Primary biliary cirrhosis

is a progressive disease of the liver caused by a buildup of bile within the liver (cholestasis) that results in damage to the small bile ducts that drain bile from the liver. Over time, this pressure build-up destroys the bile ducts leading to liver cell damage. As the disease progresses and enough liver cells die, cirrhosis and liver failure occur.

Bile is manufactured in the liver and then transported through the bile ducts to the gallbladder and intestine where it helps digest fats and fat soluble vitamins (A,D,E,K). When bile cannot be drained from the liver, it causes inflammation that leads to cell death. Scar tissue gradually replaces the areas of damaged liver and then the body cannot perform necessary functions.

Symptoms

The most common initial symptoms are fatigue or a sense of being abnormally tired and, skin that itches. Because of the intense itching and scratching, the skin may become darkened and discolored in some areas. The person may also complain of dry mouth and eyes.

Because there may be liver inflammation, some people may experience right upper quadrant abdominal pain, where the liver is located.

Causes

Should the liver damage progress, the symptoms of cirrhosis may develop. These include muscle wasting, ascites (swelling of the abdomen due to fluid accumulation), leg swelling, jaundice (yellow coloration of the skin), and confusion.

Risk factors

- There may be a genetic component to the development of PBC since a person with the disease is likely to have a family member also afflicted.
- Women are nine times more likely than men to develop PBC. It most often develops in the middle age between the ages of 40 to 60.
- The disease is seen more often in white, northern Europeans, compared to African-Americans.

Complications

If PBC is not treated or reaches an advanced stage, there may be other problems including:

- osteoporosis – a condition in which the bones become weak and brittle
- portal hypertension – increased blood pressure inside the blood vessels in your abdomen
- ascites – a build-up of fluid in your abdomen (stomach) and around your intestines
- vitamin deficiencies – including vitamins A, D, E and K
- a slightly increased risk of developing liver cancer

Diagnosis

Many people are only diagnosed with PBC after having a routine blood test for another reason. PBC can usually be diagnosed just using blood tests.

Once PBC is diagnosed, you'll also need an ultrasound scan to help rule out other problems with your bile ducts and assess your liver.

A liver biopsy is occasionally recommended to assess your liver and help doctors decide on the best treatment.

This involves safely removing a small sample of liver tissue so it can be studied under a microscope.

Treatment

PBC is a progressive condition, which means the damage to the liver can steadily get worse over time.

The rate at which PBC progresses varies between individuals. Sometimes, it can take decades.

Without treatment, the liver can become so badly damaged that it no longer works properly. This is known as liver failure and can be fatal.

Liver failure can be prevented in the majority of people being treated for PBC with current treatments such as ursodeoxycholic acid and obeticholic acid.

Other medicines can help relieve the itchiness associated with PBC. Occasionally, if the liver is severely damaged, a liver transplant may be needed.