Clinical Differences between Choledochal Cysts in Infancy and Childhood: An Analysis of 160 Patients

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Background: *Choledochal cysts (CDC) can become symptomatic at any age. Clinical features of infants and older children are different and have been documented by many authors.*
Objective: *The aim of this report was to study the differences in clinical spectrum of CDC between the infants and children treated at our hospital during a 17-year period.*
Material and Method: *A retrospective study was conducted of 160 patients with CDC treated at Queen Sirikit National Institute of Child Health between 1996 and 2012. The patients were categorized into 2 groups based on age at clinical presentation: an infantile group (up to one year old) and a childhood group (over one year old). Clinical characteristics of patients in the infantile and childhood groups were compared with statistical analysis using the Chi-square and Fisher's exact test.*
Results: *Of the 160 patients with CDC, 48 cases (30%) were categorized in the infantile group and 112 cases (70%) in the childhood group. Over three-quarters of the patients in the infantile group presented with jaundice and acholic stool, and this incidence was significantly higher than in the childhood group (77% vs. 46.4%, p = 0.001 and 50% vs. 13.4%, p<0.001). Abdominal pain was the most common symptom of patients in the childhood group, whereas it was noted in only a small number of those in the infantile group (82.1% vs. 8.3%, p<0.001). Average amylase level in CDC content was markedly elevated in the childhood group but much lower in the infantile group (43,630.5±90,234.5 vs. 79±189.9, p<0.001). Only type I and type IV CDC as defined by Todani's classification were found in our patients, and there were no statistical differences in incidences of type I and type IV in the two groups (79.2% vs. 67.8% and 20.8% vs. 32.2% p>0.05). Neonates and infants with CDC had a significantly higher risk of liver cirrhosis than did the childhood group (25% vs. 8%, p<0.001). However, surviving patients with cirrhosis in both groups were doing well at least 3 years after surgical CDC excision.*
Conclusion: *Neonates and infants with CDC were more likely to present with jaundice and acholic stool, whereas older children were more likely to present with abdominal pain. Amylase level in CDC content was markedly elevated in the childhood group but at a much lower level in the infantile group. Neonates and infants with CDC tended to develop liver cirrhosis earlier and more often than older children.*

Keywords: *Choledochal cyst, Infancy, Childhood, Todani's classification*

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A choledochal cyst (CDC) is a segmental dilatation of the biliary system, most commonly involving the common bile duct (CBD). It may involve either intrahepatic or extrahepatic bile ducts, or a combination of both biliary ductal systems. The incidence in western countries is one in 100,000-150,000 live births[1]. It is markedly higher, one in 1,000-5,000 live births, in oriental countries, especially in Japan and China[2-4]. Although patients with CDC can present at any age, most of the reports have been of children below 10 years of age[2,5-7]. O'Neill[8] has classified the patients with CDC into 2 groups based on age at clinical presentation: infantile groups (ranging in age from 1 to 3 months) and adult groups (over 2 years of age). Clinical features are clearly different between the 2 groups. CDC in infants most commonly present with jaundice and pale stool, whereas older children present

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with abdominal pain, abdominal mass and intermittent jaundice similar to CDC in adults. Many authors have used the terms "infantile" group for patients up to one year old and "childhood" or "classical pediatric" group instead of "adult" group in their reports[9-11] for patients over one year old.

The authors previously reported CDC in 74 pediatric patients during a 19-year period (1977-1995)[12]. The authors herein evaluated our experience in patients who were treated at our institute after that period. The present study has updated the clinical outcomes of Thai pediatric patients by making comparisons between the infantile and childhood groups.

Material and Method

The present study was approved by the Institutional Review Board of Queen Sirikit National Institute of Child Health (QSNICH) with Document No. 56-028. A retrospective review was conducted of 160 patients treated for CDC in the Department of Surgery at QSNICH from January 1996 to December 2012. The data were collected from existing medical records for epidemiologic studies as well as symptomatologies and clinical outcomes.

The choledochal cysts were categorised into 5 types according to Todani's classification[13]. The assignments of types of CDC were based on ultrasonography and intraoperative cholangiography (IOC) during surgery. Operative procedures included aspiration of the cyst content for bacterial culture and evaluation of amylase concentration; IOC; CDC excision with hepaticojejunostomy using 45 cm Roux-en-Y limb without antireflux procedure; and liver biopsy.

The patients were divided into two age-specific groups: the infantile group (up to one year old at clinical presentation) and the childhood group (over one year old at clinical presentation), in order to analyze the differences in clinical presentations, types of CDC and clinical outcomes. Data were analyzed using SPSS version 16 (SPSS Inc, Chicago, IL, USA). Statistical significance was evaluated using the Chi-square and Fisher's exact test. A *p*-value less than 0.05 was considered significant.

