Review article

An Institutional review on pediatric choledochal cyst

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Abstract:

Objective: To study the clinical presentation of choledochalcyst(CC), management & outcome over 5 year period.

Methods: All cases of choledochal cyst managed in our institute from 2008 to 2012 were reviewed retrospectively. Data on clinical presentation, bloodparameters, imaging and surgical management along with outcome were analyzed.

Results : Total number of cases was 56.Age at presentation ranged from 31 days to 12 years with mean age of 5.2 years.There were eight cases within one year of age(Infantile CC) and presented with abdominal fullness (50%),jaundice(37%) and fever(12%).Remaining 48cases (Pediatric CC) presented with abdominal pain (79%), jaundice (42%).Spontaneous perforation of bile duct and biliary cirrhosis wasobserved one in each.Majority of cases (52/56) were diagnosed by USGabdomen and in 4 cases diagnosis was made by MRCP. Type I CC was the most commontype seen in 40(71.4%) followed by type IV CCin 14 (25%) and type III CCin 2(3.5%).Anomalous pancreatic duct system and pancreatic divisium was present in one each.All were managed surgically by cyst excision with either Roux en Y hepaticodochojejunostomy(49 cases) or hepaticodochodudenostomy(6 cases).On follow up during the study period of one year,all were asymptomatic except one who developed idiopathic portal vein thrombosis.

Con**clusion**: Mean age of presentation was 5.2years.Clinical symptoms included abdominal pain and jaundice. Classical triad is a rare presentation in pediatric population. Type 1CC is the most common type. Surgical excision of cyst with biliary diversion is the treatment of choice.

Key words: Choledochalcyst(CC),Hepaticojejunostomy,Children,Neonatal cholestasis.

Introduction

Choledochalcyst(CC) is a congenital anomaly characterized by focal cystic or fusiform dilatation of biliary duct. The incidence ranges from 1 in 100,000 live births in West to 1 in 1000 live births in Asia.[1,2] Majority of cases are diagnosed in childhood and about 25% are diagnosed in adulthood.[3]With improved imaging options they are being diagnosed in antenatal period also.They are classified anatomically based on modified Todani et al [Fig:1]. CCs have associated anomalies like abnormal pancreaticobiliary duct junction (APBDJ) and other biliary tract abnormalities.[4]Surgical excision is the treatment of choice.Here we reviewed cases of CCs that are managed in our institute over 5 year period.

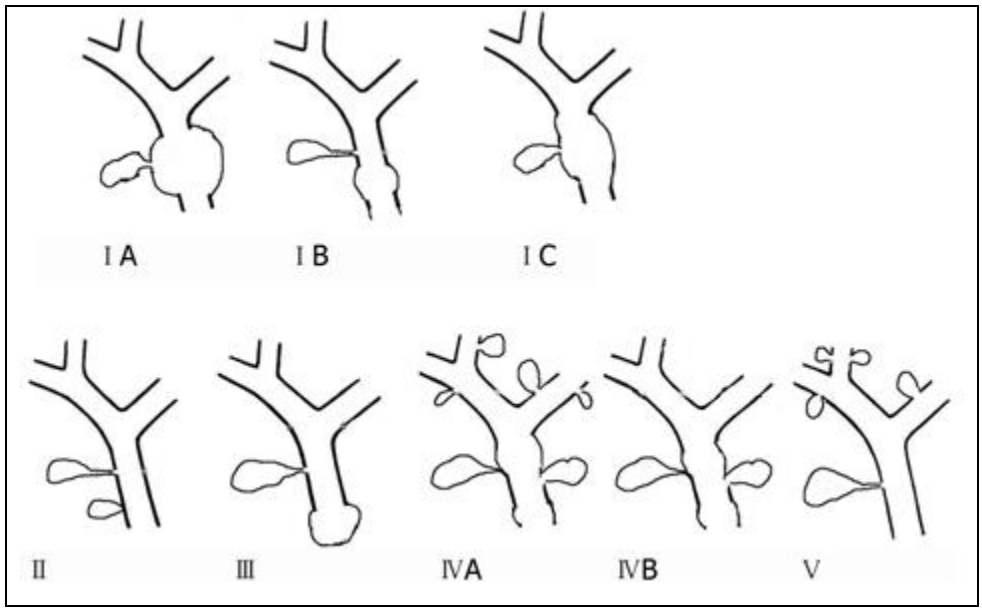

Figure 1: Modified Todani et al classification of choledochal cysts

(Source: with permission from www.wignet.com)

Materials & methods

All cases of Choledochal cyst diagnosed and managed in institute of Child Health, a tertiary pediatric care in Chennai during the period 2008 to 2012 were retrospectively analyzed. Clinical presentation, investigations,operative findings, surgical outcome and short term follow up till one year were analyzed.

