Primary sclerosing cholangitis

Susan Heggie Vice Chair, PSC Support discusses PSC and the work that PSC support does

Primary sclerosing cholangitis (PSC) is an uncommon, chronic, cholestatic liver disease caused by inflammation and fibrosis that can involve the entire biliary tree (Chapman, 2011). This progressive process destroys both intra and extra hepatic bile ducts, leading to biliary cirrhosis, portal hypertension and hepatic failure. The cause of PSC is unknown but it is closely associated with inflammatory bowel disease, particularly ulcerative colitis (UC), which occurs in approximately 70% of cases. Approximately 5–10% of patients with total UC will have co-existing PSC (Chapman, 2011).

PSC is often difficult to diagnose due to the complexity of its symptoms (European Association for the Study of the Liver, 2009). These symptoms include fatigue, intermittent jaundice, weight loss, right upper quadrant abdominal pain and pruritus. Cholangitis, sporadic episodes of inflammation within the biliary tract, often bacterial in nature cause pain, shivers, fever and distress and require immediate antibiotic therapy. Occasionally, more invasive treatment is needed.

The clinical course of PSC is variable and there is no curative treatment apart from liver transplant; medical treatment has proven unsuccessful. Fosleraas et al (2011) indicated that no treatment has confirmed any delay in progression, but studies are often conflicting (Chapman, 2009; Lindor et al, 2009). Barnabas and Chapman (2012) debated the use of ursodeoxycholic acid for PSC confirming that its role remains unclear. They found that a high dose (28–30 mg/kg/day) may be harmful, while a low-to-moderate dose may have a carcinogenic protective effect.

As diagnostic techniques are advancing rapidly, PSC is being identified earlier (Molodecky et al, 2011). Research is ongoing to try to find a treatment, cure or genetic link for this often debilitating condition. Fosleraas et al (2011) state that genome wide studies have consistently identified genetic susceptibility and environmental factors—the relation of these hopefully offer exciting opportunities for transferring knowledge and impending treatment options. UK genome-wide studies are ongoing and recruitment is continuing (UK PSC Genome Study, 2011).

The unpredictability of the condition is daunting and the uncertainty of how it will effect day to day life and future plans can bring major concerns and anxiety. Many people living with PSC describe it as very challenging: the itch and pain in particular causing daily stress which can reduce quality of life (Ponsioen, 2011). A decompensated liver can bring about many hurdles. For some, the addition of other related autoimmune conditions and immunosuppression due to prescribed therapy can be immense. As with any condition, an active healthy lifestyle is required. Good nutrition is vital for patients with a liver condition, even more important for people with associated inflammatory bowel disease. Access to dietary expertise is often limited; however, dieticians Leaper and Hamlin (2012) and the British Liver Trust (2011) produced excellent information and advice for people with various liver conditions and degrees of liver failure.

Nurses are pivotal to delivering efficient services and quality care. Gastroenterology and hepatology nurses are ideally placed to identify patients and/or carers of people who have PSC and offer them guidance on their condition, treatment (both pharmacological and non pharmacological) and strategies on how to cope and live with PSC.

PSC Support is a UK based charity that helps people affected by PSC both nationally and internationally. Run entirely by volunteers, they aim to:

- Provide information and support to those affected by PSC and the medical community
- Promote awareness of PSC, PSC Support and organ donation
- Develop effective partnerships with those involved in the treatment of and research into PSC.


Further information
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