Primary Sclerosing Cholangitis Labs and Treatment

Primary sclerosing cholangitis (PSC) is a disease of the intra- and extra-hepatic bile ducts that occurs due to an unknown cause. Progressive inflammation of the biliary tree leads to cholestasis. Labs seen in PSC include elevations of conjugated bilirubin, alkaline phosphatase, and IgM. It is associated with ulcerative colitis, cholangiocarcinoma and secondary biliary cirrhosis. The only definitive treatment is liver transplant, though stenting of the biliary tree can provide symptomatic relief.



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Labs

Increased Conjugated Bilirubin
Up-arrow Enzyme-holding Belly-ribbon dancer
Cholestasis due to inflammation and obliterative fibrosis leads to increased conjugated bilirubin. Cholestasis typically manifests as pruritus and jaundice.

Increased ALK-P
Up-arrow ELK-P
Increased alkaline phosphatase is another marker of cholestasis commonly seen in PSC. Gamma-glutamyl transpeptidase (GGT) is also similarly elevated.

Increased IgM
Up-arrow Mountain Globulin-goblin
Hypergammaglobulinemia especially involving IgM is seen with PSC. This is thought to reflect the abnormalities in humoral immunity that occur in patients with PSC.

Associated Pathologies

Ulcerative Colitis
Ulcer-volcano with Colon
Ulcerative colitis is closely associated with PSC. Up to 90% of individuals with PSC have ulcerative colitis. Ulcerative colitis typically presents with bloody diarrhea and abdominal pain.

Cholangiocarcinoma
Cola-angel-car-gnome
PSC patients have up to a 15% chance of developing cholangiocarcinoma during their lifetime. It typically presents with worsening jaundice, abdominal pain and weight loss.

Secondary Biliary Cirrhosis
(2) Tutu Bile-duck at C-roses liver
As PSC progresses it can result in scarring and nodule formation in the liver, leading to cirrhosis. When cirrhosis develops secondary to a cholestatic process, it is termed secondary biliary cirrhosis.

Treatment

Liver Transplant
Liver Train-plant
Liver transplant is the preferred treatment for advanced PSC. It is typically considered once a patient has developed cirrhosis or has other persistent and severe symptoms.

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Stent

Stent-tube

The inflammation of PSC can cause strictures within the bile ducts leading to severe symptoms. ERCP (endoscopic retrograde cholangiopancreatography) can be done to place a stent within an obstructed bile duct, opening it and providing relief. Suggested change: The inflammation of PSC can cause strictures within the bile ducts leading to severe symptoms. Initial management includes balloon dilatation via ERCP (endoscopic retrograde cholangiopancreatography) or PTC (percutaneous transhepatic cholangiography). For severe strictures, placing a short-term stent via ERCP is an option, though it is associated with adverse effects such as post-ERCP pancreatitis and cholangitis/cholecystitis. Surgery is recommended if ERCP/PTC management fails.

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