Choledochal Cyst : Review of 74 Pediatric Cases

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Abstract

Seventy-four cases of CDC were treated at the Children's Hospital from 1977 to 1995. Female to male ratio was 5:1. Forty per cent of the patients developed symptoms within one year after birth and 75 per cent within 5 years of age. About one third of the cases were treated surgically within one year of age.

Only 5 patients or 6.8 per cent had all the clinical triads of jaundice, abdominal pain and mass. Jaundice was the most common symptom in infants below one year of age but abdominal pain was the most common symptom in older children. Three newborn infants had associated biliary atresia. Established liver cirrhosis was noted during surgery in 9 patients who were operated upon within one year of age but none of the older children.

All but one had either type I or type IV CDC, while the remaining one had Caroli's disease or type V CDC.

Excision was the prefered treatment. Overall operative mortality rate after excision was 6.5 per cent. None of the patients who had internal drainage procedures succumbed. Infants below one year of age appeared to have high morbidity and mortality rates after surgery. Internal drainage procedure may be considered in high risk patients with advanced cirrhosis.

Available information suggests that the occurrence of this disease in Thailand is probably more common than in Western countries and etiology of CDC in infants may be different from that in older children or adults.

A choledochal cyst (CDC) is a segmental cystic dilatation of the bile ducts. It most commonly involves the common bile duct (CBD), but may involve any portion of the bile ducts, either extrahepatic or intrahepatic ductal system, or a combination of both. Most of the reported cases were children below 10 years of age[1-4].

It is well accepted that the occurrence of this disease in Japan is more common than in Western countries[1-7]. Reports from other Oriental

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countries also indicate that the incidence of this disease in this area is likewise higher than in Western countries[8-10]. In Thailand, only small series have been reported, and it appears as if the incidence may probably be much lower[11]. However, it has been our impression that the incidence of this disease should be much higher than what has been reported in the Thai literature[12]. This prompted us to review our experience at the Children's Hospital.

MATERIAL AND METHOD

A retrospective review of medical records of the patients who were treated in the Department of Surgery at the Children's Hospital for CDC during the period of 1977-1995 was carried out. Seventy-four cases were available for the study. The data were collected from existing medical records for epidemiologic studies as well as diagnostic and therapeutic problems. Classification of CDC in this study follows Todani's definition[5] (Table 1).

RESULTS

Age

Age at onset of symptoms varied from the first month of life to 12 years (Fig. 1). Twenty-nine patients or about 40 per cent developed symptoms within one year of age and 75 per cent within 5 years of age. About one-third of the cases were diagnosed within one year of age (Fig. 2).

Of the 29 patients who developed symptoms within one year of age, 13 did so within one month after birth, 3 between 1 to 2 months, 3 between 2 to 3 months and the remaining 10 between 3 to 12 months.

Sex

Sixty-two were female and 12 were male. The female to male ratio was 5:1. When only those who developed symptoms within one year of age were considered, the ratio was still the same.

Table 1. Classification of choledochal cyst (Todani[5]).

Type	I.	Cystic dilatation of CBD. 3 Subtypes :
	Ia :	Saccular dilatation involving both CBD and CHD
	Ib :	Segmental dilatation of CBD only
	Ic :	Diffuse or cylindrical dilatation
Type	II.	Diverticulum of extrahepaptic bile duct
Type	III.	Choledochocele
Type	IV.	Multiple cystic dilatations. 2 Subtypes :
	IV A :	Multiple cysts of both intrahepatic and extrahepatic bile ducts
	IV B :	Multiple cysts of extrahepatic bile duct
Type	V.	Intrahepatic bile duct cyst (single or multiple)

