

PRIMARY BILIARY CHOLANGITIS (PBC)

What is PBC?

PBC is a chronic (long-term) liver disease resulting from progressive destruction of the intrahepatic bile ducts (bile ducts in the liver). The liver produces bile, which aids in the digestion of fat and fat-soluble vitamins A, D, E, and K. When these ducts are destroyed, the bile builds up in the liver, causing inflammation (swelling) and scarring (fibrosis). This can lead to cirrhosis (advanced scarring), loss of liver function, and associated complications.

Women are more likely than men to develop PBC. Of people diagnosed with PBC, 90% are women. It often develops during middle age, and it is usually diagnosed between the ages of 35 and 60 years old.

What Causes PBC?

The exact cause of PBC is unknown. It is not caused by alcohol or drug use. It is most likely an autoimmune disease, meaning that the body's immune system mistakenly attacks and destroys its own cells – in this case, the cells of the intrahepatic bile ducts. Some people diagnosed with PBC may also have one or more other autoimmune diseases. Also, there appears to be a genetic component to developing PBC, because it is more common among siblings and families where one member has been affected.

Symptoms of PBC

As with most liver diseases, there may not be any symptoms of PBC in its initial stages. The most common early symptoms are fatigue and itching of the skin (pruritus). Other symptoms include abdominal pain and darkening of the skin. Small yellow or white bumps under the skin or around the eyes may appear. Other symptoms include dry mouth and eyes, and bone, muscle, and joint pain. Osteoporosis (thinning of bones) can develop, often in later stages of PBC.

Diagnosing PBC

An important test to confirm PBC is an AMA (antimitochondrial antibodies) test, which measures autoimmune response. Other tests that may be ordered include smooth muscle antibodies (SMA), antinuclear antibodies (ANA), and various enzyme and protein tests.

Treating and Managing PBC

There is no cure for PBC; however, medications can help slow disease progression and ease symptoms. Ursodiol and Ocaliva are two medications that have been found to be useful in treating people with PBC to slow the progression of the disease. People should speak with their liver specialists to determine which is the best treatment for them. They should also discuss medications available to manage symptoms, PBC-friendly diets, and a schedule of monitoring their liver health with necessary tests.

To learn more about PBC, scan the QR code below. For a complete list of all of our free resources, please visit: liverfoundation.org/resource-center.

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For questions about liver wellness and disease, or for emotional support to patients at the point of crisis and information on local resources including physician referrals, please contact our Helpline at 1-800-465-4837.

