Primary Biliary Cholangitis

liver.ca

Canadian Liver Foundation
Fondation canadienne du foie

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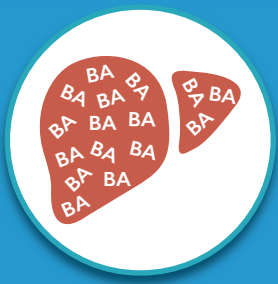
What is primary biliary cholangitis?

Primary biliary cholangitis (PBC), formerly known as primary biliary cirrhosis, is a chronic liver disease. When a person has PBC, the immune system attacks the liver causing slow, progressive damage to the small tubes through which bile flows out of the liver known as bile ducts. Bile is a yellow-green fluid produced by your liver to aid digestion. When the bile ducts are damaged, bile and other substances cannot be eliminated and accumulate in the liver. The retained substances are harmful to the liver and worsen the inflammation which leads to further damage. Over time, this can result in scarring of the liver also known as fibrosis. When there is a lot of scarring in the liver, it is called cirrhosis.

PBC is a slowly progressive disease, which means that damage to the liver typically gets worse over a long period of time.

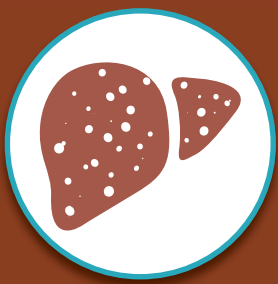
Immune response

The immune system attacks and damages bile ducts



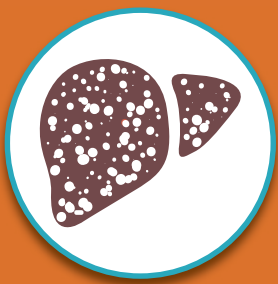
Chronic cholestasis/ inflammation

Inflammation of the bile ducts can lead to cholestasis, a build-up of toxic bile acids



Fibrosis

Chronic cholestasis can lead to fibrosis



Cirrhosis and end-stage liver disease

Further liver damage can lead to cirrcosis and liver lailure

Why this disease is called primary biliary cholangitis?

The disease is called primary biliary cholangitis (PBC) because the immune system attacks the bile ducts inside the liver, which damages them. When immune cells surround and attack parts of the body, this is called inflammation. Inflammation of the bile ducts is called cholangitis. The medical term "primary" in this case means that there is no known reason for this damage to happen. Because the disease blocks or prevents the flow of bile, PBC is known as a "cholestatic" liver disease.

Who is at risk of PBC?

PBC generally affects women between the ages of 40 and 60, but it has been diagnosed outside of this age range, as well as in men. Nine out of 10 people who get PBC are women. Although there is no evidence that PBC is inherited from mother to child, it is more common in the daughters of women with PBC.

What causes PBC?

The cause of PBC remains unknown, but current evidence suggests that PBC occurs in genetically predisposed people after exposure to an undetermined environmental factor. It is important to note that PBC is not caused by alcohol consumption.

PBC is believed to be an autoimmune disease. In the case of PBC, your immune system which is designed to protect you against infections and cancer, mistakes the bile duct cells as being not a normal part of the body, and attacks them. Something may trigger your body into making this mistake. Possible causes of this "trigger" could be an infection or some form of toxin exposure from the environment.

PBC usually develops and progresses slowly. Medication can slow its progression, especially if treatment begins early.

What are the symptoms of PBC?

Many people with PBC have no symptoms. However, when they do, symptoms may vary and may be present in any combination, including:

- mild to disabling chronic fatigue
- mild to intense and unrelenting itching of the skin (often on palms or soles of the feet)
- gradual darkening of the skin
- small white bumps under the skin, usually around the eyes.

Other symptoms, not directly related to PBC, are also often reported by patients:

- dry membranes (nose, eyes, mouth, vagina)
- thyroid problems
- arthritis
- fingers and/or toes that change colour in the cold (Raynaud's disease).

Importantly, many people with PBC never develop any symptoms related to the disease. Most commonly reported symptoms are fatigue and itching in any part of the body. Itching, also medically referred to as pruritus, may be the result of your liver's inability to process bile.

If liver damage progresses in PBC, other symptoms may appear, which can affect other parts of the body outside of the liver. These symptoms include:

- enlarged abdomen from fluid accumulation (ascites)
- easy bruising or bleeding
- yellowing of the skin and eyes (jaundice)
- internal (gastrointestinal) bleeding
- bone pain or spontaneous fracture can also occur due to osteoporosis.

How is PBC diagnosed?

Diagnosis of PBC is typically made with specific blood tests:

- Most people with PBC have something in their blood called antimitochondrial antibody (AMA). An antibody is a protein made by your body's immune system to attack an "invader". The presence of AMA in your blood is an important sign that you have PBC.
- Increased levels of alkaline phosphatase (ALP), an enzyme released into the blood by damaged bile ducts.
- Increased levels of the immunoglobulin IgM.
- The liver enzymes alanine transaminase (ALT) and aspartate transaminase (AST) are also monitored by your doctor although these enzymes are a measure of inflammation relating primarily to liver cell damage/destruction, rather than bile duct cell damage.

In addition, your healthcare provider may request other tests:

- An abdominal ultrasound scan may be used to check the condition of the bile ducts, to make sure there is nothing else causing decreased bile flow out of the liver.
- A liver biopsy may be performed to see how serious your condition is. In this procedure a small needle is inserted into the liver to take a sample of the tissue for analysis.
- A FibroScan® test (also called transient elastography), which is similar to an ultrasound test but gives information as to how stiff your liver is. The higher the stiffness (the number generated by the test), typically the more scarring there is in the liver.

PBC is a slowly progressive disease, which means that damage to the liver typically gets worse over a long period of time.

How is PBC treated?

