**Alcohol Liver Disease**

**How does alcohol affect the liver?**

Alcohol can damage or destroy liver cells. The liver breaks down alcohol so it can be removed from your body. Your liver can become injured or seriously damaged if you drink more alcohol than it can breakdown and process.

**What are the different types of alcohol-related liver disease?**

There are three main types of alcohol-related liver disease:

* fatty liver disease
* alcoholic hepatitis
* alcoholic cirrhosis

**Fatty liver disease**

Fatty liver disease is the buildup of extra fat in liver cells. It is the earliest stage of alcohol-related liver disease. There are usually no symptoms. If symptoms do occur, they may include fatigue, weakness, and weight loss. Almost all heavy drinkers have fatty liver disease. When you stop drinking, the fatty liver disease will usually go away.

**Alcoholic Hepatitis**

When alcohol damages the liver and there is swelling this is called Alcoholic Hepatitis. This may include loss of appetite, nausea, vomiting, abdominal pain, fever and jaundice. Up to 35 percent of heavy drinkers develop alcoholic hepatitis. Alcoholic hepatitis can be mild or severe. If it is mild, liver damage may be reversed. If it is severe, it may occur suddenly and lead to serious complications including liver failure and death.

**Alcoholic Cirrhosis**

Alcoholic cirrhosis occurs when alcohol damaged soft healthy tissue is the replaced by scar tissue. It is the most serious type of alcohol-related liver disease. Symptoms of cirrhosis are similar to those of alcoholic hepatitis. Between 10 and 20 percent of heavy drinkers develop cirrhosis. The damage from cirrhosis cannot usually be reversed and can cause liver failure. Not drinking alcohol can help prevent further damage.

**How does alcohol-related liver disease progress?**

Many heavy drinkers will progress from fatty liver disease to alcoholic hepatitis to alcoholic cirrhosis over time. However, some heavy drinkers may develop cirrhosis without having alcoholic hepatitis first. Others may have alcoholic hepatitis but never have symptoms. Heavy drinkers who also have a chronic liver disease such as hepatitis C are at high risk for developing cirrhosis.

**What are the complications of alcohol-related liver disease?**

Complications from alcohol-related liver disease usually occur after years of heavy drinking. The complications can be serious.

They may include:

* buildup of fluid in the abdomen
* bleeding from veins in the esophagus or stomach
* enlarged spleen
* high blood pressure in the liver
* brain disorders and coma
* kidney failure
* liver cancer

**How is alcohol-related liver disease diagnosed?**

Alcohol-related liver disease may be suspected based on medical conditions related to alcohol abuse. Blood tests may be used to rule out other liver diseases. Your doctor also may need to do a liver biopsy. During a biopsy, a small piece of liver tissue is removed and studied in the lab.

 **How is alcohol-related liver disease treated?**

Treatment for alcohol-related liver disease requires a healthy diet including avoiding alcohol. Your doctor may suggest changes in your diet to help your liver recover from the alcohol-related damage. Treatment may require you to participate in an alcohol recovery program. Medications may be needed to manage the complications caused by your liver damage. In advance cases of alcoholic cirrhosis, a liver transplant may be needed. Those with alcohol-related liver disease need to stop drinking alcohol to be considered for a liver transplant.

**Autoimmune Hepatitis**

Autoimmune hepatitis is a disease in which the body’s own immune system attacks the liver and causes it to become inflamed. The disease is chronic, meaning it lasts many years. If untreated, it can lead to cirrhosis and liver failure.

**What causes autoimmune hepatitis?**

Your immune system normally attacks bacteria, viruses and other invading organisms. It is not supposed to attack your own cells; if it does, the response is called autoimmunity. In autoimmune hepatitis, your immune system attacks your liver cells, causing long-term inflammation and liver damage. We don’t know why the body attacks itself in this way. Some medication and supplements incorrectly turn on the immune system. Also, predisposition, hereditary, and prior infections may stimulate the immune system.