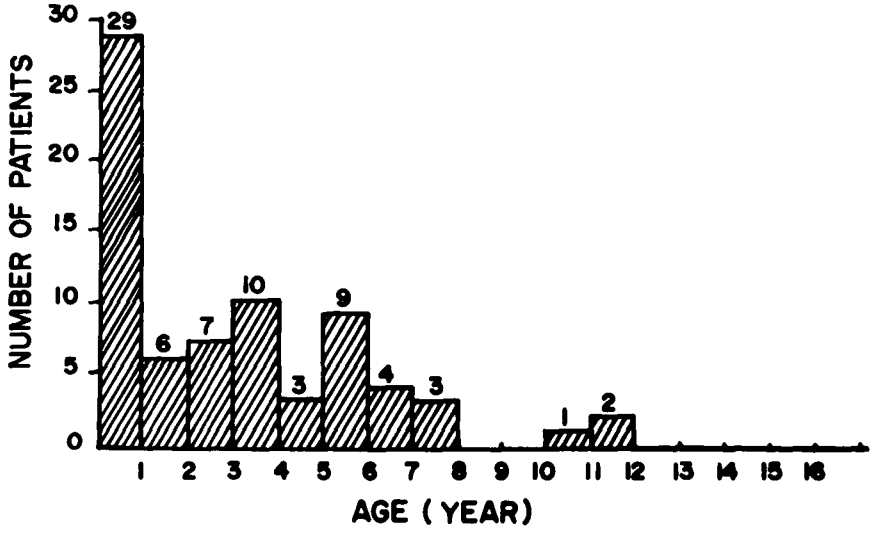


Fig. 1. Age at onset of symptoms.

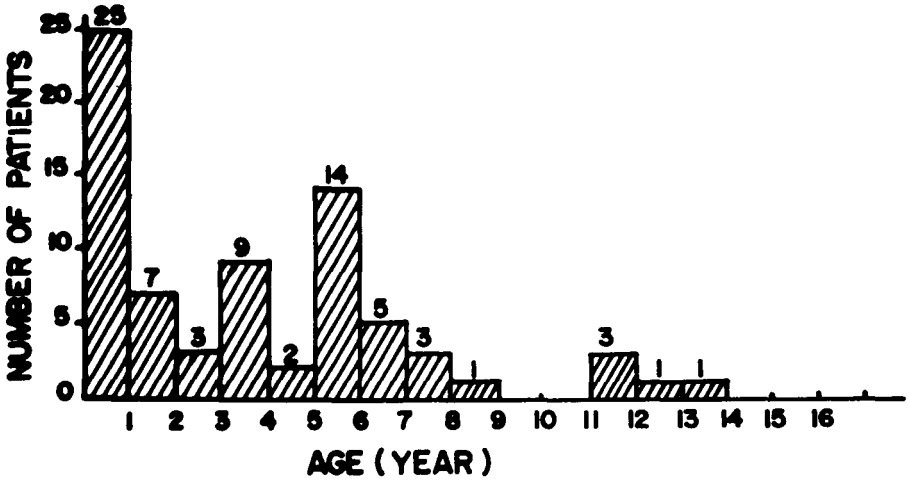
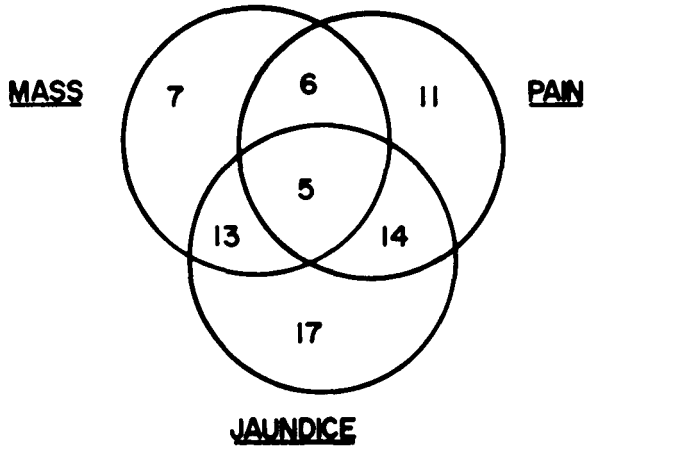


Fig. 2. Age at treatment.



