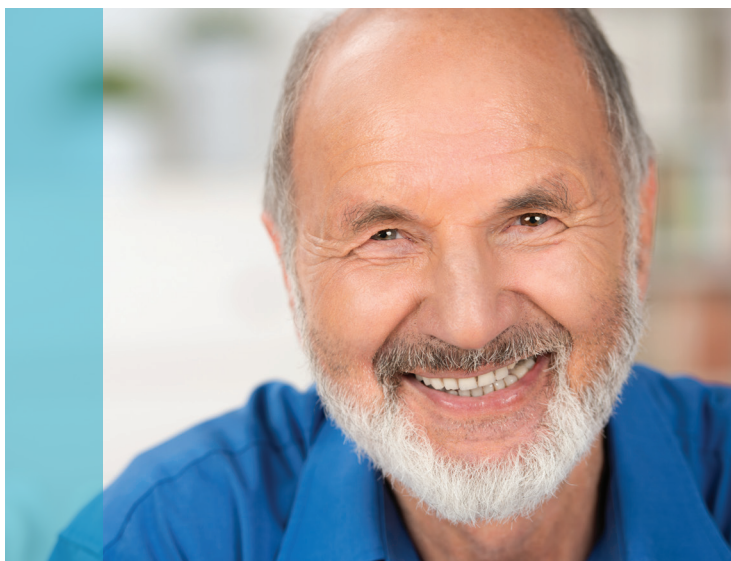
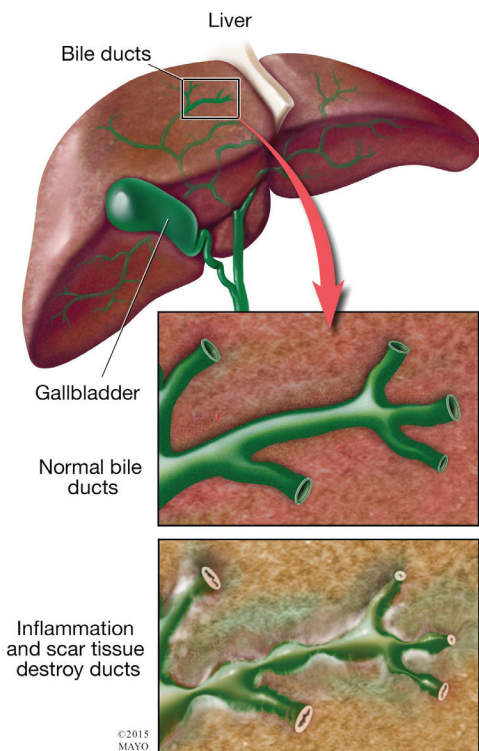


Primary Sclerosing Cholangitis



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What is PSC?

Primary Sclerosing Cholangitis (PSC) is a rare and chronic disease in which the ducts that transport bile from the liver develop inflammation and scar over time.

- Bile is made in the liver and travels to the intestine through bile ducts. It aids in metabolism and digestion of food and other substances.
- If bile cannot exit the liver due to scars and blockages in the bile ducts, liver damage can occur because of inflammation caused by the toxic

effects of retained bile.

- Scarring of the bile ducts and of the liver, called “fibrosis,” can worsen over the course of years and result in further liver damage and other complications.
- It is thought that approximately 32,000 people have PSC in the United States.

Who can get PSC?

No one knows exactly why PSC develops.

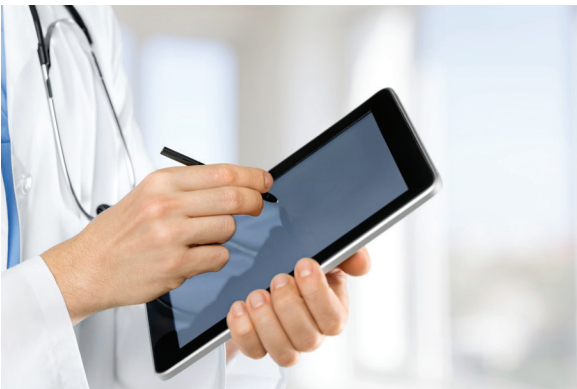
- Studies show that mutations in some genes of the immune system are more commonly found in patients with PSC.
- However, this represents only a minor portion of the risk and it is likely that unknown factors from the environment are important as well.
- The bile in PSC patients is more toxic than the bile in healthy people.
- PSC is also usually considered an autoimmune disease since the immune system causes the bile duct inflammation, and many patients with PSC also have other autoimmune diseases.
- PSC is more common in men, but it can also occur in women and children.
- The majority of patients with PSC have inflammatory bowel disease, also known as colitis, including Ulcerative Colitis or Crohn’s Disease.
- Although PSC is more common in people of Northern European heritage, it can occur in people from any background around the world.

What are the complications of PSC?

PSC is a very heterogeneous disease, which means that some patients may develop some of these complications while others may not.

Potential complications of PSC include:

- Complete blockage of the bile ducts causing jaundice
- Infections of the bile – called “cholangitis”
- Thinning of the bones – called “osteoporosis”
- Vitamin deficiency, such as vitamin D
- Severe scarring, or fibrosis, of the liver – called “cirrhosis”
- Advanced (decompensated) cirrhosis, which may require a liver transplant
- Bile duct cancer – called “cholangiocarcinoma”
- Gallbladder cancer
- Colon cancer in patients who also have inflammatory bowel disease (i.e. colitis)



What are the symptoms of PSC?

Many patients with PSC have no symptoms, especially in the early stages. However, symptoms often develop as the disease worsens.

Symptoms of PSC can include:

- Itching of the skin (pruritus)
- Fatigue
- Sudden fevers, chills and abdominal pain due to an infection of the bile (cholangitis)
- Yellowing of the skin and eyes (jaundice)

If a patient with PSC develops advanced (decompensated) cirrhosis, symptoms could include:

- Weight loss and malnutrition
- Ascites (fluid in the abdomen)
- Hepatic encephalopathy (confusion)
- Intestinal bleeding from enlarged veins

How is PSC diagnosed and managed?

PSC is diagnosed based on the finding of scars in the bile ducts, including ducts within or outside the liver.

- Bile duct scars are usually assessed by an imaging test (MRI) or by an endoscopic procedure (ERCP) where dye is injected into the bile ducts.
- Liver function tests are often abnormal in patients with PSC. Vitamin D levels may be decreased as well. It is also important to evaluate for bone density loss with an imaging test.

- Patients with PSC should have a colonoscopy to evaluate for colitis. If colitis is found, a yearly colonoscopy is needed to screen for colon cancer.
- It is not clear how to screen for bile duct cancer. However, many patients receive an imaging test yearly though this has not been studied sufficiently.
- Some patients take ursodiol (ursodeoxycholic acid) to reduce biliary inflammation in PSC, but this has not been shown to cure PSC or stop the disease from worsening.
- Patients with an infection of the bile need antibiotics, and a blocked bile duct requires an endoscopy or radiology procedure to open the duct.
- Patients with advanced disease (decompensated cirrhosis) need a liver transplant evaluation.

Is there a cure for PSC?

- While there is no cure yet for PSC, researchers around the world are working to find one.
- Several potential drugs to improve the damage from PSC will be tested in the next several years.

What should patients with PSC do?

- Patients with PSC should be seen by a liver specialist with knowledge of PSC and its management.
- Patients with PSC should ask their provider about new possible treatments and clinical trials.

Notes



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1-800-GO-LIVER

(1-800-465-4837)



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