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Choledochal Cysts in Children: Epidemiology and Outcomes

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Abstract
Background The purpose of the present study was to evaluate the epidemiology and outcomes of choledochal cysts in children.
Methods We performed a retrospective review of the records of all cases of choledochal cyst in children 0–14 years of age presenting at Chiang Mai University Hospital from May 2000 to February 2007. Demographic and clinical data, including laboratory and radiographic studies, as well as surgical and nonsurgical treatments and outcomes, were recorded.
Results There were 32 patients (25 female) with a mean age at diagnosis of 4.1 years (range: 1 month to 14.8 years). The most common clinical presentation was jaundice ($n = 17$), followed by abdominal pain ($n = 16$), nausea/vomiting ($n = 10$), and abdominal mass ($n = 8$). Based on the Tondani modification of the Alonso-Lej classification, 20 cases were type I, whereas 9, 2, and 1 of the patients had types IV, V, and II, respectively. Biliary tract infections, including cholecystitis ($n = 5$) and cholangitis ($n = 3$), were the most common preoperative complications. Twenty-six patients had definitive surgery consisting of cystectomy with Roux-en-Y hepaticojejunostomy. Postoperative complications were noted in 3 patients and included intestinal obstruction, bleeding, and cholangitis. Three patients died, all younger than 2 years of age ($p = 0.022$).
Conclusion The epidemiology of choledochal cysts in Thai children was similar to cases reported from other regions. Early diagnosis and treatment, particularly in children under 2 years of age, should be emphasized, because these patients are at a higher risk for development of chronic liver disease and poor outcome.

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Introduction

Choledochal cyst is a congenital cystic dilatation of the biliary tree, originally described by Vater and Ezler in 1723. It is more commonly found in Asian populations with a female to male ratio of approximately 3.5:1 [1]. Older children and adults typically present with the classic triad of jaundice, abdominal pain, and palpable mass. However, the clinical presentation during infancy may mimic biliary atresia and, if untreated, can progress to biliary cirrhosis and ultimately end stage liver failure. Therefore, early diagnosis and prompt surgical intervention are necessary to obtain a good prognosis. The aim of the present study was to examine the epidemiology and outcomes in a large case series of choledochal cyst in Thai children.

Materials and methods

We performed a retrospective review of the records of all cases of choledochal cyst in children 0–14 years of age presenting at Chiang Mai University Hospital from May 2000 to February 2007. Demographic and clinical data, including laboratory and radiographic studies, were recorded on a standardized form. Based on the Tondani

modification of the Alonso-Lej classification, the disease was classified into five types using the information from operative and radiographic findings, including ultrasonography, computerized tomography scan, and cholangiogram [2]. Postoperative complications were also recorded. Patients were categorized has having one of four possible outcomes: survived without residual liver disease, survived with cirrhosis or portal hypertension, deceased, or unknown. To examine the effect of age on outcomes, we compared patients less than 2 years old with patients 2–14 years old. All data were assessed with the SPSS program. Continuous data were analyzed with the Mann-Whitney *U*-test, whereas cross-tabulations were analyzed with the chi-square test. A value of $p < 0.05$ was considered statistically significant. This study protocol was approved by the Research Ethics Committee of the Faculty of Medicine, Chiang Mai University.

Results

We identified a total of 32 patients (25 female) with a mean age at diagnosis of 4.1 years (1 month to 14.8 years). The most common clinical presentation was jaundice ($n = 17$), followed by abdominal pain ($n = 16$), nausea/vomiting ($n = 10$), and abdominal mass ($n = 8$). No patient in this study had all three features of the classical triad of jaundice, mass, and pain, but 21 patients (66%) had one symptom and 10 (31%) had two symptoms. Pale-yellow and acholic stools were observed in 9 and 6 cases, respectively. Twelve patients had pancreaticobiliary complications at presentation, including cholecystitis ($n = 5$), cholangitis ($n = 3$), gallstones ($n = 2$), bleeding into the cyst ($n = 1$), and pancreatitis ($n = 1$). One patient with a diagnosis of Caroli's disease initially presented with bleeding esophageal varices. Based on the Tondani modification of the Alonso-Lej classification, 20 cases were type I, whereas 9, 2, and 1 of the patients had type IV, V, and II, respectively.

Abnormal laboratory findings included anemia and elevated levels of transaminase, alkaline phosphatase, and bilirubin. Eleven patients (34%) had coagulopathy. Children younger than 2 years of age had significantly lower levels of hemoglobin and serum albumin; significantly higher levels of alkaline phosphatase, AST, and bilirubin; and they were more likely to develop coagulopathy (Table 1).

