

Primary Biliary Cholangitis



Your Liver. Your Life.

What is Primary Biliary Cholangitis?

Primary biliary cholangitis (PBC), formerly known as primary biliary cirrhosis, is a chronic liver disease resulting from progressive destruction of the bile ducts in the liver – called the intrahepatic bile ducts. Bile produced in your liver travels via these ducts to your small intestine where it aids in the digestion of fat and fat-soluble vitamins (A, D, E and K). When the ducts are destroyed, bile builds up in the liver contributing to inflammation and scarring (fibrosis). Eventually this can lead to cirrhosis and its associated complications, as scar tissue replaces healthy liver tissue and liver function becomes increasingly impaired.



What Causes PBC to Develop?

The exact cause of PBC is unknown. It is not caused by alcohol or illegal-drug use. It's most likely an autoimmune disease that occurs in genetically susceptible individuals. 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.

Who is at Risk for PBC?

- Women are nine times more likely than men to develop PBC, meaning that women make up about 90% of PBC cases.
- The disease most often develops during middle age and is usually diagnosed in people between the ages of 35 to 60 years.
- There appears to be a genetic component to developing PBC, as it's more common among siblings and in families where one member has been affected.

What are the Signs and Symptoms of PBC?

PBC may progress slowly and many people do not have symptoms, particularly in the early stages of the disease. The most common initial symptoms are fatigue and itching of the skin (pruritus). Other symptoms may include:

- Abdominal pain
- Darkening of the skin

- Small yellow or white bumps under the skin (xanthomas) or around the eyes (xanthelasmas)

Individuals may also complain of having dry mouth and eyes, and bone, muscle and joint pain.

As the disease progresses, symptoms of cirrhosis can develop including:

- Yellowing of the skin (jaundice)
- Swelling of the legs and feet (edema)
- Enlarged abdomen from fluid accumulation (ascites)
- Internal bleeding in the upper stomach and esophagus from enlarged veins (varices)

Thinning of the bones (osteoporosis) leading to fractures is another complication of PBC. While this is more common in late stages of the disease, it can occur earlier as well. In addition, people with cirrhosis are at increased risk for liver cancer (hepatocellular carcinoma).

How is PBC Diagnosed?

Because many people with PBC have no symptoms, the disease is often discovered incidentally due to abnormal results on routine liver blood tests. Once PBC is suspected, a blood test to check for antimicrobial antibody (AMA) is done. This test is positive in nearly all people with PBC. A liver biopsy, where a small sample of liver tissue is removed with a small needle can help confirm the diagnosis. Imaging studies may be used to rule out other diseases, or to further evaluate patients once they have been diagnosed with PBC.

How is PBC Treated?

Treating the Disease

There is no cure for PBC, however, there are medications that can help slow disease progression and manage symptoms.

Ursodiol (brand names Actigall, URSO 250, URSO Forte) is a naturally occurring bile acid (ursodeoxycholic acid or UDCA) that helps move bile out of the liver and into the small intestine. If used early enough, Ursodiol can improve liver function and may keep you from needing, or delay the need for a liver transplant. People with PBC must take this medication every day for life.

UDCA is effective in more than 50 percent of patients, but up to 40 percent of patients do not achieve an adequate reduction in alkaline phosphatase (ALP) or bilirubin with UDCA, while 5-10 percent are unable to tolerate UDCA.

In May 2016, obeticholic acid (brand name Ocaliva) was approved for the treatment of PBC in combination with UDCA in adults with an inadequate response to UDCA, or as a single therapy in adults unable to tolerate UDCA. Obeticholic acid increases bile flow from the liver and suppresses bile acid production in the liver, thus reducing the exposure of the liver to toxic levels of bile acids. Side effects of obeticholic acid may include increased itching and elevations in blood lipids.

Other alternative therapies in patients who are incomplete responders to UDCA include fenofibrate. Medications to

suppress the immune system may also be prescribed including prednisone or azathioprine in PBC patients with the “overlap syndrome” with autoimmune hepatitis. Liver transplantation is considered when medical treatment no longer sufficiently controls the disease. When a person has end-stage liver disease, a liver transplant is necessary for survival.

Treating the Symptoms

- Intense itching is one of the most common symptoms of PBC. Over-the-counter antihistamines like diphenhydramine (Benadryl) may be helpful. Other agents such as rifampicin, naltrexone, cholestyramine and sertraline may be prescribed.
- Dry eyes can be relieved by using eye drops (artificial tears).
- A dry mouth may be helped by sucking on hard candy or chewing gum, both of which increases saliva. There are also saliva substitutes and some medications that can be used.

Preventing Complications

- Blood tests to monitor for deficiencies in fat-soluble vitamins are often done. As PBC progresses, some people need to replace the fat-soluble vitamins lost in fatty stools, so you may be put on vitamin A, D, E and K replacement therapy.
- Since people with PBC are at a higher risk for osteoporosis, calcium and vitamin D are usually prescribed. A baseline bone density is currently recommended at the time of initial diagnosis.

- As the ability of the liver to function decreases over time, complications associated with cirrhosis will need to be addressed and treated. Screening for varices and liver cancer is often recommended.

What Lifestyle Changes are Recommended for People with PBC?

Maintaining a healthy lifestyle can help people with PBC feel better, as well as relieve or prevent some symptoms associated with the disease. Upon diagnosis, your doctor may suggest the following:

- Start a reduced sodium diet if you have fluid overload (edema or ascites)
- Reduce fat intake if you are overweight or have fatty liver
- Drink plenty of water
- Avoid or lower intake of alcohol
- Avoid undue stress when possible
- Start exercising, particularly walking
- Stop smoking
- Maintain good skin care
- Get regular dental examinations

Keep in mind that PBC usually advances slowly over a period of years. Many people lead normal lives for years without symptoms, depending on how early the diagnosis is made. And while there is no cure, people are having good results slowing disease progression and living longer without complications by adhering to their medication regimen and maintaining a healthy lifestyle.



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