Primary Sclerosing Cholangitis

- Immune-mediated disease causing progressive inflammation, fibrosis, and stricturing of intrahepatic and extrahepatic ducts.
- High association with other autoimmune disorders and inflammatory bowel disease (especially ulcerative colitis)
- Increased risk of cholangiocarcinoma and other malignancies (including gallbladder cancer)
- Most commonly seen in young (30-40 years) males, with ↑ incidence in Europe and North America
- Usually worsens progressively with little proven benefit to various medical, endoscopic, or surgical interventions
- Liver transplantation is curative, although primary sclerosing cholangitis (PSC) may recur in liver allograft

General Features

- **Best diagnostic clue**
  - Multifocal biliary strictures, segmental ductal dilation, bile duct wall thickening, and irregular beading of intra- and extrahepatic bile ducts
- **Location**
  - Common bile duct (CBD) involved in > 90% of patients
  - Involvement of both intra- and extrahepatic ducts in 87%
    - » Isolated involvement of intrahepatic (11%) or extrahepatic (2%) ducts is unusual
  - Most severely affected segments of biliary tree are usually main right and left bile ducts
  - Strictures can affect cystic duct and pancreatic duct
- **Morphology**
  - In patients with PSC-induced end-stage cirrhosis, liver is markedly deformed (to much greater extent than with other common causes of cirrhosis)
    - » Contour is grossly lobulated and rounded with peripheral atrophy and central hypertrophy
    - » Enlargement of central liver and caudate with peripheral atrophy described as "pseudotumoral" enlargement of caudate
    - » Atrophy/hypertrophy complex may even occur in absence of cirrhosis

Imaging

- Multifocal "beaded" strictures of intra- and extrahepatic ducts with intervening sites of dilated and normal ducts
  - Pruned appearance of biliary tree develops over time
- CT/MR
  - Thickening and hyperenhancement of bile duct wall suggests active inflammation
  - Visualization of greater than expected number of peripheral ducts on MRCP is clue to presence of peripheral intrahepatic ductal strictures
  - Chronic involvement results in atrophy of peripheral liver, massive hypertrophy of central liver/caudate (pseudotumor of caudate), and lobulated, rounded liver contour
    - » Frequent periductal and perivascular fibrosis, as well as confluent fibrosis in central liver: Low density on CT and T2 hyperintense on MR
    - » Periphery of liver may show patchy T2 hyperintensity on MR due to edema/inflammation
  - Hepatolithiasis, cholelithiasis, and choledocholithiasis are common and appear as signal voids on MR
  - Extensive lymphadenopathy common

MRI

- MRCP is best noninvasive imaging technique for evaluating changes in biliary tree
- MRCP demonstrates multifocal "beaded" strictures of intrahepatic and extrahepatic ducts
  - Alternating sites of irregular strictures, mildly dilated ducts, and normal-caliber ducts
  - Visualization of greater than expected number of peripheral ducts on MRCP is clue to presence of peripheral intrahepatic ductal strictures
  - Pruned appearance of biliary tree develops as disease progresses, with obliteration of small peripheral ducts
  - Hepatolithiasis, cholelithiasis, and choledocholithiasis are common (usually pigmented stones) and appear as signal voids on all pulse sequences
- T1W C+ images (particularly on arterial phase images) demonstrates heterogeneous, patchy hyperenhancement often in periductal distribution
- Thickening and hyperenhancement of bile ducts on T1WI C+ suggests acute inflammation
  - Irregular wall thickening should raise suspicion for malignancy
- Chronic involvement results in atrophy of peripheral liver, hypertrophy of central liver, and lobulated, rounded liver contour
  - Liver periphery often shows patchy T2 hyperintensity due to parenchymal edema/inflammation
  - Periportal and periductal T2 hyperintensity also common
  - Confluent fibrosis (wedge-shaped site of parenchymal volume loss with capsular retraction) may be seen in chronic setting: Low T1WI signal, high T2WI signal, delayed enhancement
- Extensive lymphadenopathy common, particularly in precaval and aortocaval lymph nodes

