ERN RARE-LIVER

European Reference Network on Hepatological Diseases

Primary Sclerosing Cholangitis (PSC)

What is PSC?

PSC is a rare, immune-mediated liver disease that leads to scarring (sclerosing) following inflammation in the bile ducts (cholangitis). This means that bile, which normally flows through the bile ducts into the intestines, can't drain properly and it builds up in the liver. This can lead to infection and further liver damage in some people. See Figure 1.

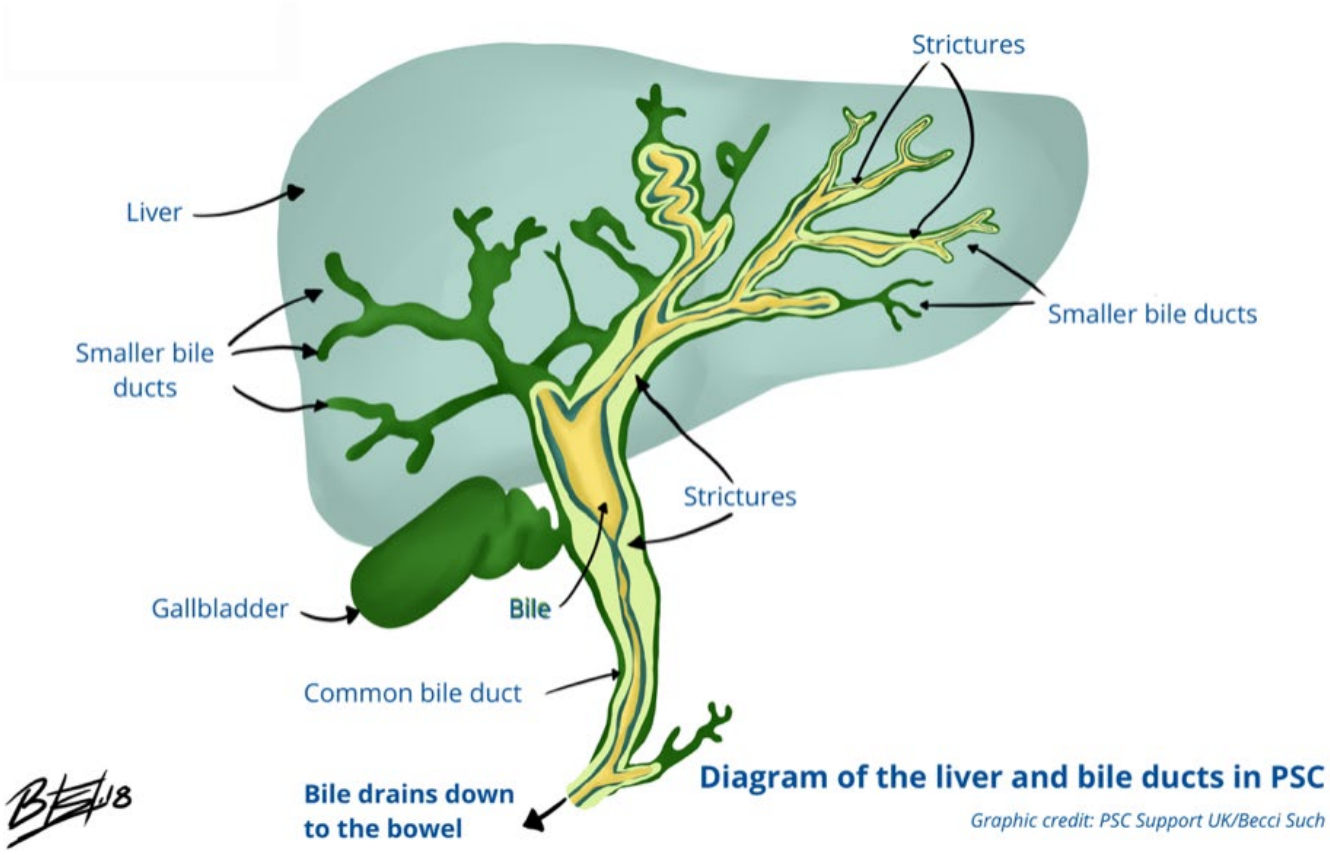

Diagram of the liver and bile ducts in PSC

Graphic credit: PSC Support UK/Becci Such

Figure 1. Diagram of liver and bile ducts in PSC

Bile ducts are tiny tubes that drain bile from the liver into the bowel. It helps to think of bile ducts as a system of tubes arranged like a tree, in which the bile drains down from the smallest bile ducts at the top (twigs), down through the large bile ducts (branches) and eventually out through the trunk (common bile duct).

In PSC, the immune system 'attacks' the bile ducts, causing hardening and narrowing (strictures) of the ducts. In PSC, both the small and large bile ducts can develop strictures, making it difficult for the bile to drain out smoothly.

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Who is affected?

PSC can be diagnosed at any age. It is most commonly diagnosed between 30 and 50 years of age but children can be affected. It affects slightly more males than females. Most people with PSC also have one or more immune-mediated diseases, in particular, inflammatory bowel disease (IBD).

