

# Primary Biliary Cholangitis (PBC)

Provider Information



*Your Liver. Your Life.*

# Primary Biliary Cholangitis (PBC)

- Formerly known as primary biliary cirrhosis
- Chronic, autoimmune disease of the liver that slowly destroys the medium-sized bile ducts (intrahepatic bile ducts)
- Eventually causes cholestasis (disruption/obstruction of bile flow), cirrhosis (scarring and deterioration of liver), and liver failure

# Primary Biliary Cholangitis: Etiology and Epidemiology

- Current research suggests PBC is an autoimmune disease; the cause is still unknown.
- Although relatively rare, PBC is the most common liver disease associated with chronic cholestasis in adults.
- Highest prevalence is adult women: approximately 95% of cases occur in women aged 35 to 70, and 90% of all cases are women.

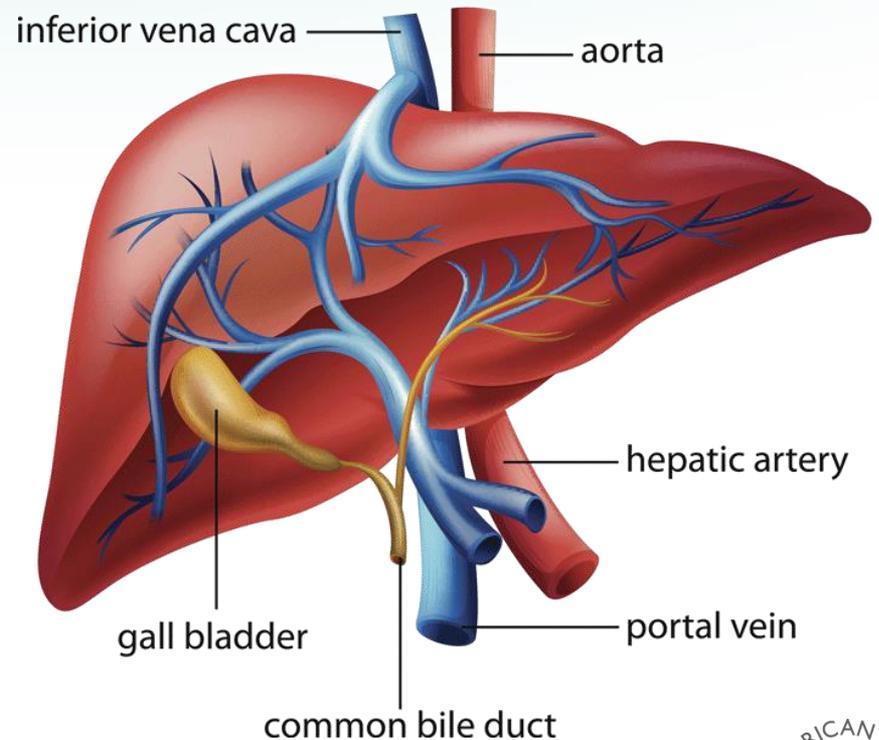
# Primary Biliary Cholangitis: Etiology and Epidemiology (cont.)

- PBC is not a hereditary disease, but patterns of occurrence appear.
- There is a higher familial rate of incidence, especially between mothers and daughters, as well as in identical twins.
- For those more genetically likely to develop PBC, environmental factors can trigger or worsen the disease—such as smoking or exposure to certain medications or toxic chemicals.

