What is Primary Sclerosing Cholangitis (PSC)?

PSC is a rare, chronic disease that causes slow damage to the drainage system (bile ducts) of the liver. The immune system normally protects our bodies against disease and attacks the bugs that cause infections. Occasionally this system becomes inaccurate and starts to target and attack parts of the body instead. In PSC, the immune system attacks the liver. This leads to inflammation and gradual scarring/narrowing of the bile ducts. Due to changes in the bile ducts, bile (which is made by the liver and helps with food digestion) builds up in the liver. Over time, this progressive damage causes normal liver tissue to be replaced by scar tissue and leads, in some patients, to cirrhosis.

Cirrhosis: Does that mean PSC is caused by alcohol?

No. People often associate the term cirrhosis with excess alcohol. However, there are lots of other causes of cirrhosis, including PSC. Cirrhosis simply describes advanced liver scarring and does not explain what causes it.

What causes PSC?

The cause of PSC is currently unknown. It is known to be an autoimmune problem, due to a faulty immune system, and likely to occur as a result of genetic and environmental factors. Research is ongoing to better understand the cause of PSC. It is more common in males (2 males to 1 female) and is commonest around the age of 30-40 years, although it can occur at any age. Approximately 80% of patients with PSC also have inflammatory bowel disease, commonly ulcerative colitis.

How common is PSC?

PSC is a rare disease and only affects 6 per 100,000 people in the UK.

What problems does PSC cause?

In early disease, patients may have no symptoms but over time people can develop symptoms which can affect quality of life. These commonly include fatigue, itch (pruritus) and abdominal pain.

The narrowed, scarred bile ducts mean that drainage of bile from the liver is reduced. This can result in infection developing within the liver (cholangitis). Cholangitis classically presents with symptoms of abdominal pain, fever and jaundice (yellowing of the eyes and skin). Patients with these symptoms **should seek medical advice immediately**. However, patients with PSC often do not develop these classic symptoms and can feel generally unwell with cholangitis. If you feel unwell you should seek medical attention and it can be helpful to take this leaflet with you to your GP or A&E department.

The liver has a huge ability to repair itself, but over time, PSC can lead to progressive liver scarring and result in cirrhosis. Eventually, the liver becomes too damaged to be able to perform its usual functions. This can result in signs and symptoms of liver failure: jaundice (yellowing of the eyes and skin), ascites (development of fluid in the abdomen), bleeding (due to increased pressure in swollen veins, varices, in the gullet), clotting problems and encephalopathy (confusion).

How is PSC diagnosed?

The first test for anyone suspected to have liver disease are liver function tests (LFTs) but there is no diagnostic blood test. Doctors will also check for other causes of liver disease. Most patients will need imaging of the liver, initially with an ultrasound (jelly scan of the abdomen similar to that used in pregnancy) and usually with an MRI scan. The MRI provides detailed pictures of the drainage system of the liver to show the narrowed, scarred bile ducts. If PSC is suspected, but not confirmed on MRI, some people need a liver biopsy. This is usually performed under local anaesthetic and involves using a biopsy needle to take a tiny piece of the liver which can then be looked at under a microscope.

How is PSC treated?

At the moment there is no cure for PSC. Many patients have been given ursodeoxycholic acid (UDCA) which has been shown to improve LFTs in some patients but not proven to improve outcome. Patients remain under long-term follow-up and monitoring focusses on controlling the symptoms of PSC and checking for possible complications. There is research ongoing to try to find appropriate treatments to slow or stop the progression of liver disease.

Episodes of cholangitis will need treatment with antibiotics and sometimes a specialised camera test (ERCP) to improve the drainage of the liver.

Itch is treatable using a variety of oral medications.

Patients with cirrhosis are known to be at increased risk of osteoporosis so should be tested, and if appropriate, treated for this.

Inflammatory bowel disease is frequently seen in patients with PSC, affecting approximately 80% of patients. Ulcerative colitis is more common than Crohn's disease. All patients with PSC should be checked for colitis (inflammation of the bowel) using a camera test (colonoscopy).

Liver Transplantation

Some patients who develop advanced liver disease eventually need a liver transplant. As PSC progresses slowly, liver transplantation can be carefully planned and can result in a dramatic improvement in quality of life.

Monitoring for Complications

Patients with PSC are at increased risk of bile duct and gallbladder cancer compared with the general population. As a result, yearly ultrasound scans are

performed to examine the liver and gallbladder.

What can I do to help myself?

Medicines
In people with liver problems, such as PSC, the liver is sometimes less able to process medicines so it is important to tell doctors and pharmacists that you have PSC.

Diet
There is no specific diet that you need to follow. However, you should eat healthily and drink alcohol sensibly.

Activity
Fatigue (tiredness) sometimes means that people find exercising difficult. It is important to try to remain active but pace yourself.

Mood
It is understandable that sometimes having PSC may get you down. It is important to have people around you for support. Having a friend to talk through how you feel can be helpful. There are also sources of support from patient groups such as LIVErNORTH and PSC Support (www.pscsupport.org.uk).

LIVErNORTH is indebted to Dr Jessica K Dyson of the University of Newcastle upon Tyne & the Liver Unit, Newcastle upon Tyne Hospitals NHS Foundation Trust for her professional contribution to this leaflet.

LIVErNORTH can help
As well as being supported by many health professionals, we have access to extensive information on all liver diseases and treatments available to us - contact details on the back cover or via our website.

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LEAFLET 10

Primary Sclerosing Cholangitis

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