Why do I have PSC?

PSC is complex and we don't fully understand what causes it. Current evidence suggests that people with a particular genetic makeup may be susceptible to some sort of environmental trigger, which causes their immune system to effectively 'attack' the bile ducts. We don't yet know what that trigger is, but it could be bacteria or a virus. See figure 2.

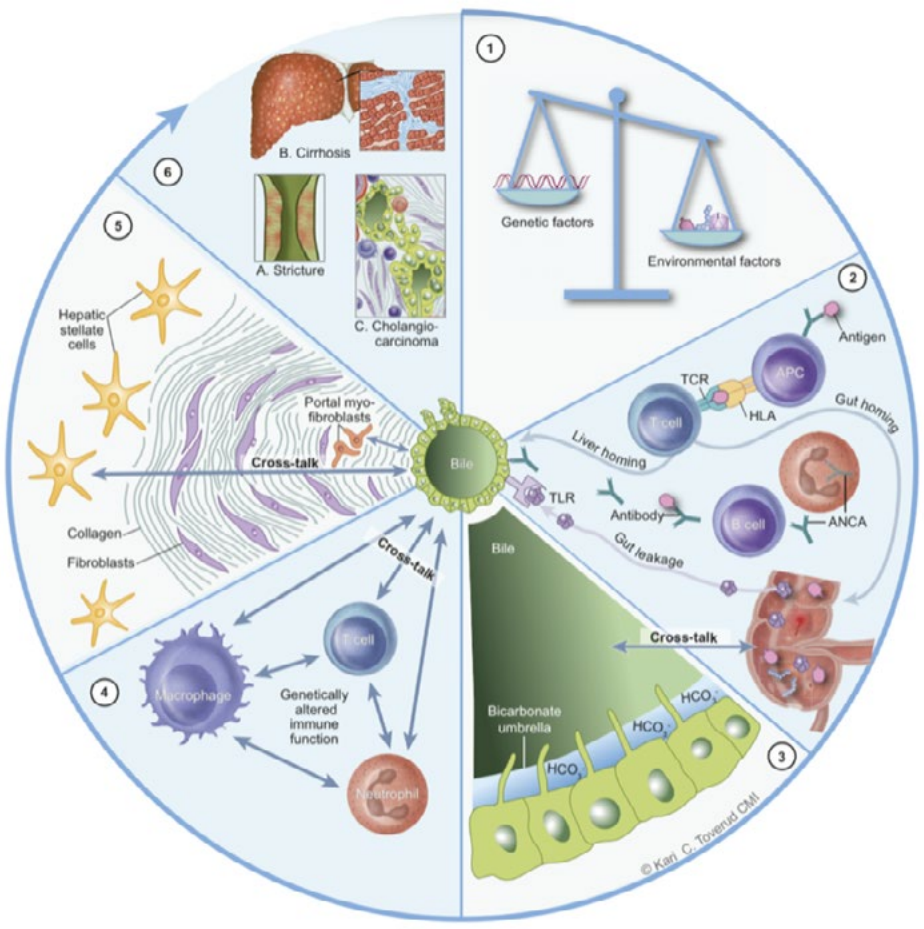

Figure 2. Factors involved in the development and presentation of PSC

1 Genetic predisposition and environmental factors
2 Immune cells activated in the gut appear in the liver
3 Loss of protective mechanism against toxic bile components
4 Altered immune system activity leads to inflammation around bile ducts
5 Injury of bile duct cells leads to scarring
6 Complications such as strictures in the bile ducts, accumulation of fibrotic tissue (cirrhosis) and an increased risk of cancer.

Thanks to Professor Tom Hemming Karlsen, Oslo University Hospital, for permission to reproduce this graphic.

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How is PSC diagnosed?

PSC can be difficult to diagnose. The diagnosis often comes unexpectedly after routine blood tests for IBD suggest bile duct injury. In other cases, people suffer with fatigue or feel generally unwell, or may show signs of liver disease such as itch or jaundice (yellowing of the eyes/skin), which triggers investigations to identify the cause.

If your doctor suspects PSC, you will be asked to have a type of MRI scan called an MRCP scan. An MRCP looks closely at your bile ducts.

Where the MRCP shows a typical appearance for PSC, invasive tests are rarely needed. However, occasionally a biopsy is taken when other conditions including associated autoimmune hepatitis are suspected. Only in rare cases is an endoscopic cholangiography (ERCP) required to establish the diagnosis.

What can my doctor do?

While there is currently no curative treatment for PSC, your doctor can help manage your symptoms using current clinical practice guidelines. This includes medication to treat itch, antibiotics to treat infections and vitamin supplements.

Some people with PSC take ursodeoxycholic acid (UDCA) at low dose (15 to 20 mg per kg body weight per day). UDCA improves liver blood tests and may help improve the flow of bile, but we do not have scientific evidence that it delays progression of PSC.

People with PSC face an increased risk of some gastrointestinal cancers, such as colon cancer and bile duct cancer. Your doctor should keep a close eye on you and monitor you regularly.