PBC is a chronic liver disease. This means that PBC is a long-lasting condition that can currently be controlled in the majority of people, but not cured. The prognosis of PBC has improved over the last two decades due to earlier diagnosis and improved treatment. Early access to treatment can significantly delay progression of the disease. The outcome for PBC patients who take treatment is better than for those who are not receiving treatment; and can even be the same as for people who do not have PBC.

- A medication called ursodeoxycholic acid (also called UDCA, and URSO) is made to mimic a naturally occurring bile acid. This medication can improve liver function and delay the development of fibrosis (scar tissue) in the liver, which in turn can delay or eliminate the potential for PBC to progress towards liver failure and/or liver transplantation.
- Vitamins A, D, E and K can be prescribed, usually when severe cholestasis and jaundice is present.
- Various medications may be prescribed by your doctor to help reduce itching. Some examples of these medications include cholestyramine, and antihistamines

- Complications of liver deterioration can often be controlled. As an example, a salt-restricted diet and medication (usually water pills) can be prescribed following accumulation of fluid in the abdomen.

A number of new medicines are currently being developed to treat PBC, typically to be added to UDCA therapy. One such new medication is obeticholic acid (OCA) which has recently been approved by Health Canada for the use either in combination or as an alternative to UDCA for patients who do not tolerate UDCA. OCA is a man-made bile acid that acts to reduce liver inflammation and cholestasis. Combination therapy with bezafibrate (a medication used to lower cholesterol) and UDCA has been evaluated in small clinical trials and has shown promising results in patients with PBC.

Liver transplantation may be recommended for eligible candidates if other treatments are no longer helpful, and liver damage and scarring has progressed to the point that the liver is no longer able to work properly. Liver transplantation works well for people with PBC, although it is possible to develop PBC in your new liver.

How do I live with PBC?

Living with chronic liver disease can be challenging. If you suffer from any or all symptoms listed below, please speak with your healthcare provider as there are a number of treatments that can help manage your symptoms.

Itchy skin

Cholestyramine may be prescribed by your doctor to help ease itching. Cholestyramine is taken orally and works by preventing re-absorption of the chemicals (bile acids) that cause the itching. Sometimes rifampicin, naltrexone or sertraline may be prescribed by your doctor. In addition to medications, a skin moisturizer may be beneficial as itching is made worse by dry skin. As well, avoid taking hot baths and hot showers which may dry out your skin further.

Dry eyes and dry mouth

These symptoms may be relieved by artificial tears, artificial saliva, lubricating gels and creams. Sugarless lozenges and gum may help with the dryness in your mouth. Good dental hygiene is very important. Biotène® products may also be helpful for dry mouth.

Alcohol consumption

If you have a liver disease, you should avoid drinking alcohol as it can damage your liver.

Dietary tips

A healthy diet is important to liver health. Specific things to keep in mind include:

- Maintain a healthy caloric intake recommended for your age, gender and activity level.
- Eat from all four food groups, including fruits and vegetables, whole grains and cereals, milk products, and meats and alternatives.
- Limit your intake of salty, sugary and fatty foods.

Taking medication

Talk to your healthcare provider before taking any medicine, including over-the-counter supplements and herbal remedies. Many medicines may have unwanted side effects and interactions.

Complementary and alternative medicines

Some of these may possibly help with the symptoms of liver disease, but many can damage the liver. More research is needed in order to assess the role of complementary and alternative medicine in liver disease. Please discuss the use of these therapies with your healthcare provider.

Dealing with fatigue

The cause of fatigue in PBC is unknown. It is important for your doctor to rule out other potential non-PBC related causes of fatigue, including anemia and thyroid problems. If you feel tired and unable to carry out day-to-day tasks, you may find that some lifestyle changes can make a difference:

- Pace your activities.
- Try daily aerobic physical activities such as walking and swimming to help make you more fit without becoming exhausted.
- Adjust your hobbies and daily activities to more closely match your energy level.

Bone health

As we get older our bones often become thinner and more brittle. This is more common among women, especially after menopause. PBC may make this worse. You can help prevent this problem by doing plenty of weight-bearing exercise and eating food with plenty of calcium (dairy products such as milk). Sometimes, despite these efforts, medicine may be needed. All Canadians should take a vitamin D supplement (1,000–2,000 IU/day). You may be advised to take calcium supplements. As well, your healthcare provider may recommend bone enhancing medication or hormone replacement therapy.

Are lifestyle changes required for people affected by PBC?

The prognosis of PBC varies greatly from one patient to another. Many lead active lives with few symptoms for more than 10 or 20 years. In some patients, however, the condition progresses more rapidly and liver deterioration may occur in just a few years. There is a healthy way to live with a chronic disease and you are encouraged to maintain an active lifestyle under the supervision of your healthcare provider. It is important to know that support is available that may help you improve your quality of life. In consultation with your healthcare provider, you should do your best to:

- Maintain a healthy lifestyle
- Reduce stress
- Be physically active.

SUPPORT SERVICES

The Canadian Liver Foundation provides support and information to individuals and families coping with liver disease through our national help line, website liver.ca, *community outreach and educational programs.*

If you are looking for a place to turn for answers after diagnosis, to help you understand your disease or to learn more about your liver, please contact us by phone: 1(800) 563-5483
or email: clf@liver.ca.

CANADIAN LIVER FOUNDATION

1 in 4 Canadians may be affected by liver disease, including everyone from newborn babies to older adults.

Founded in 1969 the Canadian Liver Foundation (CLF) is the only national charity in Canada focused on liver health, and the main source of non-profit funding for liver health research.

Today, we are *bringing liver research to life* by raising funds to promote liver health, improve public awareness, fund research and provide support to individuals affected by liver disease.

To support liver research visit **liver.ca/donate**

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