Primary biliary cirrhosis

Fighting liver disease
Primary biliary cirrhosis

The liver

Your liver is your body’s ‘factory’, carrying out hundreds of jobs that are vital for life. You only have one liver, but it is very tough. It keeps on going when badly damaged and can repair itself.

Your liver has about 500 different roles. Importantly, it:

- makes quick energy when the body needs it
- helps fight infections
- makes bile to help breakdown food in the gut
- stores sugars, vitamins and minerals, including iron
- destroys and deals with poisons and drugs
- filters and cleans the blood
- controls the amount of cholesterol
- makes and controls the amount of hormones
- makes the chemicals – proteins and enzymes - that keep our body working.

What is primary biliary cirrhosis (PBC)?

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

The bile ducts carry bile – which is yellow and made in your liver – to your gut (also known as your intestine or bowels).

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

Over the years this damage becomes widespread. The liver becomes less able to repair itself properly, leading to a condition known as cirrhosis and the failure of the liver to carry out all of its jobs properly.
Symptoms of PBC

Some people with PBC will never get any symptoms of the disease. When symptoms do develop they usually include the following:

- constant tiredness - mild or severe
- intense itching of any part of the body
- dry eyes and/or dry mouth
- constant ache or discomfort in the upper right hand side below the ribs
- indigestion, nausea or poor appetite
- arthritis
- pain in the bones
- mottled palms with red or pink blotches
- weakness in wrists and/or hands
- diarrhea
- dark urine and/or stools
- jaundice - yellowing of the skin and whites of the eyes.

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.

Why is it called PBC?

It is called primary biliary cirrhosis as the disease attacks the bile duct (biliary), which then leads to scarring (cirrhosis) of the liver. In this case primary does not mean first, but that there is no known reason for the damage to happen.

The name PBC is a little misleading, as 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.

What is bile?

Bile 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 the body to take up the fat-soluble vitamins A, D, E and K from the food passing through the gut.

What is cirrhosis?

Cirrhosis is the result of long-term and non-stop damage to the liver. The damage leads to scarring, known as fibrosis. At the same time, irregular bumps (nodules) replace the smooth liver tissue and the liver becomes harder. Together the scarring and the nodules are called cirrhosis.

Although, in the UK, alcohol abuse and hepatitis are the main cause of cirrhosis (more than half of all cases) a number of other diseases can lead to it, including PBC.

Please read our leaflet Cirrhosis of the liver or visit our website www.britishlivertrust.org.uk.
Causes of PBC

PBC is thought to be a disease where the body attacks itself, known as an autoimmune disease. The body’s defence against germs and infections - the immune system - mistakes the bile ducts as ‘foreign’ and attacks them.

Why this happens is still not known. Some researchers think that something may trigger the body into making this mistake. Possible causes of this ‘trigger’ could be:

- an infection
- some form of 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 closer 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.

Despite the strong association between alcohol and liver disease, drinking alcohol does not cause PBC.

How is PBC likely to affect you?

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, 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.

Doctors cannot cure PBC or completely stop its slow destruction of the bile ducts. Because of this PBC is a major reason for liver transplants in the UK.

Testing for PBC

Doctors can tell whether you have PBC on the basis of symptoms and blood tests. If you do have PBC you may need a biopsy (see below) to see how serious the disease is.

Antibody test

Most people with PBC have something in their blood called anti-mitochondrial antibody (AMA). An antibody is a chemical made by the body to attack an ‘invader’. Though doctors are not really sure why, if AMA is found in the blood, it is an important sign that you have PBC.

Liver function tests (LFTs)

Liver function tests are blood tests that are carried out to check how well the liver is working.
Ultrasound scan

Ultrasounds - the same technology as is used with pregnant women to check that everything is okay with the baby - send sound waves into the body. The echoes are picked up and used to build a picture of what is going on inside the body.

Ultrasounds are painless. A little gel, which may be rather cold, is smeared on the skin over the liver and then the ultrasound prod is passed over it.

Ultrasounds are used to rule out the possibility that your symptoms could be a sign of a different liver problem.

Liver biopsy

A liver biopsy is a process where a tiny piece of the liver is cut away for study in a laboratory.

During the biopsy a fine hollow needle is passed through the skin into the liver and a small sample is withdrawn with the needle. The sample is sent to a laboratory to be examined by a specialist doctor called a pathologist.

The test is usually done under a local anaesthetic and may mean that you have to stay overnight in hospital, although some people may go home later on the same day (you have to wait in bed for at least six to eight hours). The test can be uncomfortable and there is a small risk of internal bleeding or bile leakage. Get your doctors to give you more information about this.

Treatment

There are a number of treatments for PBC. Some of them help with any unpleasant symptoms, such as dry eyes, and others slow the progress of the disease. At this time there is no cure for PBC.

