

Primary sclerosing cholangitis



BRITISH
LIVER
TRUST

Pioneering Liver Health

Primary sclerosing cholangitis

This publication is for people diagnosed with primary sclerosing cholangitis and for those who would like to better understand the condition.

The British Liver Trust works to:

- support people with, and affected by, liver disease
- improve knowledge and understanding of the liver and related health issues
- encourage and fund research into new treatments
- campaign for better services and improved patient care
- increase awareness of the risk factors of liver disease and promote earlier diagnosis

All our publications are reviewed by medical specialists and people living with liver disease. Our website provides information and our Helpline gives advice and support on enquiries about liver health. Call the Helpline on **0800 652 7330**, general enquires on **01425 481320**, or visit **britishlivertrust.org.uk**

For the latest updates to this information, please refer to our website **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 able to repair itself (even renewing large sections). **However, the liver's ability to repair itself is limited and continuous harm can lead to permanent scarring.** Your liver is very tough and able to function even when some of it is damaged, which means you may not notice any symptoms until your disease is quite advanced and noticeably affecting your health.

Your liver performs hundreds of functions. Importantly it:

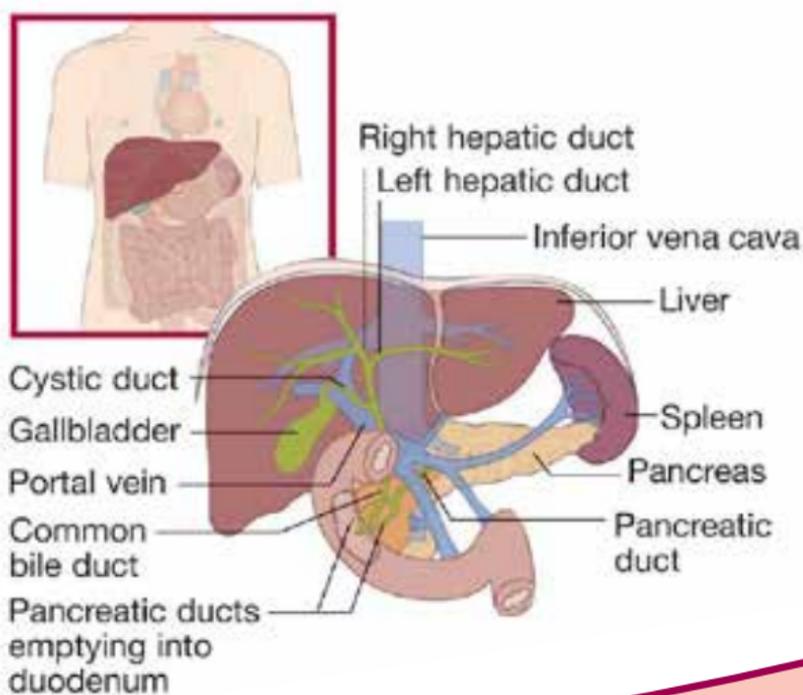
- filters and cleans the blood
- fights infections and disease
- deals with and destroys poisons and drugs
- makes vital proteins which make your blood clot when you cut yourself
- produces bile to help break down food in the gut
- processes food once it has been digested
- stores energy that can be used rapidly when the body needs it most
- regulates fat breakdown and distribution in the bloodstream
- stores sugars, vitamins and minerals, including iron
- gets rid of waste substances from the body
- produces and maintains the balance of some hormones
- produces chemicals – enzymes and other proteins – responsible for most of the chemical reactions in the body, for example repairing tissue
- repairs damage and renews itself (up to a point).

How liver disease develops

Your liver responds to harm by becoming inflamed. Any inflammation of the liver is known as hepatitis, whatever its cause. Sudden inflammation of the liver is known as acute hepatitis. When inflammation of the liver lasts longer than six months, it is known as chronic hepatitis.

Inflammation is part of the process of repairing damaged tissue. In a similar way to a scab forming over a skin wound, a temporary fibrous 'scaffold' forms while new liver cells regenerate. If your liver is repeatedly harmed, new liver cells cannot regenerate fast enough and the fibrous scaffold remains as a scar. This is called fibrosis, and can take a variable amount of time to develop.

