

Primary Biliary Cirrhosis (PBC)

Explore this section to learn more about primary biliary cirrhosis, including a description of the disease and how it's diagnosed.

What is primary biliary cirrhosis?

Primary biliary cirrhosis, or PBC, is a chronic, or long-term, disease of the liver that slowly destroys the medium-sized bile ducts within the liver. Bile is a digestive liquid that is made in the liver. It travels through the bile ducts to the small intestine, where it helps digest fats and fatty vitamins.

In patients with PBC, the bile ducts are destroyed by inflammation. This causes bile to remain in the liver, where gradual injury damages liver cells and causes **cirrhosis**, or scarring of the liver. As cirrhosis progresses and the amount of scar tissue in the liver increases, the liver loses its ability to function. Cirrhosis also prevents blood from the intestines from returning to the heart.

PBC advances slowly. Many patients lead active and productive lives for more than 10 to 15 years after diagnosis. Patients who show no symptoms at the time of diagnosis often remain symptom-free for years. Patients who have normal liver tests on treatment may have a normal life expectancy. PBC is a chronic illness and may lead to life-threatening complications, especially after cirrhosis develops.

What are the signs and symptoms of PBC?

Many people with PBC do not have symptoms, especially in the early stages of the disease. When symptoms do occur the most common is pruritus, or intense itching of the skin, often in the arms, legs and back. Other symptoms may include fatigue, jaundice, fluid build-up in the ankles and abdomen, and darkening of the skin and collection of fatty deposits in the skin around the eye. Jaundice, is an indication of how far the disease has progressed.

Several other disorders often occur with PBC. The most common is problems with the tear and salivary glands, causing dry eyes or mouth. Arthritis and thyroid problems may also be present. Renal stones and gallstones may develop. Bone softening and fragility leading to fractures can occur in late stages of the disease.

What causes PBC?

The cause of this disease is unknown. It may be related to problems in the immune system. Although PBC is technically not a hereditary disease, meaning a disease caused by a specific gene or genetic defect that is passed from parent to child, there appears to be some family link. PBC is more common among siblings and in families where one member has been affected.

How is PBC diagnosed?

Because many PBC patients have no symptoms, the disease is often discovered through abnormal results on routine liver blood tests. Doctors need to do several tests to confirm a diagnosis of primary biliary cirrhosis. One test looks for the presence of antimitochondrial antibodies (AMA) in the blood. This test is positive in nearly all PBC patients. Ultrasound exams and a liver biopsy, in which a small sample of liver tissue is removed with a needle, may also be performed.

How is PBC treated?

Patients most often take a daily dose of a drug called ursodiol. This is urseodeoxycholic acid, a naturally occurring bile acid. This drug improves the liver's ability to function in PBC patients. It also extends life expectancy and may delay the need for a liver transplant. The recommended dose is 15 mg per kilogram of body weight, and the medication must be taken every day for life, or until a liver transplant. Patients rarely experience side effects from ursodiol. Other drugs are effective at relieving the symptoms of PBC, including the itching.

If I've been diagnosed with PBC, what questions should I ask my doctor about PBC?

- "How severe is the liver damage?"
- "What treatment do you recommend? Will this slow down the progression of the disease?"
- "Will any medication be prescribed? What are the side effects?"
- "Should I change my diet?"
- "Are there any supplements you would suggest that I take?"
- "What can be done to relieve my symptoms?"
- "If cirrhosis develops, is transplantation my only option?"

What lifestyle changes may be helpful for PBC patients?

Maintaining a healthy lifestyle will help patients feel better and may relieve or prevent some of the secondary symptoms of PBC. Your doctor may recommend:

- A reduced sodium diet
- Drinking ample water
- Taking calcium and Vitamin D supplements
- Avoiding or reducing the consumption of alcohol
- Reducing stress
- Exercising, particularly walking
- Skin Care
- Regular dental examinations
- Artificial tears for dry eyes

Who is at risk for PBC?

Women account for about 90 percent of PBC cases. It is most commonly diagnosed in patients between the age of 35 and 60.

What is being done to find a cure for PBC?

PBC has been known for more than 100 years, but now doctors are able to diagnose it very early. This means that treatment can begin before the liver is severely damaged. Scientists are continuing to study the disease to find the cause and understand its development.

In addition, investigators are exploring the use of several additional medications to lessen the symptoms and control liver damage through drug therapy trials, involving a large number of patients around the world.