Primary biliary cirrhosis

Fighting liver disease
Primary biliary cirrhosis

The British Liver Trust works to:

- support people with all kinds of liver disease
- improve knowledge and understanding of the liver and related health issues
- encourage and fund research into new treatments
- lobby for better services.

All our publications are reviewed by medical specialists and people living with liver disease. Our website provides information on all forms of adult liver disease and our Helpline gives advice and support on general and medical enquiries. Call us on 0800 652 7330 or visit www.britishlivertrust.or.uk
The liver

Your liver is your body’s ‘factory’ carrying out hundreds of jobs that are vital to life. It is very tough and able to continue to function when most of it is damaged. It can also repair itself – even renewing large sections.

Your liver has around 500 different functions. Importantly it:

- fights infections and disease
- destroys and deals with poisons and drugs
- filters and cleans the blood
- controls the amount of cholesterol
- produces and maintains the balance of hormones
- produces chemicals – enzymes and other proteins – responsible for most of the chemical reactions in the body, for example, blood clotting and repairing tissue
- processes food once it has been digested
- produces bile to help break down food in the gut
- stores energy that can be used rapidly when the body needs it most
- stores sugars, vitamins and minerals, including iron
- repairs damage and renews itself.
How liver disease develops

Liver damage develops over time. Any inflammation of the liver is known as hepatitis, whether its cause is viral or not. A sudden inflammation of the liver is known as acute hepatitis. Where inflammation of the liver lasts longer than six months the condition is known as chronic hepatitis.

Fibrosis is where scar tissue is formed in the inflamed liver. Fibrosis can take a variable time to develop. Although scar tissue is present the liver keeps on functioning quite well. Treating the cause of the inflammation may prevent the formation of further liver damage and may reverse some or all of the scarring.
Cirrhosis is where inflammation and fibrosis has spread throughout the liver and disrupts the shape and function of the liver. With cirrhosis, the scarring is more widespread and can show up on an ultrasound scan. Even at this stage, people can have no signs or symptoms of liver disease. Where the working capacity of liver cells has been badly impaired and they are unable to repair or renew the liver, permanent damage occurs.

This permanent cell damage can lead to liver failure or liver cancer. All the chemicals and waste products that the liver has to deal with build up in the body. The liver is now so damaged that the whole body becomes poisoned by the waste products and this stage is known as end stage liver disease. In the final stages of liver disease the building up of waste products affects many organs. This is known as multiple organ failure. Where many organs are affected, death is likely to follow.
What is primary biliary cirrhosis (PBC)?

PBC is a chronic disease that can, little by little, destroy some of the tubes linking your liver to your gut. These tubes are called bile ducts.

The bile ducts carry a fluid called bile to your gut (also known as your intestine or bowels).

When PBC damages your bile ducts, bile can no longer flow through them. Instead it builds up in the liver, damaging the liver cells and causing inflammation and scarring.

Over the years this damage becomes widespread. The liver becomes less able to repair itself, leading to a condition known as cirrhosis. The damage caused to your liver cells by cirrhosis can result in the failure of your liver carry out all of its jobs properly.

What is bile?

Bile is a yellow-green fluid produced by your liver which contains chemicals as well as waste products to aid digestion. It plays a central role in helping the body digest fat. It acts as a detergent, breaking the fat into very small droplets so that it can be absorbed from food in your gut. It also makes it possible for your body to take up the fat-soluble vitamins A, D, E and K from the food passing through the gut.
What are the symptoms of PBC?

Some people with PBC will never get any symptoms of the disease. Clear symptoms of PBC are constant tiredness (for some people this can be severe) and intense itching in any part of the body. Itching, also known as pruritus, may be a result of your liver’s inability to process bile. It is thought that bile acids are not the cause of the itching but rather other chemicals that are retained in the body. As with tiredness, the severity of the itching will vary from person to person. Severity is not an indication of the amount of liver damage.

Other symptoms that may develop usually include the following:

- dry eyes and/or dry mouth
- constant or variable ache or discomfort in the upper right hand side, below your ribs
- indigestion, nausea or poor appetite
- arthritis (inflammation of the joints)
- pain in the bones
- mottled palms with red or pink blotches
- diarrhoea
- dark urine and/or pale stools
- jaundice – yellowing of the skin and whites of the eyes.

Tiredness and itching are generally the first symptoms to appear while jaundice is usually associated with the later stages of the disease.
Why is it called PBC?

It is called primary biliary cirrhosis because the disease attacks the bile (biliary) ducts inside the liver. This leads to scarring (fibrosis) and then to cirrhosis. In this case ‘primary’ does not mean first but that there is no known reason for this damage to happen.