**What are the symptoms and complications of autoimmune hepatitis?**

Often, the symptoms of autoimmune hepatitis are minor. When symptoms do occur, the most common are fatigue, mild case of the flu, abdominal discomfort, aching joints, itching, jaundice (yellowing of the skin and whites of the eyes), enlarged liver, nausea, dark urine, loss of appetite, pale stools and absence of menstruation. More severe complications can include ascites (fluid in the abdomen) and mental confusion. In 10%-20% of cases, autoimmune hepatitis may present with symptoms like an acute hepatitis.

**How is autoimmune hepatitis diagnosed?**

To make a diagnosis of autoimmune hepatitis, we use blood tests and sometimes a liver biopsy, in which a sample of liver tissue is removed with a needle for examination in a laboratory.

**How is autoimmune hepatitis treated?**

The goal of treatment is to stop the body’s attack on itself by suppressing the immune system. This is accomplished with a medicine called prednisone, a type of steroid. Often times, a second drug, azathioprine (Imuran) is also used. Treatment starts with a high dose of prednisone. When symptoms improve, the dosage is lowered and azathioprine may be added. In most cases, autoimmune hepatitis can be controlled but not cured. That is why most patients will need to stay on the medicine for years, and sometimes for life. Unfortunately, long-term use of steroid can cause serious side effects including diabetes, osteoporosis, high blood pressure, glaucoma, weight gain and decreased resistance to infection.

**Who is at risk for autoimmune hepatitis?**

About 70 percent of people with autoimmune hepatitis are women, usually between the ages of 15 and 40. Many have other autoimmune diseases, including type 1 diabetes, thyroiditis (inflammation of the thyroid gland), ulcerative colitis (inflammation of the colon), vitiligo (patchy loss of skin pigmentation), or Sjogren ’s syndrome (dry eyes and mouth).

**Hepatitis A**

**What is hepatitis A?**

Hepatitis A is a liver disease caused by the hepatitis A virus (HAV). HAV causes the liver to swell and prevents it from working well. HAV usually goes away on its own in almost all cases with no serious complications. However, HAV may cause some patients to suffer liver failure. In the United States, there are about 100 deaths a year due to HAV. Those at risk of serious long term effects from HAV include people with other liver diseases and people over 60.

**Who is at risk of having hepatitis A?**

Anyone who has come in close contact with someone who has HAV or who has eaten food or drank water polluted by HAV is at risk.

* Have ever lived with an infected person?
* Have ever been a sexual partner of an infected person?
* Are a man who has sex with men?
* Have ever used drugs?
* Have ever traveled to countries where HAV is common (Latin America, Middle East or Asia)?

HAV is most commonly spread by:

* Not washing hands before preparing or eating food
* Not washing hands after using the bathroom or changing a diaper
* Eating raw or undercooked shellfish that came from waters polluted by sewage

**What are symptoms of hepatitis A?**

Low energy is the most common symptom of HAV. Other symptoms include fever, tiredness, loss of appetite, nausea, headache, itchy skin, muscle soreness, pain near the liver, and jaundice (a yellowing of the skin and whites of the eyes). Symptoms of HAV can occur two to seven weeks after infection and are often mild. Children may not have any symptoms. Symptoms usually go away within two months. If you think you have HAV, it is important to see a doctor -- symptoms of HAV are similar to other more serious liver diseases.

**How is hepatitis A diagnosed?**

Hepatitis A is diagnosed by a blood test. A blood test is done to see if HAV antibodies are in the body. Antibodies are proteins created by the immune system in response to viruses.

**How is hepatitis A treated?**

HAV usually goes away on its own within six months. Doctors often recommend bed rest, drinking lots of fluids, eating a healthy diet and avoiding alcohol. Medicines are not used to treat HAV. Talk to your doctor before taking prescription or over-the-counter drugs, vitamins or herbal supplements. Itchy skin caused by HAV can be treated with non-prescription anti-itch medicine. It is important to see your doctor regularly to make sure your body has fully recovered from the virus. Also, talk to your doctor about getting vaccinated for hepatitis B.

**What is the best way to stop the spread of hepatitis A?**

Hepatitis A vaccination is the best way to prevent hepatitis A. The hepatitis A vaccine is given in 2 doses, usually about 6 months apart.

