

Primary Biliary Cirrhosis

- Name of this disease was changed from **primary biliary cirrhosis** to **primary biliary cholangitis** in 2014.
- Chronic progressive cholestatic liver disease
- Characterized by nonsuppurative destruction of interlobular bile ducts Leads to advanced fibrosis, cirrhosis, and liver failure.
- May coexist with other autoimmune disease of liver Including primary sclerosing cholangitis and autoimmune hepatitis
- ~ 95% of patients are female
 - Typical onset of symptoms in 40- to 50-year-old women
- Fatigue and pruritus are early symptoms
- Takes ~ 7-10 years to progress to liver failure
- **Serum antimitochondrial antibody** tests highly sensitive and specific (positive in 95%)

Imaging

- In less advanced disease, liver is enlarged and smooth
- Regenerative nodules are often evident
 - 3-10 mm, hyperdense on NECT; hypointense on T2W MR
- Lymphadenopathy is prominent feature
- Lace-like fibrosis is a prominent feature
 - Hypointense on T1W, hyperintense on T2W MR
- Signs of portal hypertension may precede biopsy evidence of cirrhosis
- Hepatocellular carcinoma: Less frequent in primary biliary cirrhosis than with other causes of cirrhosis

Demographics

■ Age

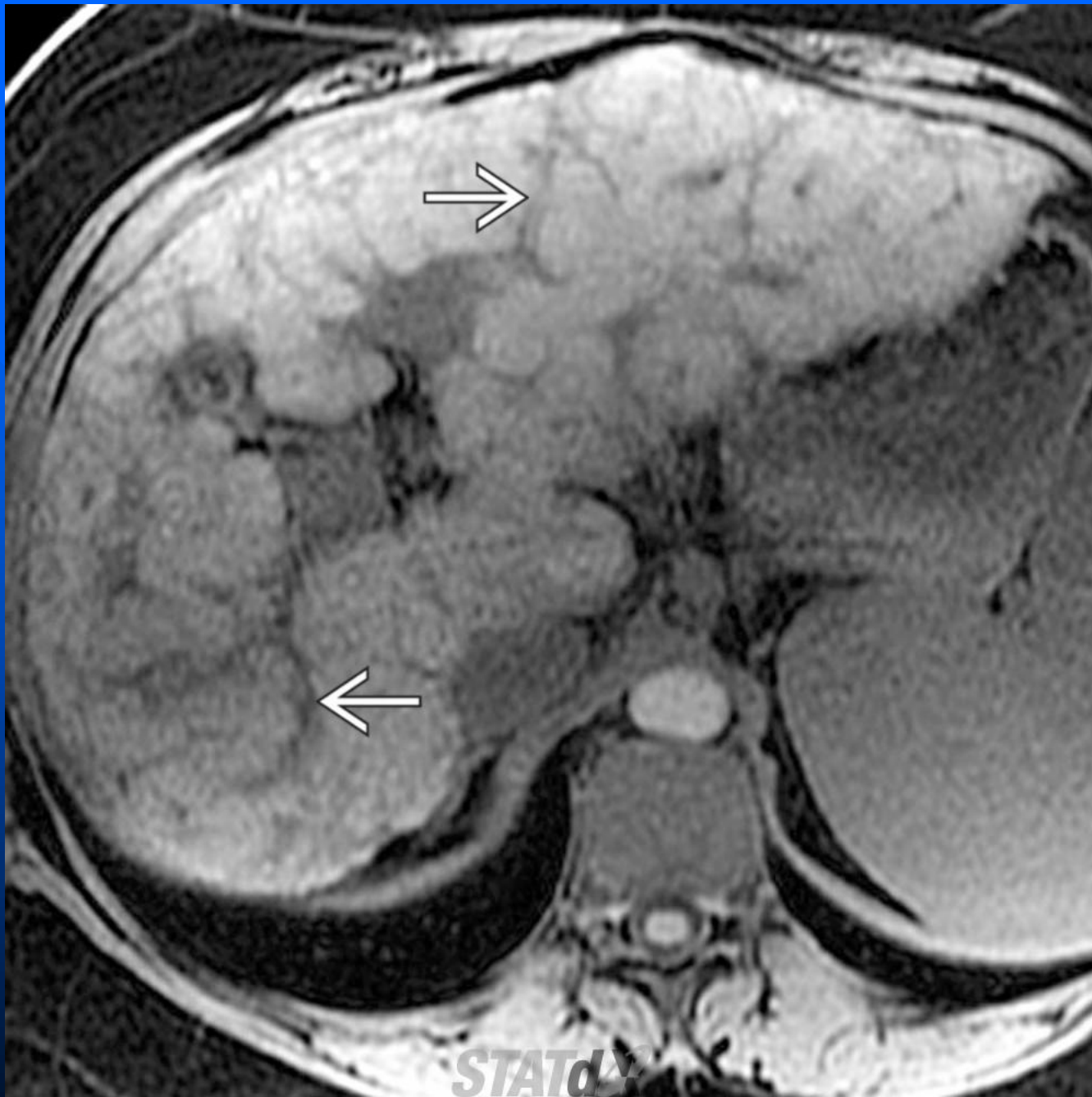
- Onset between ages 30-65 years
 - » Typical onset of symptoms in 40- to 50-year-old women
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■ Gender

