Primary Biliary Cirrhosis
What is Primary Biliary Cirrhosis?

Primary biliary cirrhosis (PBC) 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 (pruritis). Other symptoms may include:
• Abdominal pain
• Darkening of the skin
• Small yellow or white bumps under the skin or around the eyes (xanthomas)

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 can occur in late stages of the disease. 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 antimitochondrial antibody (AMA) is done. This test is positive in nearly all people with PBC. Imaging of the abdomen by ultrasound and a liver biopsy, where a sample of liver tissue is removed with a small needle, can help confirm the diagnosis.
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.

Depending on the severity of the disease, medications to suppress the immune system may also be prescribed including methotrexate, cyclosporine and prednisone.

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. A drug called cholestyramine (Questran) helps bind bile and decreases the itching associated with elevated bilirubin levels. Other agents such as rifampicin, naltrexone 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. Screening for osteoporosis with bone density testing may be recommended.

- 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 and/or low-fat diet
- Drink plenty of water
- Avoid or lower intake of alcohol
- Lower caffeine intake
- 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.

In fact, improvements in early detection and better management of PBC have led to a worldwide initiative to formally change the name of the disease to primary biliary cholangitis (inflammation of the bile ducts). This would more accurately reflect the fact that currently, 85% of people do not have cirrhosis at the time of diagnosis, and as treatments improve, many people may never progress to that stage of liver disease.