Other ways to stop the spread of HAV are:

* Always washing your hands with soap and warm water immediately after using the bathroom or changing a diaper
* Always washing your hands with soap and warm water before preparing or eating food

**Who should be vaccinated against hepatitis A?**

Those who should get vaccinated against HAV include:

* All children at age 1
* All children above age 1 who live in areas where HAV is common
* People with long-term liver disease
* People with blood-clotting disorders
* People who have had or are waiting for a liver transplant
* People who use drugs
* Men who have sex with men
* Travelers to countries where HAV is common
* Sexual partners and household members of people with HAV

If you think you have come in contact with HAV, your doctor may give you a HAV vaccination or a shot of immune globulin, which can help increase protection to HAV.

**Hepatitis B**

Hepatitis B is a liver disease caused by the hepatitis B virus (HBV). HBV causes the liver to swell and prevents it from working well. About 95% of adults who are exposed to HBV fully recover within six months (acute HBV) without medication. About 5% have HBV all their lives (chronic HBV) unless they are successfully treated with medications. Infants born to mothers infected with HBV are at high risk of developing chronic HBV. Chronic HBV can lead to cirrhosis (scarring) of the liver, liver cancer, and liver failure.

**Who is at risk of having hepatitis B?**

Anyone who has come in direct contact with HBV-infected bodily fluids (blood, semen, and vaginal secretions) is at risk.

* Were born to an HBV-infected mother
* Have ever worked with or come in contact with infected bodily fluids.
* Have ever lived with an infected person
* Have ever had unprotected sex with an infected person
* Have ever had multiple sexual partners
* Have ever had a sexually transmitted disease
* Are a man who has sex with men
* Have ever injected or inhaled drugs (even once)
* Have ever worked or been housed in a prison
* Have ever traveled to countries where HBV is common
* Have ever been on hemodialysis

**What are symptoms of hepatitis B?**

Many people with acute or chronic HBV have no symptoms. When symptoms occur, they may include tiredness, fever, loss of appetite, nausea, headache, muscle soreness, pain near the liver and jaundice (yellowing of the skin and whites of the eyes). Symptoms often begin two to five months after infection. Symptoms usually last for several weeks, but can last up to six months.

**How is hepatitis B diagnosed?**

Hepatitis B is diagnosed by blood tests.

Blood tests are done to check if HBV antibodies are in the body. Antibodies are proteins created by the immune system in response to viruses.

**How is hepatitis B treated?**

Acute HBV

Doctors often recommend bed rest, drinking lots of fluids, eating a healthy diet and avoiding alcohol. Medicines are not used to treat acute HBV. It is important to see your doctor regularly to make sure your body has fully recovered from the virus.

Chronic HBV

There are several treatment options for chronic HBV: tenofovir, interferon alfa 2b, pegylated inferon alfa 2a, entecavir, and telbivudine. These medicines may not work for all people with hepatitis B. Also, patients taking these medicines need to be monitored by their doctors for side effects.

HBV medications should not be taken by pregnant women unless recommended by their doctors. Some pregnant women with HBV can be treated to prevent transmitting HBV to their babies.

If you have HBV, it is important to talk to your doctor about treatment options and liver cancer screenings every 6-12 months. Also, talk to your doctor about the hepatitis A vaccine.

**What is the best way to stop the spread of hepatitis B?**

Hepatitis B vaccination in people who have not been exposed to HBV is the best way to prevent infection. The hepatitis B vaccine is given in 3 doses. The first 2 doses are given one month apart and the final dose is given 6 months later.

**Other ways to stop the spread of HBV are:**

* Getting tested if you are pregnant or want to become pregnant
* Not sharing needles
* Practicing safe sex
* Not sharing razors, toothbrushes, or other personal items
* Not donating blood, organs, or tissue
* Using only clean needles and equipment for tattoos or body piercings
* Telling your doctors, dentist, and other healthcare providers

**Who should be vaccinated against hepatitis B?**

Those who should be vaccinated against HBV include:

