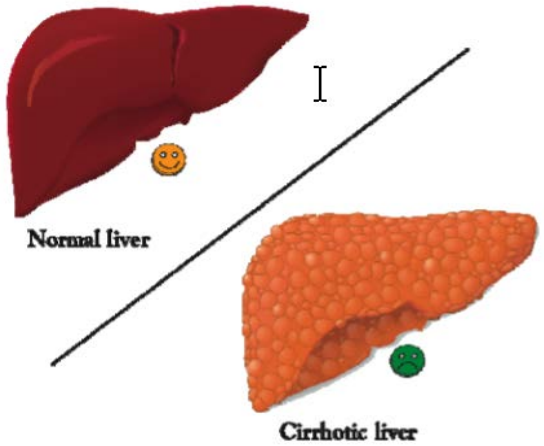
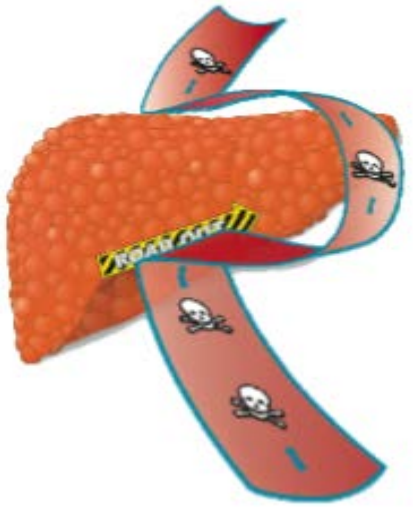
LIVER CIRRHOSIS: PATIENT HANDBOOK AND INFORMATION

Liver Cirrhosis

When something attacks and damages the liver, liver cells are killed, and scar tissues is formed. This scarring process is called **fibrosis** and it happens slowly over many years. When the entire liver is scarred, it shrinks and hardens. This is called **cirrhosis,** and this is irreversible. Any illness that affects the liver over a long period of time may lead to fibrosis and eventually cirrhosis. Heavy alcohol use and viruses such as hepatitis B and C are common causes of cirrhosis. Cirrhosis may also be caused by a build up of fat within the liver of people who are overweight, have diabetes and high cholesterol. Some people inherit genes that can cause liver disease. Other causes include medications, environmental toxins and autoimmune hepatitis which is a condition in which a person's own immune system attacks the liver.

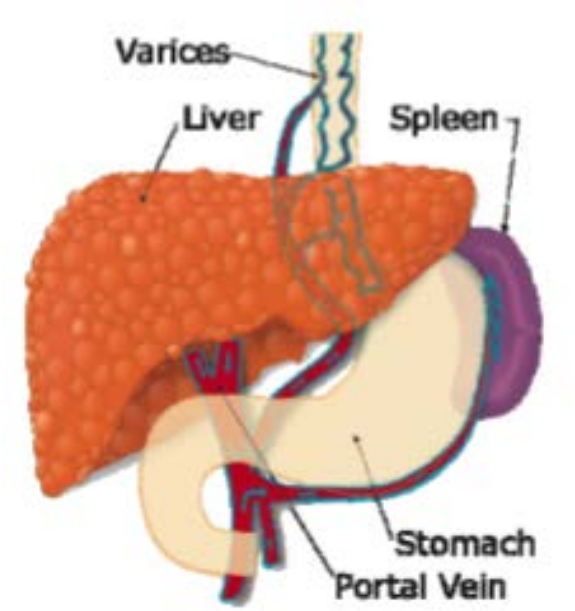


What happens when you have cirrhosis?



When a liver is cirrhotic, it shrinks and becomes stiff. As a result, blood cannot flow through the liver easily and pressure will build up in a vein that brings blood to the liver called the portal vein. When the pressure is high in the portal vein, the condition is called **portal hypertension**. To relieve this pressure, the blood goes around the portal vein and through other veins called **varices**. Varices are often found in the esophagus and/or the stomach and can be a cause for gastrointestinal bleeding. The high pressure in the portal vein can also cause changes to the size of the spleen. As the spleen enlarges, platelets which help blood to clot, can get destroyed. In addition, as cirrhosis advances, the liver is unable to produce proteins that assist with clotting. These changes can increase the risk of bleeding.

What are the symptoms of cirrhosis?



