Fibrozystic liver diseases

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Fibrocystic liver diseases - definition

- Group of related lesions of liver and biliary tract
- Caused by abnormal development of embryologic ductal plate
Fibrocystic liver diseases - embriology

- Ductal plate: a cylindrical layer of cells surrounds the portal vein branches during 1\textsuperscript{st} week of gestation
- Bile ducts are formed by partial involution + remodelling of the ductal plate
- Anomalies may develop at various stages of remodelling:
  - Timing or stage determines the clinico pathological disorder
Fibrocystic liver diseases

- Small intrahepatic bile ducts
- Congenital hepatic fibrosis
- 10%
Congenital hepatic fibrosis

Clinico-path

Results in periportal fibrosis

Usually progresses to cirrhosis by adolescence (variceal bleeding, splenomegaly)
Congenital hepatic fibrosis

- Small right lobe
- Atrophy of segment IV
- Enlarged segment IV
- Hep C cirrhosis (different patient)
Congenital hepatic fibrosis

Hyperplastic nodules
Collateral vessels

Hep C cirrhosis (different patient)
37-year-old woman
Fibrocystic liver diseases

- Small intrahepatic bile ducts
- 10%
- Congenital hepatic fibrosis
- Biliary hamartomas
Synonyms: bil. microhamartomas, von Meyenburg complexes

Clinico-path:
 Usually multiple small (<15 mm) masses lined by biliary epithelium
 Rarely symptomatic
 Often mistaken for cysts or metastases
Biliary hamartomas

80 y/o woman
-asymptomatic
Biliary hamartomas

- Epithelial-lined
- Contains bile
- Not in communication with ducts
Fibrocystic liver diseases

- Small intrahepatic bile ducts
- Medium-size intrahepatic bile ducts

40%

- Congenital hepatic fibrosis
- Biliary hamartomas
- Autosomal polycystic liver disease
Clinico-path:
Multiple (innumerable) cysts (few mm to >12 cm)
May be inherited with or without kidney or other organ involvement
Often causes huge hepatomegaly, but rarely affects liver function
Polycystic liver disease
Polycystic liver disease
Fibrocytic liver disease

- Small intrahepatic bile ducts
- Medium-size intrahepatic bile ducts
- Large intrahepatic bile ducts

- Congenital hepatic fibrosis
- Autosomal polycystic liver disease
- Biliary hamartomas

70% Caroli
Carolí’s disease

Abnormal ducts retain communication with biliary tree
Often associated with hepatic fibrosis
Often develop intrahepatic calculi
Often present with cholangitis, jaundice
Can progress to cirrhosis
Can develop cholangiocarcinoma
Carolí’s disease

Dot sign
Caroli’s disease
Carolí’s disease

Evolution to biliary cirrhosis
Fibrocystic liver disease

- Small intrahepatic bile ducts
- Medium-size intrahepatic bile ducts
- Large intrahepatic bile ducts
- Large extrahepatic bile ducts
- Congenital hepatic fibrosis
- Autosomal polycystic liver disease
- Biliary hamartomas
- Caroli
- Choledocal cyst

90%
Choledochal cyst

56-year-old woman
Choledochal cyst

note aberrant entry of CBD into side of panc duct
Fibrocystic liver disease

(47 y/o woman)
Cong Hep Fibrosis
+biliary hamartomas
+polycystic kidneys
25-year-old woman with congenital hepatic fibrosis and Caroli syndrome.