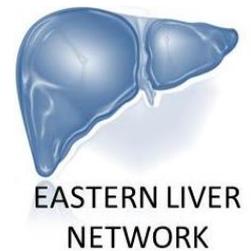


PRIMARY SCLEROSING CHOLANGITIS (PSC)



What is PSC?

PSC is a chronic autoimmune liver disease whereby the bile ducts, which are the passages that carry bile from the liver to the intestines, become narrowed from inflammation and scarring called fibrosis. Over time, the scar tissue builds up and can block parts of the bile ducts completely. The blockages can occur in any part of the biliary tree: inside the liver itself (intrahepatic) or outside the liver (extrahepatic). PSC can also lead to liver cancers and so requires careful monitoring by liver specialists at Addenbrooke's Hospital.

What causes PSC?

PSC is an auto inflammatory disease, in which the bile ducts become inflamed for no apparent reason. No one knows why this happens. Some people may have a higher genetic risk, and those with other autoimmune conditions may also be more likely to be affected by PSC. PSC more often occurs in men than women. The age at which it is diagnosed in most patients is between 25 and 60 years, though may occur at any age.

What are the symptoms of PSC?

PSC often causes no symptoms and is picked up because of abnormal liver blood tests. The symptoms that PSC can cause are tiredness, itching, yellow skin (jaundice) and fevers.

Is there a treatment for PSC?

PSC is a complex condition that affects each individual differently. There are no medications for PSC that significantly alter the outcome of the condition. The main treatment is to monitor patients for problems caused by it. You will have yearly outpatient appointments or more frequently if your disease requires it. You will need a liver ultrasound scan at least once a year. Those with inflammatory bowel disease may be recommended to have regular (usually yearly) colonoscopies. These tests are important because there is an increased risk of some cancers in patients with PSC.

Patients with PSC may be prescribed 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. Some patients with PSC may need a liver transplant.

What is bacterial cholangitis?

This is where the bile cannot drain properly and it builds up in the liver. The normally sterile bile can then become infected. When it is infected the condition is called bacterial cholangitis. Bacterial cholangitis can strike at any time, although it is less common in early stages of PSC.

Symptoms can be variable and should not be ignored.

Some PSC patients say it feels a bit like having flu and symptoms can include: itching, dark urine, temperature, shivers and chills*, fever* abdominal pain, jaundice, pale stools, nausea/vomiting and night sweats.

*Fever, shivers and chills may indicate a serious bacterial infection that needs urgent antibiotic treatment, although not all bacterial infections cause chills. Bacterial cholangitis in PSC may need medical attention even in the absence of fever or chills.

What should I do if I suspect bacterial cholangitis?

If you think you have bacterial cholangitis, urgent medical care is required so you will need to be assessed by A&E, your GP or hepatology doctor. You will be administered an antibiotic to cover likely causative bacteria. If left untreated, bacterial cholangitis can cause serious complications such as sepsis. If you are initially treated for cholangitis at A&E, we recommend that you ensure that your hepatology doctor is informed. Your doctor will consider

your individual circumstances, and in some cases, may also consider an MRI scan or endoscopy to investigate and/or help clear your bile ducts.

What research is happening in PSC

There are opportunities to be involved in clinical trials for new treatments of PSC. If you are interested in taking part in this research or would like to discuss it further, please contact Sister Abi Ford on 01223 256225 or e-mail hepatologyresearch@addenbrookes.nhs.uk

Where can I receive further information and support?

- The British Liver Trust: <http://www.britishlivertrust.org.uk> Helpline: 0800 652 7330 (10:00 to 15:00 Monday to Friday)
- UK PSC: www.uk-psc.com
- PSC support Group www.pscsupport.org.uk