



Small bile duct surrounded by white blood cells and other inflammatory cells

How do I know I have PBC?

Many individuals with PBC have no symptoms. When those with PBC do have symptoms, the two most common are fatigue and itchiness (pruritus). Itching tends to be worse at night, mostly affecting the palms and soles of the feet. Some individuals also have xanthomas, which are raised, white-yellow spots, around the eyes, palms, soles, elbows, knees or buttocks. Jaundice, a yellow discolouration to the skin and eyes, can also occur, but is now rare.

How does my doctor know I have PBC?

Patients are diagnosed with PBC through blood work, ultrasounds and liver biopsies. More than 95% of patients with PBC have antimitochondrial antibodies (AMA). Patients often have elevated levels of alkaline phosphatase (ALP), an enzyme released into the blood when the bile ducts are inflamed. Alanine and aspartate aminotransferases (ALT and AST), cholesterol and bilirubin may also be raised.



If you would like to contribute to research or would like to donate to help our clinic please contact:

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For more information regarding PBC and support groups, please look at these sites:

Canadian PBC Society

www.pbc-society.ca

PBC Foundation

www.pbcfoundation.org.uk

PBCers

www.pbcers.org

Canadian Liver Foundation

www.liver.ca



Primary Biliary Cirrhosis



What is Primary Biliary Cirrhosis?

Primary Biliary Cirrhosis (PBC) is a chronic and slowly progressive disease caused by inflammation and destruction of small bile ducts in the liver.

The liver produces bile, a yellowish liquid that is stored in the gallbladder and then transported through the bile ducts into the intestines where it is necessary for fat digestion and absorption.

When the bile ducts are destroyed, bile remains in the liver and accumulates, which becomes toxic to the liver cells themselves. When liver cells become damaged, they are replaced by scar tissue, which leads to cirrhosis.

Who has PBC?

1 in 1,000 women over the age of 40 are diagnosed with PBC. It is 9 times more common in women than in men. Most patients are post-menopausal at diagnosis but earlier diagnosis is seen more and more frequently.

Why do I have PBC?

The exact cause of PBC is unknown, but it is likely that your immune system recognizes your own bile ducts as foreign and begin to attack them. It is unclear as to the reason for the attack. Triggers may be in the environment.

PBC is not hereditary, as it is not directly passed on from parent to child; however, PBC does seem to run in certain families and has a strong genetic background.

What can my doctor do to treat PBC?

Ursodeoxycholic acid (UDCA) is the only approved treatment for PBC. UDCA is a water-soluble, naturally occurring bile acid. Treatment with UDCA causes the composition of bile to be less detergent and therefore less damaging to the liver. Although it does not cure PBC, it delays the progression of the disease significantly in the majority of patients.



UDCA is prescribed at a dosage of 13-15mg/kg and most patients take it as a single dose after dinner. It should be taken at least 4 hours apart from Questran (treatment for itching) so that they don't bind to each other. UDCA is a very safe treatment and the side effects are minimal. For the few who do experience side effects, weight gain, thinning of the hair and diarrhea are recognized. Fatigue is best treated by exercising, and for itch we have a long well tried list of treatments to use: don't despair!

What is the latest research in PBC?

Here at the Liver Centre we have led groundbreaking research. Along with collaborators throughout North America, we have discovered at least 5 genes that make people prone to developing PBC. These genes are all associated with how you respond to inflammation. Our patients contributed to this important research which is now on the cusp of leading to new medicines for PBC.



What does my future with PBC hold?

It is very difficult to predict the progression of the disease, as it varies greatly between patients. Those who are diagnosed early and respond to UDCA generally live normal lives. Many are asymptomatic and remain so. Some develop symptoms later on. Fewer and fewer of our patients are in fact ever needing liver transplants.

FAQ

What if I don't respond to UDCA treatment?

At this time, UDCA is the only treatment for PBC. However, just as this clinic did the original studies of UDCA, we are also involved in the latest new clinical trials.

What can I do to manage my PBC?

It is important that you maintain good overall health through eating healthily and exercising regularly. To help you liver, be sure to not smoke. Most patients can drink alcohol so long as they are sensible. Avoid herbal remedies, as they can often be toxic to your liver. It is also recommended that you take calcium and Vitamin D supplements. You can drink coffee, as it is associated with decreased liver fibrosis.

Tips & Reminders

Bring all of your medications in their properly labeled containers to each clinic visit. This will help your doctor in determining the best treatment options for you.

Keep a running list of questions that you have for your doctor and bring them to your next visit.

Bring a relative or friend with you for support and to help you remember and record what has been said during the consultation. We teach residents so clinics can also run late and be longer than your office MDs.

Always get repeat prescriptions before you leave. The clinic is very busy and we can't guarantee to find time to call the pharmacists at other times.