The name PBC is a little misleading. Many people do not have cirrhosis when they are first diagnosed or for many years afterwards. Indeed, many people with PBC never go on to get cirrhosis.

Because it blocks or stops the flow of bile, PBC is known as a ‘cholestatic’ liver disease.

Who is at risk of PBC?

Nine out of ten people who get PBC are women. No one knows why this is. In particular the following women are most at risk:

- women who are middle aged or older
- women who have a family history of PBC.

There is no evidence that PBC can be passed on to others by simple contact, kissing, blood or sexual contact.
What are the causes of PBC?

Over-consumption of alcohol and hepatitis are the main causes of cirrhosis in the UK, accounting for more than half of all cases. Despite the strong association between alcohol and liver disease, drinking alcohol does not cause PBC.

PBC is believed to be an autoimmune disease. This is a disease where the body attacks itself. In this case, your body’s defence against germs and infections – the immune system – mistakes the cells lining the bile ducts as ‘foreign’ and attacks them.

Why this happens is still not known. Some medical specialists believe that something may trigger your body into making this mistake. Possible causes of this ‘trigger’ could be:

- an infection
- some form of toxin (poison) taken in from the environment.

Occasionally PBC comes to light during or just after pregnancy. It is not clear whether pregnancy may spark it off or, because pregnant women are under close medical supervision, the symptoms of PBC are first noticed during pregnancy.

This external trigger probably starts PBC only in people who are already at risk because they have inherited certain genes from their parents.
Testing for PBC

Antibody test
Most people with PBC have something in their blood called antimitochondrial antibody (AMA). An antibody is a chemical made by the body to attack an ‘invader’. Although doctors are not really sure why, the presence of AMA in your blood is an important sign that you have PBC.

Liver function tests (LFTs)
This type of test is performed to gain an idea how the different parts of your liver are functioning. The liver function test is made up of a number of separate examinations, each looking at different properties of your blood.

In PBC, doctors will be looking for increased levels of both alkaline phosphatase (ALP), an enzyme released into the blood by damaged bile ducts, and the immunoglobulin IgM.

The liver enzymes alanine transaminase (ALT) and aspartate transaminase (AST) are also monitored, although these are a measure of any leakiness or damage relating primarily to liver cells rather than the bile duct cells.

Ultrasound scan
Ultrasound is the same technology used to confirm all is well in pregnancy. Gel will be applied to your skin, which may feel slightly cold. A probe, like a microphone, will be moved across your skin to send sound waves into the liver area.
The reflected sound waves, or echoes, are picked up through the probe and used to build a screen image of the liver’s condition.

This painless test is used to check the condition of the bile ducts and to rule out the possibility that your symptoms could be a sign of a different liver problem.

**Liver biopsy**

If tests show you have PBC you may need a liver biopsy to see how serious the condition is.

During a liver biopsy a tiny piece of the liver is taken for study. A fine hollow needle is passed through the skin into the liver and a small sample of tissue is withdrawn.

The test is usually done under local anaesthetic and may mean an overnight stay in hospital, although some people may be allowed home later the same day. As the test can be uncomfortable and there is a very small risk of internal bleeding or bile leakage, a stay in bed of at least six to eight hours is required. Ask your doctor for more information on this.

**Treatment**

There are a number of treatments for the symptoms of PBC. Some of them help with any unpleasant side effects, such as dry eyes, and others slow the progress of the disease.
Managing unpleasant symptoms

Itching skin: colestyramine (sold as Questran) may be prescribed by your doctor to help ease itching. Taken orally, colestyramine works by preventing re-absorption of the chemicals that cause the itching. It can take days or even weeks before this becomes effective.

Some people taking colestyramine have problems such as changed bowel habits and bloating. Your doctor may prescribe ‘Questran light’ to reduce these side effects.

If colestyramine does not help, a hospital specialist may try other medicines such as rifampicin and naltrexone.

Itching is made worse by dry skin. It is very important to use plenty of moisturiser. If you’re running a bath, don’t make it too hot.

Dry eyes and dry mouth: the combination of dry eyes and a dry mouth (sicca syndrome) might be soothed by such treatments as artificial tears and saliva, lubricating gels and oestrogen creams. You may find that lozenges from your pharmacist will help with the dryness in your mouth.

Controlling your condition

A medication called ursodeoxycholic acid (URSO, also referred to as UDCA) has helped some people. Made from naturally occurring bile acid, it seems to work through several mechanisms by protecting the liver from the harmful chemicals in the bile.
Although early studies failed to show any direct impact on survival rates, new evidence suggests that URSO may help to keep people alive for longer and delay liver transplantation.

