CONDITION

Fibrosing cholangitis

Fibrosing cholangitis is long-term inflammation and scarring that narrows the bile ducts.

WHAT IT IS
Inflammation and scarring of the bile ducts

BODY SYSTEM
Liver and digestive system

MAIN SYMPTOMS
Itching, fatigue, jaundice, upper abdominal pain

WHAT CAUSES IT
Often immune-related; sometimes bile-duct injury or infection

HOW IT IS DIAGNOSED
MRCP or ERCP imaging; sometimes liver biopsy

MAIN TREATMENT
Cause-specific care, symptom control, endoscopy, or liver transplant

CAN IT BE CURED?
Sometimes - transplant or treating a reversible cause

IS IT AN EMERGENCY?
Sometimes - fever with jaundice may mean acute infection

LONG-TERM OUTLOOK
Often slowly progressive, requiring lifelong specialist monitoring

Overview

Fibrosing cholangitis is inflammation and scarring in the bile ducts, which carry bile from the liver to the intestine. Scarring can create narrow areas called strictures. Bile may then build up, causing further inflammation and liver damage.

Primary sclerosing cholangitis , or PSC, develops without another direct cause of bile-duct injury and is thought to involve abnormal immune activity. Secondary sclerosing cholangitis follows a known injury or condition, such as severe infection, reduced blood supply, surgery, pancreatitis, or long-term blockage.

IgG4-related sclerosing cholangitis can cause similar narrowing. Identifying it is important because it may respond to treatment that reduces immune activity.

Fibrosing cholangitis can gradually lead to repeated bile-duct infections, cirrhosis, liver failure,

or bile-duct and gallbladder cancers, even when early symptoms are mild. Regular specialist follow-up is important.

Symptoms

Some people have no symptoms when the condition is found. Abnormal liver blood tests or unrelated imaging may be the first sign.

Possible symptoms include:

Symptoms do not always reflect the amount of scarring. Mild symptoms can occur despite significant disease, while severe itching may develop before advanced liver damage.

Fever, chills, jaundice, and upper abdominal pain may indicate acute cholangitis, a bacterial infection in a blocked bile duct. This needs urgent assessment.

- Persistent itching, sometimes worse at night
- Fatigue
- Yellow skin or eyes, called jaundice
- Discomfort under the right ribs
- Dark urine or pale stools
- Fever or chills during a bile-duct infection
- Reduced appetite or unplanned weight loss

Causes and risk factors

The exact cause of PSC is unknown. Genes, immune activity, gut bacteria, and environmental exposures may contribute. It is not caused by something a person ate or did.

PSC is strongly linked with inflammatory bowel disease, especially ulcerative colitis. However, most people with inflammatory bowel disease do not develop PSC, and PSC can occur without bowel inflammation.

Secondary sclerosing cholangitis has an identifiable cause, such as:

IgG4-related sclerosing cholangitis is part of an immune condition that may also affect the pancreas, salivary glands, kidneys, and other organs. It can resemble PSC or bile-duct cancer on scans, so careful testing is needed.

Fibrosing cholangitis is not generally directly inherited. A family history may increase the

chance of PSC, but there is no simple inheritance pattern.

- Long-lasting blockage from stones, injury, or a growth
- Damage after bile-duct or liver surgery
- Reduced blood flow to the bile ducts
- Severe infection or critical illness
- Recurrent pancreatitis
- Certain infections in people with weakened immunity

Diagnosis

Diagnosis usually begins with liver blood tests. Raised alkaline phosphatase, or ALP, can suggest reduced bile flow. Other tests assess bilirubin, inflammation, and the liver's ability to make proteins. Blood tests alone cannot confirm the condition.

The main imaging test is magnetic resonance cholangiopancreatography, or MRCP . This MRI scan creates detailed pictures of the bile ducts without placing instruments inside them. It may show alternating narrowed and widened sections.

An endoscopic retrograde cholangiopancreatography, or ERCP , may be needed if a significant blockage is suspected. A flexible tube is passed through the mouth into the small intestine, and dye is placed in the bile ducts for X-rays. The clinician can widen a narrowing, drain bile, or collect cells to check for cancer.

ERCP is not usually the first test because it can cause infection, bleeding, or pancreatitis. It is mainly used when treatment or tissue sampling is needed.

A liver biopsy may help when scans are normal but small-duct PSC is suspected, or when another liver condition may be present. Most people with typical MRCP findings do not need one to establish PSC.

Additional testing may check for:

Similar bile-duct changes can have different causes, so no result should be interpreted alone.

- IgG4-related disease
- Inflammatory bowel disease
- Stones or previous bile-duct injury
- Liver scarring and portal hypertension

- Possible bile-duct or gallbladder cancer

Treatment and management

Treatment depends on the type of fibrosing cholangitis, the strictures present, symptoms, and the amount of liver damage.

No medicine has been proven to reliably stop PSC from progressing. Ursodeoxycholic acid may improve liver blood tests in some people, but its effect on long-term outcomes is uncertain, and it is not suitable at all doses. A liver specialist can advise whether it has a role.

Treatment may include:

For secondary sclerosing cholangitis, removing a stone, draining a blockage, treating infection, or addressing the underlying injury may prevent further damage. Existing scarring may not fully reverse.

IgG4-related disease often responds to medicines that reduce immune activity. Specialist supervision is essential because infection and cancer must first be considered.

Liver transplantation may be considered for advanced cirrhosis, liver failure, repeated serious infections, or severe symptoms that cannot otherwise be controlled. PSC can occasionally return in the transplanted liver.

