CONGENITAL DILATATION OF INTRAHEPATIC DUCTS (CAROL

CONGENITAL DILATATION OF INTRAHEPATIC DUCTS (CAROLI’S DISEASE)

This rare congenital condition is characterised by multiple irregular saccular dilatations of the intrahepatic ducts, sepa - rated by segments of normal or stenotic ducts, with a normal extrahepatic biliary system. In Caroli’s syndrome, the biliary dilatation is associated with congenital hepatic fibrosis. The pr esentation is varied, with most patients presenting with abdominal pain, cholangitis or end-stage liver disease. The majority of patients present before the age of 30 years. The sex distribution is equal. Malignancy is a complication of longstanding disease. Management is multidisciplinary: cholangitis or jaundice are treated with appropriate antibiotic therapy and endoscopic or interventional stenting. Hepatic resection may be consid ered in patients with limited disease. Patients with di ff use dis ease and concomitant hepatic fibrosis are candidates for liver transplantation. Recurrence is common, particularly after resection, and long-term surveillance is required.

Ia Ib IVa Figure 71.24 Classi /f_i cation of choledochal cysts. Type Ia and Ib (80–90%): diffuse cystic dilatation; note the extension into the intrapancreatic portion in type Ib. Type II (3%): diverticulum of the common bile duct. Type III (5%): diverticulum within the intrapancreatic portion. Type IV (10%): extension into the liver; type IVa: fusiform dilatation of the entire extrahepatic bile duct with extension into the intrahepatic ducts; type IVb: multiple cystic dilatations involving only the extrahepatic bile duct. Type V: cystic dilatation only of the intrahepatic ducts.

Revision #1
Created 2025-12-31 15:26:35 UTC by Omar Ayman
Updated 2025-12-31 15:26:35 UTC by Omar Ayman