Initially, there may be no symptoms at all. This is called **compensated cirrhosis**. In fact, a person may have cirrhosis for many years without being aware that there is liver damage. This is because there are still enough healthy liver cells to keep up with the body's needs. However, if there is ongoing damage to the liver (continued alcohol use, increased fat build-up in the liver, untreated viral infection) then the few remaining liver cells may get damaged and stop functioning properly. At that point, symptoms such as low energy, poor appetite weight loss or loss of muscle mass can develop.

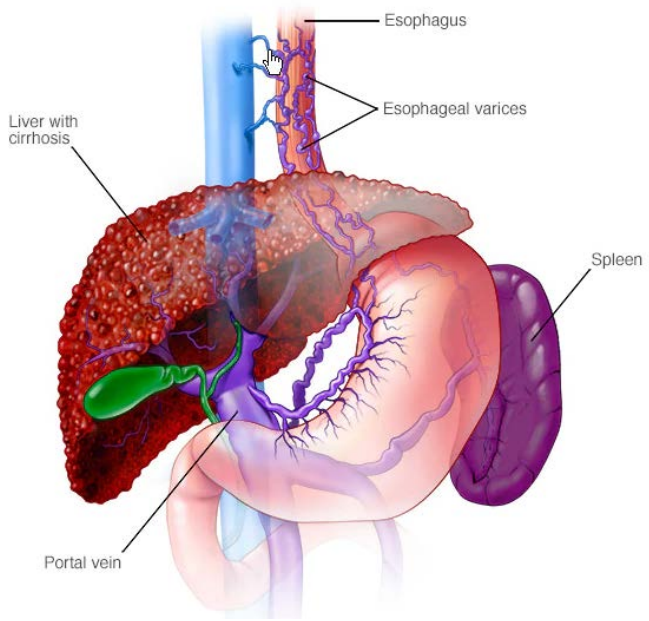
What is decompensated cirrhosis?

Patients with cirrhosis are at risk for developing the following complications:

- Internal bleeding from large blood vessels in the esophagus and stomach called **varices**
- Buildup of fluid in the belly a called **ascites**
- Confusion from the build-up of toxins in the blood called **encephalopathy**
- Yellowing of the eye and skin called **jaundice**
- **Liver cancer**

When a patient experience any of these serious problems, it is a sign that the liver disease has progressed from compensated to **decompensated cirrhosis**. The presence of any or all these complications can be life threatening.

Internal bleeding: Varices



Esophageal varices are enlarged veins that occur in the walls of the esophagus because of backup of blood from the scarred liver. The esophagus is the swallowing tube that connects the throat to the stomach. The pressure in these swollen veins is higher than normal. Increased pressure can cause the veins to become larger and eventually burst, leading to sudden, severe and even life-threatening bleeding which may present as vomiting blood or black tarry stools. The lining of the stomach may also become irritated and swollen (**portal gastropathy**) which may cause oozing of blood, dark stools and lower blood counts.

What can be done to stop variceal bleeding?

Vomiting blood is a medical emergency and requires immediate evaluation. A procedure called an upper endoscopy (EGD) can be performed to evaluate and treat varices. If varices are present, medications can be used to decrease the pressure in the varices to prevent bleeding. If varices do bleed, doctors may perform an EGD to apply rubber bands to the varices to stop the bleeding. In some cases, an internal tunnel in the liver can be constructed called a **transjugular intrahepatic portosystemic shunt (TIPS)** that reduces blood flow and pressure in varices.

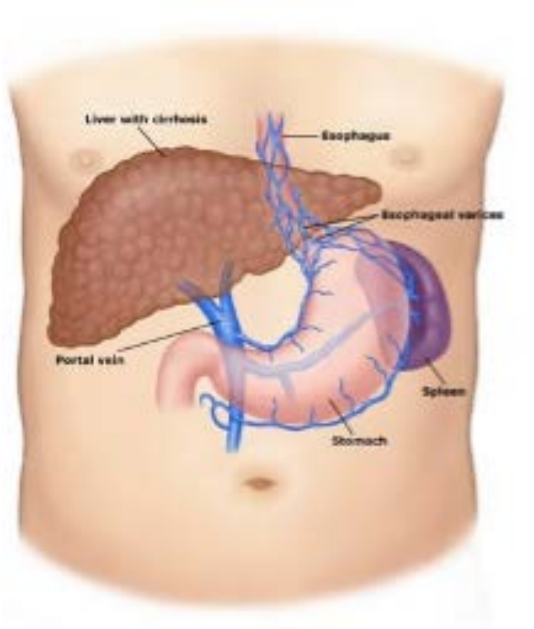
What can be done to prevent serious bleeding?

