Summary & conclusion

Choledochal cysts are congenital anomalies of the bile ducts. They consist of cystic dilatations of the extrahepatic biliary tree, intrahepatic biliary radicles, or both that occur more commonly in female than male individuals (with a ratio of 3–4:1), and the incidence in Western nations (Europe and North and South America) is 1 in 100,000 to 150,000 live births and although classically considered a disease of infancy, an increased number of patients are now being discovered in adulthood most commonly in patients of Asian descent, with more than two thirds of the reported cases occurring in Japanese patients. There is no evidence of increased incidence in the Hispanic population.

The definitive etiology of choledochal cysts is not clear. There are two actual concepts: first concept suggests that choledochal cysts arise from inequality in the vacuolization of the biliary tract in early embryonic life . The second is common channel theory, based on an abnormality of the pancreaticobiliary junction and the formation of an abnormally long common channel.

Five major classes of choledochal cysts exist (ie, types I-V), with subclassifications for types I and IV (ie, types IA, IB, IC; types IVA, IVB):

I- Type I cysts, are the most common and represent 80-90% of choledochal cysts. They consist of saccular or fusiform dilatations of the common bile duct, which involve either a segment of the duct or the entire duct.

A- Type IA is saccular in configuration and involves either the entire extrahepatic bile duct or the majority of it.

B- Type IB is saccular and involves a limited segment of the bile duct.

C- Type IC is more fusiform in configuration and involves most or all of the extrahepatic bile duct.

II- Type II choledochal cysts accounts for 2% of choledochal cysts and appear as an isolated diverticulum protruding from the wall of the common bile duct. The cyst may be joined to the common bile duct by a narrow stalk.

III- Type III choledochal cysts ,accounts for about 1 to 5% of choledochal cysts and arise from the intraduodenal portion of the common bile duct and are described alternately by the term choledochocele.

IV- (A) Type IVA cysts consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts.

(B) Type IVB choledochal cysts are multiple dilatations involving only the extrahepatic bile ducts.

V- Type V (Caroli disease) consists of multiple dilatations limited to the intrahepatic bile ducts.

Malignancies in choledochal cyst can arise from the distal CBD, the wall of the cyst (even after successful drainage at cystoenterostomy), or the intrahepatic bile ducts.

Although the classical triad composed of abdominal pain, jaundice and abdominal mass is more common presentation among children compared to adults, in infants in particular, jaundice alone presents more frequently than the classical state.

Ultrasonography, as a non-invasive procedure, is the initial method of choice being able to make the diagnosis even during the intra-uterine period and can demonstrate a choledochal cyst as early as 12 weeks gestation

The major application of CT is to define the course and status of the extrahepatic biliary tree and adjacent structures.Intravenous contrast enhancement of the biliary tract is mandatory.

MRI is a non-invasive, multiplanar imaging modality which does not involve the use of ionizing radiation, has excellent contrast resolution, and requires only minimal patient preparation.

ERCP can delineate the structure definitively, but it is an invasive procedure. Further MRCP provides good structural visualization of the pancreaticobiliary tract

Treatment of choledochal cysts is surgical, except in type V multiple intrahepatic cysts, which can benefit from medical management for variable periods of time.

Total excision of the cyst in types I, II, and IV followed by reconstruction of the biliary tree with hepaticojejunostomy in a Roux-en-Y fashion has been widely accepted as the procedure of choice in treating choledochal cysts

Recently, several groups have successfully performed laparoscopic-assisted and laparoscopic total cyst excision with Roux-en-Y hepatoenterostomy with less complication rates comparable to those of the open procedure. Recently roux-en Y hepaticojejunostomy were performed using a robotic surgical system In an attempt to overcome the the technically challenging laparoscopic biliary procedure.