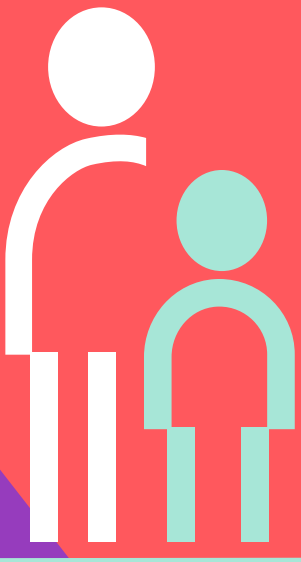
LIVER UK

The new name for the British Liver Trust and Children's Liver Disease Foundation

Primary sclerosing cholangitis in children



Help, support and guidance for children and families

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Welcome

Welcome to your guide to understanding PSC. It provides information on causes, symptoms, diagnosis and treatment.

This information has been written for:

Parents and carers of children and young people with PSC.

Others who may also find this information useful:

- young people with PSC
- healthcare professionals

You may also find it helpful to read the following booklets:

- An introduction to liver disease in children
- Portal hypertension and ascites
- Pruritus

Use this QR code to view our resources or visit **www.liveruk.org**

What is primary sclerosing cholangitis?

Primary sclerosing cholangitis (PSC) is a rare, long-term liver disease that causes inflammation and scarring of the bile ducts, both inside and outside the liver. Over time, the bile ducts can become narrow or blocked, making it difficult for bile to flow normally.

In a healthy liver, bile is made by liver cells and transported through the bile ducts to the gallbladder. When you eat, the gallbladder contracts and bile flows into the intestine. This allows toxins to be removed from the body in the poo (stool). Bile also helps the body absorb fats and essential vitamins (vitamins A, D, E and K).

In PSC, the bile ducts become progressively scarred and narrow so that bile is unable to flow normally. This causes harmful substances to build up in the body over time, which can damage the bile ducts and liver.

PSC is a rare condition affecting around 1.5 in every 100,000 children. PSC may occur alone but is more common in children with inflammatory bowel disease (IBD), affecting at least 10% of those with ulcerative colitis.



What causes PSC?

The cause of PSC is unknown, but researchers think there may be genetic and environmental factors involved.

Current research suggests that PSC may be associated with an immune reaction against the liver generated by changes in the gut (an autoimmune condition). This leads to inflammation causing scarring and damage to the bile ducts.

Other research suggests that there may be an imbalance in the microorganisms or bacteria (gut flora) in the bowel.



What are the signs and symptoms of PSC?

Some children have PSC for years before they develop symptoms. In some children it is found on routine blood tests, during tests for inflammatory bowel disease (IBD), or due to symptoms such as tiredness (fatigue).

PSC affects each child differently. Most children have no symptoms if the disease is mild. But as the disease progresses, symptoms may become more noticeable and can include:

- Yellowing of the skin and eyes (jaundice)
- Itching (pruritus)
- Dark urine and pale stools
- Infection of the bile ducts (cholangitis)
- Tiredness and reduced energy
- Loss of appetite
- Weight loss
- Abdominal pain
- A high temperature (in rare cases).



How is PSC diagnosed?

If your child displays symptoms of PSC, there are a number of tests which may be used to confirm the diagnosis:

Family history

There may be other people in the family with autoimmune diseases (such as diabetes, thyroid disease, coeliac disease or IBD).

Blood tests

Blood tests may be repeated over several days or weeks to look for changes that could suggest damage to the bile ducts. Blood tests for autoantibodies may show a positive p-ANCA result. Autoantibodies can also be found in some other autoimmune diseases.

Blood tests can also show how well the liver is working and if it has been affected by the damage. This may include tests that check how well the blood clots (such as the INR test) and tests to measure the level of a protein called albumin, which is made by the liver. Although these results may suggest PSC, blood tests alone cannot confirm the diagnosis. Further investigations may be needed.

Ultrasound scan

An ultrasound is often a key diagnostic tool in liver disease. It may show an enlarged gallbladder, areas where the bile ducts have become wider than normal, or areas of narrowing.