* All newborns and children
* People with liver disease not caused by HBV
* People with HIV
* Healthcare and emergency workers, military personnel, morticians and embalmers
* People who have ever been on hemodialysis
* People working or housed in prisons
* Staff and patients at institutions for the developmentally challenged
* People with multiple sexual partners
* Men who have sex with men
* People who have ever injected or inhaled drugs
* Sexual partners and household members of people with HBV
* Travelers to countries where HBV is common
* Members of ethnic or racial groups with a high rate of HBV infection including Asian and Pacific Islander Americans, African Americans, Latino Americans, Native Americans and Alaskan Natives

**Hepatitis C**

Hepatitis C is a liver disease caused by the hepatitis C virus (HCV). HCV causes the liver to swell and prevents it from working well. Acute HCV occurs within six months after exposure and approximately 25% of people with acute HCV fully recover during this time. About 75% of people with acute HCV develop long-term of chronic HCV. Unless successfully treated with medications, chronic HCV can lead to cirrhosis (scarring) of the liver, liver cancer, and liver failure.

**Who is at risk of having hepatitis C?**

Anyone whose blood has come in direct contact with HCV-infected blood is at risk.

* Have ever injected or inhaled drugs (even once)
* Received a blood transfusion or organ transplant before 1992
* Received a clotting factor made before 1987
* Have ever been on hemodialysis
* Have had abnormal ALT levels several times (on blood test results)
* Have ever worked or come in contact with infected needles or blood
* Have ever worked or been housed in a prison
* Were born to an HCV-infected mother
* Have HIV infection
* Have HBV infection
* Have ever had unprotected sex (with multiple partners)
* Have ever had a sexually transmitted disease
* Have ever had tattoos or body piercings

**What are symptoms of hepatitis C?**

Most people with acute or chronic HCV have no symptoms. When symptoms occur, they may include tiredness, itchy skin, dark urine, muscle soreness, nausea, loss of appetite, stomach pain and jaundice (a yellowing of the skin and whites of the eyes). Someone can have HCV for years or even decades without symptoms.

**How is hepatitis C diagnosed?**

Hepatitis C is diagnosed by blood tests. Blood tests are done to check if HCV antibodies are in the body. Antibodies are proteins created by the immune system in response to viruses.

For patients with HCV, a liver biopsy may be needed to check how much of the liver is damaged. During a biopsy, a small piece of liver tissue is removed and studied in the lab.

**How is hepatitis C treated?**

*Acute HCV*

Doctors often recommend bed rest, drinking lots of fluids, eating a healthy diet and avoiding alcohol. Medicines may be used to treat acute HCV. It is important to see your doctor regularly to have tests done to make sure your body has fully recovered from the virus.

*Chronic HCV*

Doctors may recommend taking oral medications that kill the virus directly, they are known as direct acting antivirals or DAAs for short. The main ones include Mavyret, Epclusa and Harvoni. Each of these brand names include a combination of two antivirals in a single pill. The length of treatment can vary based on the patient’s HCV genotype (different strains of the hepatitis C virus) and severity of liver disease. For most patients the duration will be between 8 and 12 weeks. The chances of curing the infection are very high (close to 100%).

If you have HCV, it is important to talk to your doctor about treatment options and liver cancer screenings every 6-12 months. Also, talk to your doctor about hepatitis A and hepatitis B vaccines.

**What is the best way to manage hepatitis C?**

Many hepatitis C patients can lead active lives.

* Eat healthy meals
* Exercise
* Rest when you feel tired
* Take only the medications recommended by your doctor
* Avoid alcohol and drugs
* See a liver doctor regularly (hepatologist or gastroenterologist)
* Keep all medical appointments
* Talk to your doctor about hepatitis A and hepatitis B vaccines

**What is the best way to stop the spread of hepatitis C?**

There is no vaccine to prevent HCV. The only way to stop the spread of HCV is to avoid direct contact with infected blood.

* Do not share needles
* Use recommended safety measures if you are exposed to blood or needle sticks at work
* Practice safe sex
* Use clean needles and equipment for tattoos or body piercings
* Do not share razors, toothbrushes, or other personal items with others
* Wear gloves if you have to touch someone’s blood

**Cirrhosis**

Cirrhosis is the severe scarring of the liver — hard scar tissue replaces soft healthy tissue. As cirrhosis becomes worse, the liver will have less healthy tissue. If cirrhosis is not treated, the liver will fail and will not be able to work well or at all.

