Cholangiocarcinoma

- Cholangiocellular or bile duct adenocarcinoma
- Malignancy arising from intrahepatic bile duct (IHBD) or extrahepatic bile duct epithelium
- Tumor types
  - Exophytic intrahepatic masses (nodular type)
  - Scirrhous infiltrating neoplasms (sclerosing type): Cause strictures
  - Polypoid neoplasms of ductal wall (papillary type): Bulge into bile duct lumen
- Risk factors
  - PSC
  - Fibropolycystic liver disease (choledochal cysts)

Demographics

- **Age**
  - Peak: 6th to 7th decade
    - » ~ 20 years earlier in patients with PSC and choledochal cyst
- **Gender**
  - M:F = 3:2
- **Epidemiology**
  - More common in Asia
  - Causes 15-33% of primary hepatobiliary cancers
  - 2nd most common primary hepatic tumor (following hepatocellular carcinoma)
  - 3% of gastrointestinal malignancies
  - Prevalence in USA: 1-2 in 100,000
  - 0.01-0.46% prevalence in autopsies

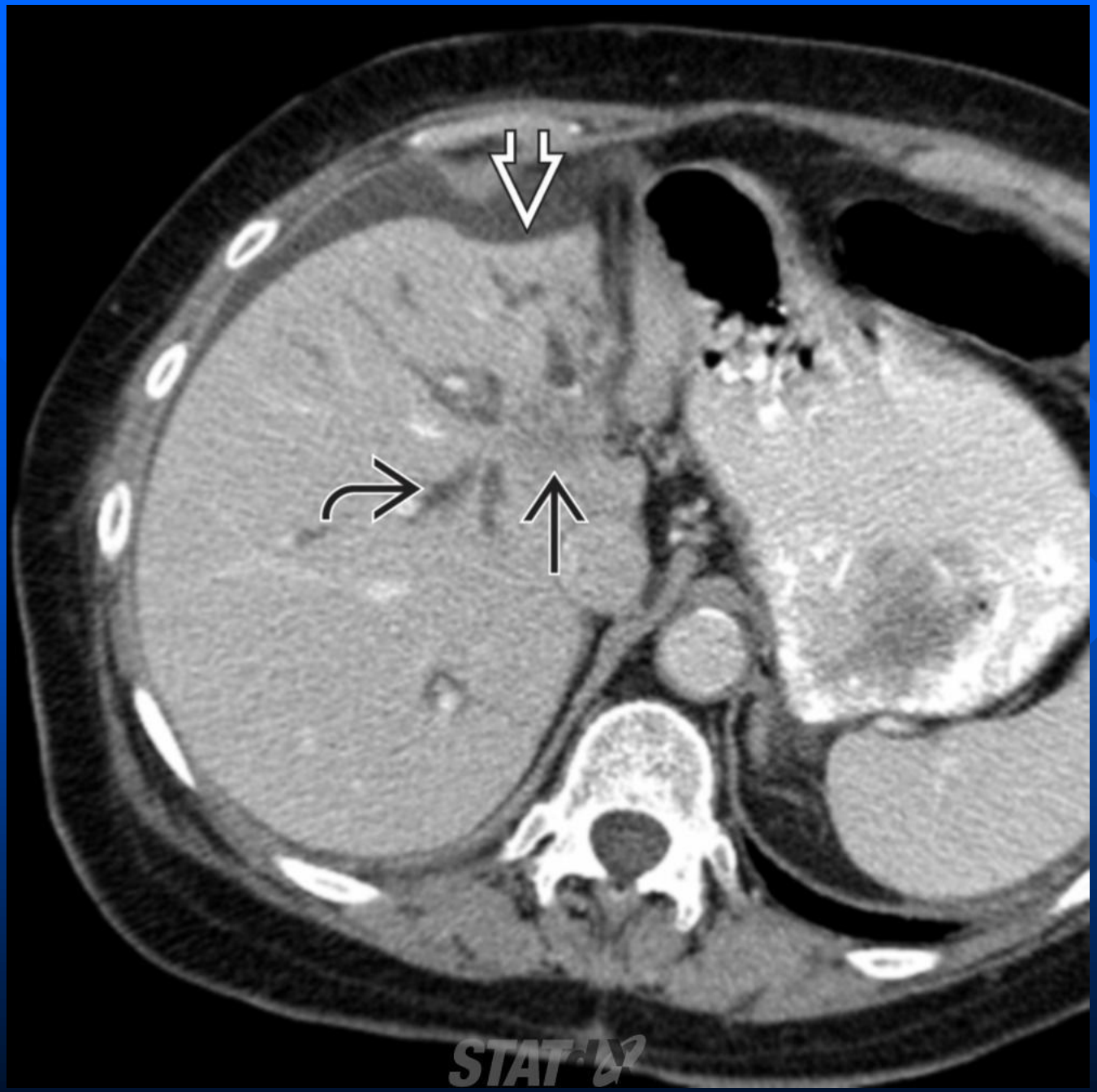
Imaging

- **Klatskin tumor**: Small hilar mass obstructing bile ducts on CT or ERCP
- Cholangiography (PTC/ERCP)
  - Important to determine extent of intra- and extrahepatic duct involvement
  - Extension up right or left duct often precludes surgical resection
- CECT
  - Portal phase: Minimal enhancement of irregularly thickened bile duct wall and distal ductal dilatation
  - Delayed phase: Persistent enhancing tumor (due to fibrous stroma)
- MR
  - Superior to CT for detection of small hilar tumors, intrahepatic & periductal tumor infiltration
  - Shows location of obstruction and IHBD dilatation