Results

Total number of cases was 56. Eight cases were diagnosed in infancy and rest 48 cases presented in more than 1 year of age.

The youngest child was a 31 day girl child who presented with neonatal cholestatic jaundice. Age at clinical presentation ranged from 31 days to 12 years with mean age of 5.2 years. There is slight female preponderance with male female sex ratio of 1:1.2.The most common presentation in infancy was abdominal fullness (hepatomegaly) in 4out of 8 cases (50%) followed by jaundice in 37.5% .In pediatric age group, abdominal pain (79%) was the most common presentation followed by jaundice in 41% of cases.None had the triad of abdominal pain, mass and jaundice. One child presented with signs of biliary peritonitis and another presented with distended abdominal veins with splenomegaly.

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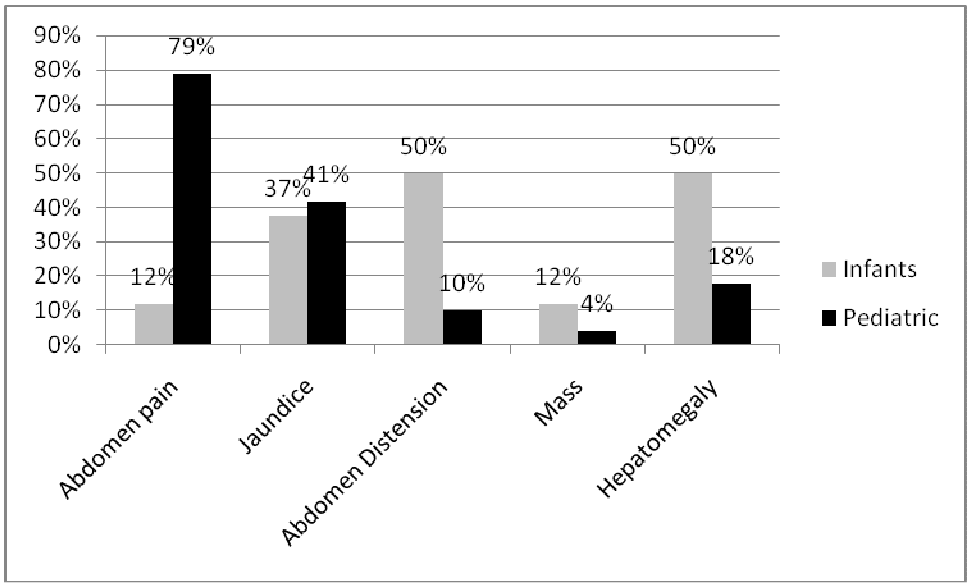

Figure 2: Frequency of clinical presentations in infantile and Pediatric CCs.

Table 1: Laboratory Parameters

Blood Investigations	N = 56(%)
Anemia	14(25)
Jaundice	23(41)
< 5mg%	12(21.4)
>5mg%	11(19.6)
Abnormal transaminases	18(32.1)
Increased SAP(>250iu)	7(12.5)

The important pretreatment blood investigations were as shown in table 1.Jaundice was present in 41% and mild abnormal transaminases in about one third of cases. Only 12.5% had elevated serum alkaline phosphatase.

Ultrasound abdomen (USG) was done in all cases and diagnosis was made in 52 out of 56 cases using this modality. In four cases USG was equivocal and diagnosis was made using MRCP.

Table 2: Frequency of types of CholedochalCysts(CCs)

Type of Choledochal cyst	N(%)
Ia	40(71.4)
Ib	1(1.8)
Ic	3(5.3)
II	0
III	2(3.5)
IVa	10(18)
IVb	0
Association APBDJ, pancreatic divisium	1 case

Based on modified Todani'sclassification type Ia was the most common type(71%) followed by type IVa(18%) and type Ic(5.3%).Type IVb or type II was not seen in our series. Type Ib and III were present in 1.8% and 3.6% respectively. Intraoperative findings were consistent with imaging studies.Thus,USG abdomen is a good diagnostic tool with high accuracy in diagnosis of CC in children. Elective surgery was done in all children except in one who presented with peritonitis.Complete excision of cyst and biliary diversion in the form of either hepaticojejunostomy(49 cases) or hepaticojejunostomy(6 cases) was done.Lilly's procedure followed by hepaticojejunostomy was done in one. Postoperative complications included minor wound infections in seven successfully treated with antibiotics,biliary leak in two was managed conservatively and requiring relaporotomyone in each..One child with cirrhosis died on fourth post-operative day.

Follow up.

All 55(one died on fourth POP) children were on follow up for mean period of 11.5 months. Except for one child whodeveloped idiopathic portal vein thrombosis, all were asymptomatic.

