Autoimmune (IgG4) Cholangitis

- **IgG4-related sclerosing disease**:
  - Spectrum of disorders characterized by tumor-like lymphoplasmacytic infiltration and varying degrees of fibrosis in different organs
- **IgG4-related sclerosing cholangitis**:
  - Biliary manifestations of IgG4-related sclerosing disease frequently associated with autoimmune pancreatitis
- Frequently associated with imaging findings of autoimmune pancreatitis and other manifestations of IgG4-related sclerosing disease
- Exact pathophysiology not well understood, but some evidence for both autoimmune and allergic mechanisms

Imaging

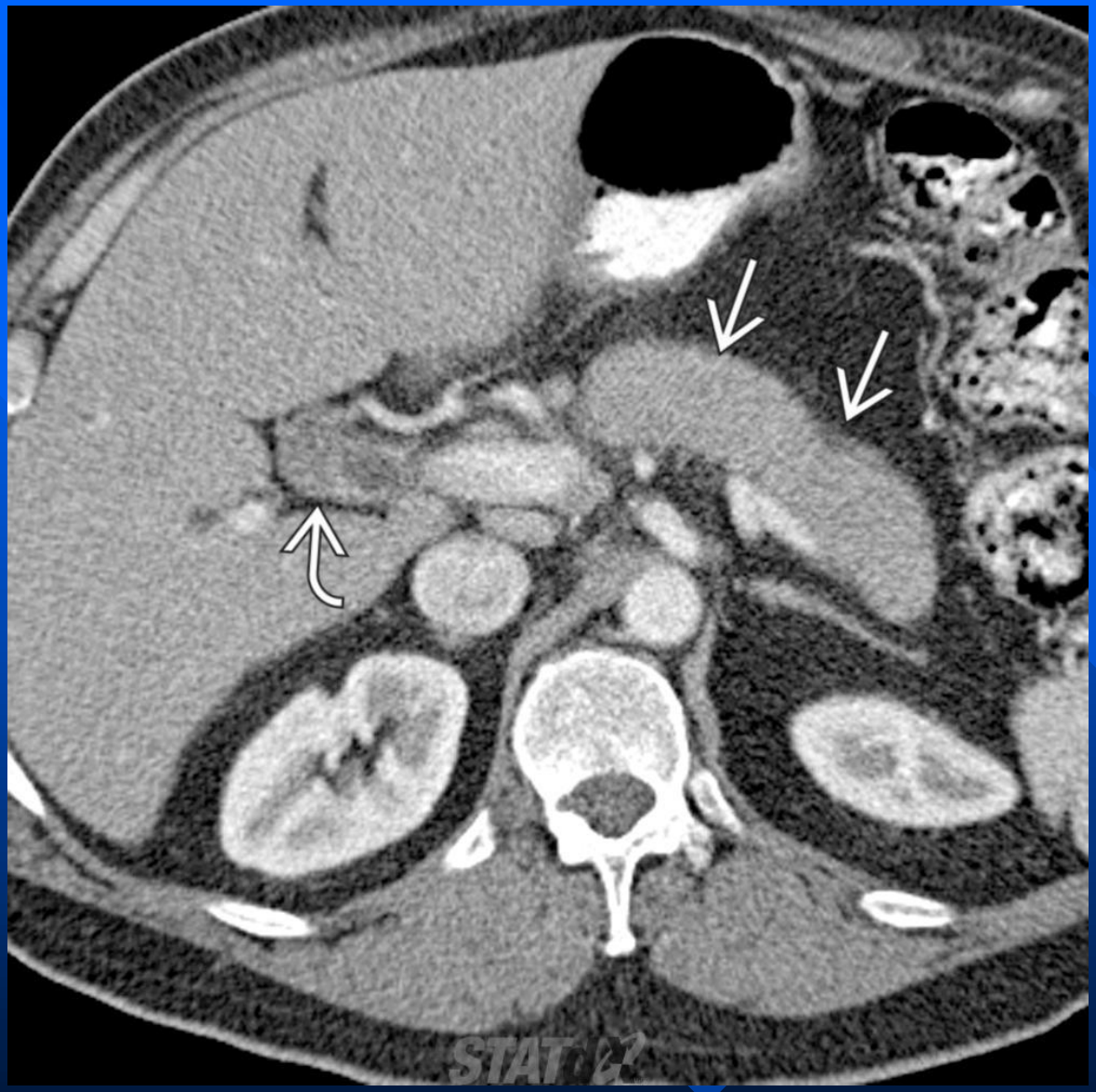
- **ERCP**:
  - Strictures can involve any portion of the biliary tree, but distal common bile duct (CBD) strictures are most common
  - Strictures long and smooth without irregularity and may cause proximal biliary dilatation
- **MR**: MRCP
  - images nicely demonstrate irregularity and strictures of intrahepatic or extrahepatic bile ducts (most commonly affecting distal CBD)
  - Affected segments demonstrate concentric ductal wall thickening with hyperenhancement on T1WI C+
  - Stricture long and smooth (without irregularity)
  - Findings of concomitant autoimmune pancreatitis
    - » Enlarged pancreas with abnormal signal (hypointense on T1WI and hyperintense on T2WI)
    - » Rim of peripheral hypoenhancement and low T2WI signal with delayed enhancement
    - » Multiple discontiguous pancreatic duct strictures on MRCP, which may resolve after administration of secretin
- **CT**: Circumferential focal or diffuse bile duct wall thickening with hyperenhancement of affected segments

