Primary Sclerosing Cholangitis

Primary Sclerosing Cholangitis (PSC) is a rare liver disease that damages the bile ducts inside and outside the liver.

Symptoms

- **Pruritus**: Intense itching, particularly on the palms of the hands or soles (bottom) of the feet, though it can occur anywhere, including in the eyes and mouth.
- **Fatigue**: Feeling run down, tired, unable to get enough sleep, or a flu-like exhaustion that does not go away with sleep.
- **Pain:** Felt in the right side or middle of the abdomen near or under the rib cage. This is often called right upper-quadrant (RUQ) pain. The pain may extend to the shoulder blade area, may be of any intensity, and may last for an indefinite period of time.

- **Jaundice**: Yellowing of eyes and skin caused by excess bilirubin that the liver cannot process. May be accompanied by dark urine.
- Chills and Fever: These may be signs of a bacterial infection in the bile ducts, a condition called cholangitis. A cholangitis attack requires immediate medical attention and treatment.
- **Depression**: Feelings of sadness, hopelessness, or loss of interest in activities. Changes in sleep, appetite, energy level, concentration, or self-esteem may also occur.

Causes

It's not clear what causes primary sclerosing cholangitis. An immune system reaction to an infection or toxin may trigger the disease in people who are genetically predisposed to it.

A large proportion of people with primary sclerosing cholangitis also have inflammatory bowel disease, an umbrella term that includes ulcerative colitis and Crohn's disease.

Primary sclerosing cholangitis and inflammatory bowel disease don't always appear at the same time, though. In some cases, primary sclerosing cholangitis is present for years before inflammatory bowel disease occurs. If primary sclerosing cholangitis is diagnosed, it's important to look for inflammatory bowel disease because there is a greater risk of colon cancer.

Risk factors

Factors that may increase the risk of primary sclerosing cholangitis include:

- **Age.** Primary sclerosing cholangitis can occur at any age, but it's most often diagnosed between the ages of 30 and 40.
- **Sex.** Primary sclerosing cholangitis occurs more often in men.

- **Inflammatory bowel disease**. A large proportion of people with primary sclerosing cholangitis also have inflammatory bowel disease.
- **Geographical location**. People with Northern European heritage have a higher risk of primary sclerosing cholangitis

Complications

Complications of primary sclerosing cholangitis may include:

- **Liver disease and failure**. Chronic inflammation of the bile ducts throughout your liver can lead to tissue scarring (cirrhosis), liver cell death and, eventually, loss of liver function.
- **Repeated infections**. If scarring of the bile ducts slows or stops the flow of bile out of the liver, you may experience frequent infections in the bile ducts. The risk of infection is particularly high after you've had a surgical procedure to expand a badly scarred bile duct or remove a stone blocking a bile duct.
- **Portal hypertension**. Your portal vein is the major route for blood flowing from your digestive system into your liver. Portal hypertension refers to high blood pressure in this vein.

- **Thinning bones**. People with primary sclerosing cholangitis may experience thinning bones (osteoporosis). Your doctor may

recommend a bone density exam to test for osteoporosis every few years. Calcium and vitamin D supplements may be prescribed to help prevent bone loss.

- **Bile duct cancer**. If you have primary sclerosing cholangitis, you have an increased risk of developing cancer in the bile ducts or gallbladder.
- **Colon cancer**. People with primary sclerosing cholangitis associated with inflammatory bowel disease have an increased risk of colon cancer. If you've been diagnosed with primary sclerosing cholangitis, your doctor may recommend testing for inflammatory bowel disease, even if you have no signs or symptoms, since the risk of colon cancer is elevated if you have both diseases.

Diagnosis

A diagnosis of primary sclerosing cholangitis is made based upon a thorough clinical evaluation, a detailed patient history, identification of characteristic findings, and a variety of specialized tests.

Clinical Testing and Work-Up

Blood tests called liver function tests may be performed to measure the activity and levels of certain chemicals produced by the liver. Certain liver enzymes may be elevated including alanine aminotransferase, aspartate aminotransferase, alkaline phosphatase, and gamma-

glutamyltranspeptidase. Elevation of these enzymes is indicative of liver disease, but is nonspecific to PSC.

Additional blood tests to detect other substances (e.g. autoantibodies) may also be performed to aid in diagnosing PSC or to rule out other conditions.

Specialized imaging tests may also be used to help obtain a diagnosis of PSC. A magnetic resonance cholangiopancreatography or MRCP is a noninvasive test used to evaluate both the intrahepatic and extrahepatic bile ducts. This exam employs MRI, an imaging technique that uses a magnetic field and radio waves to produce cross-sectional images of particular organs and bodily tissues.

In the past an exam called endoscopic retrograde cholangiopancreatography or ERCP was used to help diagnose PSC. This exam involves inserting a thin flexible tube (endoscope) into the mouth and down through the esophagus and stomach eventually reaching the bile ducts. A contrast dye is injected into the bile ducts and an x-ray is taken to evaluate the health and function of the bile ducts.

A liver biopsy, which involves the surgical removal and microscopic examination, of a small sample of liver tissue, may be performed to evaluate the liver and determine how far PSC has progressed.

Some physicians recommended a colonoscopy to evaluate the health and function of the bowels because of the high association of PSC with inflammatory bowel disease and colon cancer. Cross sectional imaging of the liver is recommended by some providers at 6 – 12 month intervals because of the high risk of hepatobiliary cancers.

Treatment

There is no specific, universal treatment for individuals with PSC. Treatment is directed toward the specific symptoms that are apparent in each individual and at slowing the progression of the disorder.

Endoscopic surgery to remove blockages and enlarge narrowed bile ducts may be of benefit to help prevent liver deterioration in certain cases. Lost vitamins should be replaced when required to prevent complications related to these deficiencies. Antibiotics may be useful in controlling inflammation or infection. Individuals with PSC are encouraged to follow a normal healthy diet and to avoid alcohol or only have alcohol in small amounts.

The drug cholestyramine may be effective in controlling itching. Cholestyramine may be given with or without antihistamines. If cholestyramine is ineffective, other medications may be recommended. Bisphosphonates, which are drugs that prevent the loss of bone mass, may be used to treat osteoporosis.