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The distal classification and management of choledochal cyst in adults

Based on the relation between cyst and pancreatic duct

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Abstract

Todani classification is extensively used to guide the surgical strategy of choledochal cysts, but no systematic investigations on the distal management of intrapancreatic choledochal cysts have been conducted. This study reports the distal classification and management of choledochal cysts in adults based on the relation between the cyst and pancreatic duct. Patients with choledochal cyst who underwent operation, including distal management, in our department from January 2009 to December 2014 were retrospectively reviewed. Patients presenting symptoms, coexisting diseases, surgical treatment, perioperative complications, and long-term follow-up according to the distal classification of choledochal cyst were analyzed. A total of 54 patients with choledochal cyst were included in the present retrospective study. Based on the distal classification of choledochal cyst, 39 patients (72.22%) were type 1, 13 patients (24.07%) were type 2, and 2 patients (3.70%) were type 3. Thirty-nine type 1 patients and 10 type 2 patients underwent excision of intrapancreatic choledochal cyst or bile duct. Three type 2 patients received excision of distal cylindrical cyst and papilla, followed by pancreatic duct plasty with duodenum mucosa. One type 3 patient underwent endoscopic sphincteroplasty, and another type 3 patient underwent transduodenal sphincteroplasty. After the operation, 11 patients (20.37%, 11/54) had short-term perioperative complications. The long-term follow-up results showed that the satisfactory rate (excellent and good outcomes) was 95.83%. Current distal classification of choledochal cysts could provide a more targeted strategy for complete excision to eliminate potential dead space within the pancreas, protect the pancreatic duct, and prevent reoperation.

Abbreviations: CT = computed tomography, MRCP = magnetic resonance cholangiopancreatography.

Keywords: choledochal cyst, distal classification, pancreatic duct, surgical management

1. Introduction

Choledochal cysts are characterized by varying degrees of congenital dilatation of the biliary system including the intrahepatic and extrahepatic duct, with an incidence of 1 in 100,000 to 150,000 individuals in Western populations and approximately 1 in 13,000 individuals in Asian populations.[1] Presence of an anomalous junction of the pancreaticobiliary duct that allows pancreatic secretions to reflux into the biliary tree is the generally accepted etiopathogenic theory of these cysts.[2,3] Nearly 20% of choledochal cysts are present in adults, and malignant transformation is the most feared complication.[4] A 6% to 30% risk of developing malignancy is estimated in patients with choledochal cysts; the risk is low in childhood (<1%) but increases to about 30% to 40% with age greater than 50 years.[5,6]

Currently, to avoid complications from remnant cyst excision and reoperation, the best surgical strategy is to excise the cyst areas completely (including gallbladder) with hepaticojejunostomy. Surgical treatment depends on the anatomic findings and extent of biliary involvement according to the widely used Todani classification.[7] Complete cyst excision includes proximal and distal excision. Complete cyst excision to the normal proximal bile duct is the standard procedure, which is accompanied with additional hepatectomy if necessary. As the cyst extends into the pancreas, its management is also crucial and sometimes difficult. Injury to the pancreatic duct and surrounding tissues may cause serious complications, such as recurrent pancreatitis, bleeding, stone formation, and carcinoma.[8] Furthermore, remnant intrapancreatic choledochal cysts may give rise to subsequent malignant transformation with an estimated risk of 0.7% to 6%.[9]

To date, no systematic investigations have been conducted on the distal management of intrapancreatic choledochal cysts. Based on the above situation and our surgical experience, we propose the following classification concerning the distal management of choledochal cysts (Fig. 1): type 1, no relation

Editor: Neil Merrett.

Funding/support: The research was supported by Shandong Provincial Natural Science Foundation of China (no. 2015ZRE2760).

The authors have no conflicts of interest to disclose.