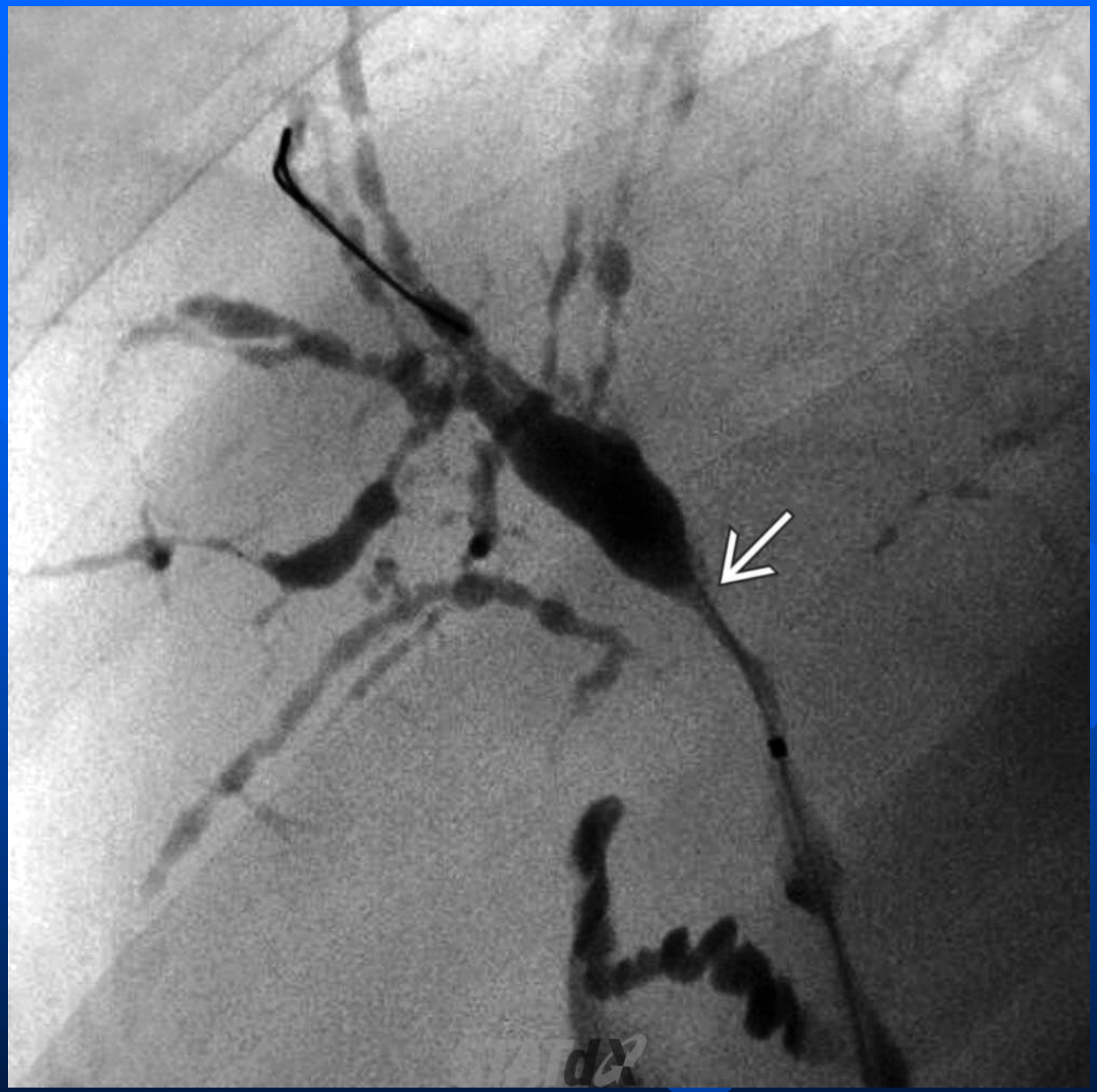
General Features

- **Best diagnostic clue**
  - Klatskin tumor: Small hilar mass obstructing bile ducts on CT or ERCP
- **Location**
  - Distribution in different segments of biliary tree
    - » Distal common bile duct (CBD) (30-50%)
    - » Common hepatic duct (14-37%)
    - » Proximal CBD (15-30%)
    - » Confluence of hepatic ducts (**Klatskin tumor**) (10-26%)
    - » Isolated left or right hepatic duct (8-13%)
    - » Cystic duct (6%)
  - Classified based on anatomy and radiography
    - » Peripheral (10%)
      - Intrahepatic; proximal to secondary biliary radicles
    - » Perihilar (50%)
      - **Klatskin tumor**: Hilar tumor involving the confluence of hepatic ducts
    - » Distal (40%)
      - Extrahepatic; distal CBD
      - May arise as short stricture or small polypoid mass
- **Size**
  - Peripheral cholangiocarcinoma (5-20 cm)
  - Perihilar and extrahepatic types much smaller
- **Morphology**
  - Infiltrative (sclerosing)
  - Exophytic (nodular)
  - Papillary

Cholangiogram shows a mass (white solid arrow) at the confluence of the main right and left ducts with marked dilatation of the intrahepatic bile duct. The common hepatic duct is involved, but the cystic and common bile ducts are not.