N.B. ONE PATIENT WITH CAROLI'S DISEASE HAD NONE OF THESE SYMPTOMS.

Fig. 3. Symptomatology.

Symptomatology

Only 5 patients, or 6.8 per cent of the 74 cases, had all of the clinical triads of jaundice, abdominal pain and abdominal mass (Fig. 3). The most common symptomatology was jaundice, appearing in 49 cases or 66.2 per cent (Table 2). Abdominal pain and mass were noted in 48.7 per cent and 41.9 per cent of the cases respectively. Acholic stool was present in about one- third of the cases and appeared more commonly in infants below one year of age. Abdominal pain, which was most commonly colicky in nature, was most frequently accompanied with vomiting.

Table 2. Symptomatology.

Symptomatology	Number of patients	%
1. Jaundice	49	66.2
2. Abdominal pain	36	48.7
3. Abdominal mass	31	41.9
4. Acholic stool	24	32.4
5. Vomiting	24	32.4
6. Fever	20	27.0
7. Ascites	4	5.4

About 30 per cent of the patients had fever. Some of these presented with a clinical manifestation of sepsis, which was probably secondary to cholangitis.

Four patients were found to have obvious ascites by physical examination. Three of these had cirrhotic ascites and one had bilious ascites due to a perforation of CDC. All four cases were infants below one year of age.

Symptomatology in various age groups

Jaundice appeared to be more common in the younger age group. It was present in 18 of the 19 infants who developed symptoms within 3 months after birth. The incidence was 70 per cent of the 10 infants who developed symptoms during the age of 3 to 12 months and 54.5 per cent of the children who developed symptoms after one year of age.

Abdominal pain appeared to be more common in older children. It was not present in infants below one year of age, but present in 64.3 per cent of children between 1 to 3 years of age and 83.3 per cent of children older than 3 years of age. In about one-third of the latter group, colicky abdominal pain and vomiting were the only symptoms without any accompanying positive finding on a physical examination. These patients most frequently had several episodes of such symptoms before the correct diagnosis of CDC was made by an ultrasonographic study.

The patient with Caroli's disease had fever, weakness and anorexia.

Associated diseases

Two patients had thalassemia. The CDC was diagnosed at the age of 14 months in one case and 38 months in the other.

Three infants had associated biliary atresia (BA). Two of these were diagnosed at 3 months of age and one at 5 months of age.

Liver cirrhosis was noted during surgery in 9 cases. All were operated upon within 12 months of age : one each at the age of 2 months, 3 months, 5 months, 8 months, 10 months and two each at 4 months and 11 months of age.

Preoperative investigations

1. Upper gastro-intestinal contrast study (UGI series)

UGI series was done in patients who were suspected to have CDC in the early period of the study, when ultrasonography and hepatobiliary radionuclide scan were not available at the Children's Hospital, but not in recent years. Nineteen cases had UGI series. Widening of the duodenal C-loop (Fig. 4) was noted in 9 cases, extrinsic duodenal compression in 6 cases and duodenal displacement without compression or widening of duodenal C-loop in 2 cases. In the remaining 2 cases, no apparent radiologic abnormality was noted.

2. Intravenous cholangiography (IVC)

This investigation was also done in the early period of the study only. Seventeen cases had IVC done. Dilatation of the CBD was reported in 15 cases. No visualization of the CBD was noted in the remaining 2 cases. Neither of the latter had jaundice during the examination.