# Liver Physiology

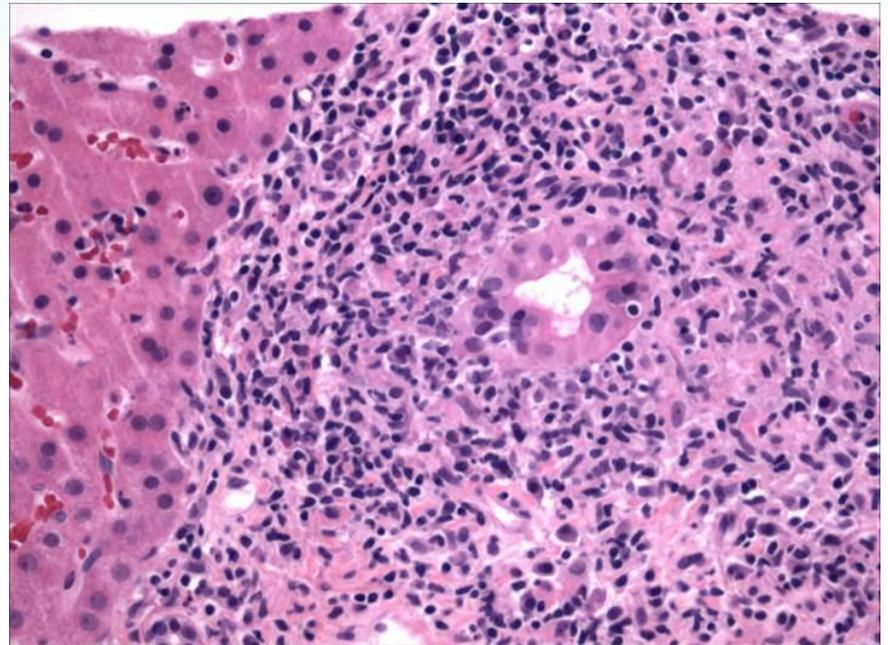
- The body's largest internal organ
- Essential Functions for the Body
  - Absorbing, storing, and processing nutrients from food
  - Making new proteins, such as albumin and clotting factors
  - Producing bile
  - Removing waste products the kidneys cannot remove (fats, cholesterol, toxins, and medications)

## Human Liver Anatomy



# Liver Physiology and Presentation of Primary Biliary Cholangitis

- In patients with PBC, the bile ducts are slowly destroyed by inflammation, which causes bile to remain in the liver, leading to scarring, cirrhosis, and liver failure.
- As many as half of patients with PBC present without symptoms.



# Primary Biliary Cholangitis: Presentation and Associated Disorders

- Most Common Symptoms
  - Fatigue
  - Pruritus
- Other Symptoms
  - Dry eyes and mouth
  - Fluid build-up in the ankles and abdomen
  - Collection of fatty deposits in the skin around the eye
  - Jaundice
- Associated Disorders
  - Problems with tear and salivary glands
  - Sjögren's syndrome, celiac disease, rheumatoid arthritis, and especially thyroid
  - Cholesterol gallstones

# Primary Biliary Cholangitis: Diagnosis

- 95% of cases occur in women aged 35 to 70.
- Often discovered through abnormal results on routine liver blood tests.
- Doctors need to do several tests to confirm a diagnosis of PBC.
- Most characteristic laboratory finding in PBC is the presence of positive antimitochondrial antibody (AMA), generally in a titer of 1:80 or higher.
  - More than 95% of patients with PBC have a positive AMA.
- Liver biopsy is often performed to confirm diagnosis.

# Primary Biliary Cholangitis: Pathophysiology and Progression

- As PBC progresses, it may become morphologically indistinguishable from other forms of cirrhosis.
- Liver biopsy helps evaluate the stage of PBC, which has 4 histologic stages.

**Stage 1:** Inflammation, abnormal connective tissue, or both, confined to the portal areas

**Stage 2:** Inflammation, fibrosis, or both, confined to the portal and periportal areas

**Stage 3:** Bridging fibrosis

**Stage 4:** Cirrhosis

# Primary Biliary Cholangitis: Treatment

- Early detection allows physicians to treat the disease before the liver is severely damaged.
- Patients most often take a daily dose of a drug called **ursodiol** (ursodeoxycholic acid or UDCA), a naturally occurring bile acid.
- In patients who do not respond or cannot tolerate UDCA, there are alternative therapies including the addition of obeticholic acid (**Ocaliva**), recently approved by the FDA for treatment of PBC in combination with UDCA. Ocaliva increases bile flow from the liver and suppresses bile acid production in the liver, thus reducing the exposure of the liver to toxic levels of bile acids.

