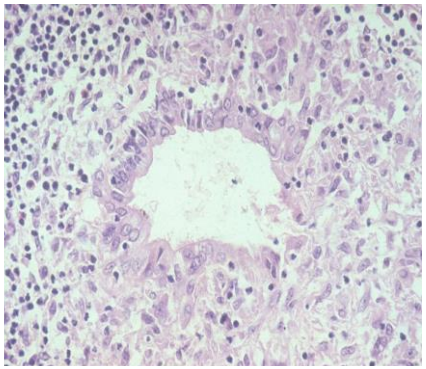
What is Primary Biliary Cholangitis?

Primary biliary cholangitis (PBC) is a chronic autoimmune liver disease where the immune system attacks the small bile ducts in the liver.

This immune damage leads to slow bile duct damage and destruction.

This means over time that bile may not leave the liver as well as it should; bile is a detergent like substance, which means that there is injury in the liver both from the immune system as well as the retained bile.

Over time if the inflammation from the immune system and retained bile is left untouched, then liver damage accumulates, and patients can develop cirrhosis.



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Resources

Canadian Liver Foundation
www.liver.ca

Canadian PBC Society
www.pbc-society.ca

PBCers
www.pbcers.org

How Can I Help?

Donate: Contact Josh Lai at 416-340-5204/ josh.lai@uhn.ca or visit tgwhf.ca

Research: Ask your physician how you can help with any research into this disease.

Primary Biliary Cholangitis (PBC): A Summary for Patients and Their Families

TORONTO CENTRE FOR LIVER DISEASE

How do I know I have PBC?

Many individuals with PBC **will not have symptoms.**

If symptoms are present, the most common ones include:

- Fatigue
- Itching
- Joint aches
- Brain fog
- Dryness (eyes, mouth, vagina)
- Abdominal discomfort
- Restless legs at night

How is PBC Diagnosed?

PBC is usually diagnosed with blood tests, and it is less common to need a liver biopsy nowadays.

Patients with PBC present with higher values of the liver enzyme alkaline phosphatase (ALP) in their blood. PBC diagnosis is then confirmed when antimitochondrial antibodies (AMAs) are detected in blood tests. An ultrasound is usually done and is usually normal.

Approx.10% of patients with PBC will be negative for AMA in their blood. When this occurs and PBC is suspected, further blood tests can help make the diagnosis, or a liver biopsy may be needed.

Many patients now also get a Fibroscan at diagnosis, and then every year. This ultrasound based test is good for measuring liver inflammation and scarring.

How is PBC Treated?

The current initial treatment for PBC is a bile acid called **Ursodeoxycholic Acid (UDCA).**

UDCA works by stimulating bile secretion, and protecting liver cells from toxic effects of more noxious bile acids. It is given by mouth at a dose based on the patient's weight (13-15mg/kg/day). Probably at least 60-70% of patients get sufficient benefit from UDCA as a sole life-long therapy.

If your blood tests on UDCA don't approach normal however, then other treatments need consideration; equally a small number of patients can't tolerate UDCA.

Obeticholic Acid (OCA) is a newer therapy that is also bile acid derived, but which works at low doses to improve bile flow, reduce inflammation and scarring.

Other treatment options include clinical trials of new drugs, or the use of non-licensed drugs such as Fenofibrate or Bezafibrate.

Symptoms of PBC are also treated: there are many options for treating itch, there are strategies for reducing the impact of fatigue, and there are approaches to managing dryness. It is key that you tell your team about symptoms important to you.

Why Do I have PBC?

The exact cause of PBC is unknown; however there may be a relationship between genetic and environmental triggers.

1 in 1000 women over the age of 40 live with PBC, and at least 90% of patients are women.

FAQ

What are the side effects of treatment?

There are minimal side effects with the use of UDCA; however there may be some weight gain within the first 12 months, hair thinning and sometimes an upset stomach. With OCA, itchiness of the skin may be triggered in some patients.

How can I manage my PBC?

It is good to maintain a healthy diet and exercise daily. Cutting out smoking and sensible alcohol intake are also sensible lifestyle choices. Your doctor may prescribe vitamin D supplements to protect your bones from thinning.

What is the outcome of PBC?

Most patients can now be treated either with current medicines, or drugs in development. A small number need liver transplantation but this is a minority nowadays.

Key Points:

1. **PBC is a treatable chronic liver disease that can be slowly progressive.**
2. **There are two approved therapies: UDCA and OCA.**
3. **Knowing whether you are a 'responder' is key in deciding if you need more therapy than just UDCA.**