CONDITION

Sclerosing cholangitis

Sclerosing cholangitis is inflammation and scarring that narrows the tubes carrying bile through and outside the liver.

WHAT IT IS
Progressive inflammation and scarring of the bile ducts

BODY SYSTEM
Liver and digestive system

MAIN SYMPTOMS
Fatigue, itching, jaundice, abdominal discomfort

WHAT CAUSES IT
Usually unknown; sometimes another injury or immune condition

HOW IT IS DIAGNOSED
MRCP bile-duct imaging, sometimes liver biopsy or ERCP

MAIN TREATMENT
Monitoring, symptom relief, duct procedures, and sometimes liver transplant

CAN IT BE CURED?
Sometimes - transplant can replace a severely damaged liver

IS IT AN EMERGENCY?
Sometimes - fever with jaundice may signal bile-duct infection

LONG-TERM OUTLOOK
Often progressive, with unpredictable timing and severity

Overview

Sclerosing cholangitis is a condition in which the bile ducts become inflamed, scarred, and narrowed. Bile ducts are small tubes that carry bile from the liver to the small intestine. Bile helps your body digest fats and remove certain waste products.

Narrowing can slow or block bile flow. This can lead to repeated infections, liver inflammation, and eventually permanent liver scarring called cirrhosis. The condition can affect ducts inside the liver, outside it, or both.

There are several forms. Primary sclerosing cholangitis , usually shortened to PSC, has no single known cause. It is strongly associated with inflammatory bowel disease, especially ulcerative colitis. Secondary sclerosing cholangitis happens after a known problem damages the ducts, such as reduced blood flow, severe infection, surgery, or long-standing blockage.

IgG4-related sclerosing cholangitis is linked to an immune condition and may respond differently to treatment.

Some people have mild changes for years. Others develop frequent infections or worsening liver damage. Regular follow-up matters even when you feel well because blood tests or scans can show changes before symptoms appear.

Symptoms

Sclerosing cholangitis may cause no symptoms at first. It is sometimes found after routine blood tests show abnormal liver results.

When symptoms occur, they may include:

Itching can range from mild to severe and may disturb sleep. Jaundice develops when bilirubin, a yellow waste substance normally carried in bile, builds up in the blood.

Symptoms do not always show how much liver damage is present. A person with few symptoms can still have important duct narrowing. New symptoms also do not automatically mean the condition has reached an advanced stage; infection, gallstones, or a treatable narrowing may be responsible.

- Persistent tiredness
- Itchy skin
- Yellow skin or eyes, called jaundice
- Discomfort under the right ribs
- Dark urine or pale stools
- Unplanned weight loss
- Fever or chills during a bile-duct infection

Causes and risk factors

The cause of primary sclerosing cholangitis is not fully understood. Genes, the immune system, bile-duct cells, and bacteria in the gut may all play a part. PSC is not caused by something you ate, and it is not contagious.

PSC often occurs alongside inflammatory bowel disease. This association is especially common with ulcerative colitis, which causes inflammation in the large bowel. Crohn's disease can also occur with PSC. Either condition may be discovered first, and bowel inflammation can

be present without obvious digestive symptoms.

Having a relative with PSC may increase risk, but PSC is not usually inherited in a simple, predictable way. Most relatives do not develop it.

Secondary sclerosing cholangitis has an identifiable source of bile-duct injury. Possible causes include:

IgG4-related sclerosing cholangitis is part of an immune condition that can also affect the pancreas and other organs. Identifying this form matters because its treatment and outlook differ from those of PSC.

- Long-lasting blockage from stones or another structural problem
- Injury during surgery or another procedure
- Severely reduced blood supply to the bile ducts
- Repeated or severe bile-duct infections
- Certain toxic or treatment-related injuries

Diagnosis

Diagnosis usually begins with liver blood tests. Sclerosing cholangitis often raises alkaline phosphatase, or ALP, which can indicate reduced bile flow. Blood tests alone cannot confirm the condition.

The main imaging test is usually magnetic resonance cholangiopancreatography , or MRCP. This is a type of MRI that creates detailed pictures of the bile ducts without placing an instrument inside them. It may show alternating narrow and widened areas, sometimes described as a beaded appearance.

An endoscopic retrograde cholangiopancreatography , or ERCP, uses a flexible camera passed through the mouth into the small intestine. Dye is injected into the bile ducts before X-rays are taken. Because ERCP can cause infection, bleeding, or inflammation of the pancreas, it is generally used when treatment or tissue sampling is needed rather than as the first diagnostic test.

A liver biopsy removes a tiny sample of liver tissue. It may be needed if scans look normal but small-duct PSC is suspected, or if another liver condition may be present.

Your assessment may also include:

No single result answers every question. The diagnosis is based on the pattern of duct changes, laboratory results, medical history, and the exclusion of other causes.

- Blood tests for liver function and possible immune conditions
- Tests for IgG4-related inflammation
- A review of previous surgery, infection, and medication exposure
- Colonoscopy to look for inflammatory bowel disease
- Tissue or cell samples if a concerning narrowing is found

Treatment and management

Treatment depends on the type of sclerosing cholangitis, the location of any narrowing, symptoms, and the amount of liver damage.

For PSC, no medicine has been proven to stop progression in everyone. Ursodeoxycholic acid, a bile acid medication, is sometimes considered, but its role is limited and it is not a cure. Decisions about it are made using liver test results, treatment response, and potential harms.

Management may include:

During ERCP, a specialist may widen a severe narrowing with a small balloon. A temporary tube called a stent is sometimes used to keep the duct open. These procedures can improve bile flow and allow samples to be checked for abnormal cells.

Secondary sclerosing cholangitis is treated by addressing the source of injury when possible. This might involve removing an obstruction, draining infected bile, or managing reduced blood flow. Existing scar tissue may not disappear even after the cause is controlled.

