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