Peripheral (Intrahepatic) Cholangiocarcinoma

- Tumor arising from intrahepatic bile ducts.
- Only minority of cholangiocarcinomas are peripheral type.
- 50-60 years, rarely occurring < 40 years
- Delayed tumoral contrast enhancement is typical but not specific feature of peripheral CCACan be seen in other adenocarcinoma metastases and in confluent fibrosis.
- In suspected cholangiocarcinoma (e.g., history of primary sclerosing cholangitis, or mass with capsular retraction, biliary obstruction) obtain delayed enhanced scans

Imaging

- Infiltrative hepatic mass with capsular retraction and delayed persistent enhancement (CECT and MR)
- Peripheral CCA is usually mass-forming tumor
  - Most common type of CCA
  - Well circumscribed, large, with lobulated margins
  - Multicentricity, especially around main tumor
- Periductal-infiltrating CCA
  - Grows along bile ducts and is elongated, spiculated, or branch-like
- Progressive, gradual, and concentric filling (centripetal) on delayed phase images
  - Usually not isodense to vessels (unlike hemangioma)
- Substantial **delayed enhancement** (i.e., greater than that of liver parenchyma) is common (74%)
  - Attributed to fibrous stroma in CCA
- ± **capsular retraction** (frequent), with parenchymal atrophy of liver segments peripheral to tumor
- Bile ducts will be dilated upstream from tumor
  - Duct lining may be thickened and enhanced
  - Rare with other types of hepatic tumors

General Features

- **Best diagnostic clue**
  - Infiltrative hepatic mass with capsular retraction and delayed persistent enhancement (CECT and MR)
- **Location**
  - Originates from interlobular bile ducts (i.e., bile ducts distal to 2nd order branches)
- **Size**
  - Usually large (> 5 cm) at diagnosis
- **Morphology**
  - Peripheral CCA is usually a mass-forming tumor
    - » Often has "satellite" nodules
- Key concepts
  - Mass-forming CCA: Well circumscribed, large, with lobulated margins
    - » Multicentricity, especially around main tumor
  - Periductal-infiltrating CCA: Grows along bile ducts and is elongated, spiculated, or branch-like
  - Intraductal-growing CCA: Small, sessile, or polypoid
    - » Often spreading superficially along mucosal surface and resulting in multiple tumors (papillomatosis) along various segments of bile ducts

