
Primary biliary cirrhosis

Summary

- Primary biliary cirrhosis is characterised by the chronic inflammation and scarring of the bile ducts within the liver.
 - For reasons unknown, the immune system attacks the cells that line the bile ducts.
 - Women are 10 times more likely to develop PBC than men.
 - There is no cure, but treatment can help slow the progression of disease and ease associated symptoms.
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The principal roles of the liver include cleansing toxins from the blood and processing food nutrients into proteins, fats and carbohydrates. The liver produces bile, which is stored in the gall bladder and added to the digestive tract via bile ducts to help break down dietary fats. Primary biliary cirrhosis (PBC) is an autoimmune condition characterised by the chronic inflammation and subsequent scarring of the bile ducts within the liver. The cause is unknown, although researchers have ruled out alcohol and diet as possible triggers. Women are 10 times more likely to develop PBC than men, for reasons unknown. The disease is usually diagnosed later in life, between the ages of 35 and 60 years. There is no cure, but treatment can slow the progression of the disease and alleviate symptoms. PBC seems to be associated with a number of other autoimmune diseases including rheumatoid arthritis, scleroderma and Sjogren's syndrome, although the reasons for this are not clear.

Symptoms of primary biliary cirrhosis (PBC)

Symptoms of PBC vary from one person to another, ranging from non-existent to mild to severe. The symptoms tend to be progressive and can include:

- increasing fatigue
- unexplained pigmentation of the skin
- jaundice (yellowing of the skin and eyes)
- chronically itching skin
- unexplained weight loss
- indigestion
- easy bruising
- aching joints
- abdominal discomfort around the liver (upper right hand side of the abdomen)
- bloated abdomen due to fluid build-up (ascites)
- osteoporosis.

Destruction of bile ducts

Bile drains into the digestive tract via bile ducts. For reasons unknown, the immune system attacks the cells that line the bile ducts inside the liver. This causes chronic inflammation, damage and scarring. Over time, accumulated scar tissue blocks the ducts and causes a build-up of bile within the liver, which in turn becomes inflamed. Functioning liver tissue is gradually replaced with non-functioning scar tissue (cirrhosis). Eventually, so much of the liver is replaced by scar tissue that it can no longer function properly. (The majority of people with PBC don't ever experience this degree of cirrhosis.)

Cause of primary biliary cirrhosis

PBC is an autoimmune disorder. The immune system attacks the bile ducts, but the trigger for this attack is so far unidentified. Alcohol and diet are not thought to play significant roles in the development of the disease. PBC seems to be slightly more common in families, which suggests a genetic susceptibility. The disease seems to be more common in Scotland, Scandinavia and North East England, for reasons unknown.

Diagnosis of PBC

By the time symptoms of PBC become apparent, the bile ducts have sustained significant damage. Early stage PBC is sometimes diagnosed during blood tests and other investigations for unrelated medical conditions. Specific tests for PBC include:

- blood tests
- liver function tests
- x-rays
- ultrasound scans
- liver biopsy.

Treatment for PBC

There is no cure for PBC, but treatment can help slow the progression of disease and associated symptoms. Options include:

- medication, particularly ursodeoxycholic acid (Ursofalk)
- fat-soluble vitamin supplements, since absorption of these vitamins from food is reduced
- low-salt diet
- diuretics to help control fluid retention
- medication to treat the itching, such as cholestyramine and rifampicin
- regular weight bearing exercise (such as walking or weight training) and calcium supplementation to reduce the risk of osteoporosis
- hormone replacement therapy, in some cases
- reduced intake of alcohol and non-essential medication
- nutritious diet high in vitamins, minerals and trace elements
- frequent small meals throughout the day, rather than three big meals spaced wide apart
- liver transplant, in severe cases.

Reducing skin itching

Chronically itching skin is a maddening symptom of PBC. Some people may even develop scars from constant scratching. Medications such as cholestyramine and rifampicin can reduce the itchiness. Other suggestions include:

- Keep fingernails short.
- Wear natural fibres like cotton rather than synthetics.
- Moderate sun exposure may help.
- Avoid overheating and hot baths.
- Avoid perfumes, scented soaps, bath oils, bubble bath and talcum powder.
- Take a cool bath infused with one cup of bicarbonate of soda before bed.

Where to get help

- Your doctor
- Gastroenterologist
- **[Australian Primary Biliary Cirrhosis Support Group](#)**

Things to remember

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- Women are 10 times more likely to develop PBC than men.
- There is no cure, but treatment can help slow the progression of disease and ease associated symptoms.

This page has been produced in consultation with and approved by:

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