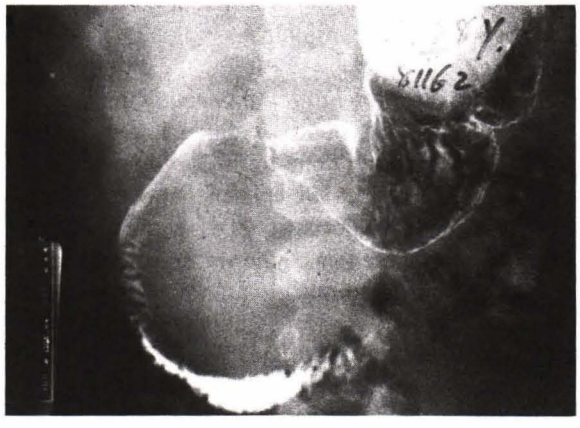


Fig. 4. Widening of duodenal C-loop in an 8-year-old girl with CDC.

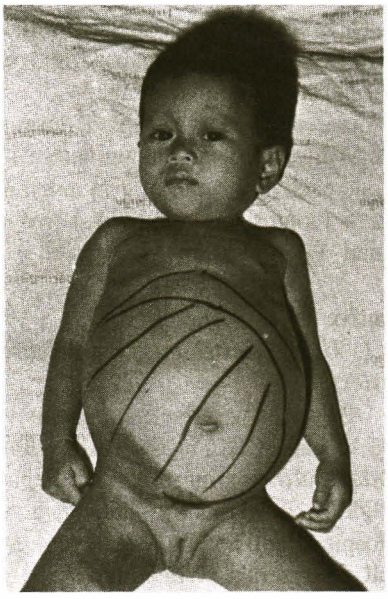


Fig. 5 A 10-month-old girl with a huge CDC.

3. Ultrasonography (US)

At the Children's Hospital, this diagnostic tool was first used to diagnose a CDC in 1983, and it has been done in every patient who was suspected to have a CDC since. Fifty-two patients had US, and the correct diagnosis of CDC was reported in 50 cases. In one patient, the CDC was so large (Fig. 5) that the ultrasonographist could not relate it to the biliary system. The CDC could not be recognized in one patient who had a perforation of the cyst with bile peritonitis.

4. Hepatobiliary radionuclide scan

The hepatobiliary radionuclide scan that was most commonly done in this group of patients was a DISIDA scan. Seventeen patients had the study. Correct diagnosis of CDC was reported in 11 cases. No excretion of the tracer into the CBD and gastro-intestinal tract was noted in 4 cases. Of these 4 cases, two were 2 months old and both were found to have associated atresia of the distal CBD, one was one month old and the other was 4 years old. In the remaining 2 cases, a small amount of the tracer in the intestine without demonstration of the CDC was reported.

Intraoperative studies

1. Intraoperative cholangiography (IOC)

This study was available for a review in only 41 cases. In several cases, especially when CDC was very large, the intrahepatic bile ducts and distal CBD were not filled by the contrast media during the study, therefore, these ducts could not be adequately studied. Poor positioning and radiologic technique also contributed to the inadequacy of the study. Identification of CDC type was inaccurate due to these limitations of the retrospective study.

Nine patients were noted to have a long common channel of the pancreatic and common bile ducts. Of these, only 2 were infants younger than 12 months of age; one was 2 months and the other was 7 months old.

2. Amylase level in CDC content

Amylase level in CDC content was available for the study in only 30 cases. Seventeen patients had an amylase level higher than 500 units/dl. The highest level was 100,000 units/dl.

Sixteen of the 18 patients above one year of age had an amylase level higher than 500 units/dl, while only one of the 12 patients below one year of age did so. The latter was a 4 - month - old girl who had an amylase level of 509 units/dl.

3. Culture of CDC centent

Culture of CDC content was done in 43 cases and was positive for bacteria in 9 cases : *E. coli* in 3 cases, *Enterobactor* in 2 cases, *Pseudomonas aeruginosa, Pseudomonas spp., Micrococcus spp.* and *Staphylococcus* coagulase negative in one case each. Only one patient with *Pseudomonas aeruginosa* in CDC content had clinical manifestation of sepsis and positive hemoculture for the same bacteria prior to surgery.

Types of CDC

The assignment of types of CDC was based on the available IOC, US and IVC during the study.

