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Choledochal cyst--a different disease in newborns and infants

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SHORT COMMUNICATION

Choledochal Cyst - A Different Disease in Newborns and Infants

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ABSTRACT

We report experience of managing Choledochal Cyst (CC) in different paediatric ages. Eleven neonates and infants (aged 0-8 months) and 24 paediatric cases (aged 2.5 - 18 years) were managed over 24 years (1988 to 2012). Neonates and infants presented with jaundice, acholic stools and abdominal mass whereas most of the paediatric cases presented with intermittent non-specific abdominal pain. Morphology of CC was mostly cystic in neonates whereas it was fusiform in majority (62%) of paediatric cases. Biliary amylase was high and correlated with the presence of abnormal pancreatico-biliary junction (PBJ) in 20 /24 paediatric patients. Obstruction at the lower end of bile duct, liver fibrosis and cirrhosis were common in neonates. In conclusion, CC in newborns and infants is different and mimic correctable Biliary Atresia (BA). Early excision of CC and biliary reconstruction is promising in neonates, infants and children and it can be performed with minimal morbidity.

Key Words: *Choledochal cyst. Choledochocal malfomation. Abnormal pancreatico-biliary junction. Children. Excision. Biliary reconstruction. Ultrasonography. Per-operative cholangiography.*

Choledochal Cysts (CC) is a rare entity with incidence of 1:10,000 - 1:150,000 live births. It is a congenital dilatation of biliary ducts. Delay in management can lead to biliary lithiasis, carcinoma and chronic liver damage with ensuing portal hypertension and cirrhosis.[1,2] Although CC can presents at any age, 80 - 90% are diagnosed in childhood. The age at diagnosis is decreasing due to widespread use of ultrasonography in evaluation of non-specific abdominal pain in children and obstetric practice. We share experience of CC from a University Hospital. The emphasis is on the differences in clinical presentation, pathology, surgical approach and outcome at different paediatric ages.

Medical records of 35 children admitted for the management of CC over a period of 24 years (1988 - 2012) were reviewed for clinical data (gender, age at presentation and clinical manifestations), laboratory investigations (serum bilirubin-direct and indirect, SGOT, gamma GT, alkaline phosphatase; biliary amylase levels and liver histology), imaging studies, operative procedure and outcome. Cysts were classified according to Alonso-Leg and Todani classification.[2] Patients were divided into two groups: Group-I having neonates and infants less than one year; Group-II having children above 1 year upto 18 years of age.

Most of the patients were females (78%) and the age at presentation ranged from day 0 to 18 years. Table I summarizes the patients' profile. Morphologically cysts were exclusively cystic in neonates whereas, majority (62%) were fusiform in older children. More than 90% neonates and infants presented with jaundice, clay-color stools and abdominal mass. Older children presented with intermittent abdominal pain and vomiting and 6 patients were initially diagnosed as acute pancreatitis (abdominal pain, and raised serum amylase and lipase levels). Classical triad of jaundice, pain and abdominal mass was uncommon. Biliary amylase levels were elevated in 20/24 patients of Group-II and correlated with the presence of abnormal pancreatico-biliary junction on peroperative cholangiogram (POC),[3] whereas, in neonates POC demonstrated obstruction at lower end of Common Bile Duct (CBD). All the patients in Group-I had abnormal liver histology: portal fibrosis (8 patients) and biliary cirrhosis (3 patients). Shortly after the diagnosis all patients had complete excision of CC and enteric biliary reconstruction. Average age at surgery was 2.2 months (range: 0 - 8 months) in Group-I and 6.4 years (range: 2.5 - 18 years) in Group-II. Follow-up ranged from 6 months to 15 years (mean: 5.4 years). Complications were seen in 5 patients (all belonging to Group-II) and included wound infection, biliary leakage, pancreatitis and cholangitis. Patient with cholangitis required revision for anastomotic stricture a year after surgery.

Choledochal Cyst (CC) can present in various ways from gestation to old age. Infantile CC is characterized as follows: (I) cystic dilatation of bile duct; (2) abdominal mass with jaundice and acholic stools; (3) no symptomatic association with acute pancreatitis; (4) low amylase levels in bile.[2,4-6] Most of these features were present in 11 neonates reported here. In contrast, the features of CC in older children include fusiform dilatation of CBD, recurrent abdominal pain and hyperamylasemia indicating pancreatitis. The classical triad of jaundice, abdominal pain and mass is rarely seen.[3-6]

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Table I: Patients profile.