**What causes cirrhosis?**

Cirrhosis is caused by chronic (long-term) liver diseases that damage liver tissue. It can take many years for liver damage to lead to cirrhosis.

**Chronic Alcohol use**

Chronic alcohol use is one of the three leading causes of cirrhosis in the United States. Drinking too much alcohol can cause the liver to swell, which over time can lead to cirrhosis. The amount of alcohol that causes cirrhosis is different for each person.

**Chronic Viral Hepatitis**

Viral hepatitis causes the liver to swell, which over time can lead to cirrhosis. Chronic hepatitis C is the third leading cause of cirrhosis in the United States. About one in four people with chronic hepatitis C develop cirrhosis. Chronic hepatitis B can also cause cirrhosis.

**Nonalcoholic Steatohepatitis (NASH)**

Fat buildup in the liver that is not caused by alcohol use, is nonalcoholic steatohepatitis (NASH). This is the most common cause of liver disease in the United States. NASH can cause the liver to swell and can lead to cirrhosis. People with NASH often have other health issues including diabetes, obesity, high cholesterol and heart disease.

**Bile Duct Disease**

Bile duct disease limits or stops bile from flowing to the small intestine. The bile backs up in the liver causing the liver to swell and can lead to cirrhosis. Two common bile duct diseases are primary schlerosing cholangitis (PSC) and primary biliary cholangitis (PBC).

**Genetic Diseases**

Some genetic diseases can lead to cirrhosis. These diseases include Wilson disease (too much copper in the body), hemochromatosis (too much iron in the body), glycogen storage diseases, Alpha-1 antitrypsin deficiency, and autoimmune hepatitis.

**What are symptoms and complications of cirrhosis?**

There are usually no symptoms of cirrhosis in its early stage. Over time, cirrhosis may cause symptoms and complications:

*Symptoms*

• Loss of appetite

• Tiredness

• Nausea

• Weight loss

• Spider-like blood vessels on the skin

• Severe itching

• Jaundice, a yellow discoloration of the skin and whites of the eyes

*Complications*

• Fluid buildup and painful swelling of the legs (edema) and abdomen (ascites)

• Bruising and bleeding easily

• Enlarged veins in the esophagus (esophageal varices) and stomach (gastropathy)

• Enlarged spleen (splenomegaly)

• Stone-like particles in gallbladder and bile duct (gallstones)

• Mental confusion (hepatic encephalopathy)

• Liver cancer (hepatocellular carcinoma)

**How is cirrhosis diagnosed?**

Cirrhosis is diagnosed by symptoms, blood tests, medical history, physical examination, and imaging tests. A liver biopsy may be needed to check how much of the liver has been damaged. During a biopsy, a small piece of liver tissue is removed and studied in the lab.

**How is cirrhosis treated?**

Treatment options for cirrhosis depend on the cause and the level of liver damage. Depending on the disease causing cirrhosis, medications or lifestyle changes may be used for treatment. The goals of treatment are to prevent further liver damage and reduce complications. When cirrhosis cannot be treated, the liver will not be able to work and a liver transplant may be needed. Doctors will determine whether a liver transplant is the best treatment option.

**What is the best way to manage cirrhosis?**

It is possible to prevent further liver damage with proper management of cirrhosis.

• Maintain a healthy lifestyle (eat a healthy diet and exercise regularly) to achieve a normal BMI (body mass index)

• You may need to reduce salt intake if you experience edema or ascites (swelling caused by a buildup of fluids)

• Avoid raw shellfish

• Stop drinking alcohol

• Talk to your doctor about all of the medications, vitamins and supplements you take

• Talk to your doctor about hepatitis A and hepatitis B vaccinations

• Talk to your doctor about an annual flu shot and the pneumococcal vaccine

• Talk to your doctor about prevention and treatment of underlying liver disease (e.g., viral hepatitis, NASH, etc.)

**How does a healthy diet help the liver?**

Eating a healthy diet helps the liver to do its functions well and to do them for a long time. Eating an unhealthy diet can lead to liver disease. For example, a person who eats a lot of fatty foods is at higher risk of being overweight and having non-alcoholic fatty liver disease.