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Medicine (2017) 96:12(e6350)

Received: 25 August 2016 / Received in final form: 18 February 2017 / Accepted: 20 February 2017

http://dx.doi.org/10.1097/MD.0000000000006350

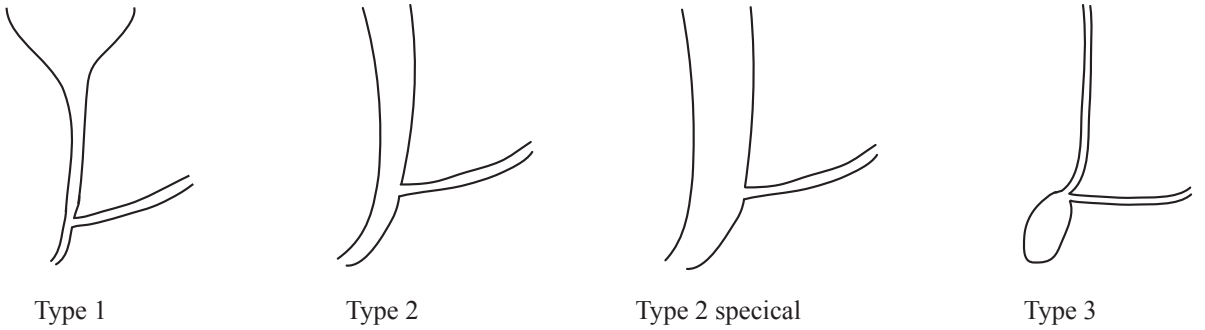

Figure 1. The current distal classification of choledochal cysts: type 1, no relation between choledochal cyst and pancreatic duct; type 2, close relation between the above twos; type 2 special subtype, the pancreatic duct converges with the cylindrical cyst; type 3, accordance with type III of Todani classification.

between choledochal cyst and pancreatic duct; type 2, close relation between the twos; and type 3, accordance with type III of Todani classification. Thus, this study aimed to report our experience with the distal management of choledochal cysts using our classification.

2. Materials and methods

2.1. Patients

From January 2009 to December 2014, the medical records of adult patients (aged ≥16 years) with choledochal cysts at the Department of Hepatobiliary Surgery, Qilu Hospital of Shandong University, were reviewed retrospectively. Clinical characteristics assessed included patient demographic information, presenting symptoms, surgical treatments, pathological results, perioperative complications, and long-term outcomes. The choledochal cyst was diagnosed by radiologic imaging including computed tomography, magnetic resonance cholangiopancreatography, and ultrasonography. Diagnosis without malignant transformation was confirmed by histopathologic findings. Todani types II and V were not included in our study profile because the present study only focused on the distal management of choledochal cysts. The patients were classified into 3 types according to the current distal classification. The study protocol was approved by the Ethics Committee of Qilu Hospital of Shandong University.

2.2. Surgical management

Given that incomplete cyst excision can lead to recurrent symptoms and malignant transformation within the remnant cyst, current surgery includes the excision of the entire cyst (including gallbladder) and restoration of biliary-enteric continuity.[10] The proximal extent should be identified and evaluated before resection. For Todani type IVa with extrahepatic and intrahepatic cysts, the extrahepatic cyst should be firmly excised; if the intrahepatic cyst is limited, partial hepatectomy with biliary-enteric reconstruction can be performed.

The distal management of choledochal cysts was performed according to our distal classification. For type 1 with no relation between choledochal cyst and pancreatic duct based on preoperative imaging, the distal cyst or seemingly normal bile duct was excised to about 5 mm from the convergence with the pancreatic duct and the stump was sutured.[11] For type 2, the same management principle as type 1 was adopted. Occasionally, a special subtype of type 2 with cylindrical cyst and insufficiency of Vater papilla can occur. The distal end of the choledochal cyst directly enters the duodenum with insufficiency of Vater papilla, and the pancreatic duct looks like an attachment that converges with the large cyst. In this situation, the distal cylindrical cyst and papilla are excised, followed by pancreatic duct plasty with duodenum mucosa, which is modified from local resection of benign neoplasm and early noninvasive adenocarcinoma in papilla.[12,13] In consideration of the low malignant rate in type 3 cysts, unroofing (endoscopic or transduodenal sphincteroplasty) or transduodenal excision (larger cysts) is optimal for choledochoceles.[1,14]

2.3. Assessment of postoperative complications and surgical outcomes

The Clavien–Dindo classification is applied to evaluate postoperative complications and consists of 7 grades (I, II, IIIa, IIIb, IVa, IVb, and V); this classification is valid and applicable worldwide in many fields of surgery.[15] In general, each complication has a corresponding grade, and the final grade of complications is adopted the highest. The patient's surgical outcome was assessed subjectively as excellent (no symptoms, attributable to the biliary tract injury or reconstruction), good (mild symptoms, not requiring invasive investigation or treatment), or failure (ongoing symptoms or stricture recurrence, requiring an invasive investigational or therapeutic procedure). Patients classified as either excellent or good were considered successful treatment.[16]

3. Results

A total of 54 patients with choledochal cyst were included in the present retrospective study. The mean patient age at diagnosis was 36.46 ± 12.44 years (range, 16–71 years). Thirty-two (59.26%) patients were in the range of aged 30 to 49 years. The presenting symptoms and coexisting diseases were shown in Table 1.

