

## WHERE CAN I FIND INFORMATION & SUPPORT?

PSCPARTNERS.ORG

For over a decade, PSC Partners Seeking a Cure, a 501(c)(3) nonprofit organization, has provided education and support to PSC patients, families and caregivers and has raised funds to research causes, treatments and potential cures for primary sclerosing cholangitis.

The PSC Partners website serves as a hub for the PSC patient and caregiver community. We welcome you to visit us at [www.pscpartners.org](http://www.pscpartners.org) and join our mailing list to receive news about new research findings, clinical trials, our newsletter and our annual PSC Partners conferences.

We welcome you to our annual patient and caregiver conferences held in collaboration with top medical centers in the U.S. Join us for our next conference.

Since 2004, we have maintained a collection of literature on PSC that is regularly updated to provide PSC patients and caregivers easy access to a vast body of information concerning PSC and connected illnesses.

By joining the PSC Partners Patient Registry, PSC patients have the power to help researchers move closer to finding a cure. We encourage you to learn more about our PSC Partners Patient Registry and participate online.

Our Facebook page offers a place to ask questions, share concerns and connect with other PSC Partners members. The community is always welcoming and helpful to new members.

[Find us on Facebook at PSC Partners](#)

We're also on Twitter [@PSCPpartners](#)

Read about PSC Partners' independent national and international support groups on our website.

For information on inflammatory bowel disease, visit the Crohn's and Colitis Foundation of America (CCFA) website at [www.cdfa.org](http://www.cdfa.org)

For information on autoimmune hepatitis (AIH), visit the Autoimmune Hepatitis Association website at [www.aihep.org](http://www.aihep.org)

## PSC PARTNERS SEEKING A CURE

PSC Partners Seeking a Cure is a 501(c)(3) nonprofit organization that was formed to give PSC patients a collective voice in healthcare issues. The mission of PSC Partners Seeking a Cure is to provide education and support to PSC patients, families and caregivers and to raise funds to research causes, treatments and potential cures for primary sclerosing cholangitis.

### FOR MORE INFORMATION

Visit our website at  
[PSCPARTNERS.ORG](http://PSCPARTNERS.ORG)

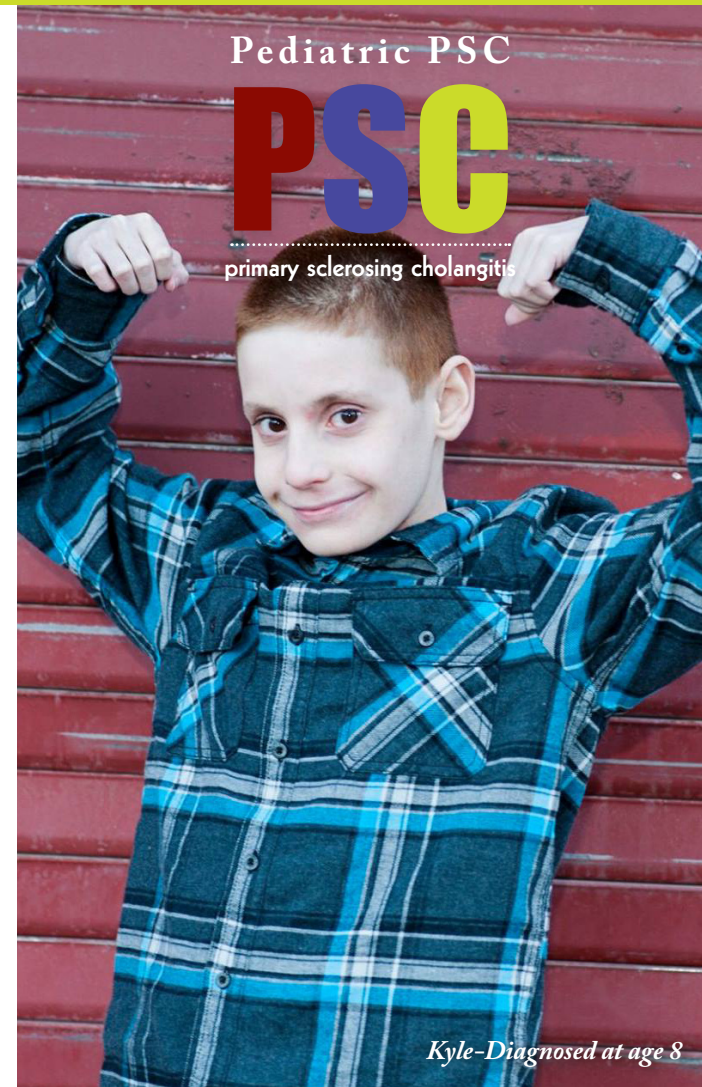
or send an e-mail to  
[contactus@pscpartners.org](mailto:contactus@pscpartners.org)

