

Sclerosing cholangitis

Definition

Sclerosing cholangitis refers to swelling (inflammation), scarring, and destruction of the bile ducts inside and outside of the liver.

Alternative Names

Primary sclerosing cholangitis; PSC

Causes

The cause of this condition is unknown in most cases.

The disease may be seen in people who have:

- Inflammatory bowel disease (ulcerative colitis and Crohn disease)
- Autoimmune disorders
- Chronic pancreatitis (inflamed pancreas)
- Sarcoidosis (a disease that causes inflammation in various parts of the body)

Genetic factors may also be responsible. Sclerosing cholangitis occurs more often in men than women. This disorder is rare in children.

Sclerosing cholangitis may also be caused by:

- Choledocholithiasis (gallstones in the bile duct)
- Infections in the liver, gallbladder, and bile ducts

Symptoms

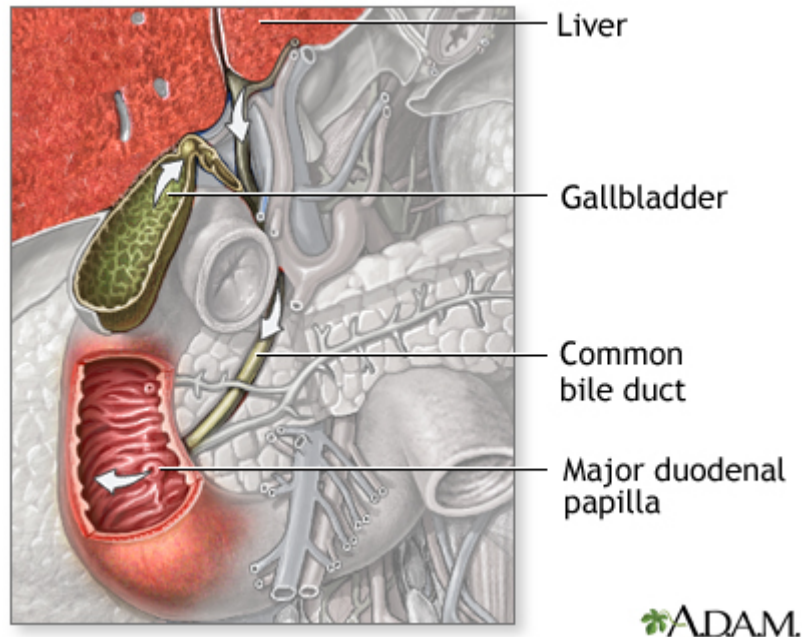
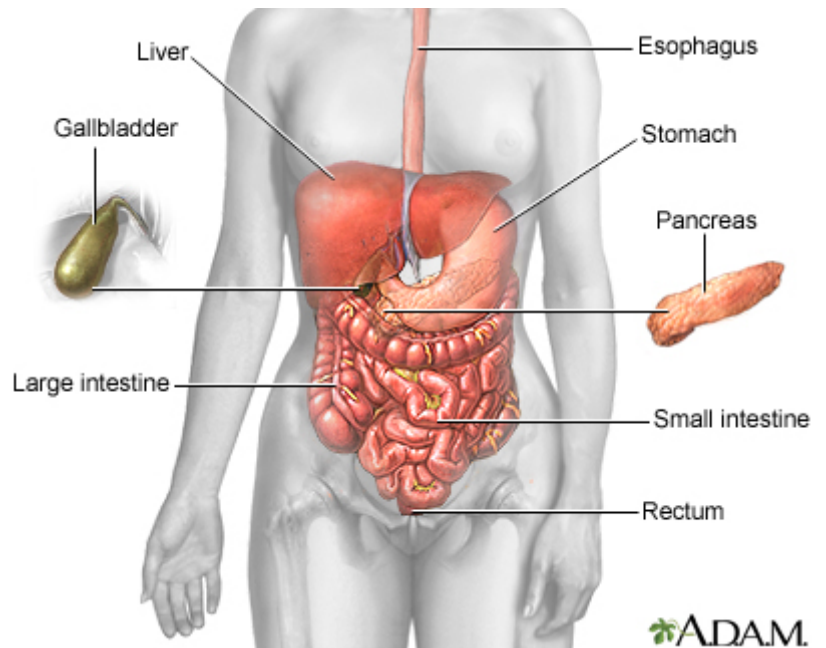
The first symptoms are usually:

- Fatigue
- Itching
- Yellowing of the skin and eyes (jaundice)

However, some people have no symptoms.