New therapies for PSC are currently being tested. Your doctor can talk to you about opportunities to take part in research trials to access potential new drugs.

What tests or procedures will I have?

An ultrasound scan every 6-12 months to look at the shape of your liver, bile ducts and gallbladder.

A colonoscopy at diagnosis to check for the presence of IBD, even if you have no symptoms. If you do have IBD, expect to have yearly colonoscopies.

A bone density scan to check your bones, as PSC is associated with an increased risk of osteoporosis.

Endoscopic treatment (ERCP) can sometimes be used to physically widen narrowed bile ducts to help the bile flow.

Regular liver blood tests to monitor your liver enzymes.

A FibroScan® every year (if available) to look at scarring in the liver.

An MRI scan of your bile ducts (MRCP) if you have new or changing symptoms or changes are seen in other tests.

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Do I need a special diet?

There is no special diet for PSC. We recommend that you eat a healthy, well-balanced diet. Avoid supplements without first checking with your doctor.

Can I drink alcohol?

PSC is not associated with drinking alcohol. However, most doctors suggest that if you have PSC, you should not drink alcohol, as it can accelerate pre-existing liver damage. You will be given alcohol advice by your doctor and this will vary depending on the severity of your disease and your general state of health.

Can I smoke?

It is highly recommended that you do not smoke. Cigarette smoking is harmful to your health, even at low levels of consumption.

Can I have a family?

Generally speaking, if your liver disease is not advanced, the risk of complications in pregnancy is no different to healthy individuals. Do speak to your doctor if you are planning a family, especially if you have cirrhosis.

What will happen to me?

It is important to remember that PSC varies from patient to patient. Many people with PSC go on to live normal lives. Others may be affected by symptoms such as fatigue, itch and abdominal pain, and some, but not all, progress to advanced liver disease. For those patients, liver transplantation is sometimes required.

If you notice a sudden change in your condition, you should let your doctor know.

Acute cholangitis

What is acute cholangitis?

Acute cholangitis (also called 'bacterial cholangitis' or a 'cholangitis attack' or a 'flare-up') may occur in people with PSC, even in those with otherwise good liver function. The exact cause is not known but strictures are thought to facilitate the colonialization of bile ducts by bacteria leading to infection of the bile ducts within the liver. Having a bout of acute cholangitis does not mean that your PSC is progressing.

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Am I having a cholangitis attack?

Occasionally PSC patients experience bacterial infections of the bile ducts (acute cholangitis) which require urgent medical attention. They can present with a broad range of symptoms and the classic features of infection can be absent. This means cholangitis attacks can be difficult to recognise.

Signs and symptoms can vary from person to person and include but are not limited to:

- new or worsening pain in the right upper part of your abdomen
- reduced appetite/ nausea
- new or worsening itch (particularly on your palms or soles of your feet)
- jaundice (yellowing of the skin and/or the whites of the eyes)*
- fever, shivers, chills*
- a high temperature*
- darkening of your urine*
- pale stools*
- sudden change in liver blood test results*

*any of these signs or symptoms may indicate a severe cholangitis attack

Do I need medical attention?

Yes. If you think you are having a cholangitis attack, urgent medical care is required. Contact your GP or out of hours medical provider for an urgent appointment about your suspected cholangitis attack (bile duct infection). Treatment may be required even if you don't have the severe signs and symptoms.

IF IN DOUBT, SEEK MEDICAL ATTENTION

If any of the following signs or symptoms of severe cholangitis are present, then it is advised to attend your local accident and emergency department immediately:

- sudden onset of jaundice
- severe pain in the right upper part of your abdomen
- unmanageable fever (for instance, not controlled with paracetamol)
- chest pain
- loss of consciousness

If you have severe cholangitis, you may need to be treated and monitored in hospital. Immediate treatment with antibiotics is important and should be guided by your doctor, taking into account severity of disease, local antibiotic resistance and potential side effects.

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Next steps

We recommend that you ensure that your PSC doctor knows about the cholangitis attack as soon as possible. They 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.

Vaccinations

Vaccinations against hepatitis A and B (viruses) and pneumococcal infections (bacteria) are recommended. We recommend vaccination against influenza as early as possible each autumn.

How can I find a PSC doctor?

PSC is a complex condition that affects each individual differently. Ideally your care should be managed by a doctor or a hospital network with experience or an interest in PSC. PSC is a rare disease and not every doctor has an interest in or experience of PSC. However, some hospitals are part of the Europe-wide network for rare liver diseases, ERN RARE-LIVER. This means that patients seen at hospitals within the network can benefit from the expertise of specialists who work at other hospitals within the network. For more information about the ERN RARE-LIVER, go to the link at the top of the page.

Research

A number of potential treatments for PSC are being tested in clinical trials across Europe. To find out more, and to find out if you could be suitable (eligible) to take part, speak to your doctor or go to Clinicaltrials.gov and search for 'primary sclerosing cholangitis'. Here you can view an interactive map displaying research trials that are recruiting volunteers.

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