

Choledochal cys: a different disease in
Newborn and infantsVijai Datta Upadhyaya^{1*}¹Department of Paediatric Surgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, India

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Editorial

Choledochal Cysts (CC) is a rare entity with incidence of 1:10,000 - 1:150,000 live births and 4 times more common in females. CC in childhood frequently categorized into an "infantile" group (patients less than one year old) and "classical pediatric" (CP) group (age more than one year but less than 18). Infantile group differ markedly from classical pediatric group in their clinical presentation and pathological anatomy. Todani *et al* [1] have characterized the infantile CDCs as follows: (1) Cystic choledochal dilatation, (2) Abdominal mass with jaundice and acholic stools, (3) No symptomatic association with acute pancreatitis and (4) A low amylase level in bile.

Clinical features of patients with choledochal cysts differ as a function of patient age and its presentation is varied. It can present with classical triad (jaundice, right hypochondric mass and pain), or in any combination or alone: with abdominal pain, jaundice, abdominal mass [2] cholangitis, pancreatitis, and history of cholecystectomy for biliary symptoms. Presentation in infants is entirely different and they tend to present with painless jaundice, hepatomegaly, and acholic stools [3].

Diagnosis of CC need high index of suspicion because of its varied presentation. Abdominal ultrasound (US) scan is the first step and had sensitivity of 71-93% [4]. Cholangiography, specifically ERCP and percutaneous trans-hepatic cholangiography, is the most sensitive technique to define the anatomy of the biliary system, but are difficult to perform in the infants given the need for general anesthesia, by and technical difficulty. Both procedures are associated with potential complications, including bleeding, cholangitis, acute pancreatitis, and perforation, as a result, noninvasive imaging; Magnetic Resonance Cholangiopancreatography (MRCP) has gained importance. MRCP is regarded as gold standard for the diagnosis of CC with a sensitivity of 70-100% and specificity of 90-100% [5], it can reliably identify APBDU (particularly with the use of secretin) as well as cholangiocarcinoma and choledocholithiasis with concurrent CC.

Etiology of CC is not clear however, there are two hypotheses to explain the development of CC. First theory relates CC to obstruction of the bile duct [6]: obstruction of bile duct leads to increased proximal bile duct pressure [7] and eventual dilatation, initially of the extrahepatic segment and subsequently the intrahepatic component. The second theory known as Babbitt's hypothesis is based on the pathophysiological consequence of reflux of activated proteolytic pancreatic enzymes on the biliary tract wall [8]. Chen *et al* [2] observed that the cystic amylase and lipase levels were significantly elevated in children and adult presenting with CC but was not elevated in cases of cysts in infantile group suggesting no reflux of pancreatic juice in common bile duct [3,9,10] in cases of infantile CC. Min-Hsuan Hung *et al* observed that most of the CC in infants has blind distal end whereas CC in children were connected distally to pancreatic duct, this demonstrate that relationship between the cyst and pancreatic duct is quite different in infants with CP. Hence the former theory backs the development of choledochal cyst in infants whereas other theory explains the development of choledochal cyst in CP. The levels of amylase in cystic bile may be very low, despite malunion due to the fact that acinar growth of pancreas is incomplete in infants. Chau-Jing Chen [2] have diagnosed CC at 23rd week of gestation, acinar development of pancreas is rudimentary at this stage, thus the choledochal cyst due to amylase reflux is unlikely during this period, further supporting the fact that development of CC in infants is different from the CP and adult.

Most widely accepted classification was reported by Todani and colleagues in 1977 [11], derived from the original Alonso-Lej classification and based on the site of cystic change. Type I cysts, the most common, are subdivided as follows: type IA, characterized by a large saccular cystic dilatation; type IB, characterized by segmental dilatation; and type IC, characterized by diffuse or cylindrical dilatation. Type II and III cyst are not reported in infants. Type IVA cysts are characterized by multiple intrahepatic and extra-hepatic cysts whereas type IVB cysts are indicated by the presence of multiple extra-hepatic cysts. Type V cysts (also known as Caroli's disease) are characterized by single or multiple intrahepatic cysts. Most of the infantile cysts are either Type-IA or Type -IV

where as in CP it is usually type I C or Type IV. Other types are usually not seen in infants and CP.

