

Primary biliary cirrhosis

Primary biliary cirrhosis (PBC) is a long-term liver disease, which affects the bile ducts in the liver. It is a so-called autoimmune condition. That means that the immune system, which is the body's natural defence against infection or illness, mistakenly recognizes the bile ducts within the liver as "foreign" and damages or destroys them. As a consequence, bile accumulates in the liver leading to further destruction of bile ducts and eventually to scarring of the liver, which may result in cirrhosis.

In the early stages of the disease some people do not have symptoms, however over the course of their illness many experience itching of the skin, tiredness and pain or discomfort in the upper right corner of their tummy.

Many patients will only be diagnosed with PBC after a routine blood test shows abnormal liver function tests indicating that there is a problem with their liver. To diagnose PBC the AMA (anti-mitochondrial antibody) blood test is used, as it is the most accurate (95%) diagnostic tool for PBC. In rare cases, AMA blood tests can be negative in PBC, in which case a liver biopsy is needed to make a reliable diagnosis.

About 90% of those who suffer from PBC are women and most cases are diagnosed at the age of 40-60. Nonetheless, PBC can occur at any age from 20 onwards.

PBC is a progressive disease, which means that the liver function can get worse over time. Therefore, maintaining a healthy life style and treatment of the condition with drugs is advised to protect the liver. The most useful drug for patients with PBC is ursodeoxycholic acid (UDCA). It can help preventing or delaying liver damage, especially when it is taken from an early stage of disease. However, UDCA usually does not improve symptoms, such as tiredness or itchy skin and therefore other drugs (for example cholestyramine for itch) can be used to treat specific symptoms.

Despite treatment and regular blood tests, liver damage progresses in a small number of patients and may result in cirrhosis. If the functions of the liver then become impaired (usually demonstrated by worsening jaundice) a liver transplant may be required. Liver transplantation is very successful in patients with PBC. Nevertheless there is a small risk that PBC occurs even in the transplanted liver.

Thus, the earlier PBC is diagnosed, the easier it is to prevent liver damage by treatment with UDCA, and the better patients can self-manage their condition in cooperation with their physician.