Helping manage unpleasant symptoms

Itching skin: cholestyramine (sold as Questran or Questran Light) may be prescribed by your doctor and can help ease itching. It can take some days before it works properly.

Some people taking Questran have problems, including changed bowel habits and bloating. If you have this problem talk to your doctor about other treatments that may suit you better.

If cholestyramine does not help, a hospital specialist may try other medicines which include rifampicin and naloxone.

Itching is made worse by dry skin. It is very important to use plenty of moisturiser, even when you take a bath.

Dry mouth: you may find that lozenges from your local pharmacist help.

Dry eyes: you may find that artificial ‘tears’ prescribed by your doctor help.
Controlling the condition

Drugs: one drug, called ursodeoxycholic (URSO for short) has helped some people, although it is unclear how effective it really is. It seems to work by protecting the liver from the harmful chemicals in the bile.

A number of other medicines are now being tried for PBC - you may be asked to try one of these by a hospital specialist.

Liver transplant: if your liver is seriously damaged by cirrhosis, a liver transplant may be the best option. This is usually only recommended if your quality of life has become very poor and other treatments are no longer helpful. This is a major operation and you will need to plan it carefully with your medical team, family and loved ones. (It is possible to get PBC in your new liver, but this rarely causes problems).

Living with PBC

Because your liver is responsible for so many tasks, if it is damaged, it can have a major impact on your life. For example:

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

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 - ask to be referred to a registered dietitian for some personal advice.

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

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

If you get heartburn you may find that eating small amounts often helps (avoid big meals, especially spicy food). A good idea is always to carry something to eat, preferably food high in carbohydrate, for example a barley sugar or boiled sweet.

For more information on what to eat, see our leaflet Diet and liver disease or visit our website www.britishlivertrust.org.uk.

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 (fatigue) is the most common symptom of PBC. It is caused because the liver is unable to provide enough energy for the rest of the body. This means that, unless you are careful, doing day-to-day tasks can be exhausting.

Some people find that making changes to their lifestyle can make a big difference, such as:
● pacing their activities
● doing gentle exercise, such as walking and swimming, which can help to make them fitter, but does not exhaust them too much
● changing their hobbies and daily activities
● giving up work or going part-time.

Other issues
PBC can impinge on your life in other ways too.

Problems with medicines: talk to your doctor before taking any medicine or treatment - whether
it is bought in the newsagent, pharmacy or is an alternative treatment.

Many medicines may have unwanted side effects that are usually dealt with by the liver. This means that if your liver is not working so well, you may have to avoid some medicines. For example, it is best to take paracetamol (but never more than four in any day) rather than aspirin for day-to-day aches and pains.

**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 (skimmed milk, for example). 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 the liver is less able to make the chemicals needed to clot blood. This means that you need to tell people, such as your dentist, before treatment that you have PBC so he or she can take special care. But remember, PBC is not infectious so there is no danger to the dentist, other healthcare professionals or your family.

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### Useful words

**Autoimmune** - a disease where our body’s defences attack another part of the body.

**Bile** - a yellowish acid made in your liver that acts as a detergent to break down fat into tiny droplets.

**Cirrhosis** - very bad (severe) scarring of the liver brought on by long-term and non-stop damage.

**Encephalopathy** - mental slowness or confusion.

**Hepatitis** - swollen and tender liver.

**Inflammation** - the body’s protective reaction to injury, involving swelling, pain, redness and heat.

**Steatorrhoea** - diarrhoea where the stools (faeces) are pale, bulky and hard to flush away. It is brought on because the body has difficulty digesting fat in the diet.
Who else can help?

**PBC Foundation**
54 Queen Street
Edinburgh, EH2 3NS
**Tel:** 0131 225 8586
**Email:** info@pbcfoundation.com
**www.pbcfoundation.org.uk**
The PBC Foundation provides support for people with Primary Biliary Cirrhosis, as well as their families and friends. The Foundation issues a regular newsletter and can put you in touch with others that have PBC.

**National Osteoporosis Society**
Camerton
Bath BA2 0PJ
**Helpline:** 0845 4500230
**Email:** nurses@nos.org.uk
**www.nos.org.uk**
The national charity for people with osteoporosis, offering support and information.
Remember, if you are seeing a hospital specialist about PBC then take this leaflet and use it to help you ask any questions you need answered.

Further reading

The British Liver Trust has a large choice of leaflets about the liver and liver problems, for the general public. See our website (www.britishlivertrust.org.uk) or order them direct from us. You can find our contact details on page 18.

Titles you may find useful having read this leaflet include:

- *Alcohol and liver disease*
- *Cirrhosis of the liver*
- *Diet and liver disease*
- *First steps - a guide to your liver*

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**British Liver Trust**
Portman House, 44 High Street,
Ringwood, BH24 1AG
**Tel:** 01425 463080  **Fax:** 01425 470706
**Email:** info@britishlivertrust.org.uk
**Web site:** www.britishlivertrust.org.uk