



# PRIMARY BILIARY CHOLANGITIS

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## What is PBC?

PBC is a chronic autoimmune liver disease in which the body's own immune system causes slow 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 may eventually lead to cirrhosis (scarring of the liver), although some people with PBC never develop this. PBC can lead to liver cancers so requires careful monitoring by liver specialists at Addenbrooke's Hospital.

## 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, however, know what the environmental trigger (or triggers) might be. PBC is more common in women, in people with other autoimmune conditions, and among those who smoke tobacco.

## What are the symptoms of PBC?

Common symptoms associated with PBC include lethargy with chronic fatigue, upper abdominal pain, dryness of the eyes or mouth (sicca complex) and itching ('pruritus'). Because of related conditions, some people experience joint pains, diarrhoea and over or under-active thyroid. Many people with PBC will live without any symptoms.

## 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 may 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.

## 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 Peterborough [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.