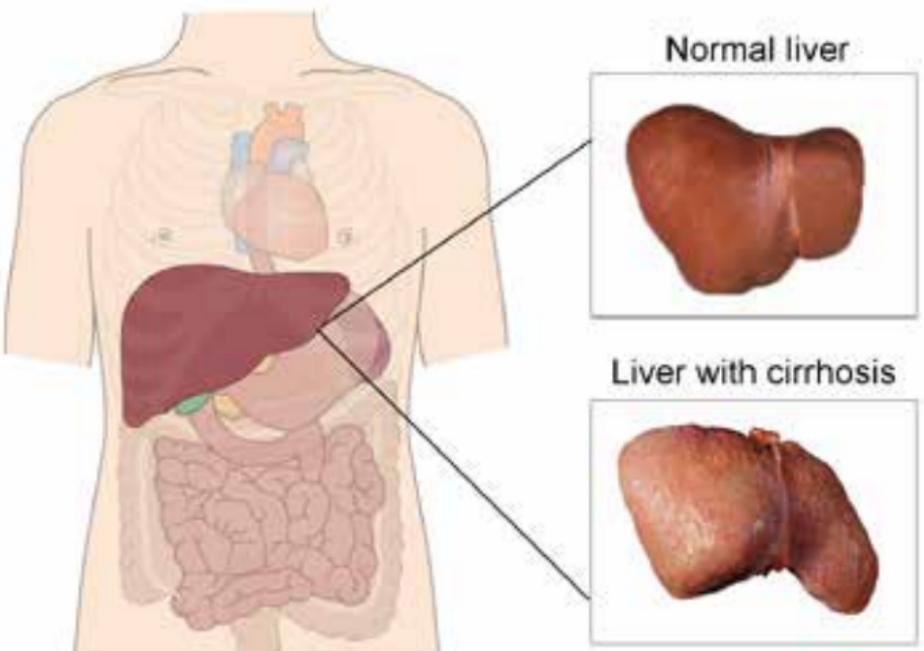
When fibrosis is present, your liver may be able to keep functioning quite well. Removing or treating the cause of the inflammation may reverse some, or all, of the fibrosis and prevent further liver damage.



If the harm to your liver continues, the inflammation and fibrosis can spread throughout your liver, changing its shape and affecting how well your liver cells work. This is known as **compensated** cirrhosis. Even at this stage, people can have no obvious signs or symptoms.

The scar tissue in cirrhosis interrupts the blood flow through the liver. As a result, the blood pressure in the veins in your abdomen is increased and may result in bleeding. Scar tissue in cirrhosis is difficult to remove and may be permanent. However, further progression can be halted and your cirrhosis stabilised, if the cause of the liver damage is removed.

Cirrhosis increases your risk of liver cancer and can lead to liver failure. If damage to your liver continues, it will become unable to function sufficiently (**decompensated** cirrhosis) and start to fail; this is sometimes referred to as 'end stage liver disease'. At this stage chemicals and waste products can build up in the body, commonly causing jaundice, ascites (a build-up of fluid in the abdomen) and hepatic encephalopathy (confusion and memory loss). In the final stages of liver disease the build-up of waste products may lead to multiple organ failure and loss of life.



What causes primary sclerosing cholangitis?

The cause of primary sclerosing cholangitis (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 and it can affect all ages.

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 and in others, symptoms do not appear for many years.

Symptoms include:

- fatigue
- some abdominal discomfort in the right upper abdomen
- itching

- occasionally jaundice – yellowing of the skin and whites of the eyes
- episodes of fever, shaking and chills.

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 MRI scan (MRCP).

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 or a hepatologist).

Liver blood tests

Liver blood tests, sometimes referred to as liver function tests (LFTs), are blood tests that are carried out to check how well the liver is functioning.

Ultrasound scan

The abdomen is examined by ultrasound annually 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. One method is a test called an endoscopic cholangiogram (ERCP). 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, after which dye (contrast media) is injected and X-rays are taken to produce a picture of the bile ducts.

A newer method is a non-invasive scan called magnetic resonance cholangiogram (MRCP) although the picture quality is not as good as ERCP. This means lying still inside a scanner, which some people may find a little claustrophobic.