- ~ 95% of patients are female

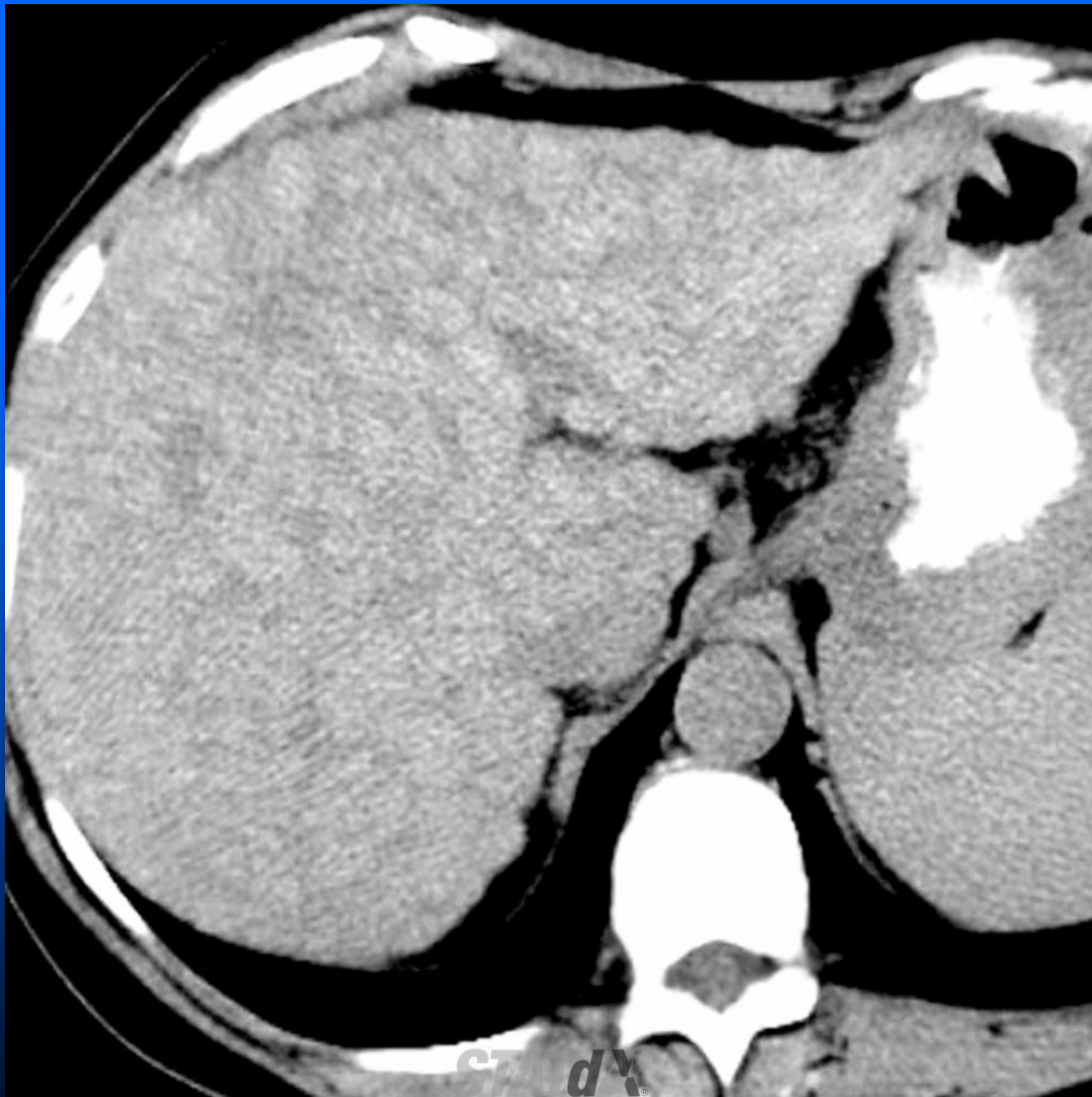
■ Epidemiology

- Accounts for 0.6-2.0% of deaths from cirrhosis
- 3rd most common indication for liver transplantation in adults
- Prevalence: 19-151 cases per million people
- Incidence: 3.9-15 cases per million people each year



In the same patient, note the widened fissures and disproportionate volume loss of the anterior and medial segments (signs of cirrhosis). The fibrotic bands (white solid arrow) are hypointense on this unenhanced T1WI.





NECT of a 53-year-old woman with PBC first diagnosed 5 years prior shows a heterogeneous liver with innumerable small hyperdense regenerating nodules separated by a lace-like network of lower density tissue, typical of the pattern of fibrosis often seen in patients with PBC.



ERCP shows pruned intrahepatic bile ducts with decreased arborization, the classic "tree in winter" appearance.