

# Primary sclerosing cholangitis (PSC)

Information for children

## Who is this leaflet for?

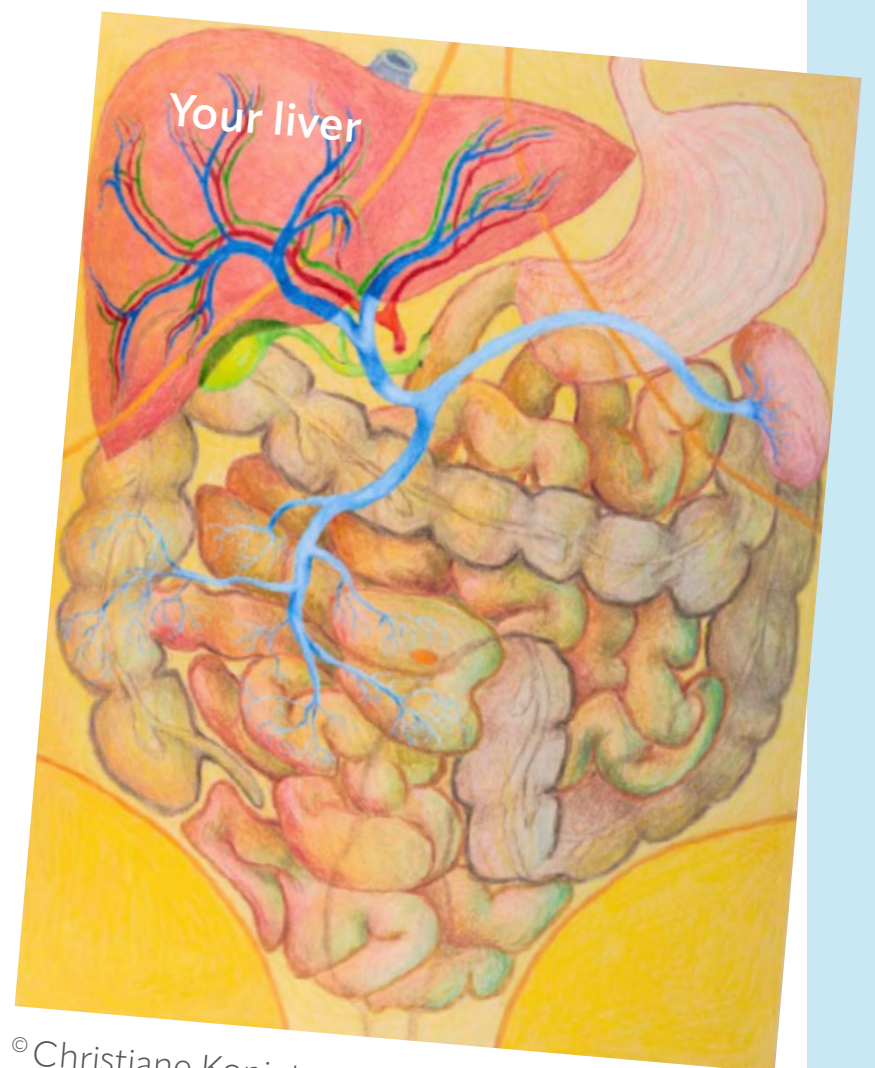
For children with PSC (around 10-15 years).

Please check out the ERN RARE-LIVER general PSC leaflet, which gives you more information about the disease: <https://rare-liver.eu> and



## What is primary sclerosing cholangitis?

- When somebody has primary sclerosing cholangitis (we call it PSC for short), their liver is diseased. Everyone has a liver. Your liver is an organ in the tummy, just under your ribcage on the right-hand side. It is a reddish-brown colour and quite large. Your liver is a very important organ and works very hard to keep you healthy.
- In fact, it has more than 500 jobs! For example, your liver helps to turn the food you eat into energy and other useful products that your body can use or store for later. It also cleans the blood which helps protect the body from too many waste chemicals building up which could make you feel unwell.
- Inside your liver are lots of thin tubes called bile ducts. These many small ducts (similar to a tree) fuse to a big main duct which transports the bile, a greenish-yellow liquid, to the gut. When someone has PSC, the bile doesn't flow very well and gets stuck. In your disease these ducts develop narrowing and the bile accumulates. The bile-accumulation causes problems which is why people with PSC sometimes feel poorly.



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### Why do I have PSC?