The majority of patients had either type Ia or IVA (Table 3). One had type V but none had type II or type III. Of the 73 cases with dilatation of the extrahepatic duct, 15 had a fusiform or cylindrical dilatation of the CBD. Eleven of these 15 patients, or 73.3 per cent, were 3 years of age or older at diagnosis.

Table 3. Types of choledochal cyst.

Type	Number of patients
Type Ia	38
Ib	0
Ic	6
Type II	0
Type III	0
Type IV A	29
Type IV B	0
Type V	1
Total	74

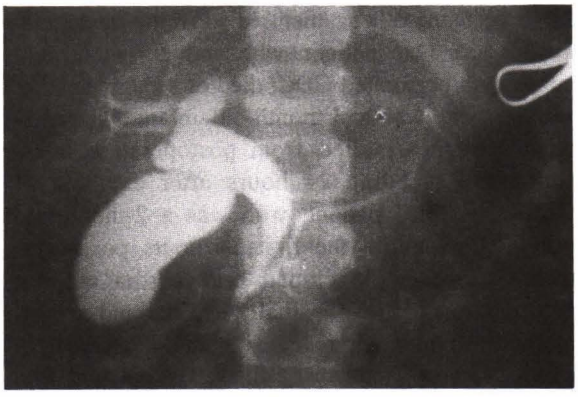


Fig. 6. IOC of a 5-month-old girl showing a long common channel of the pancreaticobiliary ductal system and cylindrical dilatation of the extrahepatic and intrahepatic ducts.

Table 4. Treatment.

Treatment	Number of patients
1. Excision	62
2. Choledochocystoduodenostomy	8
3. Roux-Y choledochocystojejunostomy	3
4. Liver biopsy only	1*
Total	74

* Patient with Caroli's disease

Treatment

Excision was the most commonly performed procedure (Table 4). Primary excision with a Roux-Y hepaticojejunostomy (RYHJ) was done in 60 cases. One patient had a T-tube choledochocystostomy done in the first operation, following which an excision with a RYHJ was done 20 days later. One patient, a 4-month-old infant who had a perforation of CDC and bile peritonitis, had an excision of CDC but, due to poor general condition during surgery, only a tube - hepaticostomy was done in the initial surgery.

Choledochocystoduodenostomy (CDCD) was performed as an internal drainage procedure in 8 cases. Six of these 8 cases had a cylindrical type of CDC. One patient was noted to have advanced liver cirrhosis with ascites, therefore, her surgeon elected to do a CDCD instead of an excision. One patient, whose CDC was a type Ia cyst, was treated with a CDCD in 1977, when an excision was not a popular procedure.

Roux-Y choledochocystojejunostomy (RYCDCJ) without an excision was performed in 3 cases. Two of these had cirrhosis and ascites when the procedure was done at the age of 2 months and 11 months respectively. The other case had the procedure done in 1977.

The patient with Caroli's disease had a liver biopsy only.

Result of Treatment

1. Postoperative complications

1.1 Excision group

Six patients or 9.5 per cent of this group developed postoperative complications. Four of these, or 6.5 per cent, succumbed.

Upper gastro-intestinal bleeding requiring a transfusion occurred in one case. The patient recovered satisfactorily after the transfusion.

Subphrenic abscess occurred in one case. The patient recovered satisfactorily after re-exploration and drainage.

Leakage of hepaticojejunal anastomosis occurred in 2 cases. One patient recovered satisfactorily after re-exploration and repair. The other, who had associated biliary atresia and liver cirrhosis at the initial surgery at the age of 5 months, was noted to have bilious drainage from the site of previous drain 26 days after the initial surgery. This required re-exploration. She had massive adhesions and diffuse bleeding during surgery, and eventually succumbed to postoperative bleeding within 24 hours after surgery.

