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## Sclerosing cholangitis

### Primary sclerosing cholangitis; PSC

Last reviewed: August 11, 2011.

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

### Causes, incidence, and risk factors

The cause of this condition is usually unknown.

The disease may be seen in patients who have:

- Autoimmune disorders
- Chronic pancreatitis
- Inflammatory bowel disease (Crohn's disease and ulcerative colitis)
- Sarcoidosis

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:

- Cholelithiasis
- 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 may have no symptoms.

Other symptoms may include:

- Enlarged liver
- Enlarged spleen
- Loss of appetite and weight loss
- Repeat episodes of cholangitis

### Signs and tests

Some people do not have symptoms, but blood work shows that they have abnormal liver function. The doctor 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

Medications that may be used include:

- Cholestyramine
- Ursodeoxycholic acid ([ursodiol](#))
- Fat-soluble vitamins (D, E, A, K)
- Antibiotics for infections in the bile ducts
- Medications that quiet the immune system ([prednisone](#), [azathioprine](#), [cyclosporine](#), [methotrexate](#))

Surgical procedures:

- 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 biliary ducts
- [Proctocolectomy](#) (for those who have both ulcerative colitis and sclerosing cholangitis)
- Liver transplant

## Expectations (prognosis)

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

- [Ascites](#) and [varices](#)
- Biliary [cirrhosis](#)
- Liver failure
- Persistent jaundice

Some patients 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.

## Complications

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

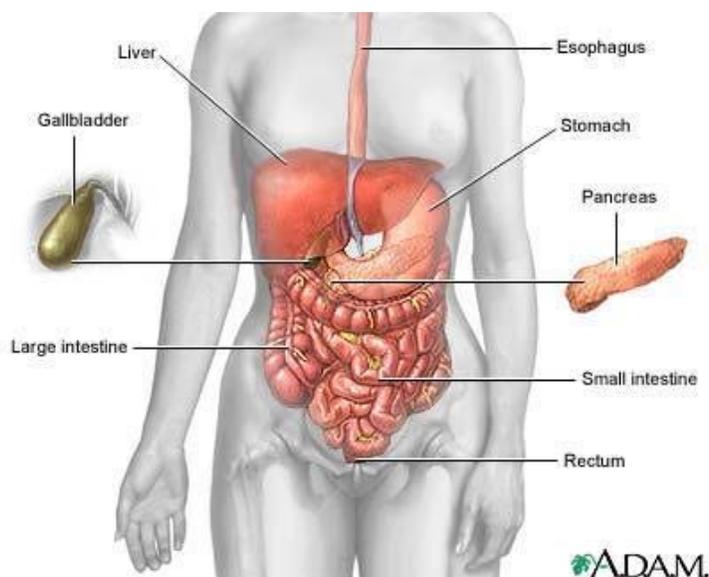
## References

1. Gordon FD. Primary sclerosing cholangitis. *Surg Clin North Am.* 2008;88:1385-1407.
2. Ross AS, Kowdley KV. Sclerosing cholangitis and recurrent pyogenic cholangitis. In: Feldman M, Friedman LS, Brandt LJ, eds. *Sleisenger & Fordtran's Gastrointestinal and Liver Disease.* 9th ed. Philadelphia, Pa: Saunders Elsevier; 2010:chap 68.

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## Figures

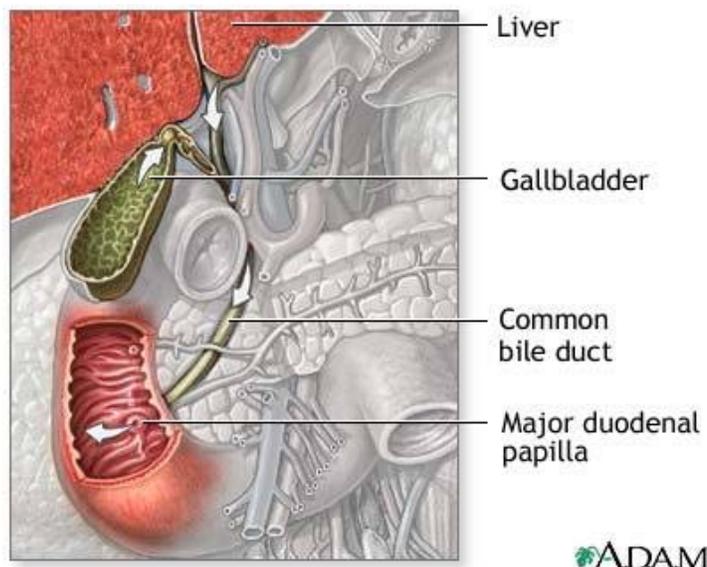


### Digestive system

The esophagus, stomach, large and small intestine, aided by the liver, gallbladder and pancreas convert the nutritive components of food into energy and break down the non-nutritive components into waste to be excreted.

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### Bile pathway

The biliary system is comprised of the organs and duct system that create, transport, store and release bile into the duodenum for digestion. Includes the liver, gallbladder and bile ducts (named the cystic, hepatic, common, and pancreatic duct).

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