Related Diseases

What Is Primary Biliary Cholangitis?

Primary biliary cholangitis (PBC) is a chronic, or long-lasting, disease that causes the small bile ducts in the liver to become inflamed and damaged and ultimately disappear. Primary biliary cholangitis used to be called primary biliary cirrhosis.

The bile ducts carry a fluid called bile from the liver to the gallbladder, where it is stored. When food enters the stomach after a meal, the gallbladder contracts, and the bile ducts carry bile to the duodenum, the first part of the small intestine, for use in digestion. The liver makes bile, which is made up of bile acids, cholesterol, fats and fluids. Bile helps the body absorb fats, cholesterol and fat-soluble vitamins. Bile also carries cholesterol, toxins and waste products to the intestines, where the body removes them. When chronic inflammation, or swelling, damages the bile ducts, bile and toxic wastes build up in the liver, damaging liver tissue.

This damage to the liver tissue can lead to cirrhosis, a condition in which the liver slowly deteriorates and is unable to function normally. In cirrhosis, scar tissue replaces healthy liver tissue, partially blocking the flow of blood through the liver. People with cirrhosis are at risk of developing liver cancer.

Studies show that about 90 percent of people with PBC are female and that about 10 percent of people with typical features of PBC will have additional features of autoimmune hepatitis (AIH). A subset, however, have no such features but go on to develop an AIH overlap syndrome.

What causes PBC?

The causes of primary biliary cholangitis are unknown. Most research suggests it is an autoimmune disease. The immune system protects people from infection by identifying and destroying bacteria, viruses and other potentially harmful foreign substances. An autoimmune disease is a disorder in which the body's immune system attacks the body's own cells and organs. In primary biliary cholangitis, the immune system attacks the small bile ducts in the liver.

Genetics, or inherited genes, can make a person more likely to develop primary biliary cholangitis. Primary biliary cholangitis is more common in people who have a parent or sibling—particularly an identical twin—with the disease. In people who are genetically more likely to develop primary biliary cholangitis, environmental factors may trigger or worsen the disease, including exposure to toxic chemicals, smoking and infections.

What are the symptoms of PBC?

The first and most common symptoms of primary biliary cholangitis are fatigue, or feeling tired; itching skin and darkened skin in itching areas due to scratching; dry eyes and mouth.

Some people may have jaundice, a condition that causes the skin and whites of the eyes to turn yellow. Health care providers diagnose up to 60 percent of people with primary biliary cholangitis before symptoms begin. Routine blood tests showing abnormal liver enzyme levels may lead a health care provider to suspect that a person without symptoms has primary biliary cholangitis.

How is PBC diagnosed?

A health care provider may use the following tests to diagnose primary biliary cholangitis: a medical and family history, a physical exam, blood tests, imaging tests or a liver biopsy.

A health care provider usually bases a diagnosis of primary biliary cholangitis on two out of three of the following criteria: a blood test showing elevated liver enzymes, a blood test showing the presence of anti-mitochondrial antibodies (AMA) or a liver biopsy showing signs of the disease.

Health care providers may order additional tests to rule out other causes of symptoms. Health care providers diagnose the majority of people with primary biliary cholangitis early in the course of the disease.

How is PBC treated?

Treatment for primary biliary cholangitis depends on how early a health care provider diagnoses the disease and whether complications are present. In the early stages of primary biliary cholangitis, treatment can slow the progression of liver damage to cirrhosis. In the early stages of cirrhosis, the goals of treatment are to slow the progression of tissue scarring in the liver and prevent complications. As cirrhosis progresses, a person may need additional treatments and hospitalization to manage complications.

Health care providers prescribe ursodeoxycholic acid (UCDA), also known as ursodiol (Actigall, Urso), to treat primary biliary cirrhosis. Ursodiol is a nontoxic bile acid that people can take orally. Ursodiol replaces the bile acids that are normally produced by the liver, which are more toxic and can harm the liver. Treatment with ursodiol can reduce levels of bilirubin and liver enzymes in the blood.

Researchers are studying the effects of several other medications on the progression of primary biliary cholangitis. For example, the U.S. Food and Drug Administration (FDA) has granted accelerated approval to obeticholic acid (Ocaliva) for the treatment of PBC to be used in combination with ursodiol for people who do not respond to treatment with ursodiol alone. After additional review, the FDA may grant approval of obeticholic acid as a monotherapy to treat PBC for people who can’t take ursodiol.

Patients who do not completely respond to UDCA may be prescribed fenofibrate. Medications to suppress the immune system, such as prednisone or azathioprine, may also be prescribed for PBC patients diagnosed with autoimmune hepatitis.

Source: U.S. National Institute of Diabetes and Digestive and Kidney Diseases

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