



The Oregon Clinic - Gastroenterology

Primary Sclerosing Cholangitis

What is Primary Sclerosing Cholangitis?

In primary sclerosing cholangitis (PSC), the bile ducts inside and outside the liver become inflamed and scarred. As the scarring increases, the ducts become congested and blocked. The ducts are important because they carry bile out of the liver. Bile is a liquid that helps break down fat in food. If the ducts are blocked, bile builds up in the liver and damages liver cells. Blockage can also cause infection with fever and pain in the right upper abdomen. Eventually, PSC can cause cirrhosis and liver failure. It may also cause bile duct cancer, or cholangiocarcinoma.

What causes PSC?

Researchers do not know what causes PSC. Among the theories under investigation are the possible role of bacteria, viruses, and immune system problems. PSC most often appears to be associated with ulcerative colitis, a type of inflammatory bowel disease.

The disease usually begins between ages 30 and 60, but the disease can also arise during childhood. PSC is more common in men than women. PSC progresses slowly, so a person can have the disease for years before symptoms develop. The main symptoms are itching, fatigue, and jaundice, which cause yellowing of the eyes or skin. Infection may also occur. There may be bone disease, such as osteoporosis, or diarrhea.

How is PSC diagnosed and treated?

PSC is diagnosed through cholangiography, which involves injecting dye into the bile ducts and taking an x ray. Cholangiography can be performed as an endoscopic procedure (endoscopic retrograde cholangiopancreatography, ERCP), through radiology or surgery, or with magnetic resonance imaging (MRI) scans specifically designed to look at the bile ducts, called MRCP (magnetic resonance cholangiopancreatography). Treatment usually includes ursodeoxycholic acid, a medication used to make bile less toxic. Other medications to relieve itching, antibiotics to treat infections, and vitamin

supplements, as people with PSC are often deficient in vitamins A, D, E and K, may be necessary. In some cases, ERCP's are required to open major blockages in the bile ducts. No medication has been found to improve survival or need for liver transplantation. Liver transplantation may be the only option if the liver begins to fail.