PRIMARY SCLEROSING CHOLANGITIS (PSC)

EASTERN LIVER NETWORK

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.

What treatment will I receive for PSC?

PSC is a complex condition that affects each individual differently. There is no proven medication for PSC. The main treatment for PSC is to monitor patients for problems caused by it. Therefore, you will have an outpatient appointment every six to 12 months. You will have an ultrasound scan of the liver every six months. Regular ultrasound scans are important because people with PSC have increased risk of cancer of the gall-bladder, bile ducts or liver – and regular scans can help to detect these cancers at an early and treatable stage. People with PSC and inflammatory bowel disease (IBD) will have a colonoscopy every year. Annual colonoscopies are important because people with PSC and IBD have increased risk of colon cancer – and regular colonoscopies can help to detect colon cancer at an early and treatable stage. Some patients with PSC may be prescribed a medication called Ursodeoxycholic Acid (UDCA), given at a dose of 13-15mg/kg/day, but the effectiveness of this medication is unproven. It may be taken in divided doses or all at once. Some patients with PSC may eventually need a liver transplant.

We will do our best to keep the monitoring of your PSC up-to-date. We strongly encourage you to become familiar with the tests you need, however, to ensure these tests happen when they should.

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

Symptoms of PSC include itching, fatigue, and abdominal pain. People with more advanced PSC might have jaundice, dark urine, and pale stools.

People with PSC can experience a condition called bacterial cholangitis. Symptoms of bacterial cholangitis include fever, shivers, chills, abdominal pain, nausea, and vomiting. Bacterial cholangitis is a serious infection that needs urgent antibiotic treatment, so these symptoms should never be ignored.

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

Contacts

If you require any further information about this leaflet or advice about your condition please contact the Addenbrooke's autoimmune liver disease advice line on 01223 216109

Where can I receive further information and support?

- The British Liver Trust: 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