Other symptoms may include:

- Enlarged liver
- Enlarged spleen
- Loss of appetite and weight loss



- Repeat episodes of cholangitis

Exams and Tests

Even though some people do not have symptoms, blood tests shows that they have abnormal liver function. Your health care provider will look for:

- Diseases that cause similar problems
- Diseases that often occur with this condition (especially inflammatory bowel disease)
- Gallstones

Tests that show cholangitis include:

- Abdominal CT scan
- Abdominal ultrasound
- Endoscopic retrograde cholangiopancreatography (ERCP)
- Liver biopsy
- Magnetic resonance cholangiopancreatography (MRCP)
- Percutaneous transhepatic cholangiogram (PTC)

Blood tests include liver enzymes (liver function tests).

Treatment

Medicines that may be used include:

- Cholestyramine (such as Prevalite) to treat itching
- Ursodeoxycholic acid (ursodiol) to improve liver function
- Fat-soluble vitamins (D, E, A, K) to replace what is lost from the disease itself
- Antibiotics to treat infections in the bile ducts

These surgical procedures may be done:

- Inserting a long, thin tube with a balloon at the end to open up narrowing (endoscopic balloon dilation of strictures)
- Placement of a drain or tube for major narrowing (strictures) of bile ducts
- Proctocolectomy (removal of colon and rectum, for those who have both ulcerative colitis and sclerosing cholangitis) does not affect the progression of primary sclerosing cholangitis (PSC)
- Liver transplant

Outlook (Prognosis)

How well people do varies. The disease tends to get worse over time, and sometimes people develop:

- Ascites (buildup of fluid in the space between the lining of the abdomen and abdominal organs) and varices (enlarged veins)
- Biliary cirrhosis (inflammation of the bile ducts)
- Liver failure
- Persistent jaundice

Some people develop infections of the bile ducts that keep returning.

People with this condition have an increased risk of developing cancer of the bile ducts (cholangiocarcinoma). They should be checked regularly with a liver imaging test and blood tests. People who also have inflammatory bowel disease may have an increased risk of developing cancer of the colon or rectum and should have periodic colonoscopy.

Possible Complications

Complications may include:

- Bleeding esophageal varices
- Cancer in the bile ducts (cholangiocarcinoma)
- Cirrhosis and liver failure
- Infection of the biliary system (cholangitis)
- Narrowing of the bile ducts
- Vitamin deficiencies

References

Bowlus C, Assis DN, Goldberg D. Primary and secondary sclerosing cholangitis. In: Sanyal AJ, Boyer TD, Lindor KD, Terrault NA, eds. *Zakim and Boyer's Hepatology*. 7th ed. Philadelphia, PA: Elsevier; 2018:chap 43.

Ross AS, Kowdley KV. Primary sclerosing cholangitis and recurrent pyogenic cholangitis. In: Feldman M, Friedman LS, Brandt LJ, eds. *Sleisenger and Fordtran's Gastrointestinal and Liver Disease*. 10th ed. Philadelphia, PA: Elsevier Saunders; 2016:chap 68.

Zyromski NJ, Pitt HA. The management of primary sclerosing cholangitis. In: Cameron JL, Cameron AM, eds. *Current Surgical Therapy*. 12th ed. Philadelphia, PA: Elsevier; 2017:453-458.

Review Date: 4/23/2017

Reviewed By: Michael M. Phillips, MD, Clinical Professor of Medicine, The George Washington University School of Medicine, Washington, DC. Also reviewed by David Zieve, MD, MHA, Medical Director, Brenda Conaway, Editorial Director, and the A.D.A.M. Editorial team.