Twenty-six patients were treated with cystectomy and Roux-en-Y hepaticojejunostomy. An intraoperative cholangiogram was performed if the anatomy had not been clearly defined preoperatively. A plane was developed between the peritoneum and the anterior wall of the cyst, keeping close to the cyst wall. The gallbladder and cystic duct were mobilized and the cystic artery was ligated. The distal common bile duct was dissected to just within the head of pancreas and transected. The common hepatic duct was divided at the level of bifurcation. The Roux loop of jejunum was passed through a window in the transverse mesocolon and anastomosed to the common hepatic duct, whereas the proximal stump of jejunum was anastomosed in an oblique end-to-side fashion to the distal jejunum. In an unresectable case from adhesion secondary to cholangitis, the mucosal resection by Lilly's technique was performed. Palliative surgical drainage was performed in two cases, one patient with common bile duct gallstones and cholecystitis and the other an unstable premature infant with tetralogy of Fallot and severe cholestasis. Medical management was recommended and provided to two patients with type V choledochal cyst, and two patients refused surgery. Postoperative complications were found in three cases, including bleeding, cholangitis, and intestinal obstruction secondary to adhesions. All were successfully managed without additional surgical intervention. Three patients died, all younger than 2 years old ($p = 0.022$). The cause of death included Pseudomonas sepsis, cardiac complication (preterm with tetralogy of Fallot), and liver failure. Although most of the patients survived without residual liver disease, three had signs of cirrhosis or portal hypertension, and in two of them this was diagnosed as

Table 1 Laboratory results and outcome in infants and children with choledochal cysts

	<2 years old	2–14 years old	*p* Value
Hemoglobin (g/dl)	9.8 (3.9–14.7)	11.5 (6.7–13.8)	0.070
Albumin (g/dl)	3.7 (3.3–4.6)	4.2 (2.3–5.4)	0.004
Globulin (g/dl)	2.9 (1.4–4.3)	2.8 (1.8–6.5)	0.457
Alkaline phosphatase (IU/l)	675 (343–2516)	279 (138–1158)	0.008
AST (IU/l)	280 (110–521)	80 (16–524)	0.001
ALT (IU/l)	141 (57–246)	68 (4–360)	0.186
Bilirubin, total (mg/dl)	13.57 (2.11–34.97)	1.04 (0.28–12.7)	<0.001
Bilirubin, direct (mg/dl)	9.86 (1.46–26.39)	0.47 (0.1–8.63)	<0.001
Coagulopathy (*n*, %)	8/12 (66.7%)	3/17 (17.6%)	0.007
Death	3/11 (27%)	0/20	0.022

Continuous values are expressed as median (range)

p values were calculated via Mann-Whitney *U*-test and the chi-square test

Caroli's disease at the age of 2.5 and 14.8 years, respectively. The third patient was diagnosed as choledochal cyst type I and was treated at the age of 6 months; however, she experienced postoperative ascending cholangitis and later on developed liver cirrhosis and ascites. No stricture formation was suspected or investigated in our study because all patients were jaundice-free after surgery.

Discussion

In this pediatric case series, clinical presentations of choledochal cyst were similar to previous reports (Table 2). The disease was predominant in females, and 38% of the patients were younger than 2 years of age. None of our patients had the typical classic triad, but most had either one or two symptoms. This may result from the inability of young children to provide an accurate history. Our results confirm Lipsett's study, in which the triad was also less likely to be seen in pediatric patients, compared with adults [3]. Additionally, preoperative complications, including cholangitis, pancreatitis, and biliary stones, were less common in children compared with adults [3, 4].

In our report, cholecystitis was the most common preoperative complication, followed by cholangitis and biliary tract stones. Laboratory abnormalities included elevated levels of alkaline phosphatase, transaminase, and bilirubin. In addition, significant deterioration of liver function was likely to be observed in the infantile type of choledochal cyst. Based on the Tondani modification of the Alonso-Lej classification, the most common was type I, followed by type IV, similar to earlier studies [2, 3, 5, 6].