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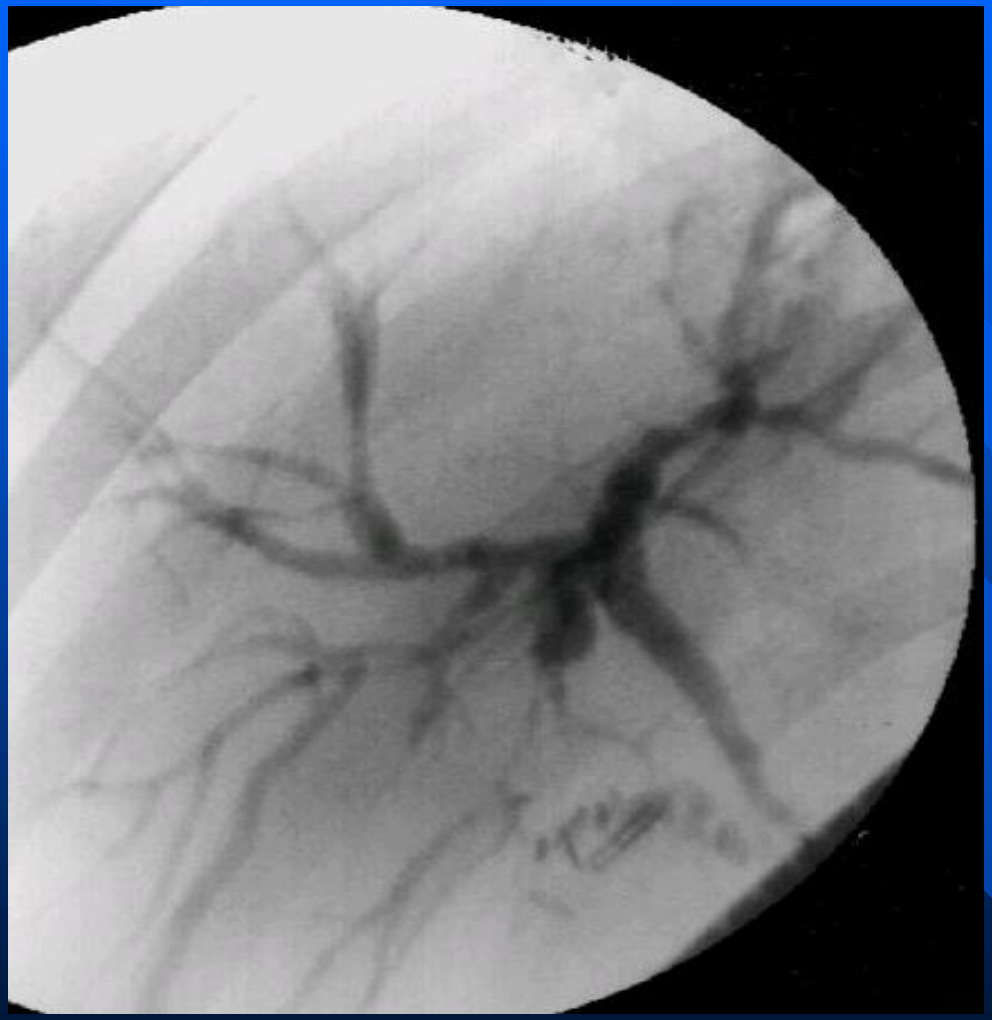
- Secondary causes include
  - stone disease, recurrent infection and previous biliary surgery
- Differential diagnosis
  - Recurrent infectious cholangitis,
  - Diffuse sclerosing cholangiocarcinoma,
  - Early Caroli disease
  - Aids
  - Chemo

