

Presentation

Women are affected ten times more frequently than men. The disease usually is first diagnosed in people 30 to 60 years old. Many patients have no symptoms of disease and are diagnosed by finding an abnormality on routine liver blood tests. Itching and fatigue are common symptoms. Other signs include jaundice, cholesterol deposits in the skin, fluid accumulation in the ankles and abdomen, and darkening of the skin. Several other disorders are often associated with PBC. The most common is impaired functioning of the tear and salivary glands, causing dry eyes or mouth. Arthritis and thyroid problems may also be present. Renal stones and gallstones may develop. Bone softening and fragility leading to fractures can occur in late stages of the disease.

Diagnosis

PBC diagnosis is based on several pieces of information. The patient may have symptoms (itching) suggesting bile duct damage. Laboratory tests, such as the alkaline phosphatase activity test, may confirm this. The test for mitochondrial antibodies is particularly useful as it is positive in nearly all patients. Infrequently, the bile ducts are X-rayed to rule out the possibilities of other causes of biliary tract disease, such as obstruction. A liver biopsy is useful in confirming the diagnosis and in giving information on the severity and extent of liver damage.

Cause

Although the cause of the initial bile duct damage in PBC is unknown, there are certain clues that may be important. Strictly speaking, the disease is not inherited, but it is more common among siblings and in families where one member has previously been affected. Multiple disturbances of the immune system have been found in persons with PBC and may be an important factor. Hormones may also play a role given that this illness is so much more common in women.

Prognosis

PBC typically advances slowly. Many patients lead active and productive lives for ten to fifteen years after diagnosis. Patients who show no symptoms at the time of diagnosis often remain symptom-free for years. Jaundice appears to be a sign of diminishing liver reserve and may be an important indication regarding the progression of the disease. The illness is chronic and may lead to life-threatening complications, especially after cirrhosis develops.

Treatment

Treatment may be useful in several ways. Proper advice will ensure the elimination of potentially harmful drugs, foods, or toxins. If the patient is deficient in vitamin D, then this should be corrected. The thyroid function should be tested and, if low, treated with a thyroid hormone. Symptoms may be successfully relieved. Itching is often reduced by using cholestyramine and rifampin. Salt restriction may be effective in reducing fluid accumulation. The diet should be well-balanced. Corticosteroids have been found ineffective in most patients. Ursodeoxycholic acid improves liver tests, symptoms, and survival. The FDA has approved only URSO 250 (manufactured by Axcan Scandipharm) for use in patients with PBC.* The recommended dose is 13-15 mg/kg once a day.

**Physicians who treat PBC patients often prescribe other ursodiol products off-label. These products include Actigall (manufactured by Novartis) and various generic formulations.*

Liver Transplantation

When medical treatment no longer controls the disease and the patient has severe liver failure, transplantation is indicated. Signs of liver failure include accumulation of fluid in the abdomen, malnutrition, gastrointestinal bleeding, intractable itching, jaundice, and bone fractures. Transplantation may be recommended before all these events occur. The outcome for patients with PBC who have undergone transplantation is excellent. The survival rate for two or more years is about 80 percent. The use of new drugs to suppress rejection has made transplantation even more successful. The disease's slow progress makes it possible to plan elective transplant surgery.

The Future

PBC has been known for more than 100 years. This knowledge has led doctors to make earlier diagnoses. Many clues to the cause have been supplied by careful observation of patients over the last 25 years, but the basic cause is unknown.

Research is following two paths:

- Basic investigation of the causes and development of the disease.
- Drug therapy trials, involving a large number of patients around the world, are exploring the potential use of several additional medications to lessen the symptoms and control liver damage.

Primary Biliary Cirrhosis

Primary biliary cirrhosis (PBC) is a chronic liver disease that causes slow, progressive destruction of bile ducts in the liver. This destruction interferes with the excretion of bile. Continued liver inflammation causes scarring and eventually leads to cirrhosis. Cirrhosis is present only in the later stage of the disease. In the early stages of the illness, the main problem is the build up of substances (like bile acids, cholesterol) in the blood, which are normally excreted into the bile. Ursodeoxycholic acid is a life-saving, safe, and approved therapy.

For further information, contact:



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The American Liver Foundation is a non-profit, national voluntary health organization dedicated to the prevention, treatment, and cure of hepatitis and other liver diseases through research, education, and advocacy.

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What is Primary Biliary Cirrhosis?