Three other patients died in the postoperative period. One patient, who was a 4-month-old infant with perforated CDC, died of sepsis. Another patient, who was a 4-month-old infant with established cirrhosis, died from postoperative intra-abdominal bleeding 24 hours after excision of CDC. The third patient, who was a 2-month-old infant, did well in the first 24 hours postoperatively, but then developed bradycardia without apparent cause. She eventually expired and a post-mortem examination revealed bilateral atelectasis.

It was noteworthy that all 4 patients who died postoperatively were infants between 2 to 4 months of age. Two of these were found to have cirrhosis during the initial surgery. Of the 6 infants below one year of age with established cirrhosis who underwent an excision of CDC, 4 developed postoperative complications. Two of these succumbed to the postoperative complications.

1.2 Internal drainage group

In the 8 patients who had CDCD, there was no immediate postoperative complication. Of the 3 patients who had RYCDCJ, one, who also had cirrhosis during initial surgery at 2 months of age, developed a massive upper gastro-intestinal bleeding. The bleeding did not respond to massive transfusion and required re-exploration, gastrotomy and ligation of the bleeding points 4 days after the first surgery. She recovered satisfactorily from the second surgery.

There was no postoperative mortality in the patients who had internal drainage procedures.

2. Delayed complications

The majority of patients lived in towns distant from Bangkok and were lost to follow-up shortly after surgery. Twenty-two patients came back for a follow-up longer than 12 months postoperatively. Of these, 8 were lost to follow-up within 36 months, while the remaining 14 patients returned for a follow-up longer than 36 months. The longest follow-up was 12 years.

The patient with Caroli's disease developed multiple episodes of hematemesis from esophageal varices. She was lost to fellow-up at the age of 6 1/2 years.

One patient, who had an excision of CDC at the age of 6 years, developed multiple stones in the intrahepatic bile ducts. She underwent re-exploration and removal of bile duct stones 6 years after the initial surgery. However, the stones recurred and required 2 additional operations to remove them. She also developed an episode of intestinal obstruction, which required another laparotomy and lysis of adhesions.

Three additional patients, who also had previous CDC excision, developed intestinal obstruction due to adhesions. Two of these patients recovered satisfactorily from a laparotomy and lysis of adhesions, but one unfortunately had inadvertent injuries to the small and large bowels during lysis of adhesions. This patient developed wound infection, wound dehiscence, small bowel fistula and eventually succumbed to sepsis.

It is noteworthy that all but one complication occurred in patients who had excision of CDC. According to our available records of follow-up, which were admittedly not complete, none of the 11 cases with internal drainage procedures returned with a cholangitis or recurrence of symptoms.

DISCUSSION

Our experience of 74 pediatric cases over a 19-year period, or approximately one in 2,400 admissions at the Children's Hospital, indicates that the incidence of this disease in Thailand is probably higher than that of Western countries, where the incidence was reported to be one in 13,000-15,000 admissions[13,14]. This report is the largest reported series in Thailand so far.

When ages at the onset of symptoms are examined, two peaks of incidence are apparent, one in the first year of life and the other at 2-6 years of age (Fig. 1). CDC in infants below one year of age appeared to be different in several aspects from that of older children and adults.

Infantile CDC most frequently presented with cholestatic jaundice and acholic stool. More than half of the infants who were diagnosed within one year of age in our series developed jaundice shortly after birth. The clinical manifestation in this group of patients is similar to that of biliary atresia and neonatal hepatitis[10,15,16]. Three of our infants had CDC with atresia of distal CBD. This association was also reported by others[4,8,10,15,17]. Histologic findings of liver biopsy specimens in patients with infantile CDC frequently could not be distinguished from those of biliary atresia[15,18]. Landing[18] proposed that both diseases, and also neonatal hepatitis, probably represented different outcomes of the same obstruc-

tive inflammatory process. In our experience, as well as other reports[4,10,15,17], infants with CDC had a poorer prognosis than patients in the older age group. They tended to develop liver cirrhosis early and had higher operative risk. In contrast, older children and adults had different clinical manifestations and better prognosis. None of our 49 older children was found to have liver cirrhosis during surgery.