For people who have liver disease, eating a healthy diet makes it easier for the liver to do its jobs and can help repair some liver damage. An unhealthy diet can make the liver work very hard and can cause more damage to it.

**What does a healthy diet include?**

• Eating foods from all the food groups: grains, proteins, dairy, fruits, vegetables, and

 fats

• Eating foods that have a lot of fiber such as fresh fruits and vegetables, whole grain

 breads, rice and cereals

**Are there diet changes for those with liver disease?**

It is important for people with liver disease to maintain a healthy weight by eating a balanced diet with foods from all food groups. Also,

• Do not eat uncooked shellfish such as oysters and clams

• Limit eating foods that have a lot of sugar or salt

• Limit eating fatty foods

**Liver Diseases and Diet**

Some liver diseases have specific diet recommendations;

*Bile Duct Disease*

Bile is a liquid made in the liver that helps break down fats in the small intestine. Bile duct disease keeps bile from flowing to the small intestine.

**Diet Recommendations:**

• Use fat substitutes

• Use kernel oil (i.e. canola, olive, corn, sunflower, peanut, flax seed oils) because it

 needs less bile to break down fats than other types of oil

*Cirrhosis*

Cirrhosis is the scarring and hardening of the liver.

**Diet Recommendations:**

• Limit salt and foods that contain a lot of salt

• Talk to your doctor about how much protein to have in your diet

*Fatty Liver Disease*

Fatty liver disease is the buildup of fat in liver cells.

**Diet Recommendations:**

• Limit foods that are high in calories

• Eat foods that have fiber

*Hemochromatosis*

Hemochromatosis is the buildup of iron in the liver.

**Diet Recommendations:**

• Do not eat foods that have iron

• Do not use iron pots and pans

• Do not take pills with iron

• Do not eat uncooked shellfish

*Hepatitis C*

Hepatitis C is a disease of the liver caused by the hepatitis C virus.

**Diet Recommendations:**

• Limit foods that have a lot of iron

• Do not use iron pots and pans

• Limit salt and foods that contain a lot of salt

*Wilson Disease*

Wilson disease is the buildup of copper in the body.

**Diet Recommendations:**

• Limit foods that have copper such as chocolate, nuts, shellfish and mushrooms

• Do not use copper pots

**How can alcohol and medicine affect the liver?**

**Alcohol**

Alcohol can damage or destroy liver cells. Liver damage can lead to the buildup of fat in your liver (fatty liver), inflammation or swelling of your liver (alcoholic hepatitis), and/or scarring of your liver (cirrhosis). For people with liver disease, even a small amount of alcohol can make the disease worse. Talk to your doctor about alcohol and your liver health.

**Medicines**

Different types of medicines are taken every day including over-the-counter and prescription medicines, vitamins, dietary supplements, and alternative medicines. Medicines can help you feel better. However, when medicines are taken incorrectly — by taking too much or the wrong type or by mixing — your liver can be harmed.

• Learn about your medicines and how they can affect your liver

• Follow dosing instructions

• Talk to your doctor or pharmacist often about all the medicines you are taking

**Alcohol and Medicines**

Mixing alcohol and medicines can be harmful even if they are not taken at the same time

**Primary Biliary Cholangitis**

Primary biliary cholangitis (PBC) is a chronic liver disease resulting from progressive destruction of the bile ducts inside the liver– called the intrahepatic bile ducts. Bile produced in your liver travels via these ducts to your small intestine where it aids in the digestion of fat and fat-soluble vitamins (A, D, E and K). When the ducts are destroyed, bile builds up in the liver contributing to inflammation and scarring (fibrosis). Eventually this can lead to cirrhosis and its associated complications, as scar tissue replaces healthy liver tissue and liver function becomes increasingly impaired.

**What Causes PBC to Develop?**

The exact cause of PBC is unknown. It is not caused by alcohol or illegal-drug use. It’s most likely an autoimmune disease that occurs in genetically susceptible individuals. The body’s immune system mistakenly attacks and destroys its own cells – in this case, the cells of the intrahepatic bile ducts. Some people diagnosed with PBC may also have one or more other autoimmune diseases.

