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PSC. Primary Sclerosing Cholangitis

NIHR BioResource – Rare Diseases study project

Lead Investigator: Dr Palak Trivedi

V1.1 20/05/2019

Dr Palak Trivedi, PSC project lead

Summary

Primary sclerosing cholangitis (PSC) is a condition leading to inflammation and fibrosis (scarring) within the bile ducts (the tubes draining bile from the liver into the bowel). As a result, multiple narrowings called 'strictures' develop throughout the bile ducts; leading to recurrent infections as a result of sluggish bile flow, jaundice, and eventually cirrhosis and liver failure.

At present, liver transplantation is the only lifesaving treatment for patients with PSC, however, the rate at which disease progresses to the point of needing a liver transplant varies immensely from one person to the next. Although rare (affecting less than 1 person per 100,000), PSC now accounts for 10-15% of all liver transplants performed within the UK and is the lead indication for transplantation in many European countries. Unfortunately, disease returns in approximately one-third of patients, and can lead to loss of the transplanted liver.

In addition, PSC significantly increases the risk of bile duct cancer (cholangiocarcinoma), which is often incurable when diagnosed. The majority of patients who have PSC also develop inflammatory bowel disease (IBD) at some point. The coexistence of both conditions is important to recognise, as this increases the lifetime risk of developing bowel cancer up to ~30%, which is more than ten-fold greater to that of IBD alone.

We want to collect more information about the condition to help

- Improve our understanding of the factors influencing disease progression.
- Help identify biological pathways for treatment with PSC.
- Identify sub-groups of patients who are at risk of the disease returning after liver transplantation.

Patients (adult) with PSC are also being recruited to the UK-PSC national platform study (http://www.uk-psc.com/) and it is hoped that patients will be recruited to both UK-PSC and the NIHR BioResource – Rare Diseases study. Children can be recruited to the PSC project as part of NIHR-BioResource – Rare Diseases study.

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Recruitment Criteria

Inclusion

Adults (16+ years)

Patients who are diagnosed with and managed as having PSC or autoimmune sclerosing cholangitis.
Diagnosis confirmed via MRCP and/or ERCP or liver biopsy.
Patients that have received an orthotopic liver transplant for PSC are also eligible.

MRCP; magnetic resonance cholangio-pancreatography. ECRP; endoscopic retrograde cholangio-pancreatography

Children (0 – 15 years)

Patients with a confirmed diagnosis of:

- Primary Sclerosing Cholangitis or
- Autoimmune Sclerosing Cholangitis or
- Autoimmune Hepatitis with possible biliary disease or
- Autoimmune Hepatitis with no biliary disease at time of recruitment

Parents or guardians and/or children must have been informed of the child's diagnosis.

The above criteria have been set because of the difficulty in obtaining a confirmed PSC diagnosis in children.

Exclusion

Sclerosing cholangitis resulting from a secondary cause, e.g. congenital biliary tree abnormalities, previous biliary surgery, bile duct carcinoma, HIV cholangiopathy, Primary Biliary Cholangitis, sarcoid, graft-versus-host disease, sickle cell disease and immunodeficiency related cholangiopathy and drug reactions.

Collaborating investigators recruiting to the PSC project should ensure that secondary causes of an abnormal cholangiogram have been excluded.