Numerous hypothesis have been proposed regarding the etiology and pathogenesis of CDC [1,7,18,19]. However, no single theory has been able to explain the pathogenesis of all CDC satisfactorily. It is possible that the etiology of this disease is multifactorial. The most plausible theory at present is the hypothesis proposed by Babbitt[19], who observed the frequent association of a long common channel of the pancreatic and bile ducts and a CDC. This anomalous pancreaticobiliary ductal system is believed to allow the reflux of pancreatic juice into the CBD. Chemical and enzymatic actions of pancreatic juice may result in inflammation, weakness of the CBD wall, narrowing and obstruction of the distal CBD and dilatation of the CBD to form a CDC. Evidence to support this theory includes high level of amylase in CDC content and demonstration of a long common channel of the pancreatic and bile ducts by an IOC and endoscopic retrograde cholangiopancreatography (ERCP)[6,7,10,20-22]. This mechanism may be responsible for the pathogenesis of most CDC in older children and adults, but is unlikely in most infantile CDC. Our experience indicated that high amylase level in CDC content was most frequently noted in older children, but rare in infants below one year of age. Antenatal diagnosis of the CDC by ultrasonographic study has been reported [23-27]. The earliest gestational age at diagnosis was 15 weeks, at which time the pancreatic acini must not be mature enough to produce enzymes [24]. Detection of CDC in such an early stage of pregnancy indicates that a long common channel of the pancreatic and bile ducts and reflux of pancreatic juice into the CBD should not be the cause of CDC in these cases. Frequent association of CDC and BA in newborn infants, similar hepatic histologic changes and a high incidence and early development of cirrhosis in infants with CDC suggest that the two diseases may be etiologically related.

In our experience, ultrasonographic study has been proved to be very accurate in the diagnosis of CDC. CT scan and DISIDA scan, though also very accurate in diagnosis, are usually not necessary in most cases. ERCP has been shown to be very accurate in the complete study of the anatomy of the biliary tree in patients with CDC[21,22]. However, this study needs special expertise and its availability is limited to a few hospitals in Thailand. Operative cholangiogram, in contrast, is readily available in most hospitals, and therefore should be done in every case during surgery, because it gives invaluable information for a surgeon to choose the appropriate procedure.

Excision is well accepted as the procedure of choice for type I or IV of CDC at present[2-7,12,15,28-31]. The intrahepatic portion is usually left alone. Todani[31] recommended that the common hepatic duct should be removed as much as possible and the anastomosis made high in the porta hepatis. If the intrahepatic ductal dilatation is confined to the left lobe, a left lobectomy may be performed[31]. The preference of excision over internal drainage procedures is based on the high failure rate after internal drainage procedures and the decreased risk for carcinoma development in the long-term follow-up. In addition, the morbidity and mortality rates have been found to be low following excisional treatment in recent years. A mortality of 6.5 per cent following excision in our series appeared to be reasonably low[2,3]. This could be lower if selection for excision was done more cautiously. A "poor risk group" in our experience included infants with established cirrhosis and infants with poor general condition due to sepsis from suppurative cholangitis or perforation of CDC. Excision in this group of patients was encountered with high morbidity and mortality rates. Infants with established cirrhosis and ascites may be better off with an internal drainage procedure. In infants with sepsis from suppurative cholangitis or perforation of CDC, a temporary external drainage or internal drainage with a subsequent excision of the CDC may offer a better chance for a successful result.

Technically, total excision of CDC may not be very difficult in most cases. However, in some cases with large CDC or CDC with previous repeated infection, a technical difficulty may be

encountered during the dissection of the cyst. Lilly's technique of excision has been very helpful to avoid injury to the neighbouring vascular structures[15]. In our experience, this technique at times resulted in considerably more blood loss during dissection than the conventional total excision technique.

Choledochocele or type III CDC is etiologically different and has different treatment options[32]. Its occurrence is rare[3].