Results

A total of 160 patients with CDC (male: female = 44:116 or 1:2.6) were treated at our institute during the study period. The age at clinical presentation varied from 7 days to 14 years (Fig. 1). Forty-eight cases (30%) were categorized in the infantile group, whereas 112 cases (70%) were in the childhood group. Females were predominant in both groups but without statistical significance (Table 1). The mean age at onset of symptoms in the infantile group was 7±4.6 months (range 0-12 months) and in the childhood group 76.7±40.6 months (range 15-168 months). Regarding the profile of clinical presentations, jaundice and acholic stool appeared to be more common in the infantile patients who mostly developed these features within 3 months of being born. In contrast, 92 patients (82.1%) in the childhood group presented with abdominal pain, whereas only 4 patients (8.3%) in the infantile group presented with this feature ($p<0.001$). The classic triad of abdominal pain, palpable abdominal mass and jaundice was identified in 14 cases (12.5%) of the childhood group but was not found at all in the infantile group. Comparison of the amylase level in the CDC content showed that the average amylase level in the childhood group was significantly higher than that of the infantile group (43,630.5±90,234.5 U/L vs. 79±189.9 U/L, $p<0.001$).

The types of CDC are shown in Table 2. Only type 1 and 4 were present in our patients. Type 1 (Fig. 2) was more common in the infantile group (79.2%) than in the childhood group (67.8%). In contrast, type 4 (Fig. 3) was more common in the childhood group (32.2%) than in the infantile group (20.8%). There was no statistical difference in the types of CDC in the two groups ($p>0.05$) (Table 2).

Cyst excision and Roux-en-Y hepaticojejunostomy were performed in all but 3 cases. Two patients in the infantile group who had marked nodular cirrhosis with severe congenital heart disease underwent cystojejunostomy and liver biopsy. Both cases succumbed within one month postoperation. The remaining case, a 5-year-old boy in the childhood group, was found to have nodular cirrhosis, cholangitis and septicemia during the operation. Only external drainage with a T-tube and liver biopsy were performed. He died within one week of the procedure.

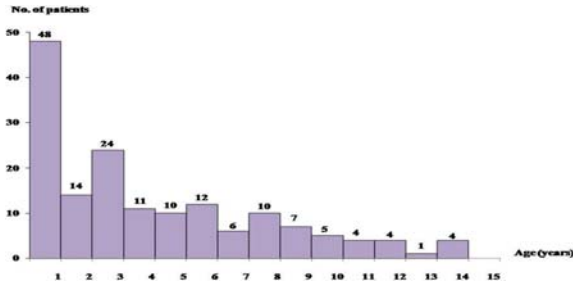

Fig. 1 Age at onset of symptoms in 160 patients.

Table 1. Demographic data, clinical presentations and amylase concentration in choledochal cysts

	Number (%)			*p*-value
	Total (n = 160)	Infantile (n = 48)	Childhood (n = 112)	
Male: female	44:116	18:30	26:86	0.067
Age at onset of symptoms (month)				
Mean ± SD	51±46.9	7±4.6	76.7±40.6	
Median (min, max)	36.0 (0-168)	6.5 (0-12)	54.0 (15-168)	
Symptoms/signs				
Jaundice	89 (55.6)	37 (77.0)	52 (46.4)	0.001
Abdominal pain	96 (60)	4 (8.3)	92 (82.1)	<0.001
Abdominal mass	46 (28.7)	19 (39.6)	27 (23.3)	0.064
Acholic stool	39 (24.4)	24 (50.0)	15 (13.4)	<0.001
Vomiting	36 (22.5)	6 (12.5)	30 (26.8)	0.047
Fever	17 (10.6)	3 (6.2)	14 (12.5)	0.240
Amylase level in CDC content (U/L)				
Mean ± SD	27,924±74,842.6	79±189.9	43,630.5±90,234.5	<0.001
Median (min, max)	2,385 (0-509,179)	4 (0-889)	10,400 (5-509,179)	

Table 2. Types of choledochal cyst based on Todani's classification*

Types	Number (%)			*p*-value
	Total (n = 160)	Infantile group (n = 48)	Childhood group (n = 112)	
1a	79 (49.3)	29 (60.4)	50 (44.6)	0.240
1b	3 (1.9)	0	3 (2.7)	
1c	32 (20.0)	9 (18.8)	23 (20.5)	
4a	46 (28.8)	10 (20.8)	36 (32.2)	