Treatment

At present there is no known cure or specific treatment for PSC although symptoms such as itching can be treated with medicines 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 the very 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.

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 improving bile flow, 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, although some people remain unaffected. 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 dietitian.

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 dietitian 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 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 herbs.

If you are aged 40 or over, 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.

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

Useful words

Acute – a short sharp illness that may be severe but from which most people will recover in a few weeks without lasting effects.

Ascites – accumulation of fluid on the abdomen (peritoneal cavity) which surrounds the bowel, leading to enlarged, swollen and tender abdomen.

Bile – a yellow-green fluid produced by your liver to aid digestion. It contains chemicals as well as waste products and plays a central role in helping the body digest fat.

Bile ducts – the tubes linking your liver to your gut (also known as your intestine or bowel). The bile ducts carry bile from your liver to your gut.

Biliary – anything to do with the bile duct or bile.

Cholangitis – an inflammation of the bile ducts.

Chronic – an illness that lasts more than six months, possibly for the rest of a person's life.

Chron's disease – a long-term condition that causes inflammation of the lining of the digestive system.

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.

Colitis – inflammation of the lining of the colon.

Diet – the range of foods a person eats.

ERCP (Endoscopic retrograde cholangiopancreatography) – a procedure which gives detailed X-rays of the pancreatic and bile ducts, to show any abnormalities in the flow of your bile. Sometimes used to remove stones which may have passed into the bile duct.

Gastroenterologist – a doctor who specialised in problems of the digestive tract including throat, stomach, bowel (gut) and liver.

Hepatic – anything relating to the liver.

Hepatologist – a doctor who specialises in liver disease.

Inflammation – the first response of the immune system to infection, commonly characterised by heat, swelling, pain and tenderness.

Jaundice – a condition in which the whites of the eyes go yellow and in more severe cases the skin does too. This is caused by a rise of bilirubin (containing yellow pigment), a waste product which is normally disposed of by the liver.

Liver blood tests – a panel of tests used to indicate whether your liver is inflamed (hepatitis), damaged or not working properly. They measure levels of certain enzyme and protein substances in your blood that may alter when liver damage is present.

Oedema – a build-up of fluid in the body which causes the affected tissue to become swollen.

Osteoporosis – a condition that weakens bones, making them fragile and more likely to break. It develops slowly over several years and is often only diagnosed when a minor fall or sudden impact causes a bone fracture.

Ulcerative colitis – a condition where ulcers and inflammation form in the rectum and the colon (part of your bowels).

Virus – a microscopic particle that infects living cells by getting inside them and reproducing (replicating). Viruses cannot reproduce by themselves and can only multiply from within the cells of their living host.

Further information

Please refer to the Trust website for details of patient organisations and support groups specialising in specific liver conditions, that you may find helpful.

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:

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

Contact us for more information:

Tel: 01425 481320

Helpline: 0800 652 7330

Email: info@britishlivertrust.org.uk

Web: britishlivertrust.org.uk

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

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PSC Support, pscsupport.org.uk

We hope you have found this publication helpful

All our publications are reviewed by medical experts and people living with liver disease. If you have any feedback on this publication please email the Trust at **info@britishlivertrust.org.uk**

The British Liver Trust is proud to be recognised as a provider of expert liver health information, but to do this we must depend on the kind donations of our supporters. The Trust receives no government aid, yet strives to fill the growing need for liver health information in the UK.

We are a small charity, and your donation can make an important difference.

A gift of £5 could help us answer patient calls to our helpline

A gift of £20 could help us to set up a new patient support group

A gift of £50 could support the costs of a new patient guide or leaflet

Gifts can be made:

Online at **britishlivertrust.org.uk/donate**

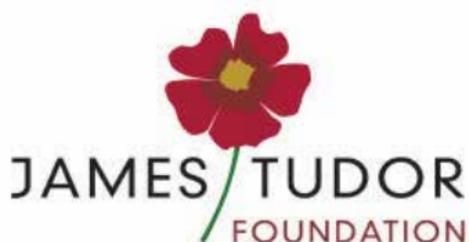
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If you have questions about making a donation, please call **01425 481320** or email **fundraising@britishlivertrust.org.uk**



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