**Who is at Risk for PBC?**

• Women are nine times more likely than men to develop PBC, meaning that women

 make up about 90% of PBC cases.

• The disease most often develops during middle age and is usually diagnosed in people

 between the ages of 35 to 60 years.

• There appears to be a genetic component to developing PBC, as it’s more common

 among siblings and in families where one member has been affected.

**What are the Signs and Symptoms of PBC?**

PBC may progress slowly and many people do not have symptoms, particularly in the early stages of the disease. The most common initial symptoms are fatigue and itching of the skin (pruritis). Other symptoms may include:

• Abdominal pain

• Darkening of the skin

• Small yellow or white bumps under the skin or around the eyes (xanthomas)

Individuals may also complain of having dry mouth and eyes, and bone, muscle and joint pain.

As the diseases progresses, symptoms of cirrhosis can develop including:

• Yellowing of the skin (jaundice)

• Swelling of the legs and feet (edema)

• Enlarged abdomen from fluid accumulation (ascites)

• Internal bleeding in the upper stomach and esophagus from enlarged veins (varices)

Thinning of the bones (osteoporosis) leading to fractures can occur in late stages of the disease. In addition, people with cirrhosis are at increased risk for liver cancer (hepatocellular carcinoma).

**How is PBC Diagnosed?**

Because many people with PBC have no symptoms, the disease is often discovered incidentally due to abnormal results on routine liver blood tests. Once PBC is suspected, a blood test to check for antimitochondrial antibody (AMA) is done. This test is positive in nearly all people with PBC. Imaging of the abdomen by ultrasound and a liver biopsy, where a sample of liver tissue is removed with a small needle, can help confirm the diagnosis.

**How is PBC Treated?**

*Treating the Disease*

There is no cure for PBC, however, there are medications that can help slow disease progression and manage symptoms. Ursodiol (brand names Actigall, URSO 250, URSO Forte) is a naturally occurring bile acid (ursodeoxycholic acid or UDCA) that helps move bile out of the liver and into the small intestine. If used early enough, ursodiol can improve liver function and may keep you from needing, or delay the need for a liver transplant. People with PBC must take this medication every day for life.

Depending on the severity of the disease, and the response to ursodiol, other medications might be prescribed. One of them is called obeticholic acid (OCA) and was recently approved to treat patients with PBC in whom the response to ursodiol was not optimal.

Liver transplantation is considered when medical treatment no longer sufficiently controls the disease. When a person has end-stage liver disease, a liver transplant is necessary for survival.

*Treating the Symptoms*

• Intense itching is one of the most common symptoms of PBC. Over-the-counter antihistamines like diphenhydramine (Benadryl) may be helpful. A drug called cholestyramine (Questran) helps bind bile and decreases the itching associated with elevated bilirubin levels. Other agents such as rifampin, naltrexone and sertraline may be prescribed.

• Dry eyes can be relieved by using eye drops (artificial tears).

• A dry mouth may be helped by sucking on hard candy or chewing gum, both of which increases saliva. There are also saliva substitutes and some medications that can be used.

*Preventing Complications*

• Blood tests to monitor for deficiencies in fat-soluble vitamins are often done. As PBC progresses, some people need to replace the fat-soluble vitamins lost in fatty stools, so you may be put on vitamin A, D, E and K replacement therapy.

• Since people with PBC are at a higher risk for osteoporosis, calcium and vitamin D are usually prescribed. Screening for osteoporosis with bone density testing may be recommended.

• As the ability of the liver to function decreases over time, complications associated with cirrhosis will need to be addressed and treated. Screening for varices and liver cancer is often recommended.

