Primary Biliary Cholangitis (Primary Biliary Cirrhosis) Diagnosis and Treatment

Primary Biliary Cirrhosis (PBC) is an inflammatory autoimmune reaction leading to destruction of intrahepatic bile ducts. As bile ducts are destroyed, cholestasis results, and over time this may lead to cirrhosis and liver failure. Patients typically have antimitochondrial antibodies present, along with elevated serum alkaline phosphatase and increased cholesterol. Treatment includes ursodiol and liver transplant.



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Diagnosis

Positive Antimitochondrial Antibodies
Positive Anti-Mitochondrial-factory Ant-tie-body
95% of patients with PBC show increased serum antimitochondrial antibodies, which are the characteristic autoantibodies in this disease. These stimulate autoreactive T cells to accumulate around bile ducts and hepatocytes.

Increased Alkaline Phosphatase (ALK-P)
Up-arrow Elk-P
Studies will also show increased serum alkaline phosphatase (ALP or ALK-P), which is a marker of cholestasis.

Increased Cholesterol
Up-arrow Cholesterol-burger
In PBC, and other situations where cholestasis occurs, the ability to transport and breakdown cholesterol is impaired. This yields increased serum cholesterol.

Treatment

Ursodiol
Usher-doll
This is the only FDA-approved medication to treat PBC, and works by reducing the rate of intestinal cholesterol absorption. It also works to nonsurgically treat gallstones, while limiting symptoms of cholestasis (jaundice, pruritis).

Liver Transplant
Liver Train-plant
The onset of this disease is insidious. It eventually leads to liver cirrhosis and an increased risk of hepatocellular carcinoma. Liver transplant is the best form of treatment in patients with end-stage liver disease.

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