An ultrasound can also show changes associated with advanced liver disease, such as fluid in the abdomen (ascites) and an enlarged spleen.

Magnetic resonance cholangio-pancreatogram (MRCP scan)

This is a specialist magnetic resonance imaging (MRI) scan of the bile ducts. It can show changes in the bile ducts inside the liver by using a special dye to identify these areas on the scan. This is better at diagnosing PSC than an ultrasound.

Liver biopsy

A liver biopsy may be useful if the diagnosis is not clear from the other tests. A biopsy can show scarring around the bile ducts and bile duct damage. The biopsy may also show how much scarring there is in the liver (fibrosis). Advanced fibrosis is known as cirrhosis.

Further information about the signs of liver disease, a glossary of terms and routine investigations can be found in the Liver UK leaflet "An introduction to liver disease in children".



Can PSC be treated?

There is currently no cure for PSC. Treatment focuses on supporting liver health, helping bile flow more easily through the bile ducts and managing symptoms.

One of these symptoms may be pruritus. This is an intense itch beneath the skin that can cause severe discomfort and can be difficult to control. The cause is not completely understood but is thought to be due to increased levels of bile acids in the blood. If your child experiences itching, the medical team will discuss ways to manage it and will recommend treatments to help relieve any symptoms.

In children where PSC is associated with IBD, improvement of the bowel disease may also improve the liver.

Medicines

- **Ursodeoxycholic acid (UDCA)**
  This can improve bile flow out of the liver and may reduce jaundice and itching.
- **Antibiotics**
  If an infection has occurred in the bile ducts (cholangitis), antibiotics are used to reduce the risk of the infection happening again.

Medication and treatment options will vary and your child's medical team will discuss the most appropriate options available.

As with other chronic liver diseases, medicines such as aspirin and ibuprofen should be avoided as they can make bleeding in the gut more likely. Paracetamol (Calpol) is a safer alternative.

Vitamins

Your child may need extra vitamins because reduced bile flow can make it harder for the body to absorb vitamins A, D, E and K from food. These vitamins are usually given by mouth, but injections may sometimes be needed.

Diet

In advanced liver disease, fluid can sometimes build up in the abdomen (tummy), causing swelling. If this happens, your child may need to follow a diet that is lower in salt to help reduce fluid build-up. Medicines called diuretics may also be prescribed to help the body get rid of excess fluid. Your dietitian will provide advice and support if any changes to your child's diet are needed.

Endoscopic procedures

In a small number of children, endoscopic procedures may be needed. These include:

- colonoscopies for children with IBD.
- gastroscopies to look for dilated blood vessels (varices) in children with portal hypertension.
- endoscopic retrograde cholangiopancreatography (ERCP) in children where an MRI scan does not provide enough information.

An ERCP is a procedure that is often used to treat blockages in the bile duct. During the procedure, a small plastic or metal tube, called a stent, may be placed in the bile duct to help keep it open. This allows bile to flow more easily.

ERCPs are rarely needed in children but are more commonly performed in adolescents.

What happens if the disease progresses?

If the disease progresses, your child may develop some of the following complications:

- Biliary strictures - narrowing of a bile duct from scar tissue. A stricture can block the release of bile and lead to infection (cholangitis). An ERCP procedure may be needed.
- Itching (pruritus)
- Portal hypertension - high blood pressure in the portal vein which may occur due to scarring of the liver or a blockage in the portal vein. The spleen may become enlarged (splenomegaly) due to increased pressure in the blood vessels around the liver. This can cause your child's tummy to become swollen.
- Varices - swollen veins in the intestine, oesophagus and stomach which can bleed if not treated. An endoscopy is usually required.
- Ascites - a build-up of fluid in the abdomen. It can be treated with medicines called diuretics. These help your child pass more urine, reducing the amount of excess fluid in the abdomen.
- Weight loss
- Development of bile duct cancer (cholangiocarcinoma). This is extremely rare in children.