Follow-up may include blood tests, imaging, and checks for complications. People with inflammatory bowel disease usually need ongoing colon examinations because their bowel cancer risk is higher than with either condition alone.

- Medicines to reduce itching
- Antibiotics like amoxicillin or doxycycline for bacterial bile-duct infections
- ERCP to open or drain an important stricture
- Treatment for inflammatory bowel disease
- Support for nutrition, bone health, and vitamin deficiencies
- Monitoring for liver failure and related cancers

Complications

Increasingly severe strictures can trap bacteria and cause acute cholangitis. Long-term obstruction and inflammation may also cause cirrhosis.

Possible complications include:

These complications do not affect everyone. Monitoring aims to identify important changes before severe symptoms develop.

- Portal hypertension, or high pressure in veins around the liver
- Abdominal fluid, internal bleeding, confusion, or reduced liver function
- Poor absorption of fats and fat-soluble vitamins
- Bone thinning
- Bile-duct stones
- Gallbladder growths or cancer
- Cholangiocarcinoma, or bile-duct cancer
- Colorectal cancer in people with inflammatory bowel disease

Prevention

PSC cannot currently be prevented. It is not caused by diet, alcohol, poor hygiene, or a failure to look after the liver.

Some causes of secondary sclerosing cholangitis are treatable, although the original injury may not have been predictable or avoidable. Prompt treatment of a known blockage or infection may limit further damage.

Discuss alcohol, medicines, and supplements with the healthcare team. "Natural" products can still harm the liver or interact with treatment.

Keeping follow-up appointments, managing inflammatory bowel disease, and reporting new symptoms cannot remove existing scarring, but they can help detect infections, strictures, liver damage, or cancer-related changes earlier.

Living with it and what to expect

The course is unpredictable. Some people remain well with stable liver function for years, while others develop repeated infections, worsening strictures, or cirrhosis. Symptoms and blood-test results may fluctuate.

Itching and fatigue can significantly affect sleep, work, and mood even when liver function is preserved. Raise these symptoms at routine appointments because treatments and practical support may help.

Long-term care usually involves a liver specialist. Follow-up continues even without symptoms because blood tests and scans can reveal hidden changes. Monitoring may become more frequent if liver function worsens or a new stricture or complication appears.

A balanced diet is often suitable. Advanced liver damage, weight loss, or vitamin deficiency may require advice from a dietitian experienced in liver disease. No specific diet can remove bile-duct scarring.

Many people continue their usual activities for long periods. Some eventually need adjustments because of fatigue, itching, infections, or advanced liver problems, and a smaller group will be assessed for transplantation. The diagnosis alone cannot predict an individual's course.

When to get medical help

Arrange a prompt, non-emergency review for new or worsening jaundice, itching, abdominal discomfort, dark urine, pale stools, reduced appetite, or unexplained weight loss. These changes may require blood tests or imaging but do not always mean the disease has advanced.

Ask for routine advice if fatigue is disrupting daily life, medicines are not controlling itching, or you are considering a new medicine or supplement.

Seek urgent medical care for:

Fever with jaundice may signal acute cholangitis. This can worsen quickly and may require hospital treatment, intravenous antibiotics, and urgent drainage of a blocked bile duct.

Call emergency services for collapse, severe confusion, difficulty staying awake, heavy bleeding, or breathing difficulty.

- Fever or shaking chills with jaundice or upper abdominal pain
- Rapidly worsening jaundice with vomiting or severe weakness
- New abdominal swelling, confusion, or unusual drowsiness
- Vomiting blood or passing black, tar-like stools

Common questions

Is fibrosing cholangitis the same as primary sclerosing cholangitis?

Not always. PSC is one form, but sclerosing cholangitis can also result from another bile-duct injury. Imaging, medical history, blood tests, and sometimes tissue samples help identify the

type.

Can fibrosing cholangitis be cured?

Some secondary forms improve when the underlying blockage, infection, or immune condition is treated, although established scarring may remain. PSC has no reliable medical cure. Transplantation replaces the affected liver and ducts, but PSC can occasionally return.

Does fibrosing cholangitis mean I have cancer?

No. It is not cancer, but it increases the risk of bile-duct and gallbladder cancers, particularly in PSC. New strictures or symptoms may therefore need investigation.

Why am I being checked for bowel inflammation?

PSC is closely associated with inflammatory bowel disease, especially ulcerative colitis. Bowel inflammation may be mild or cause no obvious symptoms. Finding it affects treatment and colonoscopy monitoring.

Can I drink alcohol?

Alcohol does not cause PSC, but it can add strain to an injured liver. A safe amount cannot be assumed for everyone. Ask for advice based on liver function, scarring, medicines, and current alcohol use.

Will I need a liver transplant?

Not everyone does. Assessment is generally considered for advanced liver failure, repeated serious bile-duct infections, or severe symptoms that cannot be managed in other ways. Follow-up helps identify whether and when assessment is appropriate.

Sources

- American Association for the Study of Liver Diseases: AASLD Practice Guidance on Primary Sclerosing Cholangitis and Cholangiocarcinoma , Hepatology , 2023
- European Association for the Study of the Liver: EASL Clinical Practice Guidelines on Sclerosing Cholangitis , Journal of Hepatology , 2022
- British Society of Gastroenterology and UK-PSC Guidelines for the Diagnosis and Management of Primary Sclerosing Cholangitis , Gut , 2019
- National Institute of Diabetes and Digestive and Kidney Diseases: Primary Sclerosing Cholangitis , diagnosis, symptoms, and treatment information
- European Guideline on IgG4-Related Digestive Disease , United European Gastroenterology Journal , 2020