Choledochal Cyst

- Congenital segmental cystic dilatation of intrahepatic or extrahepatic bile ducts, most commonly affecting main portion of extrahepatic duct (ED).

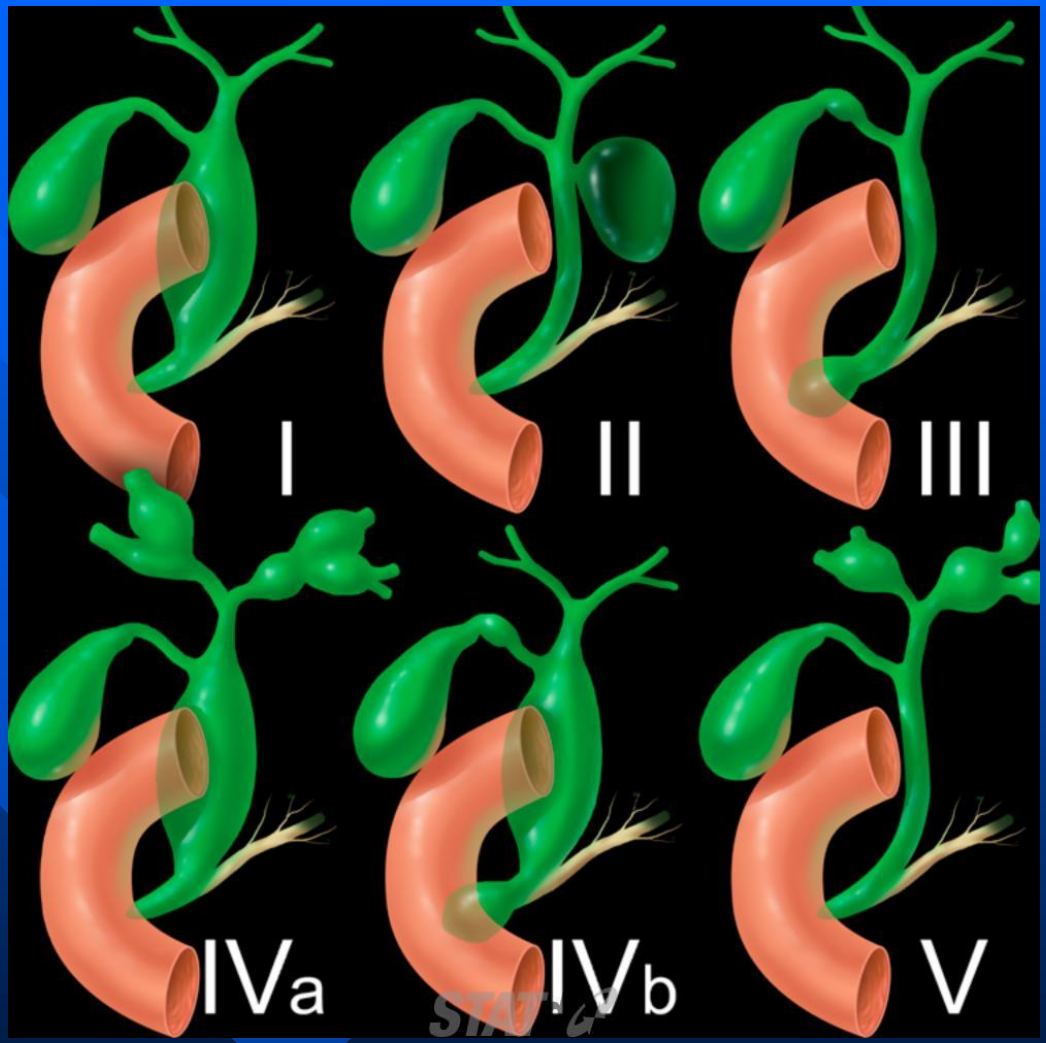
- Segmental cystic dilatation of bile ducts with dilated segments maintaining communication with biliary tree
  - Diagnosis requires excluding other causes of biliary dilatation, including tumor, stone, or stricture
  - Commonly associated with cholelithiasis, cystolithiasis, choledocholithiasis, and hepatolithiasis

- Classified into 5 types based on Todani classification

Types

- **Type I:**
  - Fusiform/cystic dilation of extrahepatic duct
- **Type II:**
  - True diverticulum of supraduodenal ED
- **Type III:**
  - Dilation limited to intraduodenal segment of ED (a.k.a. choledochocele), with dilated segment of duct located within duodenal wall
- **Type IV:**
  - Presence of multiple biliary cysts, at least 1 of which must involve extrahepatic bile duct
- **Type V:**
  - Single or multiple intrahepatic biliary cysts, with multiple intrahepatic cysts known as Caroli disease

- Type I: is fusiform dilation of the extrahepatic duct (ED).
- Type II is a true diverticulum of the supraduodenal ED.
- Type III: is an isolated choledochocele.
- Type IV: is fusiform dilation of theextrahepatic duct and intrahepatic ducts
- Type V: is synonymous with Caroli disease



Clinical Issues

- Most common in female patients, often of Asian descent
- Usually diagnosed in infancy and childhood
- Classic triad of symptoms: Recurrent RUQ pain, jaundice, and palpable mass
- Complications: Stones, cholangitis, pancreatitis, or malignant degeneration (cholangiocarcinoma or gallbladder cancer)
- Treatment varies depending on type of choledochal cyst, but type I and IV cysts typically undergo surgical excision and reconstruction by Roux-en-Y hepaticojejunostomy
- **Consider**
  - Rule out other conditions that can cause marked biliary dilatation
- **Image Interpretation Pearls**
  - MRCP or ERCP: Cystic or fusiform dilation of bile ducts without obstructing lesion