Fibrolamellar hepatocellular carcinoma

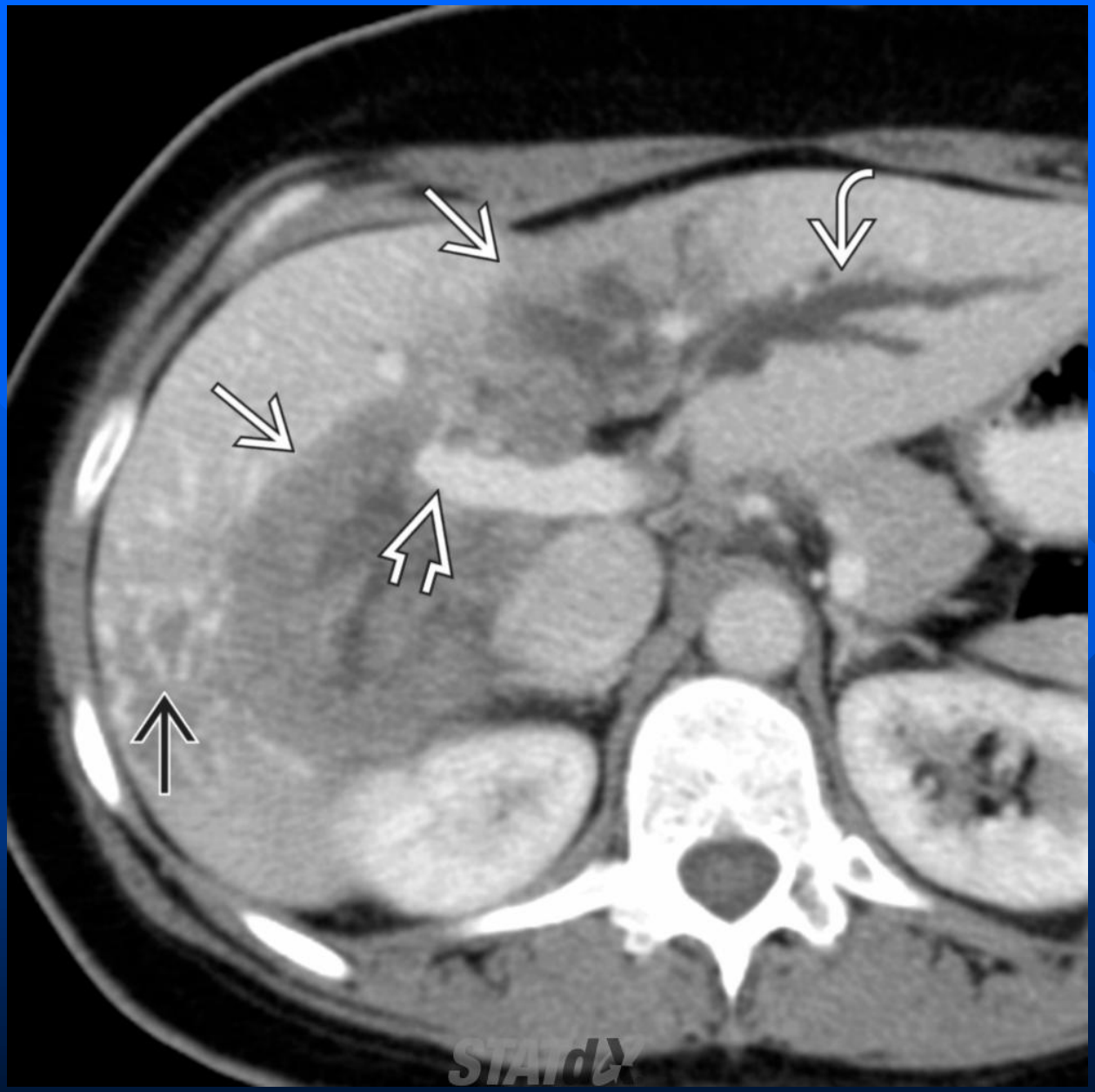
- No specific risk factors
- Usually no underlying cirrhosis or liver disease
  - Hepatitis and cirrhosis may be present (< 5%)
- Healthy young adult with large liver mass.
- May have gynecomastia or other estrogenic effects.
- **Age**
  - Adolescents/young adults
    - » Age range of 5-69 years (mean: 23 years)
  - Most patients present in 2nd-3rd decade of life