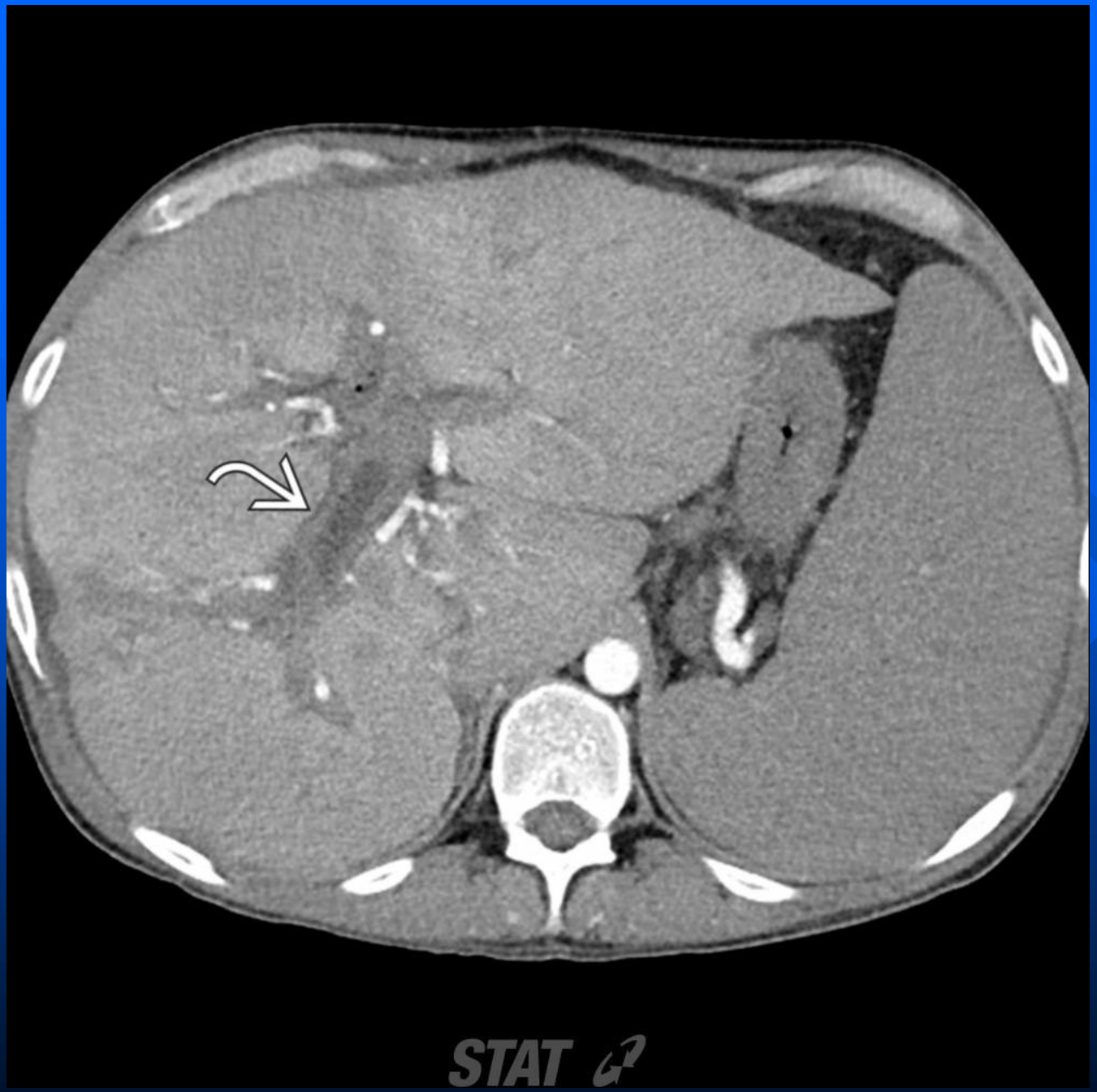
Primary Sclerosing Cholangitis



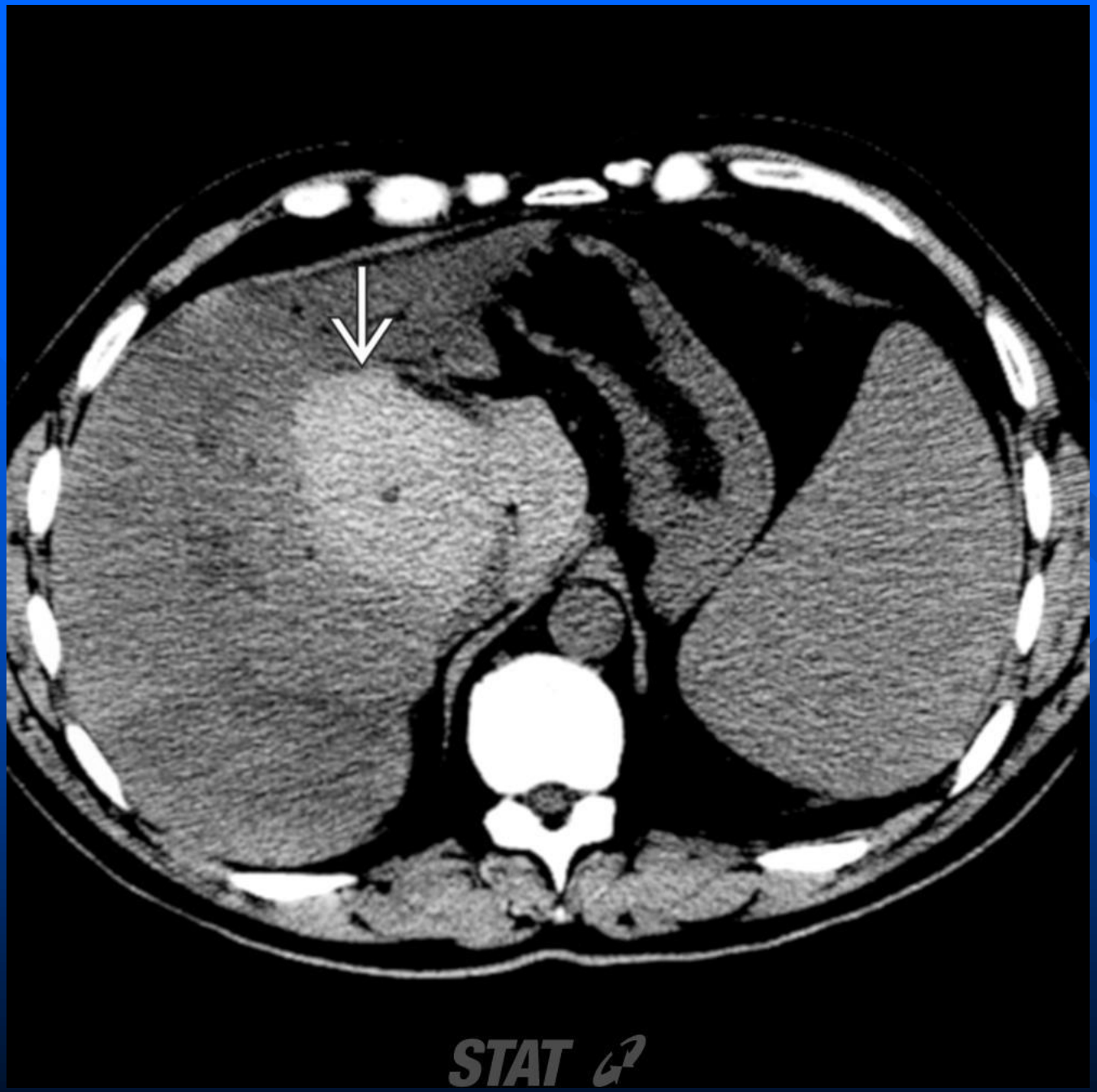
70% have underlying UC
10-15% chance of cholangiocarcinoma

Chemo – “Flurourical” can occasionally mimic PSC

AIDS cholongitis can mimic However can get papillary stenosis with AIDS, rare with PSC



Axial CECT in a patient with PSC demonstrates a cirrhotic, lobulated liver and ductal dilatation. The right hepatic duct (white curved arrow) is dilated, thickened, and hyperenhancing, suggesting active ductal inflammation.



Axial NECT shows massive hypertrophy and increased density of the caudate (white solid arrow) in a patient with longstanding PSC compared with the atrophic, hypodense, peripheral portions of the liver. Because this is often misinterpreted as a caudate tumor, this finding is sometimes called "caudate pseudotumor."