

# Primary sclerosing cholangitis



BRITISH  
LIVER  
TRUST

Fighting liver disease

# Primary sclerosing cholangitis

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 **0870 770 8028** or visit **[www.britishlivertrust.org.uk](http://www.britishlivertrust.org.uk)**

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## The liver

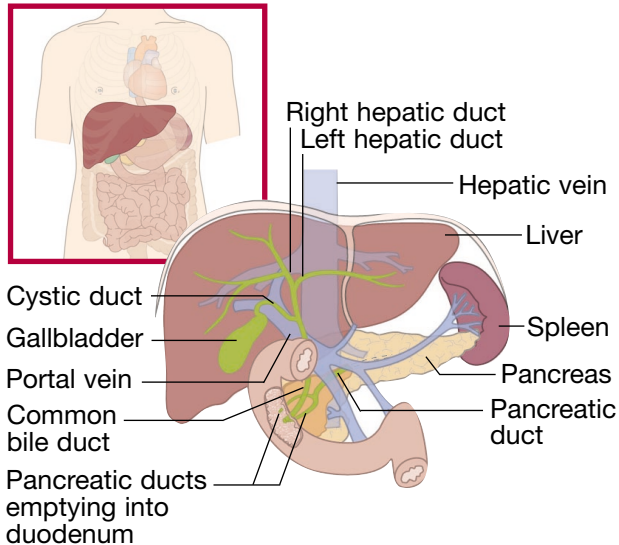
Your liver is your body's 'factory', carrying out hundreds of jobs that are vital to life. It is very tough and able to continue to function when most of it is damaged. It can also repair itself – even renewing large sections.

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.

## What is primary sclerosing cholangitis?

This information leaflet has been written to help you understand more about primary sclerosing cholangitis, often referred to as PSC. At the end of the leaflet there is a list of other British Liver Trust services and sources of further information, that may be useful. If after reading this leaflet you have more questions or concerns, it is important that you discuss these with your doctor.



PSC is an uncommon chronic liver disease in which the bile ducts inside and outside the liver progressively decrease in size due to inflammation and scarring (fibrosis). The disease may occur alone, but frequently is associated with inflammatory diseases of the colon, especially chronic ulcerative colitis.

Bile ducts are tubes which carry bile (a greenish yellow liquid made by the liver) into the upper part of the bowel. Bile acts as a detergent breaking up fat from the food we eat into small droplets that can then be absorbed into the body. It also enables the body to absorb vitamins A, D, E and K from our diet.

As a consequence, bile that is normally carried by these ducts accumulates within the liver. This blockage to bile flow also causes damage to liver cells causing inflammation and scarring. Over many years the scarring can affect the whole liver and the system of bile ducts. A damaged liver can re-grow without scarring but in PSC, the re-growth goes wrong and the healing process is incomplete. The combination of scar tissue and irregular growth is known as cirrhosis.

## What causes PSC?

The cause of PSC remains unknown. Liver damage and cirrhosis is often presumed to be caused by drinking too much alcohol, however PSC is not related to alcohol in any way.

Current evidence suggests that the disease may be triggered by an unknown bacteria or virus in people who are genetically programmed to get the disease. The common viruses known to cause hepatitis have not been associated with it.

The frequent occurrence of PSC in association with inflammatory bowel disease suggests that a common cause for both diseases may exist or that the inflamed colon allows toxins or infections to be absorbed into the body and this can cause the bile duct inflammation. The disease affects both genders, although two male patients are affected for every female patient. It can affect all ages, but it is most commonly found in young adults.

## Signs and symptoms of PSC

Many people have no symptoms at first and the disease is only discovered because of abnormal results of routine blood tests in patients with ulcerative colitis or Crohn's disease. In some people PSC does not produce any symptoms. Most people have few or no symptoms for many years.

Common early symptoms are:

- tiredness
- some abdominal discomfort in the right upper abdomen.

Late symptoms are:

- itching
- jaundice – yellowing of the skin and whites of the eyes
- episodes of fever, shaking and chills can be distressing but are uncommon.

Liver failure may ultimately develop.

PSC may be occasionally complicated by the development of bile duct cancer. PSC is closely associated with inflammatory bowel disease, usually ulcerative colitis, but also Crohn's disease. The course of the ulcerative colitis is often very mild, with few flare-ups. However bowel cancer does appear to develop more frequently in patients with colitis who also have PSC and regular yearly examination of the colon by colonoscopy (a tiny video camera) is recommended.

## Tests

PSC is diagnosed on the combination of symptoms, blood tests and a picture of the bile ducts, called a cholangiogram. A liver biopsy may be needed not only to confirm the diagnosis, but also to assess either how early or advanced the disease is.

If your GP suspects you may have the condition you will be referred to a hospital specialist for tests, expert advice and treatment. The specialist you may see could be a gastroenterologist (digestive disease specialist).

## Liver function tests

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

## Ultrasound scan

Often the bile ducts are examined by ultrasound to exclude the possibility of other diseases.

Ultrasound is a quick examination and completely painless. A special jelly, which may feel cold, is smeared on the skin over the liver and a small probe like a microphone is passed over the area. Echoes from sound waves are sent into the body and are used to build up a picture of the liver.

## Liver biopsy

A liver biopsy is the procedure by which a tiny piece of the liver is taken for examination under a microscope. It is usually performed under a local anaesthetic and patients usually go home later on the same day. A fine hollow needle is passed through the skin into the liver and a small sample is withdrawn.

## Cholangiography

There are two main methods of obtaining a picture of the biliary tree. Traditionally, an endoscopic cholangiogram (ERCP) is carried out. Under sedation a thin tube (endoscope) containing a small camera is passed through the mouth into the small bowel (the duodenum) via the stomach. A tiny tube is then passed through the endoscope into the bile ducts, dye (contrast media) is injected and X-rays are taken to produce a picture of the bile ducts.