# Primary Biliary Cholangitis: Treatment

- Ursodiol and Ocaliva improve the liver's ability to function in PBC patients.
- Extends life expectancy and may delay the need for a liver transplant.
- Patients rarely experience side effects from ursodiol.
- Obeticholic acid (Ocaliva) may aggravate or cause itching in some cases and can cause elevations in plasma lipids. Both of these side effects may respond to other medications.
- Fenofibrate may also be considered as an alternative therapy, although it is not yet FDA approved for PBC.



# Primary Biliary Cholangitis: Complications and Treatment

- Health care providers can treat symptoms and side effects of PBC
  - Itching: antihistamines; bile acid binding resins (cholestyramine); rifampicin.
  - Dry eyes and mouth: artificial tears and saliva substitutes
  - Portal hypertension: beta-blocker or nitrate
  - Varices: beta-blockers
  - Edema and ascites: diuretics
  - Osteoporosis: bisphosphonate medications
  - Gallstones and bile duct stones: surgery
- A health care provider may consider a liver transplant when cirrhosis leads to liver failure or treatment for complications is ineffective.

# Primary Biliary Cholangitis: Lifestyle Changes and Health

- A healthy lifestyle will help patients with PBC feel better—and may relieve or prevent some of the secondary symptoms of PBC.
- A reduced sodium diet
- Drinking ample water
- Taking calcium and Vitamin D supplements
- Avoiding or reducing the consumption of alcohol
- Reducing stress
- Exercising, particularly walking
- Skin care
- Regular dental examinations
- Artificial tears for dry eyes

# Primary Biliary Cholangitis: References and Further Resources

- **American Liver Foundation**  
Connecticut Division  
127 Washington Avenue  
North Haven, CT 06473  
(203) 234-2022  
[liverfoundation.org/chapters/CTALF](http://liverfoundation.org/chapters/CTALF)  
<http://www.liverfoundation.org/abouttheliver/info/pbc>
- **National Organization for Rare Disorders (NORD)**  
National Headquarters  
55 Kenosia Avenue  
Danbury, CT 06810  
(203) 744-0100  
[www.rarediseases.org](http://www.rarediseases.org)
- **Yale Liver Center**  
300 Cedar Street, Room S241  
New Haven, CT 06520  
(203) 785-5279  
[www.livercenter.yale.edu](http://www.livercenter.yale.edu)
- **Hartford Hospital Comprehensive Liver Center**  
85 Seymour Street  
Suite 320  
Hartford, CT 06106  
(860) 972-4219  
[www.hartfordhospital.org/contact-us/comprehensive-liver-center](http://www.hartfordhospital.org/contact-us/comprehensive-liver-center)
- **The National Institute of Diabetes and Digestive and Kidney Diseases**  
[www.niddk.nih.gov/health-information/health-topics/liver-disease/primary-biliary-cirrhosis](http://www.niddk.nih.gov/health-information/health-topics/liver-disease/primary-biliary-cirrhosis)
- **Interconnect Support Services**  
(844) 622-4278  
[www.interconnectsupport.com](http://www.interconnectsupport.com)

# Primary Biliary Cholangitis: References and Further Resources (cont.)

- **PBCers**  
(Largest PBC online support group, with almost 3000 members worldwide)  
PBCers Organization  
1430 Garden Road  
Pearland, TX 77581  
PBCersOrg96@aol.com  
[www.PBCers.org](http://www.PBCers.org)
- **PBC Foundation**  
(UK-based patient organization with nearly 10,000 members that provides support to people around the world who are living with PBC, as well as their families and friends)  
The PBC Foundation  
2 York Place  
Edinburgh EH1 3EP  
info@pbcfoundation.org.uk  
[www.PBCFoundation.org.uk](http://www.PBCFoundation.org.uk)
- **PBC Society**  
(Canadian organization with regional groups that provide support and encouragement, education and information, and fundraising for research on PBC)  
Canadian PBC Society  
4936 Yonge Street, Suite 221  
Toronto, Ontario M2N6S3  
info@pbc-society.ca  
[www.PBC-Society.ca](http://www.PBC-Society.ca)
- **National Institutes of Health**  
[www.nih.gov](http://www.nih.gov)
- **Mayo Clinic**  
[www.mayoclinic.org](http://www.mayoclinic.org)