Editorial

Choledochal cys: a different disease in Newborn and infants

Vijai Datta Upadhyaya

*Corresponding author
Vijai Datta Upadhyaya, Department of Paediatric Surgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, India

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 group suggesting no reflux of pancreatic juice in common bile duct [3,9,10] in cases of infantile CC. The levels of amylase in cystic bile may be very low, despite malunion due to the development of choledochal cyst in infants whereas other theory explains the development of the cyst and pancreatic duct is quite different in infants with CP. Different Disease in New Born and Infants. SM J Pediatr Surg. 2016; 2(2): 1012.

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 Cholangiopancreatograpy (MRCP) has gainedimportance. 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.

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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...
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