

Choledochal cysts

1723 Vater & Elzer

1959 Alonso-Lei

1977 Todani

IA cystic dilation of common duct 70-80%

IB saccular

IC fusiform

II supraduodenal diverticulum

III intraduodenal (choledochocele)

IVA multiple intra and extrahepatic

IVB extrahepatic cysts only

V intrahepatic cysts only (Caroli's)

Etiology

genetics (Caroli's)

embryology

viral

environment

obstruction (Oddi, dysautonomia, web)

anomalous pancreaticobiliary duct junction (>15mm)(80-100%)

Presentation

West: 1:13,000; East (Japan): 1:1,000

F:M 8:1

70% 1st decade, increasing adult presentation

child: jaundice (71%), pain, abdominal mass

adult: pain (97%), pancreatitis, cholangitis, choledocholithiasis

Dx: US, MRCP, ERCP

Complications

Cholangiocarcinoma: 15-30% lifetime risk

increase with age

posterior wall

found in 10-30% of resected cysts

type III lower incidence

Pancreatitis 30-70% of adult cases

Cholangitis

Portal hypertension

Hepatic failure/transplant

Treatment

I complete excision/Roux-Y

II diverticulum resection/T-tube

III <3cm endoscopic sphincterotomy
>3cm transduodenal resection

IV excise extrahepatic

stones, stricture, abscess: resect intrahepatic

bilobar type IV: transplant

V Caroli's

unilobar: resection

bilobar/failure: transplant

Edil B et. al.:Choledochal cyst disease in children and adults: a 30-year single-institution experience; JACS, 206(5); May 2008, pp1000-1005