duct, but this does not explain type II and type V cysts, which may have a genetic component [18].

Regarding presentations, the classical triad is found to be rare in adults (only 0–17% of cases). However, at least two elements of the triad were present in ~85% of children and 25% of adults. Pancreatitis, cholangitis, or biliary peritonitis owing to cyst rupture may also be a presentation [19]. In our study, the most common symptom was right hypochondrium abdominal pain presented in all patients. Classical triad was presented in 69.23% of children and 11.76% of adult patients. Jaundice was presented in more than 80% of our patients, and more than 70% of adult patients were presented with recurrent attacks of cholangitis.

In our study, type I choledochal cyst was presented in 63.3% of our series. This was nearly the same finding by the study by Jordan *et al.* [8], presenting in 56% of the cases. However, it presented in 85% of the cases in the study by O'Neill [20]. Ultrasound is the normal first step imaging [19]. CT is less accurate than MRCP, whereas ERCP is good in managing complicated cases. MRCP provides images similar to ERCP, without the risk of its complications, especially pancreatitis [16,21].

Surgical treatment of choledochal cyst has changed over the years. Cystoenterostomy was the start, but it is no longer considered a way of management owing to its complications (cholangitis, pancreatitis, intrahepatic stones, and malignant transformation). Now, the standard treatment involves excision of the cyst and bilioenteric derivation [3,22].

So cyst excision with hepaticoenterostomy was the treatment of choice for choledochal cyst, but it is no longer used as well, owing to its complications especially in cases of choledochal cyst with intrahepatic involvement (type IVA cyst) [23,24]. The main cause of its complications resulted from bile stasis. Bile stasis may occur from anastomotic strictures, residual intrahepatic ductal dilatation, residual intrapancreatic cysts, and intrahepatic bile duct strictures [25].

Accordingly, excision of the extrahepatic bile duct from the confluence of the hepatic duct to near the level of the pancreatobiliary junction, together with wide hilar hepaticojejunostomy, is recommended as the treatment for choledochal cyst [11,26]. However, long-term follow-up is mandatory to assess the outcomes of biliary reconstruction for choledochal cyst [12], as it still remains debatable whether or not wide hepaticojejunostomy decreases the risk of postoperative complications. In our study, we did total excision of the cyst including the intrapancreatic portion with a standard 60–60-cm-wide hilar hepaticojejunostomy.

Total cyst excision with Roux-en-Y hepaticojejunostomy is the standard surgical procedure for most type I, IVB, and many IVA, but owing to variations in cyst anatomy (especially intrahepatic dilatation), more extensive and demanding operations may be required. Liver transplantation is rarely necessary for treatment of type IVA cysts unless the entire liver is involved. So with Caroli's disease, where cystic involvement is diffuse and malignant transformation is almost always present, partial hepatectomy and transplantation may be required [27,28]. In our series, we did left lateral hepatectomy in four patients of type IVA cyst with wide hilar Roux-Y hepaticojejunostomy. Moreover, we did left formal hepatectomy in the two patients of type V (Caroli's disease) to perform complete excision of the intrahepatic biliary dilatation.

Kasai *et al.* [29] first found an obvious relation of choledochal malformation with malignancy in 1970, but it remains unknown what percentage of biliary cancers arise in cysts. Then Watanabe *et al.* [30] reviewed data from 73 Japanese institutions in 1987 and found 154 cases of malignancy from 881 collected choledochal cysts (i.e. an incidence of malignant change of 17%). However, still the incidence of malignancy malformation is rare in the general population, with an incidence of less than three per 100 000 population (including gallbladder, intrahepatic and extrahepatic ducts) [31]. Jordan *et al.* [8] calculated that the development of bile duct carcinoma in residual choledochal walls and was significantly higher (121.5 times) than in the normal population.

The incidence of late complications after cyst excision ranges from 5 and 30% [32,33]. The study by Urushihara *et al.* [12] evaluated the long-term outcomes in 120 cases with choledochal who were managed by cyst excision with hepaticojejunostomy (group A), or excision of extrahepatic bile duct with

wide hilar hepaticojejunostomy (group B). They found overall late complication rate of 15.8%, with late complications being more frequent in group A than in group B patients (21.1 vs. 4.5%). In our study during the long-term follow-up, no recurrence or malignancy was detected. Only one patient with type IVB developed stricture at the hepaticojejunostomy after 18 months from the operation. One session of percutaneous balloon dilatation was sufficient to solve the problem with no recurrence during the follow-up afterward.

Conclusion

In conclusion, the standard technique used to treat choledochal cyst is excision of the extrahepatic bile duct from the confluence of the hepatic duct to near the level of the pancreatobiliary junction, together with wide hilar hepaticojejunostomy, which is essential for the prevention of postoperative complications. Although a longer follow-up period is necessary, our study revealed satisfactory long-term results.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1 Abbas H, Yassin N, Ammori B. Laparoscopic resection of type I choledochal cyst in an adult and Roux-en-Y Hepaticojejunostomy: a case report and literature review. Surg Laparosc Endosc Percutan Tech 2006; 16:439–444.

2 Soares K, Kim Y, Spolverato G, Maithel S, Bauer T, Marques H, *et al.* Presentation and clinical outcomes of choledochal cysts in children and adults: a multiinstitutional analysis. JAMA Surg 2015; 150:577–584.

3 Fonseca-Neto O, Albuquerque-Neto M, Miranda A. Surgical managements of cystic dilatation bile ducts in adults. Arq Bras Cir Dig 2015; 28:17–19.

4 Akaraviputh T, Boonnuch W, Watanapa P, *et al.* Surgical management of adult choledochal cysts. J Med Assoc Thai 2005; 88:939–943.

