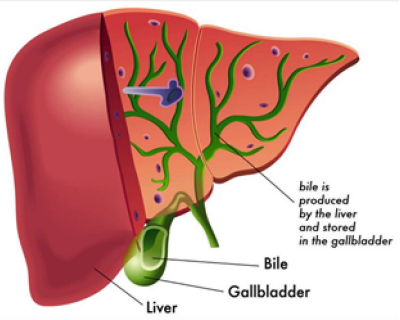
Primary Biliary Cholangitis (PBC)

Overview

- What is PBC?
  Primary Biliary Cholangitis (PBC) is a chronic liver disease that is characterized by inflammation and progressive destruction of the bile ducts. PBC is usually diagnosed in patients between the ages of 35 to 60 years.

  About 90 to 95 percent of patients are women. Liver inflammation over a long period of time may cause scarring which leads to cirrhosis. The cause of PBC is unknown, but it is most likely an autoimmune disease. An autoimmune disease is a process where the immune system attacks a specific tissue in the body. In PBC, it is thought that the immune system attacks and destroys small bile duct cells in the liver. It is not uncommon for patients with PBC to have other associated autoimmune diseases including pernicious anemia, scleroderma, or thyroid disorders. In addition, there are environmental factors such as infections, which may trigger the disease. Due to widespread laboratory screening, the number of patients being diagnosed early in the asymptomatic stage has risen dramatically over the past decade. More than 60 percent of newly diagnosed PBC patients now are asymptomatic.



- What are the signs and symptoms of PBC?
  Many patients may not have symptoms for years, and some may never have complaints relating to their disease. Because of the varying and sometimes subtle symptoms, doctors may have difficulty initially making the diagnosis. Signs and symptoms may include the following:

  - Hypercholesterolemia (elevated cholesterol levels)
  - Intense itching of the skin (pruritus)
  - Dry eyes and mouth (sicca syndrome)

  Most patients however are asymptomatic, and the diagnosis is made on the basis of an elevated alkaline phosphatase level in the blood that is ABOVE 3-4 times the normal level. .
- What type of tests are needed to evaluate PBC?
  Many patients are now diagnosed before symptoms are present. Typically, screening level blood tests show a higher than normal alkaline phosphatase (AP) level with a normal bilirubin. Usually, the bilirubin does not increase until a late stage of the disease. Your doctor may decide to conduct one or more of the following tests.

  - *Anti-mitochondrial antibody (AMA) blood test* - Up to 95% of patients with PBC will have a positive AMA test. This test is used to help confirm a diagnosis of PBC. However, the AMA tends to remain stable, and has no value in following disease progression, or in monitoring the response to treatment.
  - *Liver function tests* - Blood tests that are specific for liver disease are total bilirubin, alkaline phosphatase, aminotransferase levels (ALT, AST), albumin and prothrombin time.
  - *Cholesterol and triglyceride levels* - These blood tests are often increased.
  - *Ultrasound exam of the liver* - Images of the liver and bile ducts are taken to assess for liver abnormalities. These imaging studies are usually normal in PBC.

  Liver biopsy - A small sample of liver tissue helps to confirm the diagnosis and determines the severity of the liver damage. A liver biopsy is not always necessary, especially if the AMA is positive, the AP is 3-4 times normal, and the imaging studies are normal.
- Treatment of underlying disease
  • Ursodiolor Ursodeoxycholic acid (UDSA)

  Ursodiol as a drug is a "gentler" bile acid than those normally made by the body and is well tolerated. Studies have shown that ursodiol delays the time to liver transplantation. In addition to improving blood tests, there are some patients with PBC whose disease appears to become inactive (remission) on ursodiol.

  • Obeticholic Acid

  Obeticholic Acid is now an FDA approved medication for PBC in patients who are intolerant to Ursodiol or have inadequate response to Ursodiol

- Treatment of complications of PBC
  Some patients have complications of PBC that require therapy. In many cases, doctors find that symptoms can be controlled by lifestyle modifications and medications.

  - Itching
    Itching can be a very troublesome symptom of PBC. Itching may range from mild to severe, where a patient's lifestyle may be impaired. Itching may be worse with fatty meals or at night, interfering with sleep. The initial medications typically used by doctors include diphenhydramine,

cholestyramine and colestipol. Rifampin is used in patients who fail or are intolerant to cholestyramine and colestipol. In resistant cases, opioid antagonists such as nalmefene, naloxone and naltrexone are used. Plasmapheresis and ultraviolet light have been utilized in extreme cases. Liver transplantation is indicated for patients with uncontrolled itching.

- Osteoporosis
  When the diagnosis of PBC is made, a bone mineral density scan (DEXA scan) should be performed to determine if a patient has osteoporosis or bone thinning. In many cases, doctors may suggest that their patients start exercising if possible, stop smoking and take calcium with vitamin D. Hormone replacement therapy (estrogen) is safe in PBC and is recommended where appropriate. When osteoporosis is evident, therapy with medications such as Fosamax® (alendronate) and Actonel® may be helpful in PBC.
- Fat-soluble vitamin deficiencies
  As PBC progresses, some patients lose the ability to absorb fat-soluble vitamins A, D, E and K. Measurement and replacement of these vitamins is recommended.
- Undiagnosed and Untreated PBC
  Undiagnosed and untreated PBC can lead to liver cirrhosis, which can then have complications like Esophageal varies, ascites, hepatic encephalopathy or hepatocellular carcinoma. Patients with PRBC who have advanced fibrosis or cirrhosis should undergo EGD for screening of varies.
- Fatigue is the most common and clinically relevant complication in PBC
- Obeticholic Acid
  Obeticholic Acid is now an FDA approved medication for for PBC in patients who are intolerant to UDCA or have inadequate response to UDCA. We should add a section on Fibrates as an investigational drug for use in PBC.
- Liver Transplantation
  For those people who reach the late stages of PBC, liver transplantation may be considered. The signs and symptoms of liver failure, which may or (depending on the specific aspects of the symptoms / disease) may not prompt consideration of any potential role for liver transplant, are the following:

  - Persistent jaundice
  - Fluid retention with swelling of hands, feet and abdomen
  - Variceal bleeding (internal bleeding from swollen intestinal blood vessels, commonly in upper stomach and esophagus)
  - Bone thinning and/or fractures
  - Hepatic coma or encephalopathy

  The survival for PBC patients after liver transplantation is excellent although some patients may develop recurrent PBC over time. With better understanding of the nature of PBC and its complications, many patients will have improved outcomes.

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