* Type 1: Cystic dilatation of CDC 3 subtypes: A) Saccular dilatation involving both CBD and CHD, B) Segmental dilatation of CBD only, C) Diffuse or cylindrical dilatation.
Type 2: Diverticulum of extrahepatic bile duct
Type 3: Choledochocele
Type 4: Multiple cystic dilatation 2 subtypes: A) Multiple cysts of both intrahepatic and extrahepatic bile ducts, B): Multiple cysts of extrahepatic bile ducts
Type 5: Intrahepatic bile duct cyst (single or multiple)

Preoperative and postoperative morbidities in the two groups were not significantly different, with the exception of the presence of cirrhosis confirmed by pathohistology (Table 3). Liver biopsy was performed in 36 of the 48 cases in the infantile group and 50 of the 112 cases in the childhood group. Cirrhosis of the liver was present in 25% of the infantile group, significantly higher than the 8% found in the childhood group (odds ratio = 3.8, 95% confidence interval = 1.076-13.65, $p<0.05$). Of the 9 patients with cirrhosis in the infantile group, the youngest was 2 months old. Three of the 9 infantile cases with cirrhosis postoperatively died within one month (2 cases) and 4 months (one case) because of ascending cholangitis, sepsis and congestive heart failure from tetralogy of Fallot. The 6 surviving patients with cirrhosis were doing well at follow-up at least 3 years later. One of the 4 cases with cirrhosis in the childhood group died within one week of T-tube drainage because of ascending cholangitis and septicemia. The 3 remaining childhood cases were doing well at follow-up at least 3 years later with normal findings of liver function test and ultrasound. The

mortality rate of the infantile group was higher than that of the childhood group (6.2% vs. 0.9%, $p = 0.081$). The presence of liver cirrhosis, ascending cholangitis and associated severe congenital heart disease (tetralogy of Fallot) were the major causes of death in the present study.

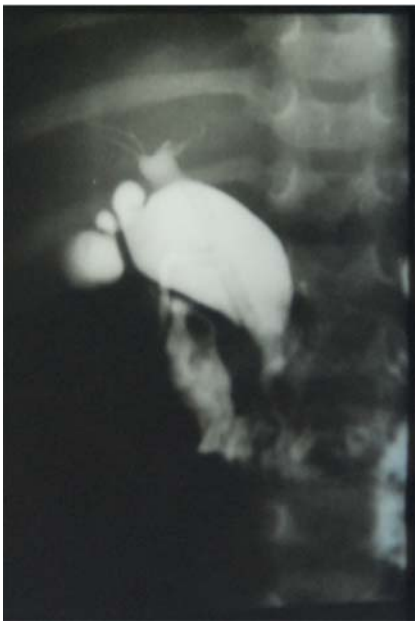

Fig. 2 IOC of a 4-month-old girl showing a saccular dilation of CBD (CDC, type 1a).

Discussion

CDC in pediatric patients are frequently categorized into those occurring in an infantile group and those occurring in a childhood or classical pediatric group[9-11]. The 2 groups have marked differences in clinical presentations and pathological anatomy. Todani[14] has described CDC in the infantile group as having the following features: cystic dilatation of CBD; palpable abdominal mass with jaundice and acholic

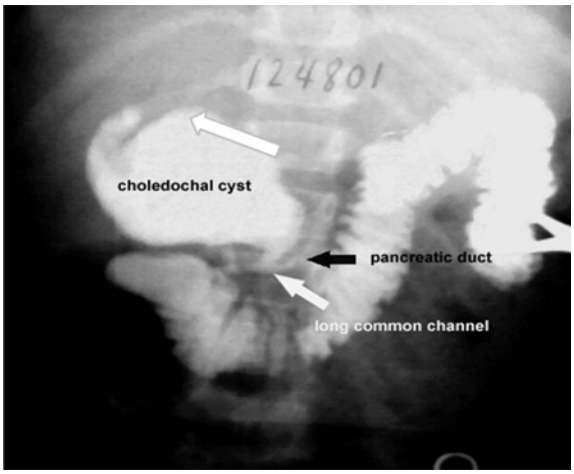

Fig. 3 IOC of a 7-year-old girl showing fusiform dilation of extrahepatic and intrahepatic duct (CDC, type 4a) with long common channel of the pancreaticobiliary ductal system in a 7-year-old girl.