3.1. Classification based on the Todani and distal classification of choledochal cyst

According to the Todani classification of choledochal cyst, 37 patients (68.52%) were type I, 2 patients (3.70%) were type III, and 15 patients (27.78%) were type IVa. Meanwhile, based on the distal classification of choledochal cyst, 39 patients (72.22%) were type 1, 13 patients (24.07%) were type 2, and 2 patients (3.70%) were type 3. Among the Todani type I patients, 29 patients were type 1 and 8 were type 2 (Table 2).

Table 1

The characteristics of patients with choledochal cysts in present study.

Characteristics	n (%)
Age at admission, y	
6–19	7 (12.96)
20–29	9 (16.67)
30–39	16 (29.63)
40–49	16 (29.63)
50–59	4 (7.41)
60–69	1 (1.85)
70–79	1 (1.85)
Gender	
Male	17 (31.48)
Female	37 (68.52)
Symptoms	
Abdominal pain	25 (46.30)
Fever	14 (25.93)
Jaundice	9 (16.67)
Abdominal mass	11 (20.37)
Coexisting diseases	
Cholecystitis	29 (53.70)
Cholecystolithiasis	20 (37.04)
Choledocholithiasis	13 (24.07)
Hepatolithiasis	7 (12.96)
Cholangitis	10 (18.52)
Pancreatitis	4 (7.41)

3.2. Surgical treatments

All the patients underwent surgical procedures with cyst excision and bilioenteric anastomosis. Among them, 43 patients (79.63%) underwent extrahepatic cyst excision (including cholecystectomy) and hepaticojejunostomy, 9 patients (16.67%) received extrahepatic cyst excision (including cholecystectomy), partial hepatectomy, and hepaticojejunostomy (Fig. 2D and E). Two Todani type III or type 3 patients underwent endoscopic and transduodenal sphincteroplasty (Fig. 2F–H).

For distal management, 39 type 1 patients and 10 type 2 patients underwent excision of the distal cyst or seemingly normal bile duct to about 5 mm from the convergence with the pancreatic duct, that is, the intrapancreatic cyst or bile duct (Fig. 2A). Three patients received excision of both distal cylindrical cyst and papilla, followed by pancreatic duct plasty with duodenum mucosa (Fig. 2B and C).

3.3. Perioperative complications and long-term follow-up

After operation, 11 patients (20.37%, 11/54) had short-term perioperative complications. Followed by the Clavien–Dindo classification, 6 patients presented with grade I complications (3 patients with pancreatic leak, 2 with abdominal infection, and 1 with delayed wound healing) and 5 patients exhibited grade II complications (2 patients with pancreatic leak, 2 with intestinal obstruction, and 1 with delayed wound healing). The last follow-up time was July 31, 2016, and the complete follow-up rate was 88.89% (48/54) with a mean follow-up of 49.69 months (range, 19–90 months). Using the surgical outcome criteria, 31 patients (31/48, 64.58%) were excellent, 15 patients (15/48, 31.25%) were good, and 2 patients (2/54, 4.17%) were fair outcomes. The satisfactory rate (excellent and good outcomes) was 95.83%. No patients were found with malignant transformation during the follow-up period.

Table 2

The patient classification based on the Todani and distal classification of choledochal cyst.

	Distal classification			
Todani classification	Type 1	Type 2	Type 3	
Type I	29	8	–	37
Type III	–	–	2	2
Type IVa	10	5	–	15
	39	13	2	54

4. Discussion

The concept of complete cyst excision is accepted by surgeons. Proximal cyst excision is a relative programatic procedure with caution to protect the portal vein and hepatic artery. For distal management, the cyst extends into the pancreas, and excision may be difficult because of surrounding adhesion to pancreatic tissue and portal vein during recurrent episodes of cholangitis. Based on these, surgeons may not perform complete excision in certain circumstances because of the risk of postoperative pancreatic leakage, bleeding, and peritoneal infection.[17–19] However, the unresected intrapancreatic choledochal cyst will result in the formation of a dead space within the pancreas. Pancreatic secretions will be forced into this space due to the presence of the anomalous pancreatobiliary junction.[2] Backflow of intestinal secretions results in the activation of pancreatic enzymes in the remnant cyst, which may lead to infection, stone formation, and an increased risk of malignancy.[20] A previous study demonstrated that no patients developed cholangiocarcinoma after complete cyst excision.[5] Thus, complete excision distal of the intrapancreatic cyst is suitable for eliminating dead space and subsequent complications, but the pancreatic duct must be treated with great care.