### TO HELP FUND THE CURE

Click on the DONATE NOW button at  
[PSCPARTNERS.ORG](http://PSCPARTNERS.ORG)

or send a tax-deductible donation to  
PSC Partners Seeking a Cure  
6900 E. Belleview Ave., Suite 202  
Greenwood Village, CO 80111

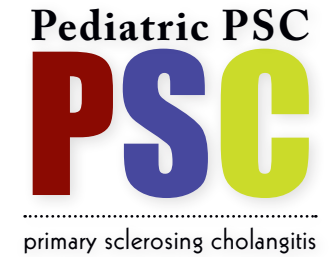
Canadian donors can receive a tax credit  
by donating directly to [www.pscpartners.ca](http://www.pscpartners.ca)



*Kyle—Diagnosed at age 8*



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*Naomi—Diagnosed at age 5*

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

## WHAT IS PSC?

Primary sclerosing cholangitis (PSC) is a rare chronic disease of inflammation and scarring that affects the bile ducts outside and/or inside the liver. The liver makes bile that is carried by bile ducts to the small intestine. Bile is important for the absorption of fats and Vitamins A, D, E and K. With a PSC patient, bile sometimes can't get to the intestines and stays in the liver where it damages liver cells, leading to scarring and possibly cirrhosis.

## WHY DID MY CHILD GET PSC?

Although no one knows the cause of PSC, there appear to be genetic, environmental and autoimmune components. As a parent, there is nothing you could have done to prevent PSC in your child.

## WHAT ARE THE COMMON SYMPTOMS?

When diagnosed, most children with PSC are in early stages of the disease process and don't exhibit symptoms. However, these are the initial symptoms that may develop:

- Fatigue
- Intense persistent itching
- Pain on right side or middle of abdomen
- Jaundice (yellowing of eyes and skin)
- Elevated liver blood tests
- Hepatomegaly (an enlarged liver)
- Splenomegaly (an enlarged spleen)

Most children with PSC have inflammatory bowel disease (Crohn's disease or ulcerative colitis), which may present with:

- Chronic diarrhea
- Blood in the stool
- Poor growth

## WHEN DO I TAKE MY CHILD TO THE HOSPITAL?

- Fever, chills and jaundice (signs of bacterial infection in the bile ducts)
- Vomiting blood (sign of burst blood vessel in the esophagus) or seeing dark black blood in the stool (sign of bleeding in the stomach or intestines).

## HOW IS PSC DIAGNOSED?

### Blood tests:

Liver tests, including conjugated and unconjugated bilirubin; gamma-glutamyltranspeptidase (GGT); alanine aminotransferase (ALT); aspartate aminotransferase (AST); prothrombin time (PT) and partial thromboplastin time (PTT) and international normalized ratio (INR).

### Imaging tests:

Magnetic resonance cholangiopancreatography (MRCP), noninvasive, with no radiation exposure; endoscopic retrograde cholangiopancreatography (ERCP), for bile duct stent placement; CT scan of the liver and bile ducts; ultrasound of the liver and bile ducts.

### Liver biopsy:

To test for autoimmune hepatitis (AIH) in the AIH/PSC overlap syndrome, as well as degree of fibrosis or scarring.