Cysts in the infants are usually spherical whereas in CP they are usually fusiform in shape [11]. Relation between distal end of CC with pancreatic duct is different in infantile CC and choledochal cyst seen in CP. Existence of a connecting duct results in pancreatic reflux and ensuing pathological changes, symptoms, and laboratory alterations of biliary pancreatitis as it is reflected in clinical presentation of CC in CP. Cholestatic jaundice acholic stools in infantile CC are 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 and infantile CC resemble each other and points towards common pathogenesis hence be been classified as correctable form of BA by few authors and it points towards a common pathogenesis and some authors have classified it as correctable form of BA. Though in majority of infants the distal end is blind but in few cases distal end CC was found to be connected with pancreatic duct [2,10]. Severe stenosis or obliteration of cysto pancreatic connection leads to rapid progression to cirrhosis due to back pressure changes and cyst rupture due to high intra-cystic pressure.

On histology CC in children and adult has wall fibrosis, mucosal autolysis and denudation caused by reflux of the bile juice where as in cases of infants the CC usually had columnar epithelial lining and wall fibrosis, denudation of epithelial lining is uncommon. Liver histology at the time of presentation is quite different in infantile and CP choledochal cysts. Obstructive cholangiopathy can lead to liver fibrosis in 35-66.7% infants although progression to cirrhosis is known to occur only in 2.1-11.8%. Gong *et al.* [12] noted a higher fibrosis score in the infantile group (2.5 ± 0.9) compared to pediatric group (1.5 ± 1.2) ($P < 0.05$) similar finding was observe by Neel *et al* [13] where they have reported 57% of infants had cirrhosis in comparison to just 6.5% in CP group.

The main objective of surgery is: complete excision with restoration of adequate bilioenteric communication to avoid long-term consequences. If left untreated in CP, choledochal cysts can cause morbidity and mortality from recurrent cholangitis, pancreatitis, sepsis, liver abscesses, and cirrhosis in long term, where as in infantile group even a slight delay in adequate management can results in rapid progression of liver fibrosis to cirrhosis or rupture of the cyst owing to near complete obstruction of the distal end of the cyst. Management of Type I and IV CC consists of complete extrahepatic bile duct cyst excision down to the level of communication with the pancreatic duct, cholecystectomy, and restoration of bilioenteric continuity. Hepaticoduodenostomy and Roux-en-Y hepaticojejunostomy (RYHJ) bilioenteric reconstruction had been described after excision of the cyst. Hepaticoduodenostomy is associated with increased rates of gastric cancer (due to bile reflux) and biliary cancer. Bilioenteric reconstruction using RYHJ is safe and associated with low incidence of postoperative complications. Development of malignant tumor tends to increase as the age increases, and has not been reported in CC presenting in infants.

Infantile CC is entirely different from the CC of CP or adults in clinical presentation, etiology, pathology and outcome .It is very difficult to differentiate infantile CC with biliary atresia, and can be differentiated with the fact that cysts are larger, IHD are dilated and gall bladder is not atretic in infantile CC in comparison to cystic variant of biliary atresia. The key issue in infantile CC is to differentiate it from cystic variety of biliary atresia and appropriate timing of surgery to avoid dreadful complication like development of cirrhosis leading to portal hypertension and rupture of the cyst. Excision of CC and biliary can be performed safely in neonates and infants but if they present with complications, temporary drainage procedure in form of percutaneous transhepatic drainage or cholecystectomy is safe to overcome acute stage before definitive procedure to avoid morbidity.

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