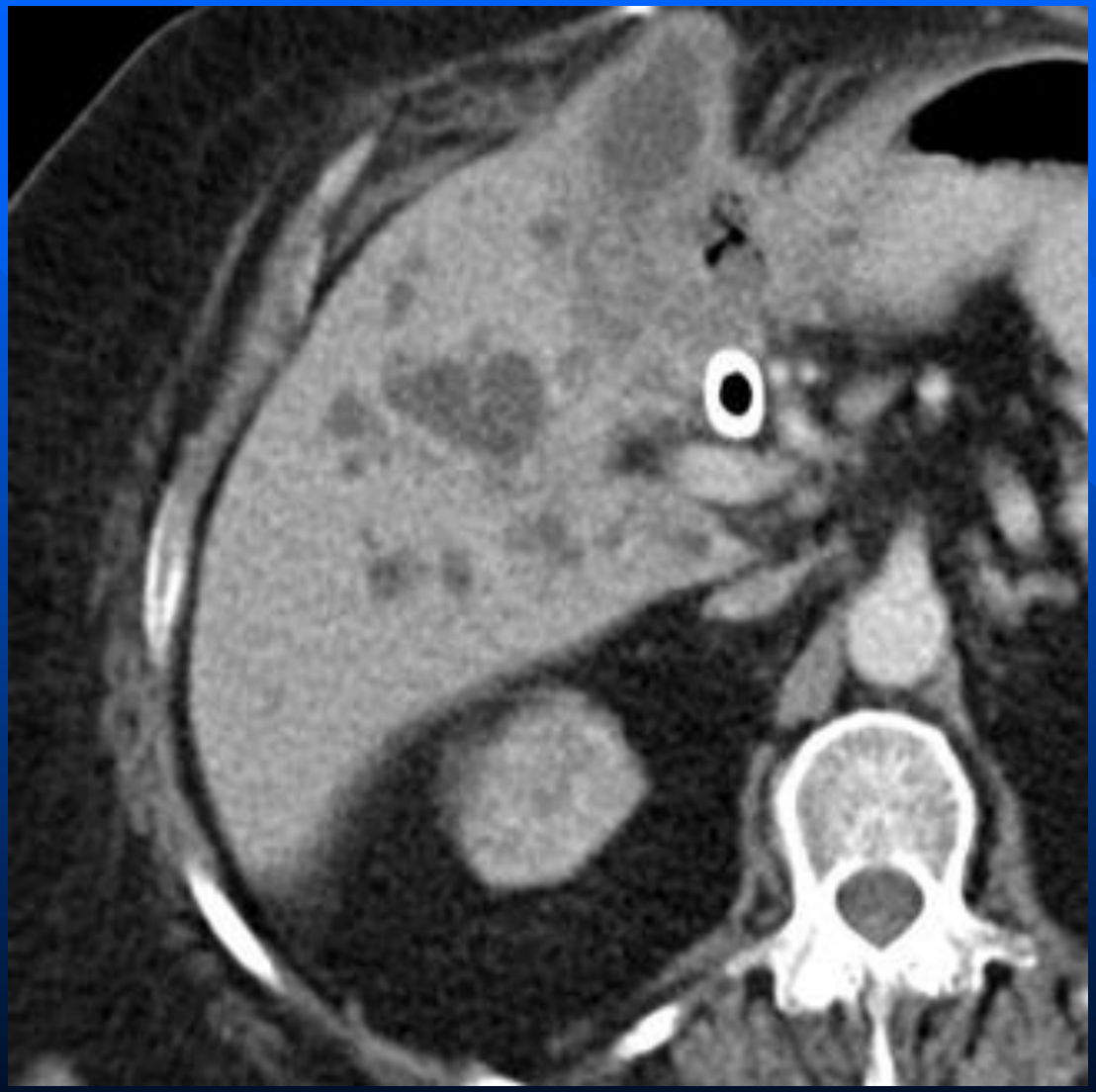


Axial CECT shows intrahepatic bile duct dilation (black curved arrow) ending abruptly at a small mass (black solid arrow) near the hepatic hilum. The lateral segment atrophy (white open arrow) is a secondary sign of longstanding obstruction.



ERCP in the same patient with PSC shows irregular biliary strictures and abrupt narrowing of the common hepatic duct (white solid arrow), with little or no filling of the left hepatic ducts, due to cholangiocarcinoma. Detailed anatomic depiction of involvement of common hepatic, right, and left ducts is critical to determine operability.

Cholangiocarcinoma



Cholangiocarcinoma of the common duct

Cholangiocarcinoma with satellite lesions

Cholangiocarcinoma