Choledochal cysts: a different disease in newborn and infants

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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. 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 choledocholeithiasis 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 Carolii’s disease) are characterized by single or multiple intrahepatic cysts. Most of the infantile cysts are either Type-IA or Type –IV.
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.

References