Discussion

Cystic dilation of extrahepatic duct, intrahepatic duct, or both is termed as choledochal cyst. The term 'choledochal cyst' is in fact a misnomer because this condition is not confined to extra hepatic bile duct and 'bile duct cyst' or 'biliary cyst' is probably a better terminology. This uncommon congenital anomalies of bile ducts have varying incidence ranging from 1 in 100,000 -150,000 live births in western population to as high as 1 in 1000 in Asian population, particularly Japan.[1,2] In India, only case series have been reported. Diagnosis of CCs is usually made in childhood period while about 25% are detected late in adult life.[3]There is unexplained female:male preponderance - 4:1 to 3:1 in various studies[3] while we observed 1.2: 1 in our series. Theetiology of choledochal cysts remains unknown. It is speculated that presence of an anomalous pancreatico-biliary ductal confluence (APBDJ) predisposes to reflux of pancreatic secretions into bile duct and induces chronic inflammation resulting in

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weakening and abnormal dilatation (Babbitt's theory).[5] This also explains the higher risk of malignancy of biliary tree.The incidence of APBDJ was reported to be 65% in Japanese studies [6, 7] but we observed only one case (≈ 2%) in our study.The reason may be difficulty in diagnosis usingUSG abdomen and none of the child had ERCP done which may be needed for diagnosis of APBDJ.

In 1959, Alonso-Lej et al [8] first described the initial classification of 3 types of CCs. Later in 1977, Todani et al [9] made modifications and this modified classification is most commonly used which is based on location of biliary duct dilatation as type I – V.[fig: 1]

Childhood CCs have wide spectrum of presentation from asymptomatic to symptomatic rupture of cysts. In general childhood CCs are categorized into "infantile group"(less than 1 year) and "classical pediatric group" (more than 1 year).This 2 groups differ in clinical presentation and pathological anatomy.[11, 12] In infantile group, presence of jaundice with hepatomegaly makes differentiation from biliary atresia a difficult task clinically and needs imaging to study the configuration of intrahepatic bile ducts to make diagnosis.[12]On the other hand, in classical pediatric group CCs the classical triad of jaundice, abdominal pain and right hypochondrial mass is a rare presentation[10, 13] but 85% of children have any 2 features of classical triad.[14]

In our series, out of 8 infants, four (50%) presented with abdominal distension (hepatomegaly) as the presenting symptom and 37% as jaundice.The youngest infant was a 31days old girl child presented with abdominal distension, jaundice (8mg %) and fever mimicking neonatal cholestasis syndrome. Abdominal pain (79%) was the most common presentation in pediatric group followed by jaundice and fever in nearly one third.Thus,CCs should be considered as one of the differential diagnosis in child with biliary colic, recurrent cholangitis and evidence of dilated CBD.

Diagnosis of CCs is confirmed by imaging techniques.Ultrasound abdomen with sensitivity of 71-97% [15] is the preferred initial investigation of choice followed by ERCP or MRCP for better visualization of both biliary and pancreatic ducts prior to surgery that helps in complete excision of cysts and detection of associated anomalies.

Advantage of ERCP is good accuracy in detection of APBDJ and therapeutic option of papillotomy simultaneously in case of type III cyst.ERCP need large amount of dye to fill cyst thereby increase the risk of cholangitis and pancreatitis[16]along with risk of radiation precluding its use in children making MRCP a non-invasive procedure as the "gold standard" option for the diagnosis of CCs with sensitivity as high as 90–100%. [17]However sensitivity for delineating pancreatic duct and common pancreaticobiliary channel is 46% and lacks therapeutic benefits. Hepatobiliaryscintigraphy usingtechnitium-99 hepatobiliaryiminodiacetic acid (HIDA) scan is useful to differentiate biliary atresia from CCs in the newborns. It can also diagnose spontaneous rupture of CCs.Demonstration of stagnation of bile by HIDA scan in a dilated bile duct in the absence ofintraductal calculi or biliary obstruction has been reported as a diagnostic sign. [18]

As reported by many studies[19, 20]we also observed type I CCs were the common CCs and type II and III were uncommon. Initially CCs were managed with internal drainage but studies later[21]revealed that there is high risk of carcinoma from the lining of cyst if internal drainage alone was done.This warranted

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complete excision of cyst with bilioenteric anastomosis as the treatment of choice. Hepaticojejunostomy is the preferred anastomosis as evidenced by various studies.[22,23,24] We had one child presented with biliary peritonitis(12%) due to rupture of CC which was comparable to the previous report of 18% in one series.[25] Follow-up of operated children showed CCs has excellent prognosis. Retrospective analysis and short term follow up are the major limitations of our study.

Conclusion

Any children presenting with abdominal pain and jaundice with or without mass diagnosis of CC should be kept in mind and USG abdomen is the preferredinitial screening tool. Surgical complete excision of cyst offers excellent short term prognosis.

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