In this study, the distal management of intrapancreatic choledochal cysts is expounded in detail given that no systematic investigations have been reported. The Todani classification system, modified from the Alonso–Lej classification, is now widely adopted for choledochal cysts, and the clinical treatment strategies are also determined according to this classification system.[7,21] Five types are included in its classification: type I, a solitary extrahepatic cyst; type II, an extrahepatic supraduodenal diverticulum; type III (choledochocele), an intraduodenal cyst; type IV, extrahepatic and/or intrahepatic cysts; and type V (Caroli disease), multiple intrahepatic cysts. Although the 5 types summarize the possible morphologies with important instruction for surgical approach, the distal situation of choledochal cysts still needs to be elucidated. Concerning this situation, the classification only aimed at the correlation between intrapancreatic bile and pancreatic duct. In this classification, since irrelevant with pancreatic duct, Todani types II and V are excluded from the present study and classification.

To eliminate potential dead space within the pancreas, protect the pancreatic duct, and maintain unobstructed flow of pancreatic fluid, the current classification of distal intrapancreatic choledochal cysts can function as a highly targeted strategy for the following surgical approach with different types. Given the presence of the anomalous pancreatobiliary junction, the reflux of duodenal juice, and the erosion of activated pancreatic enzymes, cholangitis is common in patients with remnant cysts and may develop in the progression of choledochal cyst.[11] Recurrent episodes of infection may cause further damage to the