Table 3. Morbidity and mortality of the two-age-specific groups

	Number (%)		*p*-value
	Infantile group (n = 48)	Childhood group (n = 112)	
Morbidity			
CDC perforation before operation	2 (4.2)	2 (1.8)	0.584
Postoperative intestinal obstruction	3 (6.3)	2 (1.8)	0.160
Anastomotic leak	2 (4.2)	1 (0.9)	0.214
Wound dehiscence	2 (4.2)	0	0.089
Ascending cholangitis	2 (4.2)	1 (0.9)	0.214
Pneumonia	2 (4.2)	1 (0.9)	0.214
Postoperative IHD* stone	0	1 (0.9)	1.000
Positive bacterial growth from bile culture	2 (4.2)	7 (6.3)	0.725
Presence of cirrhosis from liver biopsy	9/36 (25.0)	4/50 (8.0)	0.030
Mortality	3 (6.3)	1 (0.9)	0.081
Cirrhosis, ascending cholangitis and sepsis	1	1	
Cirrhosis and CHF**	2	0	

* intrahepatic duct, ** congestive heart failure

stool; no symptomatic association with acute pancreatitis; and a low amylase level in bilious content of the cyst. In our present study, some neonates and infants with CDC had a highly significant incidence of jaundice, acholic stool, and abdominal mass similar to features found in correctable biliary atresia[15]. Patients in the childhood group mostly presented with abdominal pain. Classic triad of CDC including abdominal pain, abdominal mass and jaundice were noted in only 12.5% of the childhood group, and in none of the infantile group. The authors found marked elevation of amylase levels in the cystic content of the childhood group and significantly lower levels in the infantile group. These clinical presentations might be indicative of the differences in pathogenesis of CDC between the 2 specific age groups. Landing's hypothesis probably explains the development of CDC in neonates and infants. Landing[16] proposed that neonatal hepatitis, biliary atresia and CDC may originate from antenatal viral infection of the hepatobiliary system. The inflammatory process is continuously progressive to the postnatal period. These 3 diseases probably represent different outcomes of the same obstructive inflammatory process. This is the reason why the clinical presentation in infantile CDC is similar to that of biliary atresia and neonatal hepatitis. In our experience, infants with CDC had a poorer prognosis than patients in the childhood group. Nine patients in the infantile CDC group developed liver cirrhosis and had a higher operative risk, similar to the findings of other reports[2,12,17-19]. In contrast, pathogenesis of CDC in older children and adults was based upon the hypothesis of Babbit[20] who proposed the theory of a long common channel of the pancreaticobiliary ductal system. Pancreatic juice can reflux into the CBD, and pancreatic enzymatic actions produce inflammation, weakness of the CBD wall, and narrowing and obstruction of the distal CBD. These inflammatory processes result in dilatation of the proximal CBD to form a CDC. Most cases of CDC in the childhood group had a high level of amylase in CDC content and a long common channel of the pancreatic duct, and CBD was demonstrated by an IOC (Fig. 3) and endoscopic retrograde cholangiopancreatography[17,21-23].

Only type 1 and 4 CDC were found in the present study. Type 1 cysts were more common than type 4 cysts in both the infantile and childhood groups but there was no statistically significant difference. The occurrence of some type 4 cysts might be a result of back pressure from chronic obstruction and inflammation of the distal bile ducts of type 1 cysts over a long period of time. It is possible that asymptomatic type 1 cysts could become type 4 cysts in older patients[24]. However, there was no obvious clinical data to support this hypothesis.

Liver cirrhosis, confirmed by pathohistology, was commonly seen in infants with CDC but rarely in older children. This finding has been reported by other previous studies[10,12,25-27]. Our youngest patient with liver cirrhosis was 2 months old, whereas Dewbury[25] reported a case as early as 10 days old. In our experience, patients with nodular cirrhosis during operations had a high risk of postoperative complications. Four cases in the present study died in the immediate postoperative period due to ascending cholangitis, sepsis and congestive heart failure. However, 9 patients with cirrhosis (6 in the infantile group and 3 in the childhood group) survived and had normal liver function test and ultrasonography. All of them were doing well at least 3 years after CDC excision.

Conclusion

Clinical differences in CDC between infants and older children in the present study included clinical presentations, amylase concentration in CDC content, and liver cirrhosis. The typical presentations of infantile CDC are characterized by persistent jaundice and acholic stool, whereas abdominal pain is the most common symptom in childhood CDC. The amylase level in CDC content is mostly normal in infants, but markedly higher in older children. Liver cirrhosis is often seen in infantile CDC, while this complication is rarely seen in childhood CDC. Patients with liver cirrhosis of the infantile and childhood group who survived after operative CDC excision were found to have recovered well at follow-up at least 3 years later.

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Potential conflict of interest

None.

