CONDITION

Primary biliary cholangitis

Primary biliary cholangitis is a long-term immune condition that slowly damages the liver's small bile ducts.

WHAT IT IS
Chronic immune damage to small bile ducts

BODY SYSTEM
Liver and bile ducts

MAIN SYMPTOMS
Fatigue, itching, dry eyes, upper abdominal discomfort

WHAT CAUSES IT
Immune reaction with genetic and environmental influences

HOW IT IS DIAGNOSED
Cholestatic blood tests plus PBC-specific antibodies

MAIN TREATMENT
Prescription bile acid and specialist monitoring

CAN IT BE CURED?
No, but treatment can slow damage

IS IT AN EMERGENCY?
Sometimes, with vomiting blood or sudden confusion

LONG-TERM OUTLOOK
Often good when found early and treatment works

Overview

Primary biliary cholangitis, or PBC, is a long-term condition affecting the small bile ducts inside the liver. These ducts carry bile, a fluid that helps digest fat and remove waste from the body.

In PBC, the immune system mistakenly attacks the bile ducts. The ducts become inflamed and are gradually damaged. Bile can then build up in the liver, causing further inflammation and scarring.

PBC often develops slowly over years. Many people have no symptoms when it is found through a routine blood test. Without effective treatment, some people develop severe liver scarring, called cirrhosis, and eventually liver failure. Early diagnosis and treatment can greatly reduce this risk.

The condition used to be called "primary biliary cirrhosis." The name was changed because many people do not have cirrhosis when diagnosed and may never develop it.

Symptoms

PBC can be present for years without causing noticeable symptoms. Abnormal liver blood tests may be the first sign.

When symptoms occur, the most common are:

Tiredness does not always match the amount of liver damage. It can remain troublesome even when treatment is controlling the condition. Other causes, such as anaemia, thyroid problems, sleep disorders, depression or medicine side effects, may also contribute.

Itching can affect one area or the whole body. It may come and go, and scratching can damage the skin. PBC itching usually happens without a rash, although scratch marks may be visible.

Signs of more advanced liver damage can include yellow skin or eyes, called jaundice, a swollen abdomen, swollen legs, easy bruising, muscle loss or weight loss. Pale stools and dark urine can occur when bile flow is reduced.

Small fatty deposits may appear around the eyelids or under the skin. These are related to changes in cholesterol levels.

- Persistent tiredness that may not improve with sleep
- Itching, which may be worse at night
- Dry eyes or a dry mouth
- Discomfort under the ribs on the upper right side

Causes and risk factors

PBC is an autoimmune condition. This means the immune system attacks healthy tissue by mistake. It is not caused by alcohol, diet or anything you have done.

The exact trigger is not known. Genes appear to affect susceptibility, while environmental factors may help start the immune reaction. PBC is not contagious.

The condition is much more often diagnosed in women, usually during middle age, but people of any sex or age can develop it. Having a close relative with PBC raises your risk, although most relatives do not develop the condition.

PBC can occur alongside other autoimmune conditions. These include autoimmune thyroid

conditions, Sjögren's syndrome, which causes dry eyes and mouth, systemic sclerosis and coeliac disease.

PBC is different from primary sclerosing cholangitis, or PSC. Both affect bile ducts, but they have different patterns of damage, associated conditions and treatments.

Diagnosis

PBC is usually diagnosed using a combination of blood tests and the overall clinical picture. No single symptom confirms it.

A liver blood test commonly shows a raised alkaline phosphatase, or ALP. This enzyme often rises when bile ducts are inflamed or blocked. Other tests measure bilirubin, liver enzymes, albumin and blood clotting. These help assess liver function and the amount of damage.

Most people with PBC have antimitochondrial antibodies, known as AMA, in their blood. These are immune proteins strongly associated with the condition. Some people who do not have AMA have other PBC-specific antibodies.

An ultrasound or another scan is usually used to look for gallstones, narrowed larger bile ducts or other causes of reduced bile flow. A scan does not normally confirm PBC by itself.

A liver biopsy involves removing a small sample of liver tissue with a needle. It is not needed for everyone. It may be considered when blood tests are unclear, another liver condition is suspected or more information about liver damage is required.

You may also have a non-invasive test such as transient elastography. This uses ultrasound-like vibrations to estimate liver stiffness, which can help assess scarring without a biopsy.

Treatment and management

Treatment aims to improve bile flow, reduce ongoing liver damage and lower the risk of cirrhosis and liver failure. It also addresses symptoms and complications.

Ursodeoxycholic acid, often shortened to UDCA or called ursodiol, is the usual first treatment. It is a prescription bile acid taken long term. It can improve liver blood tests and reduce the chance that PBC will progress, especially when treatment begins early.

Blood tests over the following months show how well treatment is working. If the response is not strong enough, or UDCA cannot be tolerated, a liver specialist may consider another medicine. Newer options include elafibranor and seladelpar in countries where they are

approved. Suitability is affected by liver function, other medicines and local availability.

Treatment may also include:

Some itching medicines can interfere with the absorption of UDCA and other medicines. Your prescriber or pharmacist can explain how to separate them safely. Do not start herbal or "liver detox" products without checking first, because some can damage the liver or interact with treatment.