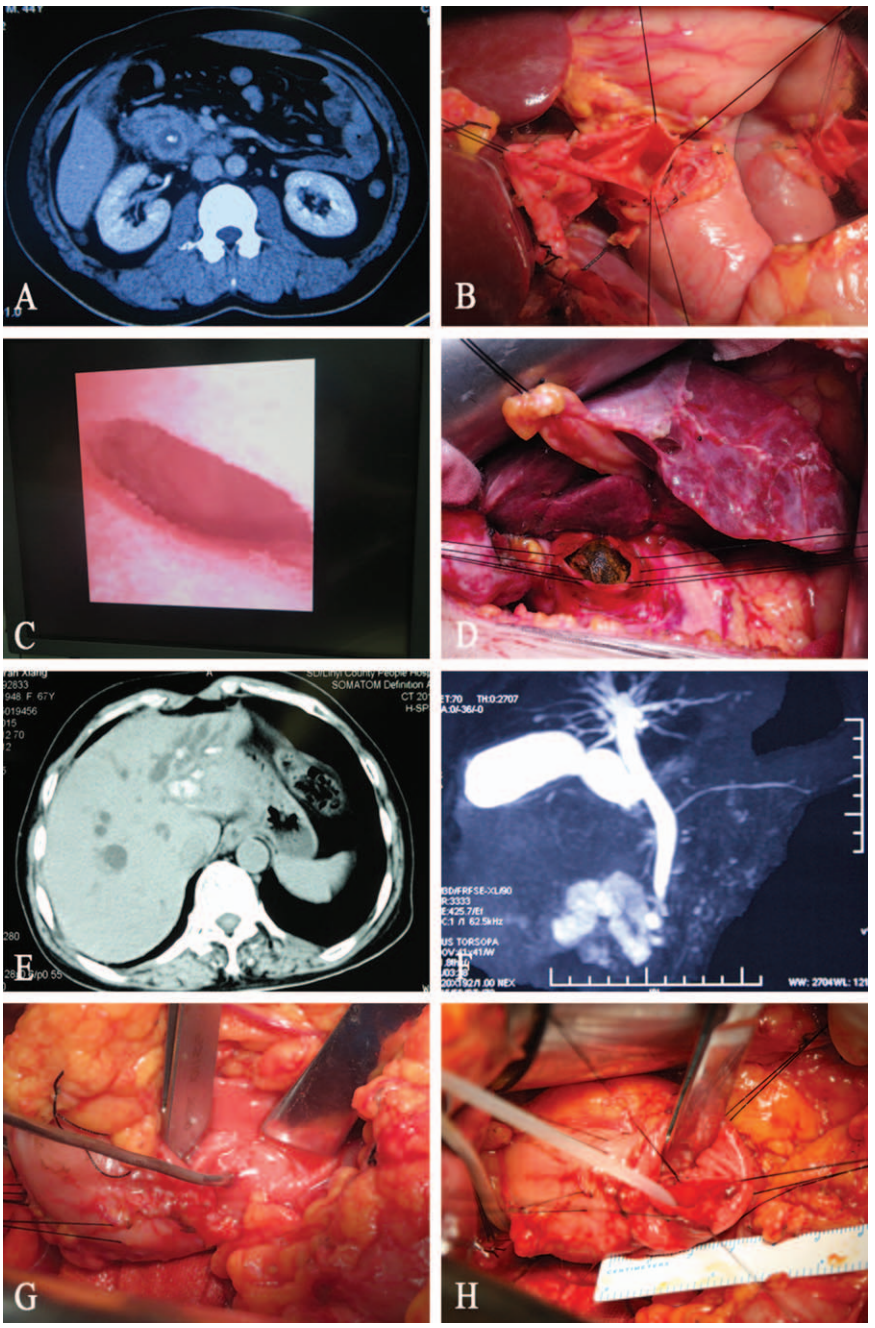

Figure 2. The typical cases in present study. Case 1: (A) shows intrapancreatic choledochal cyst with infection and stone formation in it. Case 2: a special type 2 cyst with cylindrical cyst in operation (B) and papilla insufficiency of Vater demonstrated by intraoperative choledochoscope (C). Case 3: a patient with type IV a choledochal cyst with left hepatatrophy, choledocholithiasis (D) and intrahepatic biliary stone and dilatation (E). Case 4: a patient with type 3 (choledochocele) cyst (F) was performed with transduodenal sphincteroplasty. A metal probe was used to explore the cyst through duodenal papilla (G) and a temporary urinary catheter was applied as a support for sphincteroplasty (H).

dysfunction and stenosis of duodenal papilla, resulting in recurrent pancreatitis and cholangitis.[17,20] Thus, in type 1 classification, which has no obvious relation to pancreatic duct, the intrapancreatic cyst or the seemingly normal bile duct is excised and the stump is sutured. The pathological changes in choledochal cysts from inflammatory bile duct are difficult to differentiate, and the excision range of distal cysts mainly depends on radiology and intraoperative finding. To prevent cyst recurrence, cancer of postoperative residual bile duct, and reoperation occurrence, the excision range is rational and appropriate but slightly radical. In the present study, 39 type 1 patients and 10 type 2 patients underwent such kind of surgery with careful operation to prevent the bleeding of pancreatic tissues and protect pancreatic duct. All the patients unevenly passed through the perioperative period, and the long-term effects were satisfactory, except for 2 patients with recurrent cholangitis.

For type 2 classification, the same management principal with type 1 is adopted since eliminating the dead space and keeping the pancreatic fluids flowing into the duodenum.

Nevertheless, the surgical management of special subtypes of type 2 with cylindrical cyst and papilla insufficiency of Vater differs. In this retrospective study, only 3 patients received this surgical procedure with the excision of cyst and papilla, followed by pancreatic duct plasty with duodenum mucosa. The long-term follow-up results showed that pancreatic exocrine function did not result in injury. Even so, more cases and experience need to be accumulated in the following study. Type 3 choledochal cysts (choledochoceles) can be treated by endoscopic sphincterotomy, sphincteroplasty alone, sphincteroplasty with cyst excision, or pancreaticoduodenectomy. Considering the low malignant rate in type 3 cysts, unroofing (endoscopic or transduodenal sphincteroplasty) or transduodenal excision (larger cysts) is optimal for choledochoceles.[1,14] In the study, 1 patient received endoscopic sphincteroplasty, and another received transduodenal sphincteroplasty. This finding showed that the endoscopic approach may be a better choice with minimal trauma. In the near future, more patients with choledochal cyst should be enrolled to strengthen our present study.

This study had several limitations. Owing to a respective study, selection and information bias exist, and this may lead to less-strong evidence. In addition, the study sample size is relatively small, and the operations were performed in 1 center. Further cases and collaboration with other centers are necessary for further studies.

In conclusion, the current classification of distal intrapancreatic choledochal cysts could provide a more targeted strategy for complete excision to eliminate potential dead space within the pancreas, protect the pancreatic duct, ensure unobstructed flow of pancreatic fluid, and prevent reoperation.

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