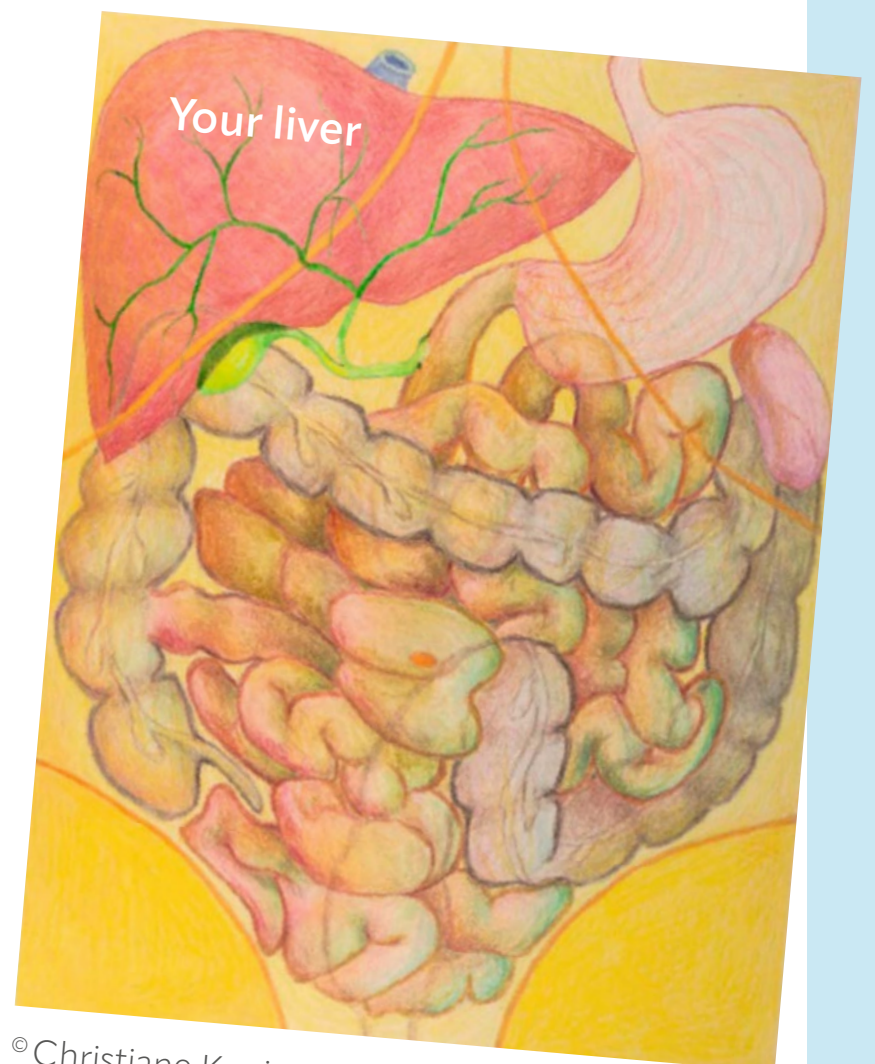
- People with PSC have not done anything wrong which has caused them to become unwell.
- The reasons why the bile ducts develop the narrowing are not clear. Inflammation and your immune system may partly be responsible. Normally, your "body police" (immune system) helps the body to fight off germs that cause you to become unwell. When you have PSC your immune system is hyperactive and mistakenly treats your bile ducts like they would germs and keeps trying to fight them off. This leads to the bile ducts and liver not working properly and can make you feel unwell.

### Can you catch PSC from someone, or is it infectious?

- PSC isn't something which you catch. It is caused by a faulty immune system.
- PSC is not a contagious disease. Some people associate yellow skin with diseases which are caused by infections and can be transmitted from one person to another person. But this is absolutely not the case for your disease! This means that people with PSC have not done anything wrong which has caused them to become unwell.

### How will my doctor help?

- There isn't a medicine that gets rid of PSC, but there are some ways which doctors can help people with PSC and how they feel. These might be medicines, or in some cases they might need an endoscopy or operation.
- Compared to PSC in adults, the signs and symptoms of PSC in children and young people tend to be milder. However, PSC in younger people can involve more inflammation in the liver and a stronger autoimmune response than seen in adults. For this reason, medicines that dampen the immune system can be a good way to treat PSC in children and adolescents. Liver biopsies are critical when taking this approach. A liver biopsy is a procedure where a tiny slice of the liver is taken and evaluated under the microscope.



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- Your doctor will do blood tests to check how well things are working inside your body and to check your vitamin levels. This does not mean that anything is wrong – everyone with PSC has regular blood tests.
- In children sometimes medication which suppresses your “body-police” (immune system) or antibiotics can help.
- Sometimes fat soluble vitamin supplements (vitamin A, E, D, K) are recommended.

### What can be symptoms of PSC?

- Not everyone experiences symptoms but for those who do, fatigue, itching and jaundice are most commonly reported. Jaundice is where your skin and whites of the eyes have a yellow tinge. Always let your liver doctor know if you start to experience new symptoms. Your liver doctor will want to know and will help you manage them.
- The itching can be a really annoying symptom. Don't downplay it, especially if it is disturbing your sleep. Talk to your parents and your doctor about it.
- Let's talk about poo! Many people with PSC also have a bowel condition called inflammatory bowel disease (IBD). This means that your doctor will routinely ask you about abdominal (tummy) pain and your poo. You may also need to have a special procedure in hospital called a colonoscopy (where a camera goes in your bottom to examine your large bowel). Don't be shy or embarrassed to talk about diarrhoea and other symptoms. It is very important that your IBD is well-controlled because untreated IBD can have a negative impact on your liver and day-to-day life. Luckily, there are many effective treatment options to manage IBD symptoms so it is always best to talk about them openly with your doctor.

### Can I go to school and do sports?

- Yes, you can do everything your friends and classmates do.
- With PSC in general you can play any sport you want. There are no restrictions, unless your spleen is enlarged. If this is the case, then your doctor will guide you about activities you can safely do.

### Do I have to follow a special diet?

- No, you can eat everything you want and this should include healthy food 😊 like for all children.