Cont

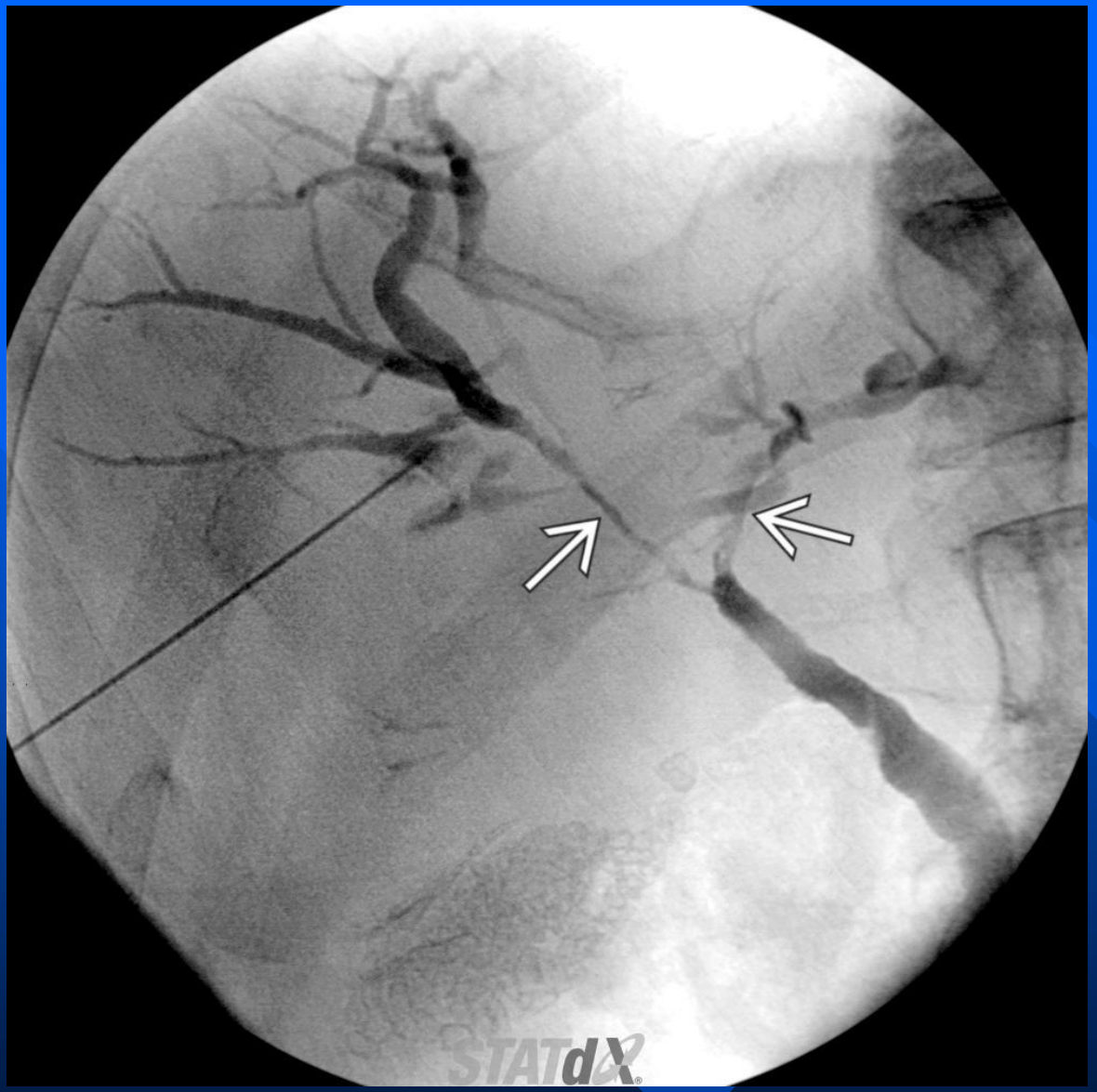
- Pathology
  - Frequently associated with imaging findings of autoimmune pancreatitis and other manifestations of IgG4-related sclerosing disease
  - Exact pathophysiology not well understood, but some evidence for both autoimmune and allergic mechanism
- Clinical Issues
  - Typically affects middle-aged and elderly males
  - Diagnosis based on a combination of imaging, histopathology, serologic (↑ IgG4) markers and clinical response to steroids/azathioprine
  - May improve spontaneously, progress, or relapse, although relapse rate after therapy may be high (> 50%)
  - Treatment with steroids typically effective, although other immunomodulators utilized in refractory cases

Image Interpretation Pearls

- Imaging features of ISC and PSC may overlap, although demographics and course of disease are different for eachPSC seen in younger patients with more indolent course
- ISC has more acute presentation, is seen in older patients, responds to steroids, and may be associated with extrabiliary manifestations of IRSD
- Some authors suggest that ISC and PSC are not completely separate conditions and indeed may be variations of same disease spectrum



CECT of a jaundiced patient shows a thickened, slightly dilated common bile duct (CBD) (white curved arrow) and an enlarged, sausage-shaped pancreas (white solid arrow). The appearance of the pancreas is consistent with autoimmune pancreatitis (AIP), and an elevated IgG4 indicates that the bile duct thickening is due to IgG4-related sclerosing cholangitis (ISC).



Cholangiogram of the same patient shows strictures (white solid arrow) of the proximal right and left hepatic ducts. The patient's serum IgG4 was not elevated, but the strictures improved with empiric steroid therapy. An elevated IgG4 is a highly sensitive and specific marker for ISC, though IgG4 levels may vary widely during the disease course.