Table 2 Comparison of clinical presentations and anatomical classifications

	Reference				Present study
	A[a]	B[a]	C[a]	D[a]	
Total, number	46	42	37	39	32
Pediatric, number	21	11	17	15	32
F:M ratio	8:1	6:1	2.4:1	2.5:1	3.6:1
Clinical presentation (%)					
Jaundice	43	48	NA	36	53.1
Pain	78	74	NA	79	50
Mass	33	30	NA	54	25
Complete triad	15	21	NA	NA	0
Anatomical classification (%)					
Type I	57	52	46	NA	62.5
Type II	0	2	0	NA	3.1
Type III	4	4	0	NA	0
Type IV	39	40	43	NA	28.1
Type V	0	2	11	NA	6.3

NA not available

[a] *A* Chijiiwa and Koga [5]; *B* Lipsett et al. [3]; *C* Todani et al. [2]; *D* Todani et al. [6]

Recently, two hypotheses have become widely accepted as describing the pathogenesis of choledochal cyst. One posits an anomalous arrangement of the pancreatobiliary junction, with a long common channel located outside the duodenum, a finding that has been generally observed [6]. This type of anomaly allows reflux of pancreatic enzymes into the biliary tree, demonstrated by the presence of biliary amylase in about one-half of patients [6, 7]. However, patients with prenatally diagnosed choledochal cyst do not have significant biliary amylase, and therefore the second hypothesis suggests a congenital structural defect with distal bile duct obstruction as the underlying etiology [7].

Prior to 1980, internal drainage procedures had been frequently used in choledochal cyst. However, this approach was likely to be asscociated with postoperative complications, such as recurrent cholangitis, biliary stones, pancreatitis, and malignancy, often requiring reoperation [5]. Importantly, Voyles et al. demonstrated that the incidence of carcinoma in choledochal cysts increased with age, with three-quarters diagnosed in adulthood [8]. Fu et al. reported subsequent cholangiocarcinoma in 1.3% of patients who underwent a bypass procedure [9]. Consequently, cyst excision with hepaticoenterostomy has been increasingly accepted as the standard surgical treatment [1]. In our study, 3 of 26 patients developed complications after cyst excision and Roux-en-Y hepaticojejunostomy, a result comparable to a 9% postoperative complication rate reported from Japan. That study also showed that the overall postoperative complication rate was lower in children than in adults (9% vs. 43%), and a risk of stone formation significantly decreased if the patients were treated before the age of 5 years [10]. An association between intrahepatic/biliary stones and recurrent cholangitis resulted in a recommendation of creating a wide anastomosis, which is best accomplished at the hepatic hilum [11]. Owing to a high incidence of intrahepatic stones and recurrent cholangitis in type IV choledochal cyst, excision of the extrahepatic portion of the biliary tract accompanying lobectomy is recommended only if left intrahepatic duct dilatation is observed.

Although the long-term outcome of our patients was generally good, three deaths occurred in patients younger than 2 years of age. Additionally, one of three patients with residual chronic liver disease presented at 6 months of age. These findings highlight the importance of early diagnosis and proper management, particularly in infants presenting with prolonged cholestasis.

In summary, the presentation and outcomes of large series of Thai children with choledochal cyst were similar to reports from other regions of the world. Early diagnosis

and appropriate treatment, particularly in children younger than 2 years of age, should be emphasized, because these patients are at a higher risk of developing chronic liver disease and poor outcome.

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References

1. Lipsett PA, Pitt HA (2003) Surgical treatment of choledochal cyst. J Hepatobiliary Pancreat Surg 10:352–359
2. Todani T, Watanabe Y, Narusue M et al (1997) Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 134:263–269
3. Lipsett PA, Pitt HA, Colombani PM et al (1994) Choledochal cyst disease: a changing pattern of presentation. Ann Surg 220:644–652
4. Schmid C, Meyer HJ, Ringe B et al (1993) Cystic enlargement of extrahepatic bile ducts. Surgery 114:65–70
5. Chijiiwa K, Koga A (1993) Surgical management and long-term follow up of patients with choledochal cysts. Am J Surg 165:238–242
6. Todani T, Watanabe Y, Fujii T et al (1984) Anomalous arrangement of the pancreatobiliary duct system in patient with a choledochal cyst. Am J Surg 147:672–676
7. Davenport M, Stringer MD, Howard ER (1995) Biliary amylase and congenital choledochal dilatation. J Pediatr Surg 30:474–477
8. Voyles CR, Smadja C, Shands WC et al (1983) Carcinoma in choledochal cysts: age-related incidence. Arch Surg 118:986–988
9. Fu M, Yang Y, Zang J (2000) Evolution in the treatment of choledochal cysts. J Pediatr Surg 35:1344–1347
10. Yamataka A, Ohshiro K, Okada O et al (1997) Complications after cyst excision with hepaticoenterostomy for choledochal cysts and their surgical management in children versus adults. J Pediatr Surg 32:1097–1102
11. Todani T, Watanabe Y, Urushihara N et al (1995) Biliary complications after excisional procedure for choledochal cyst. J Pediatr Surg 30:478–481