PRIMARY BILIARY CHOLANGITIS (PBC)

Provider Information

Primary Biliary Cholangitis (PBC)

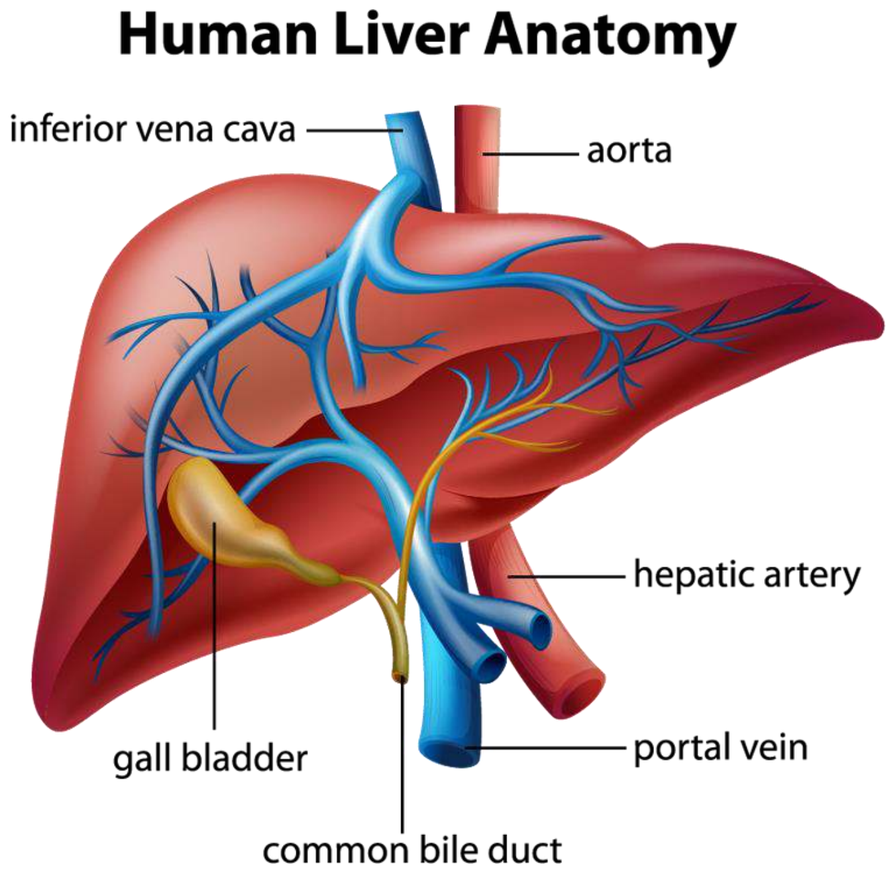
- Formerly known as primary biliary cirrhosis
- Chronic, autoimmune disease of the liver that slowly destroys the medium-sized bile ducts (intrahepatic bile ducts)
- Eventually causes cholestasis (disruption/obstruction of bile flow), cirrhosis (scarring and deterioration of liver), and liver failure

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.
- 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

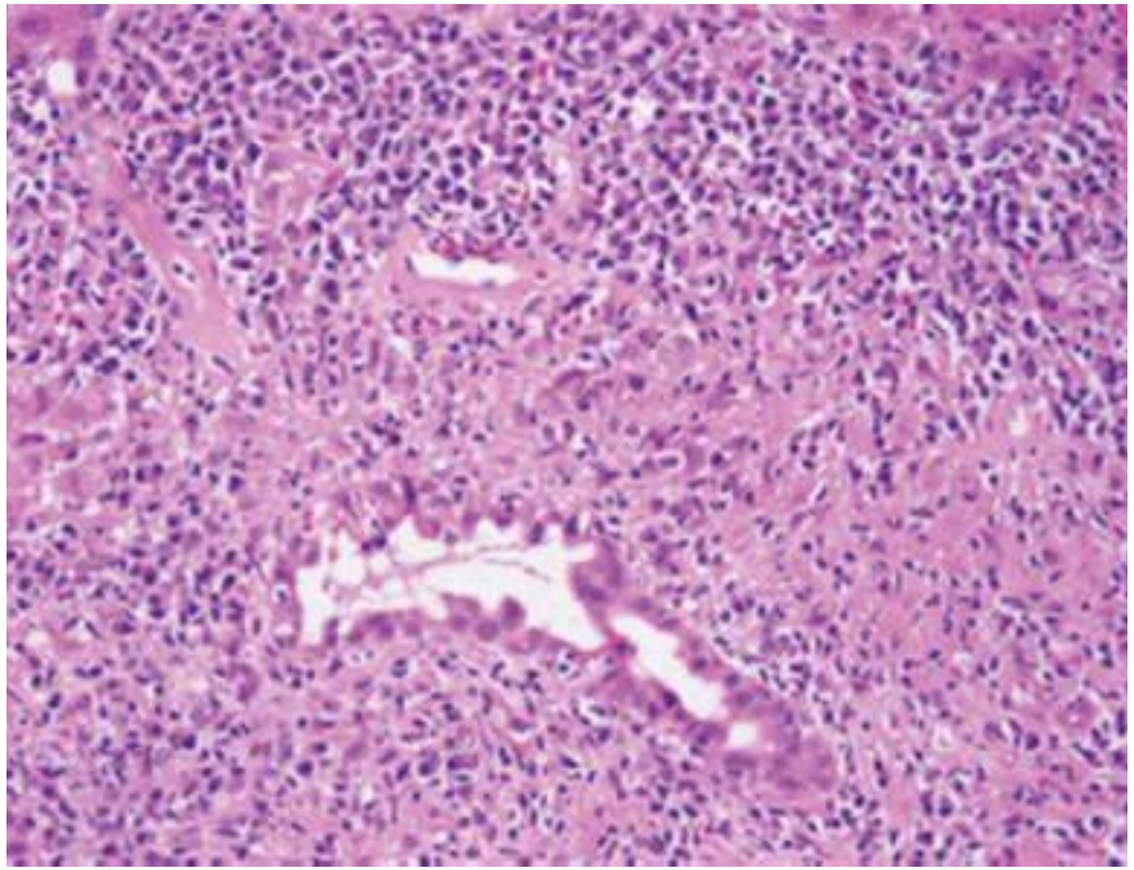
- Your liver is your body's largest internal organ
- The liver is the only solid organ that can regenerate.
- Your liver performs essential functions in the body:
  - Absorbing, storing, and processing nutrients from food
  - Making new proteins, such as albumin and clotting factors
  - Producing bile
  - Removing waste products the kidneys cannot remove (fats, cholesterol, toxins, and medications)