Regular monitoring may include liver blood tests, assessment of liver stiffness and checks of bone health. If cirrhosis develops, monitoring may also include scans for liver cancer and a camera test to check for enlarged veins in the gullet, called varices.

A liver transplant may be considered for liver failure, serious complications of cirrhosis or severe itching that cannot be controlled. It replaces the damaged liver, although PBC can sometimes return in the transplanted liver.

- Medicines that reduce itching or remove bile acids from the bowel
- Artificial tears, saliva substitutes and dental care for dryness
- Assessment and treatment of osteoporosis, or fragile bones
- Treatment of vitamin deficiencies when testing confirms them
- Support for sleep, mood or other causes of fatigue

Complications

Ongoing inflammation can cause fibrosis, meaning scar tissue in the liver. More extensive scarring is called cirrhosis. Cirrhosis can increase pressure in the blood vessels around the liver, known as portal hypertension.

Portal hypertension can cause fluid in the abdomen, an enlarged spleen or swollen veins in the gullet and stomach. These veins can bleed. Advanced liver damage may also allow waste products to affect the brain, causing confusion called hepatic encephalopathy.

Other possible complications include:

Raised cholesterol in PBC does not always carry the same heart risk as other forms of high cholesterol. Your overall risk should still be assessed using factors such as blood pressure, smoking, diabetes and family history.

- Osteoporosis and a higher risk of fractures
- Poor absorption of vitamins A, D, E and K in advanced cholestasis

- Raised cholesterol and fatty deposits around the eyes
- Gallstones or bile duct stones
- Liver cancer, mainly in people with cirrhosis

Prevention

There is no proven way to prevent PBC. It results from an immune reaction influenced partly by inherited susceptibility, and there is no screening programme that can stop it from developing.

If you have PBC, taking prescribed treatment and attending monitoring can help prevent or delay further liver damage. Avoid smoking and discuss alcohol with your healthcare team. Alcohol did not cause PBC, but it may add to liver damage, particularly if significant scarring is already present.

Check before using supplements, herbal remedies or non-prescription medicines. Keep recommended vaccinations up to date, especially those advised because of chronic liver problems.

Living with it and what to expect

PBC usually changes slowly over months and years rather than from day to day. Many people who are diagnosed early and respond well to treatment remain active and do not develop liver failure. Others have continuing inflammation despite treatment and need closer monitoring or additional medicines.

Blood test results and liver stiffness are more useful for predicting progression than symptoms alone. Severe fatigue does not necessarily mean that the liver is failing. Likewise, feeling well does not make follow-up unnecessary.

Itching may improve with PBC treatment or need separate management. Fatigue can persist, so pacing activities, protecting sleep and investigating other causes may help. Dry eyes and mouth often need ongoing symptom treatment and regular dental care.

Most people can work, exercise, travel and maintain relationships. Adjustments may be needed during periods of severe fatigue or itching. Support from a liver clinic, primary care team and patient organisation can help with work, sleep and emotional wellbeing.

Pregnancy is possible for many people with PBC. Planning is important because medicines may need review and liver-related complications can affect monitoring during pregnancy.

Routine appointments remain important even when you feel well. They allow treatment

response, bone health and signs of liver scarring to be checked over time.

When to get medical help

Arrange a routine medical appointment if you develop persistent unexplained itching, fatigue, dry eyes or dry mouth, or if blood tests repeatedly show a raised ALP. These symptoms have many possible causes and do not by themselves mean you have PBC.

If you already have PBC, contact your healthcare team promptly for new jaundice, increasing abdominal swelling, leg swelling, unexplained weight loss or a clear change in symptoms. Fever with worsening upper abdominal pain also needs urgent assessment.

Seek emergency help for:

These can be signs of bleeding, infection or advanced liver complications and should not wait for a routine appointment.

- Vomiting blood or material that looks like coffee grounds
- Black, tar-like stools
- Sudden confusion, unusual drowsiness or difficulty waking
- Fainting, severe weakness or breathing difficulty
- Rapidly increasing abdominal swelling with severe pain

Common questions

Is primary biliary cholangitis cancer?

No. PBC is an autoimmune liver condition, not cancer. People who develop cirrhosis have an increased risk of liver cancer and may be offered regular surveillance scans.

How long can you live with PBC?

Many people diagnosed early who respond to treatment have a life expectancy close to that of people without PBC. The outlook is less favourable when treatment does not control the condition or advanced scarring is already present. Blood tests and liver stiffness help estimate individual risk.

Is PBC hereditary?

PBC is not inherited in a simple, predictable way. Certain genes can increase susceptibility, so it may occur more often in some families. Most children and siblings of someone with PBC will not develop it.

Can I drink alcohol if I have PBC?

Alcohol does not cause PBC. Whether any amount is safe depends on your liver function, the amount of scarring and other health factors. People with cirrhosis are generally advised to avoid alcohol. Ask your liver team for advice based on your results.

Can PBC go into remission?

Treatment can bring liver tests under good control and greatly slow progression, but PBC is considered a lifelong condition. Medicines and monitoring usually continue even when tests improve and you feel well.

Is PBC contagious?

No. You cannot catch PBC through contact, food, sex, blood or sharing a home. It develops because of an abnormal immune response, not an infection.

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