**What Lifestyle Changes are Recommended for People with PBC?**

Maintaining a healthy lifestyle can help people with PBC feel better, as well as relieve or prevent some symptoms associated with the disease. Upon diagnosis, your doctor may suggest the following:

• Start a reduced sodium and/or low-fat diet

• Drink plenty of water

• Avoid or lower intake of alcohol

• Lower caffeine intake

• Avoid undue stress when possible

• Start exercising, particularly walking

• Stop smoking

• Maintain good skin care

• Get regular dental examinations

Keep in mind that PBC usually advances slowly over a period of years. Many people lead normal lives for years without symptoms, depending on how early the diagnosis is made. And while there is no cure, people are having good results slowing disease progression and living longer without complications by adhering to their medication regimen and maintaining a healthy lifestyle. In fact, improvements in early detection and better management of PBC have led to a worldwide initiative to formally change the name of the disease to primary biliary cholangitis (inflammation of the bile ducts). This would more accurately reflect the fact that currently, 85% of people do not have cirrhosis at the time of diagnosis, and as treatments improve, many people may never progress to that stage of liver disease.

**Primary Sclerosing Cholangitis**

Primary sclerosing cholangitis (PSC) is a chronic, or long-term, disease that slowly damages the bile ducts. Bile is a digestive liquid that is made in the liver. It travels through the bile ducts to the gallbladder and the small intestine, where it helps digest fats and fatty vitamins.

In patients with PSC, the bile ducts become blocked due to inflammation and scarring or fibrosis. This causes bile to accumulate in the liver, where it gradually damages liver cells and causes cirrhosis, or fibrosis of the liver. As cirrhosis progresses and the amount of scar tissue in the liver increases, the liver slowly loses its ability to function. The scar tissue may block drainage of the bile ducts leading to infection of the bile. PSC advances very slowly. Many patients may have the disease for years before symptoms develop. Symptoms may remain at a stable level, they may come and go, or they may progress gradually. Liver failure may occur 10-15 years after diagnosis, but this may take even longer for some PSC patients. Many people with PSC will ultimately need a liver transplant, typically about 10 years after being diagnosed with the disease. PSC may also lead to bile duct cancer. Endoscopy and MRI tests may be done to monitor the disease.

**What are the symptoms of PSC?**

Many people with PSC do not get symptoms, especially in the early stages of the disease. When symptoms do occur the most common are fatigue, pruritus, or itching of the skin, and jaundice, a yellowing of the skin and eyes. These symptoms may come and go, but they may worsen over time. As the disease continues, the bile ducts may become infected, which can lead to episodes of fever, chills and abdominal pain.

**What causes PSC?**

The cause of this disease is not known. About 70 percent of patients are men. It may be related to bacterial or viral infections, as well as problems in the immune system. Genetic factors may also play a role. PSC is considered an uncommon disease, but recent studies suggest that it may be more common than previously thought.

**How is PSC diagnosed?**

Because many PSC patients have no symptoms, the disease is often discovered through abnormal results on routine liver blood tests. Formal diagnosis is usually made by cholangiography, an X-ray test involving injection of dye into the bile ducts, or by a MRI.

**How is PSC treated?**

There is no cure or specific treatment for PSC. The itching associated with the disease can be relieved with medication, and antibiotics are used to treat bile duct infections when they occur. Most people with PSC must take vitamin supplements. In some cases, bile duct surgery or endoscopy may be useful to temporarily improve bile flow.

If I’ve been diagnosed with PSC, what questions should I ask my doctor about PSC?

* How severe is the liver damage?
* What treatment do you recommend? Will this slow down the progression of the disease?
* Will any medication be prescribed? What are the side effects?
* Should I change my diet?
* Are there any supplements you would suggest that I take?
* What can be done to relieve my symptoms?
* Does screening for bile duct cancer need to be done now?

**When is a liver transplant necessary?**

Over time, many PSC patients will continue to suffer a gradual loss of liver function. If liver failure becomes severe, a liver transplant may be necessary. The outcome for patients with PSC who have undergone transplantation is excellent. The survival rate for two or more years is about 80 percent, with a good quality of life after recovery

**Who is at risk for PSC?**

PSC is slightly more common in men than women. It usually affects people between the ages of 30 and 60. As many as 75 percent of patients with PSC also have inflammatory bowel diseases, usually ulcerative colitis.

**What is being done to find a cure for PSC?**

PSC has been known for 100 years, but now doctors are able to diagnose it very early. This means that treatment can begin before the liver is severely damaged. Scientists are continuing to study the disease to find the cause and understand its development.

In addition, 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.