Use this QR code to view our booklet on portal hypertension and ascites or visit **www.liveruk.org/portal-hypertension-in-children**

The role of liver transplantation

A liver transplant may be discussed as a treatment option if the liver begins to fail or if there is severe recurrent cholangitis. If a transplant is the best treatment option, the medical team will focus on preventing complications and treating symptoms while waiting for an organ.

Outcomes after transplantation for PSC are positive and many children go on to have a good quality of life. However, there is a chance that PSC can reoccur in the transplanted liver. The underlying reasons for this are not yet fully understood. Risk factors taken from studies of adult patients include a younger age at transplant and the co-existence of IBD.

Use this QR code to view our leaflet on liver transplantation or visit **www.liveruk.org/liver-transplant-children**



Are there any restrictions on lifestyle?

Many children with PSC can live a relatively normal life.

However, severe itching may disturb sleep and can contribute to fatigue, tiredness and irritability. Reduced appetite, nausea and vomiting may also impact your child's daily life.

There is a Liver UK leaflet called "Pruritus" that has further information and tips from parents on how to manage this.

If your child experiences fatigue (extreme tiredness), this may affect their ability to take part in everyday activities and may also impact their school life. It is helpful to speak with your child's school about their needs so that appropriate support can be put in place.

If your child has an enlarged spleen, it is advised that contact sports should be avoided due to the risk of bleeding. Please talk to your medical team about this as it depends on the individual.

Liver UK has an Education Pack to help families discuss the needs of their child with schools. Contact our Family Support Team for further information at **families@liveruk.org** or by calling **0800 652 7330**.

Living with a chronic illness and dealing with the side effects of treatment can be distressing. Specialist psychology teams at liver centres may be able to provide help and support. The Liver UK Support Team can also signpost and refer you to the most appropriate services

Use this QR code to view our leaflet on pruritus or visit **www.liveruk.org/pruritus**

What happens in the future?

Your child's growth and development will be monitored during regular hospital appointments. How often your child needs to attend will depend on their individual needs and how their condition is progressing. Appointments may be more frequent if there are signs that the disease is active or getting worse.

Liver UK is in touch with many families who have a child with PSC. PSC does not have to hold your child back from reaching their full potential. You can contact Liver UK for further support.

You may also benefit from contacting PSC Support at **www.pscsupport.org.uk**. They are a UK based charity and although they work more widely with adults, they are a good source of safe information and have useful videos and events specifically for patients with PSC.

Is research taking place?

Because PSC usually progresses slowly, it can be challenging to carry out clinical trials to develop new treatments. However, research is ongoing, with international clinical trials involving children taking place to help improve our understanding of the condition and find new treatments.

Several promising treatments are also being investigated, with encouraging results from studies involving adults with PSC. In the future, treatment may be increasingly tailored to each individual child's needs.



Notes



Useful resources



Useful websites

Liver UK
www.liveruk.org
Information and support for everyone living with a liver condition.

Medicines for children: advice and guides
www.medicinesforchildren.org.uk/advice-guides
A website with general advice on medicines in children.

How Liver UK can help

A diagnosis of liver disease can be worrying, and you may have a lot of questions.

The Liver UK Children's and Families Team are here for you and for your family and friends.

Whether you have questions or just need someone to listen, we can help.

Find out how we can help you:
www.liveruk.org/support

Call our helpline to talk to one of our team: **0800 652 7330**

How we produce information

Feedback and information services

Your feedback is important to us and will help us improve our information. To provide feedback, email **patient-info@liveruk.org**

For more information on how our information was developed, visit our website: **www.liveruk.org**

Thanks

All our information resources are reviewed by medical experts and families.

We would like to thank all our clinical experts and parent and family reviewers.



Disclaimer

This resource provides general information but does not replace personal medical advice. It is important to contact your medical team if you have any worries or concerns.

LIVER UK

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To make a gift in support of our work today, please scan the QR code or visit

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Liver UK is the UK's leading charity dedicated to supporting adults, children and young people affected by liver disease or liver cancer. Our mission is to transform liver health by raising awareness, promoting prevention, and improving care and support for everyone affected.

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