Primary Biliary Cirrhosis

What is Primary Biliary Cirrhosis (PBC)?

PBC is a chronic liver disease that is caused by slow, ongoing destruction of bile ducts in the liver.

- Bile is a substance that helps digest fat in the gut. It is produced in the liver and enters the gut through the bile ducts. When the ducts are damaged, bile builds up in the liver and damages liver tissue.
- In the early stages of the illness, the main problem is that bile and cholesterol build up in the body, instead of leaving through the gut. Over time, the disease can cause cirrhosis (severe scarring) of the liver and may lead to liver failure.
- Women are affected ten times more frequently than men are. PBC is usually diagnosed in people 30 to 60 years old.

What are the symptoms?

The symptoms vary from one PBC patient to another and may be present in any combination.

- Intense, constant itching and chronic fatigue are the most common symptoms.
- Other signs may include gradual darkening of the skin or changes in skin texture due to cholesterol deposits, dry eyes or mouth, thyroid problems and arthritis.
- As the condition progresses, other symptoms may appear, including jaundice (yellowing of the eyes and skin), fluid build up in the ankles and abdomen, easy bruising or bleeding, osteoporosis (bone thinning leading to fractures), and internal bleeding.
- Some patients have no symptoms of disease and are diagnosed by finding an abnormality on routine liver blood tests.

How is PBC diagnosed?

The diagnosis of PBC is made by looking at symptoms and by some tests.

- The person may have symptoms (severe itching) suggesting bile duct damage. Laboratory tests, such as the alkaline phosphatase activity test, may confirm this.
- The test for anti-mitochondrial antibodies (AMA) is positive in nearly all patients.
- Sometimes, the bile ducts are X-rayed to rule out other causes of bile duct disease, such as obstruction.
- A liver biopsy (a simple operation to remove a small piece of liver tissue) will confirm the diagnosis and give information on the severity and extent of liver damage.

What causes PBC?

The cause of the initial bile duct damage in PBC is unknown, but it appears that a combination of both genetic and environmental factors play a role. These have not yet been confirmed; however, there are certain clues.

- PBC is an autoimmune disease, which means that there are changes in the immune system so that it reacts against the body. Usually, the immune system fights infections by viruses and bacteria while ignoring things that are normally found in the body. In an autoimmune disease like PBC, the immune system also fights and destroys part of the body. Lupus, rheumatoid arthritis and Sjögren's syndrome are other examples of autoimmune diseases.
- PBC is more common in families where one member has previously been affected. In general, out of 100,000 people, 40.2 people (or 0.04%) will have PBC. But if a person has one parent, brother or sister with PBC, that person's chance of also being affected is 4-6%. This suggests that some people are genetically more likely to develop PBC.
- PBC is not caused by alcohol or chemicals, although these may make it worse. PBC is not contagious.

What is the prognosis for patients?

The outcome of PBC varies greatly from one person to another.

- PBC usually is slow and many patients lead active and productive lives with few symptoms for ten to twenty years after diagnosis. Patients who show no symptoms at the time of diagnosis often remain symptom-free for years.
- However, in some patients, the condition progresses more rapidly and liver failure may occur in a few years.

How is PBC treated?

Treatment may include taking vitamin and mineral supplements and medicines to relieve symptoms.

- Medication known as Ursodiol (ursodeoxycholic acid) slows down the progression of PBC although it does not cure the disease.
- Itching is treated with Questran (cholestyramine) or Rifampin.
- Thyroid hormone pills can ease fatigue.
- Artificial tears help relieve dry eyes.
- Vitamins A, D and K reduce bleeding problems and jaundice. Vitamin D and calcium supplements are used for osteoporosis.
- Other complications of liver failure may be controlled with a salt-restricted diet and diuretics (water pills).
- Finally, liver transplantation is a common treatment option for people with advanced PBC.

What precautions should be taken?

Before taking any other drugs or vitamins, a person with PBC should consult their gastroenterologist. The liver is responsible for breaking down drugs and if it is already under stress from PBC, extreme caution should be taken to avoid other possible damage. Daily use of alcohol should also be avoided.

Where can I get more information on PBC?

- The PBCers Organization is the largest PBC support group with members worldwide. They have an excellent website with an e-mail daily digest, quarterly newsletter, annual conference, chatroom, message board, a section where questions are answered by doctors, information on PBC research and clinical trials, dietary guidelines and many other items of interest.
  Website: http://www.pbcers.org/
  E-mail: pbcers@aol.com

- The Canadian Primary Biliary Cirrhosis Society is a Canadian group based on Ontario, which provides current information about liver disease and related issues.

Address:	168 Redpath Avenue	Tel: 416-440-0917
	Toronto, Ontario	Toll Free: 1-866-411-3643
	M4P 2K6	Website: www.pbc-society.ca
		E-mail: pbcsociety@yahoo.ca

- MedicineNet.com is a website with information written by doctors. It has more detailed information about the causes of PBC, its diagnosis, progression and treatments.
  Website: www.medicinenet.com/Primary_Biliary_Cirrhosis/article.htm

If you, or one of your family members, have a diagnosis of PBC and are of First Nations heritage, we invite you to take part in a research study. Dr. Laura Arbour at the University of Victoria and her colleagues are carrying out research on this condition, in consultation with the involved First Nations groups. Please call Dr. Laura Arbour at 250-472-5544 or her assistant Sarah McIntosh toll-free at 1-888-853-8924.