A newer method is a tubeless test called magnetic resonance cholangiogram (MRCP) although the picture quality is not quite as good as ERCP. This means lying in a scanner which can be a little claustrophobic for some people.

## Treatment

At present there is no known cure or specific treatment for PSC although preliminary trials have suggested that the natural bile acid, ursodeoxycholic acid, may slow the progression of the disease possibly by increasing bile flow and reducing liver inflammation. Symptoms such as itching can be treated with agents such as cholestyramine, rifampicin or naltrexone.

Cholestyramine (trade name Questran® or Questran Light®) may be prescribed to help relieve itching. It works better when taken before and after meals, especially breakfast, but it may take some days before the treatment is effective. Some people taking Questran® experience side effects such as altered bowel habits or bloating. If this is a problem, tell your doctor as there are other treatments such as albumin dialysis that may suit you better.

Many people suffer with a dry mouth and dry eyes, but this can be helped by taking lozenges and artificial tears prescribed by your doctor.

For a few people who eventually go on to get advanced cirrhosis, a liver transplant may be recommended when their quality of life has deteriorated and medical treatment can no longer control their symptoms. Because PSC usually develops slowly, transplantation can generally be carefully planned well ahead.

People with advanced PSC are often deficient in vitamins A, D and K and replacement fat-soluble vitamins are given. Endoscopic treatment of the bile ducts is aimed at reducing or halting the progression of the disease, usually by stretching or dilating the narrowed bile ducts. In later stages of the disease liver transplantation is often an option. Survival after this operation is good, although in some patients PSC can recur in the new liver.

## Diet

When someone has a long-term liver problem such as PSC, the liver's ability to carry out its many functions may not be as good as in a healthy person.

In particular, the liver's ability to break down alcohol and medicines may be impaired, however this is often not a problem. If in doubt, check with your doctor.

Many people with PSC can eat a normal diet, while others may need more detailed advice. If you are well with few symptoms you may not need to make any changes, although it is important to eat as healthily as possible.

If you have cirrhosis or other complications such as fluid retention (ascites and oedema) or mental slowness or confusion (encephalopathy), you may need specialist advice from a State Registered Dietician.

A few people have problems digesting fat and can develop a type of diarrhoea called steatorrhoea, in which stools are bulky, pale and difficult to flush away. There may also be nausea. If this occurs, it may help to reduce the amount of fat in the diet under the

supervision of a dietician. A low fat diet should be followed only if steatorrhoea is causing problems.

The aim of a low fat diet is to improve the diarrhoea, abdominal pain and discomfort associated with steatorrhoea. As fat is an important source of energy, anyone following a low fat diet should eat extra carbohydrate, such as starch and sugar.

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

A few people experience heartburn and an unpleasant taste in the mouth, usually caused by acid from the stomach going back up into the gullet. Eating small amounts often helps. A good idea is always to carry something to eat, preferably food which contains carbohydrate.

It is important to talk to your doctor or dietician before making any changes in your diet.

## Alcohol

Many people with PSC find that they can no longer tolerate alcohol. Some may be advised to drink only a little on special occasions while others should not drink at all.

Sensible drinking advice varies from person to person and will depend on many factors, such as the severity and stage of the disease, as well as your general health. Your specialist is the best person to advise.

## Looking after yourself

Tiredness is the commonest symptom of PSC. Some people may need to consider making changes to their lifestyle, such as giving up work or a particular activity.

Some people find that pacing their daily activities helps to preserve stamina and energy. Gentle exercise such as walking and swimming can be beneficial.

It is important to tell your dentist that you have PSC as there may be an increased risk of bleeding.

Also, there are a few medicines that are best avoided, and the dose of others may need to be reduced. For example, it is better to take paracetamol rather than aspirin to combat aches and pains. However, you should check this with your doctor first; they are best equipped to advise you.

### Important

Talk to your doctor before taking any medicine (not prescribed by him or her), including paracetamol, herbal remedies and Chinese medicine.

If you are female and middle aged, it is important to discuss with your doctor the best way of reducing your risk of developing osteoporosis.

Your doctor may advise various measures which might include increasing the intake of calcium in your diet, taking calcium supplements and HRT (hormone replacement therapy) if appropriate.

Most doctors will also advise not smoking.

If you are concerned by any of the symptoms mentioned in the leaflet, it is important that you consult your doctor.

## Who else can help?

### The PSC Support Group

39 Belvoir Road, Dulwich, London SE22 0QY

Tel/fax: 020 8693 8789

Email: pscsupport@aol.com

Website: www.psc-support.demon.co.uk

A voluntary organisation supporting people with PSC and their families.

### National Osteoporosis Society

Camerton, Bath BA2 0PJ

Tel: 0845 130 3076

Email: info@nos.org.uk Website: nos.org.uk

## Further information

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

Leaflets that you may find particularly helpful include:

- *Cirrhosis of the liver*
- *Diet and liver disease*

This leaflet is for information only. Professional, medical or other advice should be obtained before acting on anything contained in the leaflet as no responsibility can be accepted by the British Liver Trust as a result of action taken or not taken because of the contents.

## Special thanks

**We would like to thank:** Dr Roger Chapman  
BSc (Lon) MD (Lon) MBBS FRCP, John Radcliffe  
Hospital, Oxford.

## Can you make a difference?

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 through 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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**0870 770 8028**

**Email us at**  
**info@britishlivertrust.org.uk**

**Make a donation via our website at**  
**www.britishlivertrust.org.uk**

**or write to**  
**British Liver Trust**  
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Ringwood, BH24 1HY

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