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ความแตกต่างทางคลินิกของโรคท่อน้ำดีรวมโป่งพองระหว่างกลุ่มเด็กทารกและเด็กโต: การศึกษาวิเคราะห์ในผู้ป่วย 160 ราย

รังสรรค์ นิรามิษ, รจิต นฤมิตสุธน, สุขวัฒน์ วัฒนาธิษฐาน, ไมตรี อนันต์โกศล, วีระ บูรณะกิจเจริญ, อัจฉริยา ทองสิน, วราภรณ์ มหธราดล, สุรเนตร ลออวงศ์

ภูมิหลัง: ท่อน้ำดีรวมโป่งพองเป็นโรคที่เกิดขึ้นได้ในทุกอายุ อาการในเด็กทารกและเด็กโตมีความแตกต่างกันและถูกนำเสนอโดยแพทย์หลายราย

วัตถุประสงค์: เพื่อศึกษาความแตกต่างทางคลินิกระหว่างท่อน้ำดีรวมโป่งพองในเด็กทารกและเด็กโตที่รักษาที่โรงพยาบาลของเราในช่วงระยะเวลา 17 ปี

วัตถุและวิธีการ: เป็นการศึกษาย้อนหลังในผู้ป่วย 160 รายที่เป็นโรคท่อน้ำดีรวมโป่งพองที่รักษาในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี ระหว่างปี พ.ศ. 2539 และ พ.ศ. 2555 ผู้ป่วยถูกแบ่งเป็น 2 กลุ่ม ขึ้นอยู่กับอายุที่เริ่มมีอาการคือ กลุ่มเด็กทารก (อายุภายใน 1 ปี) และกลุ่มเด็กโต (อายุมากกว่า 1 ปี) ลักษณะทางคลินิกของผู้ป่วยในกลุ่มเด็กทารกและเด็กโตถูกนำมาเปรียบเทียบด้วยการวิเคราะห์ทางสถิติโดยใช้ Chi-square และ Fisher's exact test

ผลการศึกษา: ในจำนวนผู้ป่วย 160 รายที่มีท่อน้ำดีรวมโป่งพองผู้ป่วย 48 ราย (ร้อยละ 30) ถูกจัดอยู่ในกลุ่มเด็กทารกและ 112 ราย (ร้อยละ 70) จัดอยู่ในกลุ่มเด็กโต ผู้ป่วยส่วนใหญ่ในกลุ่มเด็กทารกมักจะมีอาการดีซ่านและอุจจาระสีซีดมากกว่าในกลุ่มเด็กโต (ร้อยละ 77:46.4, p = 0.001 และร้อยละ 50:13.4, p<0.001) อาการปวดท้องพบได้บ่อยที่สุดในกลุ่มเด็กโต ขณะที่พบได้น้อยมากในกลุ่มเด็กทารก (ร้อยละ 82.1:8.3, p<0.001) ค่าเฉลี่ยของระดับ amylase ในซีสต์ของท่อน้ำดีรวมสูงมากในกลุ่มเด็กโตและต่ำมากในกลุ่มเด็กเล็ก (43,630.5±90,234.5:79±189.9, p<0.001) มีซีสต์เพียง 2 ชนิดเท่านั้นคือชนิดที่ 1 และ ชนิดที่ 4 ตามการแบ่งชนิดของ Todiani ที่พบในการศึกษาครั้งนี้และไม่มีความแตกต่างทางสถิติในซีสต์แต่ละชนิดของผู้ป่วยทั้งสองกลุ่ม (ร้อยละ 79.2:67.8, และร้อยละ 20.8:32.2, p>0.05) ในเด็กแรกเกิดและเด็กทารกที่มีท่อน้ำดีรวมโป่งพองมีความเสี่ยงในการเกิดตับแข็งได้มากกว่ากลุ่มเด็กโต (ร้อยละ 25:8, p<0.001) ถึงอย่างไรก็ตามผู้ป่วยทั้งสองกลุ่มที่เป็นตับแข็งและรอดชีวิตยังเป็นปกติดีอย่างน้อย 3 ปี ภายหลังผ่าตัดแก้ไขโรคท่อน้ำดีรวมโป่งพอง

สรุป: เด็กแรกเกิดและเด็กทารกที่มีโรคท่อน้ำดีรวมโป่งพองมักจะมีอาการดีซ่านและอุจจาระสีซีด ขณะที่เด็กโตมักจะมีอาการปวดท้องค่า amylase ในซีสต์ของท่อน้ำดีรวมมีค่าสูงมากในกลุ่มเด็กโต แต่จะต่ำมากในกลุ่มเด็กทารก กลุ่มเด็กทารกที่เป็นโรคนี้มีแนวโน้มจะเกิดอาการตับแข็งได้มากกว่าเด็กโต