2015

Primary Biliary Cirrhosis

Capital Health
Primary Biliary Cirrhosis

What is primary biliary cirrhosis?

Primary biliary cirrhosis (PBC) is a chronic autoimmune disease which affects the liver.

If you have PBC, your immune system will attack your liver and your bile ducts. This causes inflammation (swelling) in the bile ducts. Bile ducts are the pathways that move bile out of the liver. Bile is used for digestion and getting rid of toxins from your body.

When the bile ducts and liver are damaged by the immune system, it is harder for harmful substances to be removed. The result of this is a back-up of toxins causing additional liver damage. This cycle of inflammation, damage, build-up, more inflammation, and more damage leads to permanent scarring of the liver tissue. Permanent scarring of the liver is known as cirrhosis.

This disease develops very slowly and people are often diagnosed before they show any symptoms. If not treated, scar tissue will continue to replace healthy liver tissue which will prevent the liver from working well. Scarring makes it harder for the liver to carry out its important jobs, and liver failure can happen.
What are the causes?

PBC is an autoimmune condition in which the body turns against its own cells. However, the exact causes are not clear.

Genetics as well as environmental factors such as smoking, alcohol use, chemical exposure, and infection may increase your chances of getting this illness.

Who is at risk?

The current population of patients with PBC are generally aged 30 to 60. The disease more commonly affects women than men.

The exact cause of PBC is unknown but there is some data suggesting a serious bacterial, fungal, or parasitic infection may also be a potential trigger for the disease.
What are the symptoms?

Many patients have no symptoms for years after being diagnosed. However, as the disease gets worse, symptoms may come on slowly and may include:

- Fatigue (tiredness)
- Itchy skin
- Pain in upper right part of abdomen
- Dry eyes or mouth
- Jaundice (yellowing of the eyes or skin)
- Darkening of skin (not because of sun exposure)
- Swollen feet or ankles
- Build-up of fluid in abdomen (ascites)
- Fatty deposits on skin around eyes, eyelids, hands, soles, elbows, and knees
- Diarrhea
Can there be complications?
This is a disease which has a large impact on the liver. Because the liver is such an important organ, there are more complications that may happen over the long term. These may include:

› Cirrhosis
› Portal hypertension (high blood pressure)
› Enlarged veins
› Enlarged spleen
› Osteoporosis (soft or weak bones)
› Vitamin deficiencies
› Malnutrition
› Memory problems
› Liver cancer
› Gallstones and bile duct stones
› Increased risk for other disease

How is it diagnosed?
Tests for PBC can vary. Almost 60% of people are diagnosed before symptoms even show. Your health care provider may take your medical and family history to see whether you are at risk for this disease.

Additionally, a physical exam may be performed but the more conclusive tests are blood tests, imaging tests, or a liver biopsy.
Blood tests
Health care providers look for either 1 of 2 things to find out whether you have the disease: a positive AMAs test or a higher level of liver enzymes. Tests may come back showing anti-mitochondrial antibodies (AMAs). A positive test for AMAs is a strong sign that the disease is present, as these antibodies are uncommon in people who do not have PBC. The other element your blood test may show is a higher level of liver enzymes. This is often a sign for a general liver disease or injury to the bile ducts.

Imaging tests
Imaging tests include ultrasound, magnetic resonance imaging (MRI), or a computerized tomography (CT) scan. These can be used to inspect the liver and bile ducts to identify the disease and get rid of other possible diagnoses.

Liver biopsy
A biopsy (tiny piece) of your liver may be taken to prove the disease is there, and to find out the stage of the disease. This test is done with a tiny needle in a very small incision (cut). A small piece of the liver is taken and used for testing.
The disease and your liver

It is important to know what PBC can do to your liver. As your largest internal organ, it has many important jobs. These include:

- Taking up, storing, and processing nutrients and delivering them throughout the body when needed
- Building proteins
- Making bile
- Getting rid of waste products

The liver is a complicated organ that works non-stop. Nearly all of the blood returning from the gastrointestinal (GI) tract is filtered through the liver. Everything you eat and drink comes in contact with the liver and it detoxifies (cleans), converts medicines to be used by the body, creates proteins to resist infection, and keeps a healthy balance in your body. Because of the many roles the liver plays, recognizing the potential damage that can happen is important.

How is it treated?

PBC is a disease which currently has no known cure. The aim of treatment is to slow its progression, ease any symptoms, and avoid complications.

When started early on, medication works very well in slowing down the growth of the disease. The most common form of treatment is ursodeoxycholic acid (UDCA), or ursodiol (known as URSO®). UDCA is
often the first line of treatment for the disease and can work very well when started early. For patients with chronic damage, it isn’t always as helpful.

UDCA is a natural bile acid which works by helping bile move through the liver. It increases bile flow as well as prevents the making and build-up of toxins. It is a drug taken orally with a single dose between 10-15 mg per kilogram of body weight each day. Once on medication, it is likely you will be on it for life. UDCA has very minimal side effects as it is a naturally-occurring acid in the body. However, some patients may have weight gain, hair loss, and diarrhea while on treatment.

When damage is severe and there is potential for liver failure, a liver transplant will often be considered. A liver transplant involves the removal of the damaged liver to replace it with a healthy one from a donor. In these cases, PBC often comes back; however, it takes many years to grow.

**Treating symptoms**

Itching is a major symptom of PBC. Both cholestyramine (known as Olestyr™) and rifampin (Rifadin®) can be used to help with itching. Cholestyramine comes as a powder which can be mixed into food or drinks. Rifampin comes as a pill and is also helpful to control itching.

Artificial tears and saliva are sometimes used to help get rid of dry eyes and mouth. You can get these treatments usually without a prescription.
To help in the prevention of osteoporosis, calcium and vitamin D supplements are usually recommended as well as plenty of exercise to increase bone density. In the case of malnutrition or vitamin deficiency, other vitamin supplements will be added.

If deemed necessary, your health care providers will also continue to screen for other complications such as portal hypertension and liver cancer.

**What lifestyle changes should I make?**

One of the most important things to consider is keeping a healthy diet. It is strongly recommended to reduce (cut down) sodium (salt) intake and be careful if eating raw shellfish. It has bacteria which can cause serious infection. Alcohol is a major stress on the liver because of the amount of toxins which need to be removed. Limiting alcohol or removing it altogether is an easy way to remove unnecessary stress on the liver.

Finding time for regular exercise increases bone density and reduces the risk of bone loss through osteoporosis.
Helpful points

• Learn about your condition!

• It helps to be educated about the disease so you fully understand symptoms, effects of treatments, and what you can do to stay healthy.

• Seek help and support if you need it.

• Your doctors and nurses are there to answer your questions and make you feel comfortable.

• Before any appointment:
  › Be aware of any pre-appointment restrictions (for example, if your doctor told you to stop taking a certain medication 1 week before your appointment).
  › Take note of any symptoms you may be experiencing.
  › Write down questions you may have for your health care provider.

• Do not start any new medications or treatments without talking to your health care professional.

If you have any questions, please ask.

We are here to help you.