CT

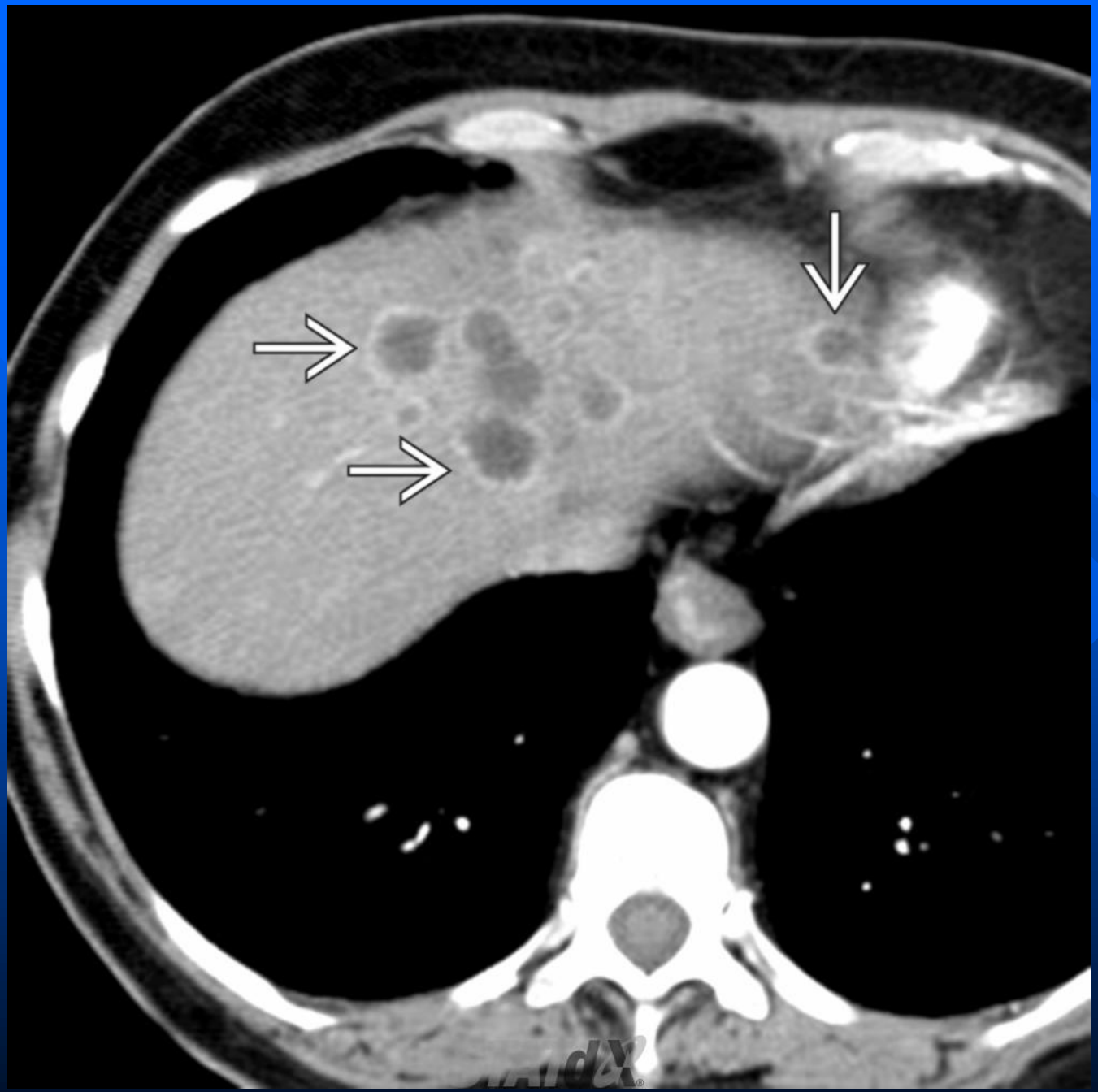
- **NECT**
  - Well-defined, predominantly homogeneous, hypodense mass
    - » Lobular margins
    - » Hypodense satellite nodules (65% of CCAs)
    - » Punctate, stippled, chunky calcifications (18% of CCAs)
- **CECT**
  - Mass-forming PCC
    - » Thin or thick rim-like enhancement frequently seen around periphery of tumor on arterial phase images
    - » Progressive, gradual, and concentric filling (centripetal) on delayed phase images
      - Usually not isodense to vessels (unlike hemangioma)
    - » Substantial **delayed enhancement** (i.e., > that of liver parenchyma) is common (74%)
      - Attributed to fibrous stroma in CCA
    - » Homogeneous or heterogeneous hyperattenuating enhancement
      - Entire mass may be enhanced only on delayed-phase images
      - May be only evidence of tumor
    - » ± **capsular retraction** (frequent), with parenchymal atrophy of liver segments peripheral to tumor
  - Bile ducts will be dilated upstream from tumor
    - » May not be evident in very peripheral CCA
    - » Duct lining may be thickened and enhanced
      - Rare for hepatic metastases or other hepatic primary tumors