Patient's characteristic	Group I (Neonates) N=11	Group II (Children) N=24
Age	0 - 8 months (mean 2.2 ± 2.0)	2.5 - 18 years (mean 6.4 ± 3.4)
Gender:		
Male	3 (27%)	4 (16%)
Female	8 (73%)	20 (84%)
Type of cyst:		
Cystic	11	8
Fusiform	0	16
Abnormal pancreatico-biliary junction (PBJ)	Obstruction at PBJ in all patients	Anomalous PBJ in 20/24 patients
Clinical features:		
Prenatal diagnosis	3 (27%)	0
Jaundice	11 (100%)	7 (29%)
Abdominal pain	0	24 (100%)
Abdominal mass	9 (82%)	5 (21%)
Acholic stools	11 (100%)	0
Pancreatitis	0	6 (25%)
Classical triad	0	2 (8%)
Biliary amylase	< 25 IU/dl	> 25,000 - 45,000 IU/dl
Liver histology:		
Portal fibrosis	8	2
Cirrhosis	3	0
Hepatico-biliary reconstruction:		
Hepaticoduodenostomy	5	0
Roux-en-y hepaticojejunostomy	6	24

The pathogenesis of CC is debated. It seems that pancreatico-biliary reflux caused by anomalous junction of pancreatic and common bile ducts results in fusiform dilatation of CBD in children and adults.[3] However, this theory does not explain the CC diagnosed prenatally or those presenting early in infancy. Cholestatic jaundice with acholic stools in infantile CC is attributed to stenosis or obliteration of lower Common Bile Duct (CBD) during embryonic development of bile duct. The obstruction of CBD also leads to progressive hepatocellular damage ranging from variable grades of fibrosis to cirrhosis. It appears that the clinical and pathological features of type-I cystic Biliary Atresia (BA) and infantile CC resemble and it points towards a common pathogenesis. Some authors have classified it as correctable form of BA.[4]

Improved quality of ultrasonography and its widespread use in obstetric and paediatric practice has improved detection of CC and *in-utero* diagnosis of biliary dilatations. CC has been diagnosed as early as 15 to 16 weeks of intrauterine life.[2,9] A focused ultrasound examination of hepatobiliary region can diagnose and differentiate BA and CC with reasonable sensitivity and specificity. Diagnostic modalities for further delineation of pancreatic and biliary ducts include Magnetic Resonance Cholangio-Pancreaticography (MRCP), endoscopic retrograde cholangio-pancreaticography (ERCP), DICIDA scintigraphy and peroperative cholangiography (POC). Ultrasonography combined with POC is all that is necessary for cost effective and optimal management of CC. POC confirms the diagnosis delineate the intrahepatic bile ducts, abnormal pancreaticobiliary junction and differentiate precisely between the CC and type-I cystic biliary atresia. DCIDA scintigraphy is useful in follow-up of neonatal cases.[2]

Timing of surgical intervention in neonatal CC is controversial probably due to difficult surgery on thin walled cysts in neonates and fear of complications. Options include definitive surgery immediately after diagnosis, close monitoring with serial ultrasound scans and Liver Function Tests (LFT) and surgery at 6 months of age.[7-9] Delay in treatment can cause rapid enlargement of the cyst and progressive liver damage. Liver fibrosis of variable grades and cirrhosis is present in all the reported cases of infantile CC irrespective of age at surgery. In authors' opinion, infantile CC can be operated safely soon after the diagnosis. Patients reported in this series were operated soon after the diagnosis. Favorable outcome after early surgery is well documented.[7-9] In addition, surgery offers the opportunity to distinguish between CC and type-I cystic BA.

Complete cyst excision and *Roux-en Y* Hepatico-Jejunostomy (HJ) is currently the procedure of choice, though concerns are raised about cholangitis, peptic ulcer disease and adhesive intestinal obstruction. Appendico-duodenostomy, jejunal interposition hepatico-duodenostomy with or without valve have been reported as an alternative but are not popular.[2,10]

In conclusion, Choledochal Cyst (CC) in newborns and infants is different when compared to the disease in children and adults and seems to have clinical and pathological features similar to correctable biliary atresia. Prompt surgical treatment is imperative because of difficulty in differentiating this entity from cystic BA and early onset of liver fibrosis. Excision of CC and biliary reconstruction can be performed safely in neonates and infants and it may prevent the end stage liver disease.

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