5 Atkinson H, Fischer C, Jong C, *et al.* Choledochal cysts in adults and their complications. HPB (Oxford) 2003; 5:105–110.

6 Balbbitt D. Congenital choledochal cysts: new etiological concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol 1969; 12:231–240.

7 Benjamin I. Biliary cystic disease: the risk of cancer. J Hepatobiliary Pancreat Surg 2003; 10:335–339.

8 Jordan P, Goss J, Rosenberg W, Woods K. Some considerations for management of choledochal cysts. Am J Surg 2004; 187:672.

9 Ronnekleiv-Kelly S, Soares K, Ejaz A, Pawlik T. Management of choledochal cyst. Curr Opin Gastroenterol 2016; 32:225–231.

10 Todani T, Watanabe Y, Urushihara N, Noda T, Morotomi Y. Biliary complications after excisional procedure for choledochal cyst. J Pediatr Surg 1995; 30:478–481.

11 Stringer M. Wide hilar hepaticojejunostomy: the optimum method of reconstruction after choledochal cyst excision. Pediatr Surg Int 2007; 23:529–532.

12 Urushihara N, Fukumoto K, Fukuzawa H, Mitsunaga M, Watanabe K, Aoba T, *et al.* Long-term outcomes after excision of choledochal cysts in a single institution: operative procedures and late complications. J Pediatr Surg 2012; 47:2169–2174.

13 Park D, Kim M, Lee S, Lee S, Choi J, Lee Y, *et al.* Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts? Gastrointest Endosc 2005; 62:360–366.

14 Hung M, Lin L, Chen D, Huang C. Choledochal cysts in infants and children: experiences over a 20-year period at a single institution. Eur J Pediatr 2011; 170:1179–1185.

15 Sacher V, Davis J, Sleeman D, Casillas J. Role of magnetic resonance cholangiopancreatography in diagnosing choledochal cysts: case series and review. World J Radiol 2013; 5:304–312.

16 Kim S, Lim J, Yoon H, Han B, Lee S, Kim Y. Choledochal cyst: comparison of MR and conventional cholangiography. Clin Radiol 2000; 55:378–383.

17 Lipsett PA, Henry AP. Surgical treatment of choledochal cysts. J Hepatobiliary Pancreat Surg J 2003; 10:352–359.

18 Singham J, Schaeffer D, Yoshida E, Scudamore C. Choledochal cysts: analysis of disease pattern and optimal treatment in adult and pediatric patients. HPB (Oxford) 2007; 9:383–387.

19 Visser B, Suh I, Way L, Kang S. Congenital choledochal cysts in adults. Arch Surg 2004; 139: 855–862.

20 O'Neill J. Choledochal cysts. Curr Probl Surg 1992; 29:371–410.

21 Adamek H, Shilling D, Weitz M, Riemann J. Choledochocele imaged with magnetic resonance cholangiography. Am J Gastroenterol 2000; 95:1082–1083.

22 Yamaguchi M, Sakurai M, Takeushi S, Awazu S. Observation of cystic dilatation of the common bile duct by ultrasonography. J Pediatr Surg 1980; 15:207–210.

23 Urushihara N, Fukumoto K, Fukuzawa H, Morimichi Tani M, Matsuoka T, Suzuki K, *et al.* Hepaticojejunostomy and intrahepatic cystojejunostomy for type IV-A choledochal cyst. J Pediatr Surg 2007; 42:1753–1756.

24 Tsuchida Y, Takahashi A, Suzuki N, Kuroiwa M, Murai H, Toki F, *et al.* Development of intrahepatic biliary stones after excision of choledochal cysts. J Pediatr Surg 2002; 37:165–167.

25 Kobayashi S, Asano T, Yamasaki M, Kenmochi T, Nakagohri T, Ochiai T. Risk of bile duct carcinogenesis after excision of extrahepatic bile ducts in pancreaticobiliary maljunction. Surgery 1999; 126:939–944.

26 Fei F, Da-Peng X, Zheng-Xiang X, Hai-Jia L, Hai-Bei X, Huan Z, Jin-Wei Z. Clinical significance of intrapancreatic choledochal cyst excision in surgical management of type I choledochal cyst. Int Med Res J 2018; 46: 1221–1229.

27 Kawarada Y, Das B, Tabata M, Isaji S. Surgical treatment of type IV choledochal cysts. J Hepatobiliary Pancreat Surg 2009; 16:684–687.

28 Pal K, Singh V, Mitra D. Partial hepatectomy and total cyst excision is curative for localized type IV-a biliary duct cysts – report of four cases and review of management. Eur J Pediatr Surg 2009; 19:148–152.

29 Kasai M, Asakura Y, Taira Y. Surgical treatment of choledochal cyst. Ann Surg 1970; 172:844–851.

30 Watanabe Y, Toki A, Todani T. Bile duct cancer developed after cyst excision for choledochal cyst. J Hepatobil Pancreat Surg 1999; 6:207–212.

31 West J, Wood H, Logan R, Quinn M, Aithal G. Trends in the incidence of primary liver and biliary tract cancers in England and Wales 1971–2001. Br J Cancer 2006; 94:1751–1758.

32 Shah O, Shera A, Zargar S, Shah P, Robbani I, Dhar S, Khan A. Choledochal cysts in children and adults with contrasting profiles: 11-year experience at a tertiary care center in Kashmir. World J Surg 2009; 33:2403–2411.

33 Ono S, Fumino S, Shimadera S, Lwai N. Long-term outcomes after hepaticojejunostomy for choledochal cyst: a 10- to 27-year follow-up. J Pediatr Surg 2010; 45:376–378.