

## Primary Sclerosing Cholangitis

is more common in men than women. Initially, many individuals have no symptoms and the disease is detected because of abnormal laboratory test results, particularly an enzyme test called alkaline phosphatase. It usually begins in the 30s, 40s, and 50s, and is commonly associated with fatigue, itching, and jaundice. Episodes of fever and chills from superimposed infection in the bile ducts occasionally occur and can be distressing symptoms. The diagnosis of primary sclerosing cholangitis is made by cholangiography, an X-ray test involving injection of dye into the bile ducts. This is usually accomplished by an endoscopic procedure called ERCP (endoscopic retrograde cholangiopancreatography) but also may be done radiologically, surgically, or, more recently, with magnetic resonance imaging (MRI) scans.

## Course of Disease

The course of the disease is unpredictable for the patient, but is generally slowly progressive. The patient may have the disease for many years before symptoms develop. Symptoms may persist at a stable level, be intermittent, or progress gradually. Liver failure may occur after 7-15 years of disease or even longer. Patients who have the disease on a longstanding basis may develop a superimposed tumor of the bile ducts called cholangiocarcinoma, but this occurs in only 0.5 to 1% of patients per year.

## Treatment

There is currently no specific treatment for primary sclerosing cholangitis. Research is under way to determine the effectiveness of a number of medications. The various symptoms of primary sclerosing cholangitis often respond effectively to medications that control itching, antibiotics when recurrent infections occur, and vitamins to replace those that are deficient. In some instances, endoscopic, radiologic, or surgical techniques may be employed to open major blockages in the common bile duct and improve bile flow. When progressive liver failure occurs in spite of these measures, liver transplantation may be indicated. Liver transplant patients have about an 87% one-year and 77% three or more year survival rate and a good quality of life after recovery.

# Primary Sclerosing Cholangitis

Primary sclerosing cholangitis is a disease in which the bile ducts inside and outside the liver become narrowed due to inflammation and scarring. This causes bile to accumulate in the liver and can result in damage to liver cells. Although the exact cause of primary sclerosing cholangitis is unknown, genetic and immunologic factors appear to play a role. Primary sclerosing cholangitis has been considered a rare disease, but recent studies suggest that it is more common than previously thought. It may occur alone, but approximately 70% of patients have associated inflammatory bowel disease, particularly ulcerative colitis.

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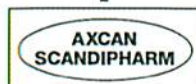
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The American Liver Foundation is a nonprofit, national voluntary health organization dedicated to the prevention, treatment, and cure of hepatitis and other liver diseases through research, education, and advocacy.

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What is

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