A number of other medicines are also now being tried and your medical team may ask for your assistance in this. At present, however, a liver transplant is the only option for people who have advanced PBC.

**Will I need a liver transplant?**

A liver transplant is usually only recommended if other treatments are no longer helpful and your life is threatened by end stage liver disease. It is a major operation and you will need to plan it carefully with your medical team, family and friends.

However, liver transplantation works well for people with PBC. It is possible to get PBC in your new liver, but it may take up to 15 years before the disease becomes significant.

**Looking after yourself**

PBC affects people in very different ways. Because of this it is very difficult to say what impact PBC may have on your life.

For example, many people with PBC may live with very few problems for many years or even decades. On the other hand, PBC can have a major impact on people’s day-to-day lives early on by causing unpleasant symptoms such as very itchy skin.
If your liver has become damaged it can have a major impact on your health. For example:

- it may reduce your body’s ability to digest food properly, especially fats
- it may mean your body does not get enough energy, making you always feel tired (fatigue)
- you may not cope very well with poisons such as alcohol and some medicines.

**Diet**

Whether you have PBC or not you will help yourself by eating a healthy diet. This means:

- plenty of fruit and vegetables (at least five portions a day)
- lots of carbohydrates (pasta, potatoes, wholemeal bread and rice)
- only a little fat (as little saturated animal fat as possible).

Some people with PBC may need to eat a special diet. If you have any questions about your diet, talk to your GP. You can ask to be referred to a registered dietician for some personal advice.

If you have any of the following you should definitely see a registered dietician about the right food and drink.

- cirrhosis
- fluid building up in your abdomen (ascites) or leg (oedema)
- mental slowness or confusion (encephalopathy)
- diarrhoea where the stools are pale, bulky and difficult to flush away (steatorrhoea).
Problems with fat
Some people with PBC have difficulty digesting fat. This leads to a type of diarrhoea called steatorrhoea (see above). Some people may also feel sick and bloated. If this happens to you, apart from seeing a registered dietician, you may find that a low fat diet helps to reduce the diarrhoea, abdominal pain and any other discomfort from the steatorrhoea.

Fat is essential and should not be cut out of your diet completely without proper advice from a dietician. Fat is an important source of energy, so if you do have to cut it out you may need to eat more carbohydrates for more energy.

Some people may need to have energy supplements and injections of fat-soluble vitamins. Others are prescribed medium chain triglycerides (MCT) which are fats that are easier to digest.

Heartburn
PBC may cause some people to experience an unpleasant taste in their mouth, or heartburn – a severe pain in the chest that can be mistaken for heart problems. Stomach acids escaping into your food pipe (oesophagus) are the usual cause of this sensation.

If you get heartburn you may find that eating small amounts often helps. Avoid big meals and spicy food. It is a good idea to always carry something to eat, preferably food high in carbohydrate such as a barley sugar or boiled sweet.
Alcohol
Alcohol is actually a poison dealt with by the liver. If your liver is damaged it may mean that you have difficulty coping with alcohol and many people with PBC find they can no longer drink alcohol. Others may find they can only drink a little alcohol now and then, perhaps on a special occasion.

Sensible drinking advice varies from person to person. It will depend on many different things, such as how serious your liver problems are and on your general health. Your doctor will be able to advise you.

Energy levels
Always feeling tired is caused by your liver being unable to provide enough energy for the rest of your body. This means that carrying out day-to-day tasks can be exhausting. You may find that making changes to your lifestyle can make a big difference, such as:

- pacing your activities
- doing gentle exercises such as walking and swimming to help make you fitter without becoming exhausted
- changing your hobbies and daily activities
- if you are able, giving up your work or doing it part-time.

PBC can also affect your life in the following ways.

Caution with medicines
Talk to your doctor before taking any medicine or treatment. Many medicines may have unwanted
side effects that are usually dealt with by your liver, which may not be working very well. For example, it is best to take paracetamol (but never more than four 500mg tablets in any day) rather than aspirin for day-to-day aches and pains.

However, most drugs are well-handled by the liver in PBC even in end stage disease.

**Thinning bones (osteoporosis)**
As we get older our bones often become thinner and weaker. This is more common among women, especially after the menopause. PBC may make this worse. You can help to stop this problem by doing plenty of weight-bearing exercise and eating food with plenty of calcium, such as skimmed milk.

