



# PRIMARY BILIARY CHOLANGITIS (PBC)

---

## What is PBC?

PBC is a chronic autoimmune liver disease in which the body's own immune system causes slow but progressive damage to the small bile ducts inside the liver. (The bile ducts are the channels through which bile flows.) Over the course of many years, PBC can eventually lead to cirrhosis (scarring of the liver), although this does not happen in everyone with the disease.

## What causes PBC?

PBC is an autoimmune disease, in which the body's own immune system mistakenly attacks the cells that line the small bile ducts inside the liver. No one knows why this happens. Some people with PBC have relatives with the condition. This suggests that people with PBC may have an inherited predisposition to the disease, which is finally triggered by something in the environment. We do not know what the environmental trigger (or triggers) might be. PBC is more common in women, in people with other autoimmune conditions, and amongst those who smoke tobacco.

## What are the symptoms of PBC?

Common symptoms associated with PBC include fatigue, itching ('pruritus'), dryness of the eyes or mouth (sicca complex), and upper abdominal pain. Because of related conditions, some people experience joint pains, diarrhoea and over or under-active thyroid. Many people with PBC have no symptoms at all.

## Is there a treatment for PBC?

There is no cure for PBC but there are very effective treatments. All patients with PBC begin treatment with a medication called Ursodeoxycholic Acid (UDCA), given at a dose of 13-15mg/kg/day. It may be taken in divided doses or all at once. It is very well tolerated. Most people taking UDCA have no side effects at all. The most common side effects reported by patients are weight gain and loose bowel motions.

In most people with PBC, UDCA is highly effective at controlling the disease. In some people, however, UDCA is less effective. These patients may receive treatment with obeticholic acid (OCA) or bezafibrate, in addition to UDCA. OCA has recently been licensed for the treatment of PBC and is only available via specialist centres (such as Addenbrooke's Hospital). It is generally well-tolerated, but might cause itching which can be controlled using other medications. Bezafibrate is a medication licensed for treatment of high cholesterol, which is used 'off licence' for treatment of PBC.

It is generally well-tolerated and has few side effects but does require close monitoring with blood tests when it is first started. ('Off licence' means that the medication is used for a different condition than it is licensed for.)

## What should I expect from Addenbrooke's Hospital?

You will be under the care of a hepatology consultant. Following your first consultation with the doctor you will have a baseline ultrasound scan (USS), fibroscan, and DEXA scan, and you will start treatment with UDCA. Your subsequent management will depend on the stage of the PBC and how you respond to treatment with UDCA. Patients without cirrhosis who respond well to treatment with UDCA (with or without second-line treatment) will be seen every one or two years. Patient with cirrhosis will have an ultrasound scan of the liver every six months, and a clinic appointment every six to 12 months. Regular ultrasound scans are important for people with cirrhosis because they have an increased risk of liver cancer – and regular scans can help to detect liver cancer at an early and treatable stage. Other investigations may be required as the disease progresses.

We will do our best to keep the monitoring of your PBC up-to-date. We strongly encourage you to become familiar with the tests you need, however, to ensure these tests happen when they should.

## Contacts

If you require any further information about this leaflet or advice about your condition please contact the Addenbrooke's autoimmune liver disease advice line on 01223 216109

## Where can I receive more information and support?

- The PBC Foundation: [www.pbcfoundation.org.uk](http://www.pbcfoundation.org.uk) Tel: 0131 5566811
- The British Liver Trust: [www.britishlivertrust.org.uk](http://www.britishlivertrust.org.uk) Helpline: 0800 652 7330 (10:00 to 15:00 Monday to Friday) UK-PBC: [www.uk-pbc.com](http://www.uk-pbc.com)
- Psychological Wellbeing Service offers treatments across Cambridgeshire and Peterbough [www.cpft.nhs.uk](http://www.cpft.nhs.uk) Tel: 0300 300 0055

## What research is happening in PBC?

There are also opportunities to be involved in clinical trials for new treatments of PBC. Please call 01223 256225 or e-mail Sister Abi Ford on [Hepatologyresearch@addenbrookes.nhs.uk](mailto:Hepatologyresearch@addenbrookes.nhs.uk) if you would like to discuss this option.