IgG4-related sclerosing cholangitis often responds to treatment that reduces immune activity. It must be distinguished from PSC and bile-duct cancer before treatment is chosen.

A liver transplant replaces the damaged liver and bile ducts. It may be considered for liver failure, repeated serious infections, severe symptoms that cannot be controlled, or selected cancers. PSC can occasionally return after transplantation, but many recipients have substantial long-term benefit.

- Regular blood tests and liver imaging
- Medicines to reduce itching
- Antibiotics like amoxicillin or doxycycline for bacterial bile-duct infections
- Endoscopic treatment of an important narrowing
- Care for inflammatory bowel disease, if present
- Nutrition support when poor bile flow affects vitamin absorption

- Liver transplant assessment for advanced complications

Complications

Ongoing narrowing can cause bile to collect behind a blockage. This raises the risk of acute cholangitis , a bacterial infection of the bile ducts. It can become serious and needs prompt treatment.

Long-term inflammation may lead to fibrosis, meaning scar tissue, and then cirrhosis. Advanced cirrhosis can cause fluid in the abdomen, internal bleeding, confusion, or reduced liver function.

Other possible complications include:

Most symptoms have causes other than cancer. However, worsening jaundice, weight loss, or a new severe narrowing needs assessment because distinguishing inflammation from cancer can require several tests.

- Gallstones or stones within the bile ducts
- Poor absorption of fats and fat-soluble vitamins
- Thinning bones
- Cancer of the bile ducts, called cholangiocarcinoma
- Gallbladder cancer
- Increased colorectal cancer risk when PSC and colitis occur together

Prevention

Primary sclerosing cholangitis cannot currently be prevented. There is no proven diet, supplement, or lifestyle change that prevents PSC in someone at risk.

Some causes of secondary sclerosing cholangitis may be preventable through safe medical procedures and prompt treatment of bile-duct obstruction or infection. These causes are often related to serious medical events and are not something you could reasonably have prevented yourself.

If you already have the condition, avoiding unnecessary liver injury may help protect the liver you have. This can include discussing alcohol, medicines, and supplements with your healthcare team. "Natural" products can also affect the liver, so they should not automatically be assumed safe.

Recommended vaccinations, bowel care, and cancer monitoring are based on your liver health and any inflammatory bowel disease. They do not prevent PSC itself, but they may reduce avoidable complications.

Living with it and what to expect

Sclerosing cholangitis usually needs follow-up over many years. The course is difficult to predict from one scan or blood test. Some people remain active with few symptoms for a long time. Others experience infections, troublesome itching, worsening strictures, or progressive liver scarring.

Follow-up commonly involves blood tests, imaging, and appointments to review symptoms. If you have inflammatory bowel disease, colonoscopy monitoring is especially important because PSC changes the long-term bowel cancer risk. Your follow-up schedule should reflect your subtype, bowel health, and previous test results.

Fatigue may continue even when liver tests are stable. It can help to pace demanding activities and address other possible causes, such as poor sleep, anaemia, low mood, thyroid problems, or medication effects. Itching should be discussed rather than endured; several management approaches are available.

There is no special diet that removes bile-duct scarring. Most people can follow a balanced diet unless they have cirrhosis, weight loss, vitamin deficiency, or another digestive condition. A dietitian familiar with liver conditions can help when eating becomes difficult.

Transplant is not inevitable. If it becomes appropriate, assessment usually happens before an emergency develops. It includes tests of your overall health and discussions about benefits, risks, recovery, and long-term anti-rejection medication.

When to get medical help

Arrange routine follow-up if you develop increasing tiredness, persistent itching, new abdominal discomfort, darker urine, paler stools, or gradually worsening jaundice. Report unexplained weight loss or a change in your usual bowel symptoms as well.

Seek urgent same-day medical assessment for fever, chills, new jaundice, or increasing pain under the right ribs. These symptoms can indicate an infected or blocked bile duct.

Go to an emergency department if fever or chills occur with jaundice and you feel very unwell. Emergency help is also appropriate for confusion, fainting, severe weakness, vomiting blood, black stools, difficulty breathing, or a rapidly enlarging abdomen.

Do not wait for a routine appointment if symptoms suggest an acute infection or a complication of advanced liver scarring.

Common questions

Is sclerosing cholangitis the same as PSC?

Not always. PSC is the form without a known direct cause. Secondary sclerosing cholangitis results from an identifiable bile-duct injury. IgG4-related sclerosing cholangitis is an immune-related form with different treatment.

Is sclerosing cholangitis cancer?

No. It is inflammation and scarring of the bile ducts. However, PSC increases the risk of bile-duct and gallbladder cancers. PSC with inflammatory bowel disease also increases colorectal cancer risk, so planned monitoring is important.

Will I need a liver transplant?

Not everyone does. Transplant may be considered if the liver stops working properly, infections keep returning, or severe symptoms cannot be controlled. Your test results and course over time are more useful than one result for judging this risk.

What should I eat if I have sclerosing cholangitis?

There is no diet proven to reverse the condition. A balanced diet is generally appropriate. Individual advice may be needed if you lose weight, have cirrhosis, cannot absorb fats properly, or have inflammatory bowel disease.

Can I drink alcohol?

Alcohol does not cause PSC, but it can add to liver injury. A safe amount cannot be assumed for everyone with a liver condition. Discuss your liver stage and current alcohol use with your healthcare team.

Can sclerosing cholangitis go away?

Bile-duct scarring is usually permanent. Treating a secondary cause or IgG4-related inflammation may prevent further injury and improve narrowing. For advanced PSC, liver transplantation can replace the damaged liver, although PSC can occasionally return.

Sources

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