## HOW DOES PSC IN CHILDREN DIFFER FROM ADULT PSC?

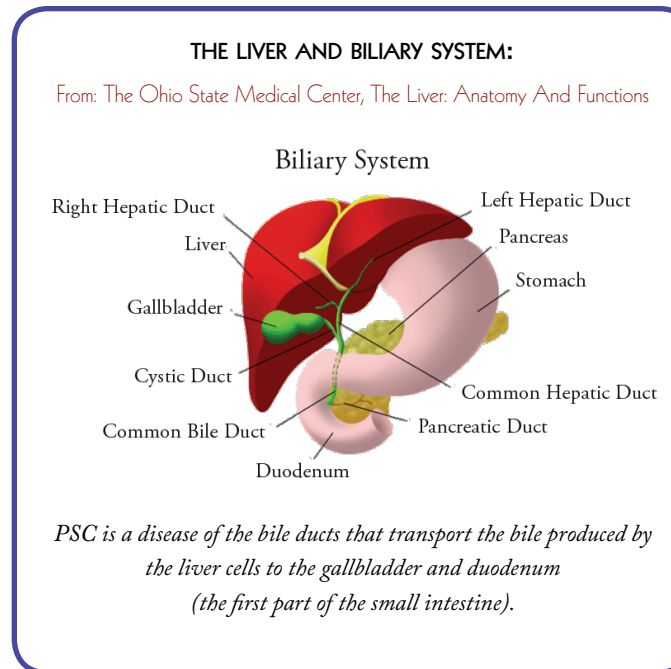
PSC is less common in children than in adults. Overlap of autoimmune hepatitis with PSC is more common in children, and children have markers of autoimmune disease more often than adults. Children rarely have dominant strictures of the common bile duct or cancers of the bile ducts and colon. Experts question whether childhood PSC is a different disease than adult PSC or an earlier phase of the adult disease.

## WHAT IS THE TREATMENT FOR PSC?

There is currently no medical cure for PSC. Most therapies are directed at managing symptoms rather than the underlying cause of PSC:

Ursodeoxycholic acid (Ursodiol, Urso Forte, UDCA, Actigall) may improve liver enzymes and itching in pediatric PSC; oral vancomycin may improve laboratory tests

and clinical symptoms; immunosuppressants (steroids, azathioprine) since patients with PSC and autoimmune hepatitis overlap, and elevated IgG4 may respond to immunosuppression; rifampin, cholestyramine, colestevlam, antihistamines and anti-opiates (naltrexone) are prescribed for itching; ERCP (balloon dilatation and/or stent placement for dominant strictures); endoscopy procedures



to prevent bleeding from esophageal varices; liver transplantation.

Researchers' understanding of the efficacy of drugs for PSC is constantly changing. Several promising new drugs for the treatment of PSC are in various stages of development in adults, and may be ready for trials in pediatric PSC patients in the near future. [pscpartners.org/clinical-trials/](http://pscpartners.org/clinical-trials/)

## WHO WILL NEED A LIVER TRANSPLANT?

About 15% of children may need a liver transplant for PSC before age 18. There is no reliable way to predict who will have progressive disease.

## IS THERE A SPECIAL DIET FOR PSC AND WHAT SHOULD BE AVOIDED?

Special considerations must be given to maintain normal nutrition and growth in children with PSC. Although there is no special diet for PSC, the diet should be a healthy balance of protein, carbohydrates and fat. Fatty foods, processed foods and preservatives should be avoided. Fat-soluble vitamins (A, D, E, and K), calcium and phosphorus may require supplementation. Medications that may harm the liver, as well as herbal supplements, alcohol and recreational drugs need to be avoided. A knowledgeable nutritionist can be of help.

## HOW WILL PSC AFFECT THE LIFE OF MY CHILD?

The majority of children with PSC have a normal quality of life and do not experience any liver-related symptoms or pain. Children with progressive disease may require more frequent monitoring, clinic visits, admissions and occasionally liver transplantation.