Liver Physiology and Presentation of PBC

- 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



PBC Presentation and Associated Disorders

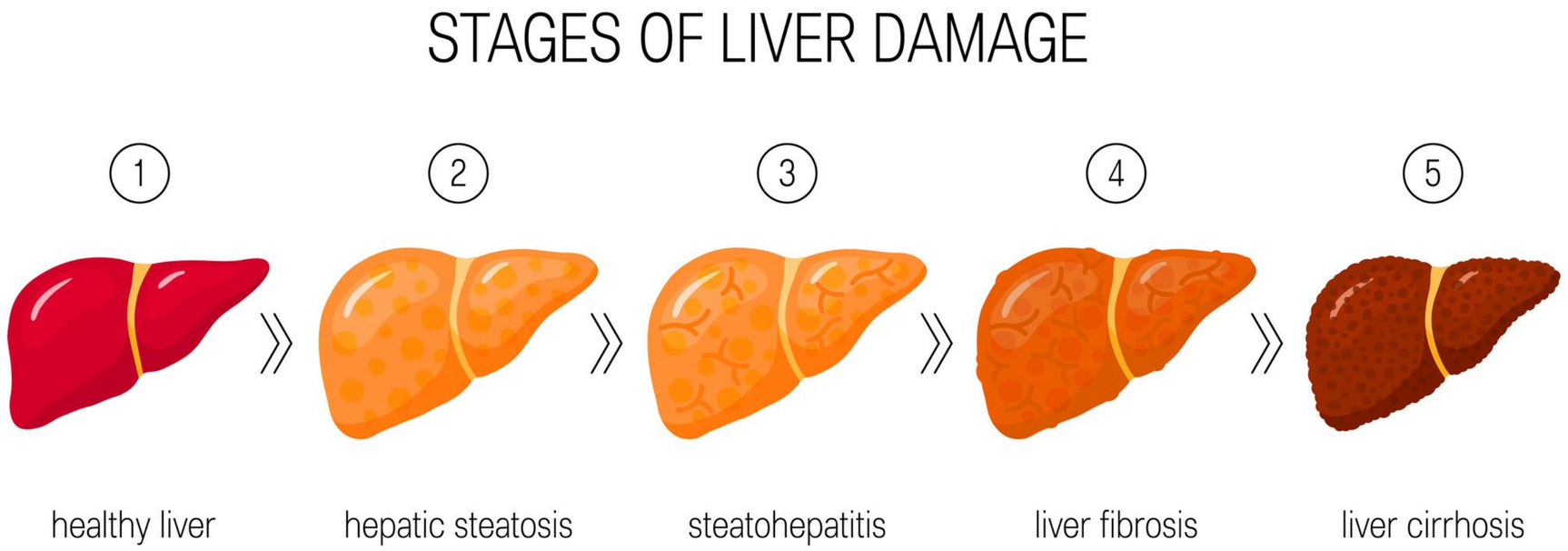
- Most Common Symptoms
  - Fatigue
  - Itching (or Pruritus)
- Other Symptoms
  - Dry eyes and mouth
  - Fluid build-up in the ankles and abdomen
  - Collection of fatty deposits in the skin around the eye
  - Jaundice
- Associated Disorders
  - Problems with tear and salivary glands
  - Sjögren's syndrome,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

PBC 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 positive AMA.
- Liver biopsy is often performed to confirm diagnosis.

PBC 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.



PBC 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.

PBC Treatment and Complications

- 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.

PBC Treatment and Complications

- 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

About ALF

Mission

ALF is the nation's largest patient advocacy organization for people affected by liver disease. Their mission is to promote education, advocacy, support services and research for the prevention, treatment, and cure of liver disease.

Programs

We offer a variety of free liver health education programs for patients, caregivers, and professionals on a variety of liver diseases and complications. To view upcoming free programs and community events visit liverfoundation.org/events.

Support Services

ALF also offers a free national Helpline (phone and chat), online support groups, interpretation services for non-English speakers and medically reviewed literature for patients. 1-800-GO-LIVER (1-800-465- 4837) or liverfoundation.org.

Get Involved

Getting involved is easy and we offer a variety of volunteer opportunities for patients, the public and healthcare professionals.

- Patients can explore volunteer opportunities at liverfoundation.org/how-you-can-help/get-involved
- Healthcare professionals can learn more about volunteer opportunities at liverfoundation.org/medical-professionals/ways-to-get-involved

We encourage patients to visit liverfoundation.org and hope healthcare professionals will include the link in patient follow-up and post-appointment summaries. ALF programs, support and resources are free and available nationwide.

For a complete list of free resources, visit liverfoundation.org/resource-center.