Chris was diagnosed with PSC in the early 1990s. He received a liver transplant in July 2000. Just 18 months later, Chris achieved a lifetime ambition, winning the Bronze Medal at the Olympic Games. He is an inspiration to all those diagnosed with PSC.

“PSC can be beat. After my transplant, I’m healthier and stronger than ever before.”

– Chris Klug

Chris Klug, snowboarder, Olympic medal winner, author, PSC patient and liver transplant recipient.
PRIMARY SCLEROSING CHOLANGITIS

Primary sclerosing cholangitis (PSC) is a disease that causes the bile ducts inside and outside of the liver to become scarred, narrowed, and eventually blocked. As more and more ducts are blocked, bile becomes trapped and damages the liver. This damage causes liver cell injury which can ultimately lead to cirrhosis and require a liver transplant.

PSC SYMPTOMS

Although many patients report few symptoms, if any, some of the symptoms of PSC include the following:
- **Fatigue**: Feeling run down, unable to get enough sleep, flu-like exhaustion
- **Pruritus or intense itching**: Particularly on the soles of hands or feet, though it can occur anywhere, including in the eyes and mouth
- **Pain**: On the right side or middle of the abdomen towards the rib cage; may be of any intensity and may last for an indefinite period
- **Jaundice**: Yellowing of the eyes and skin caused by a blockage of bile flow in the bile ducts or by excess bilirubin which cannot be processed by the liver
- **Chills and fever**: Signs of bacterial infections in the bile ducts requiring immediate medical attention

Additionally, signs that PSC has progressed to cirrhosis, and that liver transplantation may be indicated, include:
- **Ascites**: Buildup of fluids in the abdomen
- **Encephalopathy**: Personality changes, confusion, and sleep disturbances caused by a buildup of toxins such as ammonia in the blood
- **Varices**: Swollen veins prone to bleeding, usually in the esophagus. When bleeding occurs, it can cause vomiting of blood or passage of black tarry stool. Medical attention should be sought immediately in these cases.
- **Splenomegaly**: An enlarged spleen
- **Abnormal blood tests**: Several blood tests including bilirubin, creatinine and INR (International Normalized Ratio) may become abnormal and are used to determine if liver transplant is indicated.

PSC DIAGNOSIS

Patients diagnosed with PSC often do not have symptoms, but on routine blood tests are found to have elevated liver function numbers. PSC should be suspected in patients with inflammatory bowel disease (IBD) and elevated liver tests because over 75% of PSC patients also have IBD, usually ulcerative colitis. Diagnostic liver tests called liver function tests or LFTs include:
- alkaline phosphatase (ALP)
- alanine aminotransferase (ALT)
- aspartate aminotransferase (AST)
- gamma-glutamyltransferase (GGT)

Elevated ALP and GGT usually indicate an injury to the bile ducts and are almost always elevated in PSC. The ALT and AST may also be elevated in PSC, but usually not as much. The diagnosis of PSC requires an image of the bile ducts called a cholangiogram. This can be done by a special MRI called an MRCP (magnetic cholangiopancreatography) or sometimes a procedure called an ERCP (endoscopic retrograde cholangiopancreatography). Sometimes a liver biopsy is needed. Other blood tests may be performed to exclude other diseases which can look similar to PSC.

Although the average time from diagnosis to liver transplantation is 15 to 20 years, some patients need transplants sooner, and some will never need a transplant.

FREQUENTLY ASKED QUESTIONS

HOW DID I GET IT? IS IT CONTAGIOUS?

Currently, no one knows what causes PSC. Although PSC is a liver disease, it is not caused by alcohol consumption. There appears to be a complex interaction of many genetic and environmental factors at work. Although there is a genetic predisposition to PSC, very rarely are family members, including children of patients, affected with PSC. PSC is not contagious.

WHO GETS PSC?

PSC is a rare liver disease affecting about 1 in every 10,000 people. Although PSC can occur at any age in both men and women, PSC is more common in men and is typically diagnosed between the ages of 30 and 40 years. PSC is often accompanied by inflammatory bowel disease (IBD), most often ulcerative colitis (UC), and sometimes Crohn’s disease. Over 75% of PSC patients have ulcerative colitis, while only 2-5% of patients with ulcerative colitis have PSC. PSC can sometimes be associated with other auto-immune diseases.

WHAT OTHER RISKS ARE INVOLVED WITH PSC?

There are mainly two types of cancer that occur with increased frequency in PSC patients: colon cancer and bile duct cancer (cholangiocarcinoma). The increased risk of colon cancer is mainly in PSC patients who also have underlying inflammatory bowel disease (ulcerative colitis or Crohn’s).