Information about

Primary Biliary Cholangitis (PBC)

What is primary biliary cholangitis (or PBC)?

Primary biliary cholangitis (PBC), which used to be known as primary biliary cirrhosis, is a rare autoimmune liver condition where the immune system attacks the cells that line the small internal (or "intrahepatic") bile ducts of the liver.

This leads to chronic inflammation ("cholangitis") and scarring of the internal bile ducts, which interferes with the flow and drainage of bile.

Bile is a fluid that's made in the liver. It flows from the liver's internal bile ducts to "extrahepatic" bile ducts that exit the liver, then drains into the small intestine. Bile helps with digesting fat and removing waste products from the body.

Over time, the damage PBC does to the internal bile ducts and the way it disrupts the flow of bile gradually inflames the liver cells. This can lead to scarring ("fibrosis"), which may develop into advanced scarring, known as cirrhosis. However, many people with PBC do not develop cirrhosis.

What causes PBC?

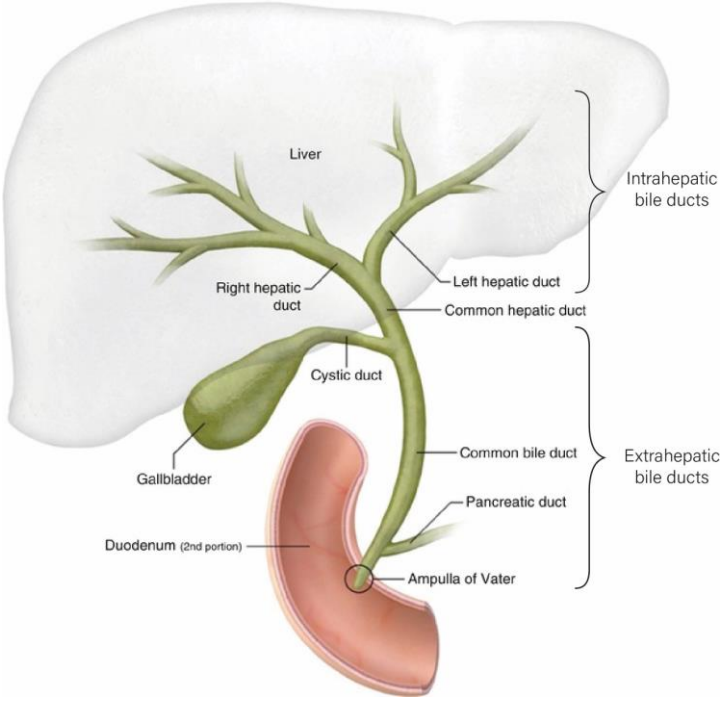
It's not yet known what causes PBC, but it's likely an autoimmune condition. People with PBC often have other autoimmune disorders, like Sjögren's disease, autoimmune thyroid disease or Raynaud's syndrome.

PBC can be diagnosed at any age but is most common in people older than 40 years. Women are nine times more likely than men to develop PBC. First-degree relatives of someone with PBC, like their siblings or children, are also at higher risk of PBC.

How is PBC diagnosed?

Your doctor might suspect you have PBC if you have a persistent abnormality in your liver enzyme tests (particularly in the level of an enzyme called alkaline phosphatase, or ALP) or if you have symptoms of PBC.

PBC is usually diagnosed by a specialist gastroenterologist based on test results:



1. **Blood tests**
   - High levels of the enzyme ALP are common in people with PBC, and this is often the first problem noticed
   - 95% of patients with PBC will have a positive anti-mitochondrial antibody (AMA) blood test, although people without PBC can also have this

2. **Liver biopsy**
   - A liver biopsy can show bile duct damage

3. **Imaging**
   - Imaging tests, like an ultrasound, FibroScan or magnetic resonance imaging (MRI) scan, may be needed to rule out other liver conditions or to check for damage in the liver.

In most cases, liver enzyme test results that remain abnormal (like having a high ALP level for at least 6 months) and a positive AMA test are enough to diagnose PBC. If there's any doubt about the diagnosis, or if your doctor suspects you may also have a second liver problem, a liver biopsy may be needed too.

What are the symptoms of PBC?

Many people have no symptoms of PBC, particularly when the disease first starts. Typical symptoms, when they do occur, can include fatigue (feeling tired) and itchy skin ("pruritus"). Some people develop jaundice (yellowing of the skin) and abdominal (belly) pain.

What are the treatment options for PBC?

Unfortunately, there's no cure for PBC. However, there are treatments that can slow it down and treat its symptoms. If you have PBC, you will need to keep seeing a specialist for the rest of your life.

Ursodeoxycholic acid (UDCA) is a naturally occurring bile acid that is normally found in small amounts in humans. As a medicine, UDCA is approved as the first-line therapy for PBC, which means it works well and will be the first treatment you're given. Regularly taking UDCA will improve your liver test results (like ALP level) and slow down the progression of PBC. As PBC is a lifelong disease, you will usually need to keep taking UDCA indefinitely.

Obeticholic acid (OCA) is a second-line treatment that you might be given if you have intolerance to UDCA or if UDCA doesn't work well for you after using it for at least 12 months.

Fibrates are medicines that are usually used for lowering cholesterol levels, but they can also be used "off-label" (meaning for a different disease) for people with PBC who don't respond well to other treatments.

General health recommendations for people with PBC include keeping up to date with vaccinations and avoiding heavy drinking of alcohol. PBC can make your bones more fragile and lead to osteoporosis, so you may be screened with a bone density (DEXA) scan and given vitamin D supplements.

Clinical trials of possible new treatments are running around the world, including in Australia, and your treating specialist may recommend you join one of these.

This fact sheet gives general information only and is not intended to replace medical advice. If you have any questions or need advice, please speak with your doctor.

What are the treatment options for complications of advanced liver disease?

Some people with PBC will develop advanced liver disease and cirrhosis. Cirrhosis can cause complications like hepatic encephalopathy, which is an altered mental state due to a build-up of toxins, or portal hypertension, which is high pressure in the blood vessels within and around the liver. Portal hypertension can lead to ascites (fluid in the abdomen) and varices (abnormally dilated blood vessels that are at risk of bleeding).

Lactulose (which is a laxative to help you have regular bowel motions) and rifaximin (an antibiotic) are used to treat hepatic encephalopathy. You may need an endoscopy to check your oesophagus (food pipe) and stomach for varices. Medications known as beta-blockers, such as propranolol or carvedilol, are often prescribed to help lower portal hypertension and the risk of bleeding from varices. People with cirrhosis may be at risk of primary liver cancer, which can occur without any symptoms, so regular screening with ultrasound imaging every 6 months is recommended for these people.

With the treatments available, liver transplantation is rarely needed for people with PBC. For patients who go on to have liver failure, a liver transplant can be a good option if they are medically fit for transplantation.

What's my prognosis?

The availability of treatments like UDCA has greatly improved the prognosis for people with PBC, and many people with early-stage PBC have a normal life expectancy. People with normal levels of ALP and bilirubin (one of the components of bile) are likely to have a good prognosis. Those who develop complications like cirrhosis and advanced liver failure may have a worse prognosis.

Acknowledgements

This resource was created by the following health professionals in 2024:

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