Sometimes, despite these efforts, medicine is needed. You may be advised to take calcium supplements, a ‘bone enhancing’ medicine called a biophosphate or a course of hormone replacement therapy (HRT), for example.

Bone loss can be detected with a special type of X-ray called a ‘Dexa Scan’.

**Higher risk of bleeding**
With PBC you may be more at risk of bleeding because your liver is less able to make the chemicals needed to clot blood. This means that before treatment you need to tell people such as your dentist that you have PBC so they can take special care. But remember, there is no evidence that PBC can be passed on from contact with blood.
Complementary and alternative medicines

Many complementary and alternative medicines are available that may ease the symptoms of liver disease. But certain medications used in non-liver related disease can damage the liver. At present, healthcare professionals are not clear on the role and place of some therapies in managing liver disease. More research needs to be done on the use of these therapies. You may wish to discuss the use of these therapies with your doctor.
Useful words

**ALP** – alkaline phosphatase, an enzyme found in certain membranes of the liver. Increases in ALP and another liver enzyme called Gamma GT (GGT) can indicate obstructive or cholestatic liver disease, where bile is not properly transported from the liver because of obstruction (blockage) of the bile duct.

**Autoimmune** – a type of disease causing the body’s immune system to attack another part of the body.

**Bile** – a yellow/green fluid made by the liver to help digest foods containing fat and cholesterol.

**Cholestasis** – a condition where the flow of bile from the liver is reduced.

**Chronic** – an illness that lasts a long time (more than six months), possibly for the rest of a person’s life.

**Cirrhosis** – where inflammation and fibrosis have spread to disrupt the shape and function of the liver. Even with no signs or symptoms of liver disease, the working capacity of liver cells has been badly impaired and they are unable to repair the liver. This is permanent cell damage and can lead to liver failure or liver cancer.

**Encephalopathy** – disturbed brain function leading to mental confusion and memory loss. Encephalopathy can follow the development of cirrhosis, for example.

**Inflammation** – the first response of the immune system to infection, commonly characterised by heat, swelling, pain and tenderness.
**Immunoglobulins** – also known as Ig, these are large proteins that act as antibodies found in body fluids and cell tissues. They will bind to invading organisms, such as bacteria or viruses, to destroy them.

**Steatorrhoea** – diarrhoea where the stools (faeces) are pale, bulky and difficult to flush away. This is caused by the body’s problems with digesting fat in the diet.
Who else can help?

PBC Foundation (UK)
54 Queen St
Edinburgh EH2 3NS
Tel: 0131 225 8586
Fax: 0131 225 7579
Email: info@pbcfoundation.org.uk
Web: www.pbcfoundation.org.uk
The PBC Foundation (UK) Ltd is a charity offering support and information to PBC sufferers, their families and friends. The Foundation issues a quarterly newsletter and can put you in touch with others that have PBC.

National Osteoporosis Society
Camerton
Bath BA2 0PJ
Tel: 0845 130 3076
Fax: 01761 471 104
Email: info@nos.org.uk
Web: www.nos.org.uk
The National Osteoporosis Society is the only national charity dedicated to improving the prevention, diagnosis, and treatment of this fragile bone disease.

UK Transplant
Tel: 0117 975 7575
Email: enquiries@uktransplant.nhs.uk
Web: www.uktransplant.org.uk
Part of NHS Blood and Transplant and the official NHS transplant site. It provides news, information and resources about organ transplantation, real life stories from those who have benefited from transplants and a facility for potential donors to sign on to the organ donor register.

Transplant Support Network (TSN)
6 Kings Meadow Drive
Wetherby
West Yorkshire LS22 7FS
Telephone support line: 0800 027 4490/4491
Email: tsnetwork@tiscali.co.uk
Web: www.transplantsupportnetwork.org.uk
Nationwide network and registered charity supporting patients, family and carers of all solid organ transplants and mechanical implants.
Further information

The British Liver Trust publishes a large range of leaflets about the liver and liver problems written for the general public.

Leaflets that you may find particularly helpful include:
- Autoimmune hepatitis
- Cirrhosis and liver disease
- Diet and liver disease
- Liver disease tests explained
- Liver transplantation
- Primary sclerosing cholangitis

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Liver disease is increasing alarmingly and the need to do more is greater than ever before…

For the British Liver Trust to continue its support, information and research programme, we need your help. We raise funds from many sources and a large proportion is donated by voluntary contributions. If you would like to send a donation it will enable us to continue providing the services that people need.

If you can help, please fill in the form on the page opposite.

If you wish to help us further with our work by organising or participating in a fundraising event or becoming a “Friend of the British Liver Trust” please:

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