

WHERE CAN I FIND SUPPORT AND INFORMATION?

An online PSC support group was established in 1998. This is a message board (forum) on Yahoo/Health, where PSC patients and caregivers can exchange information and lend support/advice to one another:

<http://health.groups.yahoo.com/group/psc-support/>

In the United Kingdom, there is a similar group that publishes a newsletter and holds an annual meeting in Oxford with Dr. Roger Chapman, a leading PSC expert:

www.psc-support.demon.co.uk

There is also a parent's support group, especially necessary for parents of young children, as PSC presents itself differently in pediatric patients, and is treated accordingly.

This group is found at:

<http://health.groups.yahoo.com/group/pscmoms/>

For post-transplant PSC patients, there is a support group with discussions based on the special needs of being post-surgical, immunosuppressed, and their "at-risk" status.

You may join this group at:

<http://health.groups.yahoo.com/group/Livertx-PSC/>

For post-transplant PSC patients, there is a support group with discussions based on the special needs of being post-surgical, immunosuppressed, and their "at-risk" status.

You may join this group at:

<http://health.groups.yahoo.com/group/Livertx-PSC/>

Facebook: For social networking, sharing and learning about living life in your 20s and 30s with PSC:

<http://www.facebook.com/group.php?gid=39847120173>

To complement the support groups, a PSC literature website was developed in 2004 to provide PSC patients and caregivers easy access to a vast body of information concerning PSC and allied illnesses:

www.psc-literature.org

PSC Partners Seeking a Cure publishes a free quarterly newsletter and holds an annual conference on PSC for patients and caregivers:

www.pscpartners.org

For detailed information on inflammatory bowel disease, please visit the Crohn's and Colitis Foundation of America (CCFA) web site at:

www.ccfa.org

PSC PARTNERS SEEKING A CURE

PSC Partners Seeking a Cure is an organization that was formed to give PSC patients a collective voice in healthcare issues, and to raise funds to research the origins and cure for the disease.

PSC Partners Seeking a Cure is a 501(c)3 nonprofit foundation whose mission is to:

1. Raise funds with which to research the causes and cures for PSC,
2. Promote PSC and organ donation awareness, and
3. Provide education and support to PSC patients and their families.



FOR MORE INFORMATION, OR TO HELP FUND THE CURE:

Visit our website at:
www.pscpartners.org

or contact Ricky Safer by e-mail at:
Contactus@pscpartners.org

or send a tax-deductible donation to:
PSC Partners Seeking a Cure
5237 S. Kenton Way
Englewood, CO 80111

To order more brochures, please write to us at:
Brochures@pscpartners.org

living with
PSC
primary sclerosing cholangitis



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

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

A Publication of **PSC** Partners Seeking a Cure

PSC DIAGNOSIS

PSC is often diagnosed in otherwise asymptomatic patients by observing elevated serum liver enzyme levels in routine blood tests (liver function tests).

These enzymes include:

- alanine aminotransferase (ALT),
- aspartate aminotransferase (AST),
- alkaline phosphatase (ALP), and
- gamma-glutamyltranspeptidase (GGT).

Elevated ALT and AST generally indicate liver cell damage. Elevated serum ALP and GGT usually indicate a cholestatic liver disease or bile duct blockage. PSC might be suspected in patients presenting with inflammatory bowel disease (IBD) and elevated ALT, AST, and ALP, because over 75% of PSC patients also have IBD. Other tests may be performed, such as antibody tests, ERCP (endoscopic retrograde cholangiopancreatography), MRCP (magnetic resonance cholangiopancreatography), and/or a liver biopsy. For more information on diagnosis, refer to the “Diagnosing PSC” brochure at <http://www.pscpartners.org/pdfs/PSCDiagnosis.pdf>.

PSC SYMPTOMS

Although many patients report few, if any, symptoms, some of the symptoms of PSC include the following:

- **Pruritus or intense itching:** particularly on soles of hands or feet, though it can occur anywhere, including in eyes and mouth.
- **Fatigue:** feeling run down, unable to get enough sleep, flu-like tiredness.
- **Pain:** in right side or middle of abdomen towards rib cage. Pain may be of any intensity and may last for indefinite period.

- **Jaundice:** yellowing of eyes and skin caused by excess bilirubin that liver cannot process.
- **Chills and fever:** signs of bacterial infections in 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 abdomen.
- **Encephalopathy:** personality changes, intellectual impairment, and sleep disturbances caused by buildup of neurotoxins, such as ammonia, in blood.
- **Varices:** swollen veins in gastrointestinal tract prone to bleeding. When gastrointestinal bleeding occurs, medical attention should be sought immediately, since this condition can be life-threatening.
- **Splenomegaly:** swollen spleen.
- **Reduction in ability of blood to form clots:** increase of prothrombin time or International Normalized Ratio (INR).
- **Impaired kidney function:** increased serum creatinine levels.

FREQUENTLY ASKED QUESTIONS

WHAT'S WRONG WITH MY LIVER?

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, if left unchecked, causes liver cell death leading to cirrhosis, and may eventually require the need for a liver transplant.

HOW DID I GET IT? IS IT CONTAGIOUS?

Currently, no one knows what causes PSC. There appear to be genetic, autoimmune, and

environmental variables at work. PSC is not contagious. It cannot be transmitted to another person by touching, kissing, sexual activity, or even blood transfusion. Although there is a genetic predisposition to PSC, most children of patients with PSC are healthy and unaffected.

WHAT IS THE TREATMENT?

At present, the only definitive treatment for PSC is liver transplantation. Although the statistics for long-term survival with PSC are bleak, many live with the disease for years before experiencing symptoms or facing a liver transplant.

Therapy with UDCA (ursodeoxycholic acid, Ursodiol) has been a main component in the management of PSC. Although Ursodiol is not FDA-approved for treatment of PSC in the United States, several studies suggest that it may improve liver biochemistry.

It is not known whether UDCA slows progression in PSC or reduces risk for colon cancer or cholangiocarcinoma; existing studies have yielded conflicting results. Studies are ongoing, but presently there is no definitive answer, so your physician will make a decision based on your individual case. Endoscopy (e.g. balloon dilatation, or placement of stents in the bile ducts) can be used to open up significant bile duct strictures and temporarily restore bile flow. Treatments for pruritus (itching) associated with PSC include

the use of various skin lotions, or medications that include hydroxyzine, rifampin, naltrexone, or cholestyramine.

WHO GETS PSC?

PSC is a rare liver disease that predominantly affects males ages 30-40 years old. At diagnosis, 43% are asymptomatic. The disease has an estimated prevalence of 20.9 per 100,000 men and 6.3 per 100,000 women. 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. PSC can sometimes be associated with other autoimmune diseases.

WHAT OTHER RISK FACTORS 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 in colon cancer is mainly in PSC patients who also have underlying inflammatory bowel disease. For more information on cancer and PSC, refer to the “Diagnosing PSC” brochure at <http://www.pscpartners.org/pdfs/PSCDiagnosis.pdf>.