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ซีสต์ของท่อน้ำดีร่วม : การศึกษาในผู้ป่วยเด็ก 74 ราย

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ผู้ป่วยเด็ก 74 รายได้รับการรักษาเพราะโรคซีสต์ของท่อน้ำดีร่วมในโรงพยาบาลเด็ก ในช่วง พ.ศ. 2520 ถึง 2538. อัตราส่วนเพศหญิงต่อเพศชาย คือ 5 : 1 ร้อยละ 40 ของผู้ป่วยทั้งหมดเริ่มมีอาการภายในอายุ 1 ปีแรก และร้อยละ 75 เริ่มมีอาการภายในอายุไม่เกิน 5 ปี ประมาณหนึ่งในสามของผู้ป่วยทั้งหมดได้รับการวินิจฉัยโรคที่ถูกต้องและการรักษาภายในอายุไม่เกิน 1 ปี

มีผู้ป่วยเพียง 5 รายหรือร้อยละ 6.8 ของผู้ป่วยทั้งหมดเท่านั้นที่มีลักษณะทางคลินิกครบทั้งสามอย่างคือ อาการตัวเหลือง อาการปวดท้องและคลำพบก้อนในท้อง ในเด็กที่มีอายุต่ำกว่า 1 ปีอาการที่พบบ่อยที่สุดคืออาการตัวเหลือง แต่ในเด็กที่อายุมากกว่านั้นอาการที่พบบ่อยที่สุด คืออาการปวดท้อง ทารก 3 รายมีโรคท่อน้ำดีตันร่วมด้วย ในกลุ่มเด็กที่ได้รับการผ่าตัดเมื่ออายุไม่เกิน 1 ปีมีถึง 9 รายที่แพทย์พบว่าตับมีโรคตับแข็งแล้วในขณะผ่าตัด แต่ในเด็กที่ได้รับการผ่าตัด เมื่ออายุมากกว่านั้นไม่มีรายใดเลยที่แพทย์พบว่ามีโรคตับแข็งในขณะผ่าตัด

ผู้ป่วย 44 รายมีซีสต์ของท่อน้ำดีร่วมแบบที่ 1 ผู้ป่วย 29 รายมีซีสต์แบบที่ 4 และ 1 รายมีซีสต์แบบที่ 5 การตัดเอาซีสต์ออกได้รับความนิยมทำมากที่สุดและมีอัตราตายหลังผ่าตัดร้อยละ 6.5 การผ่าตัดเปลี่ยนทางเดินน้ำดีจากซิสต์ลงสู่ลำไส้ ไม่มีอัตราตายหลังผ่าตัดเลย ผู้ป่วยที่ได้รับการผ่าตัดเมื่ออายุไม่เกิน 1 ปีมีอัตราการเกิดภาวะแทรกซ้อนและอัตราตายหลังผ่าตัดสูงกว่าผู้ป่วยที่ได้รับการผ่าตัดเมื่ออายุมากกว่า 1 ปี คณะผู้รายงานเชื่อว่าการตัดเอาซีสต์ออกเป็นการรักษาที่เหมาะสมที่สุดในผู้ป่วยส่วนใหญ่ แต่การผ่าตัดเปลี่ยนทางเดินน้ำดีจากซิสต์ลงสู่ลำไส้ อาจมีความเหมาะสมในรายที่มีอัตราเสี่ยงสูงบางรายโดยเฉพาะอย่างยิ่งในรายที่มีโรคตับแข็งแล้ว

ข้อมูลจากการศึกษาผู้ป่วยกลุ่มนี้และการทบทวนวารสารบ่งชี้ว่าอุบัติการของโรคนี้ในประเทศไทยคงจะสูงกว่าในประเทศทางตะวันตก สาเหตุของการเกิดโรคนี้ในเด็กเล็กอาจจะต่างจากสาเหตุในเด็กโตหรือผู้ใหญ่

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