MRI

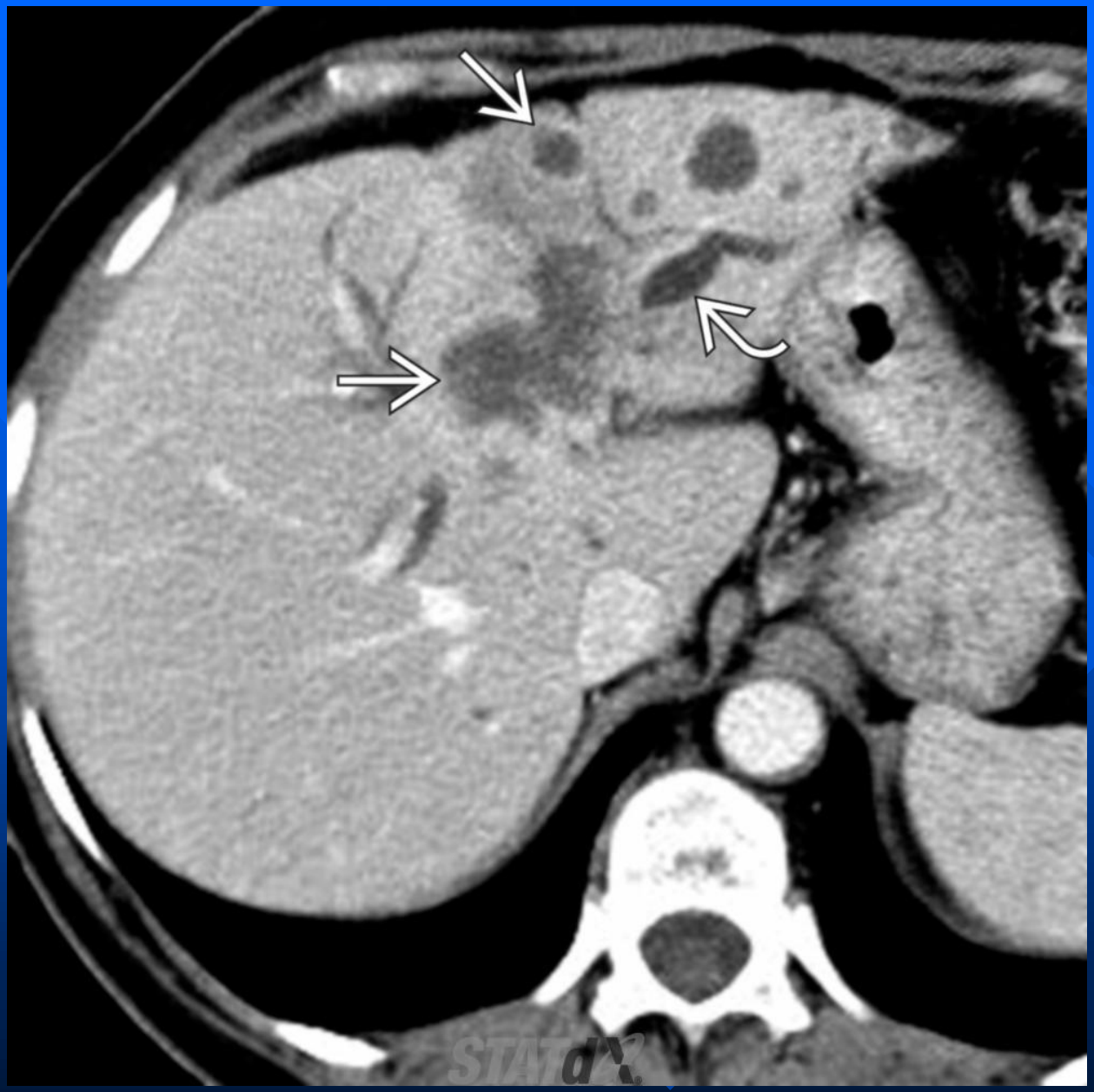
- **T1WI**
  - Heterogeneous hypointense mass
- **T2WI**
  - Hyperintense periphery (cellular tumor) + large central hypointensity (fibrosis)
  - Hyperintense foci in center may represent necrosis, mucin
- **T1WI C+**
  - Central hypointense areas exhibit homogeneous, heterogeneous, or no enhancement
    - » Regions of fibrosis display enhancement (delayed) while those of coagulative necrosis and mucin show no enhancement
  - Dynamic MR: Minimal or moderate rim enhancement and progressive and concentric filling with contrast material
    - » Intratumoral fibrous stroma displays marked or prolonged enhancement on delayed phase scans
    - » Some cases of PCC exhibiting little fibrosis may show early enhancement on dynamic studies
- **MRA**
  - Displacement or encasement of adjacent vessels



Axial CECT of a 46-year-old woman with jaundice shows the portal vein (white open arrow) and bile ducts (white curved arrow) encased and obstructed by the tumor (white solid arrow), accounting for the altered perfusion of the right hepatic lobe. Hepatic veins were encased as well, resulting in collateral blood vessels seen within the right lobe (black solid arrow).



Axial CECT in arterial phase of a 55-year-old woman with jaundice shows multifocal masses with continuous peripheral ring enhancement (white solid arrow).



The portal venous phase in the same patient shows little enhancement of the tumors (white solid arrow). The intrahepatic bile ducts are dilated (white curved arrow), and the left lobe of the liver is atrophic. Parenchymal atrophy of liver segments affected by peripheral cholangiocarcinoma is common and may be evident as lobar atrophy or capsular retraction.

Cholangiocarcinoma with satellite lesions