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# Cystic Diseases Of The Liver

## What is the liver and what does it do?

Situated beneath the diaphragm in the upper right part of the abdomen, the liver is the largest internal organ in the body (weighing 1-1.5 kg in adults). The liver receives an arterial blood supply from the hepatic artery, as well as blood from the intestinal tract via the portal vein. All digested and absorbed materials and nutrients from the intestinal tract are processed, stored or excreted by the liver for transport into the blood. Harmful materials, also carried to the liver by portal blood, are transformed into compounds that are less toxic for the body.

Among hundreds of other tasks, the liver produces bile. This green-yellow fluid flows into the intestine through the bile ducts in response to digested food in the small intestine. Bile is needed for absorption of fat and some vitamins. The bile ducts in the liver are like the branches on a tree, that come together just below the stomach. These bile ducts are often referred to as the biliary tree. A side branch leads to a sac for storing bile, called the gall bladder.

## What is cystic disease of the liver?

Cystic disease of the liver is rare and can take several forms. Cysts in the main trunk of the biliary tree are called choledochal cysts. Cysts (lakes) that occur in the small branches of bile ducts within the liver are referred to as Caroli's syndrome. The other cysts in the liver that do not occur in the biliary tree are referred to as polycystic liver disease.

## What is a choledochal cyst?

This condition typically occurs in the main part of the biliary tree (common bile duct) and typically occurs because the bile duct is structurally abnormal (dilated), probably from the time of birth. Eventually, usually by age two or three, but sometimes not until adolescence or adulthood, bile accumulates in the duct resulting in damage to the duct and formation of a sac or cyst which then prevents bile from reaching the intestine. When this happens, bile backs up into the liver and the patient develops jaundice (a yellowing of the skin and eyes). Occasionally, this accumulated bile becomes infected, causing abdominal pain and fever. In some patients the cyst can be felt by the doctor examining the abdomen. In most patients the diagnosis can be confirmed by ultrasound or by injecting a radioactive substance which gives an "image" of the abnormal duct. Treatment is surgical. The abnormal bile duct is removed and a piece of intestine is used to replace it. In most cases, surgery permanently corrects the disease. Rarely, infection in the newly formed biliary tree can reoccur. If the condition is not correctly diagnosed, the blockage of bile may result in scarring in the liver (cirrhosis).

## What is Caroli's syndrome?

Caroli's syndrome (intrahepatic ductal ectasia) is another rare congenital disease (existing from the time of birth). It is probably inherited. In this condition, the small branches of the biliary tree in the liver are abnormal. Small lakes alternate with narrowed segments of the bile ducts, instead of the normal smooth contour. Some ducts can be dilated. These abnormalities may be present throughout the liver, or may be limited to only a small area. If the biliary tree becomes infected, the patient develops fever, abdominal pain and, rarely, jaundice. This complication may first appear in childhood or may not occur until middle age. This disease is usually diagnosed by using radioisotopes to "image" the biliary tree. Other procedures include injecting dye by inserting a needle through the skin into the liver (percutaneous transhepatic cholangiogram) or by using a tube to pass dye through the intestine up into the bile duct (endoscopic retrograde cholangiography).

## What is congenital hepatic fibrosis?

In patients with congenital hepatic fibrosis, there is abnormal growth of fibrous (scar) tissue around the small branches of the bile ducts in the liver. As a result, the liver becomes enlarged and hard and blood can no longer flow freely through the liver. This can result in damage to the liver and create high amounts of pressure (portal hypertension) within the blood system. Blood from the intestines, is then forced to find a new way around the liver through new vessels. Some of these new blood vessels called "varices" which form primarily in the stomach and esophagus become quite large. These varices may rupture due to high blood pressure (portal hypertension) and thin vessel walls, causing bleeding in the upper stomach or esophagus. Patients with this condition are usually discovered in childhood, either because they get an enlarged liver or because of bleeding. The diagnosis is proven by liver biopsy and x-rays of blood vessels. There is no specific treatment for this condition but many patients require rerouting of blood from the intestines (shunt operation) to prevent bleeding.

## What is polycystic liver disease?

In patients with polycystic liver disease, large lakes (cysts) separate from the biliary tree form in the liver. In severe cases, the liver looks like a sponge. These cysts may cause pain in some people, but do not affect liver function. In most patients, the kidneys are similarly affected with cysts, which may cause high blood pressure and kidney failure. The tendency to form cysts is probably present at birth in these patients, but usually the cysts do not enlarge and give problems until adulthood. This condition may be detected using ultrasound or CAT scan and x-rays of the kidney (intravenous pyelogram). Polycystic disease is inherited and once it has been detected in one member of a family, all the patient's relatives should be tested for it. There are two major categories of polycystic disease of the liver and kidney. In the more benign form, the cysts are mostly in the liver and kidney function is near normal. These patients have normal life expectancy. However, patients who have kidney damage need treatment for the equivalent of polycystic kidney disease.

*This information is current as of December 2015*