Non-selective beta blockers can be used to reduce blood flow and pressure in the varices. The medications proven to be effective are:

Propranolol (Inderal): **Given twice a day**
Nadolol (Corgard): **Given once a day**
Carvedilol (Coreg): **Given twice a day**

Your doctor will generally start you on a very low dose of one of these medications and check your heart rate (pulse). The goal of treatment is to reduce your heart rate by 25% and ideally within the range of **55-60 beats per minute.** One side effect of lowering your heart rate may be symptoms of tiredness and decreased exercise tolerance.

Patients with cirrhosis usually have low blood pressures. However, the addition of beta blockers are usually well tolerated even in patients with low blood pressure.



- Unless there is severe dizziness due to the use of this medication, these medications should not be discontinued with low blood pressures.
- Sudden discontinuation of beta blockers can cause abrupt and even life-threatening bleeding.
- Some male patients with cirrhosis may also have problems with an enlarged prostate that can make it difficult to urinate. Often medications called "alpha blockers" are prescribed to improve urine flow. Examples of these medications are Hytrin (terazosin), Flomax (tamsulosin) and Cardura (doxazosin). These medications should be avoided as they may decrease the effectiveness of beta blockers.

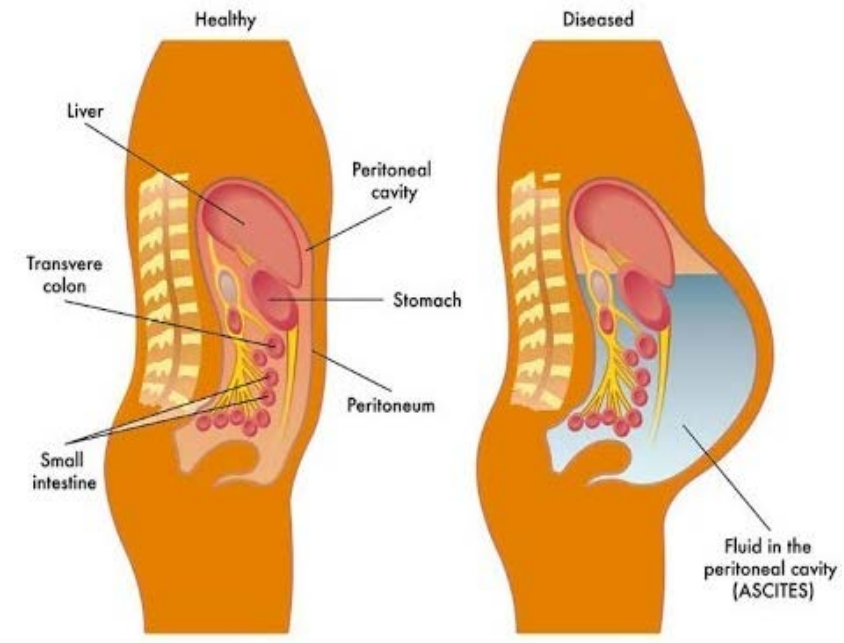
If you vomit blood or your stool turns black and tarry, you should seek immediate medical attention. These are signs that varices may have begun to bleed, and this can be life threatening. If this occurs, you should seek medical attention immediately and go to the nearest Kaiser Urgent Care or to the nearest hospital

Ascites (Fluid in the belly)

Another problem caused by high pressure in the veins of the liver is **ascites**. Ascites is the abnormal collection of fluid in the abdominal cavity.

Cirrhosis or “scar tissue" within the liver is a common cause for ascites. This scar tissue changes the normally smooth liver surface to a lumpy and hard surface which then blocks the blood from exiting the liver. If the blood cannot flow freely, pressure will build up in the liver and this is called **portal hypertension**. This causes the surface of the liver to leak fluid into the abdominal cavity, which accumulates and causes ascites. The liver then sends signals to the kidney to help regulate the loss of fluid by absorbing and holding on to salt. This ultimately results in additional fluid buildup in the legs and abdomen.

In mild cases of ascites, there are often no symptoms. As more fluid collects, the abdomen swells. There may be a loss of appetite, frequent heartburn, fullness after eating, or abdominal pain. Both legs can also get swollen and this can be very uncomfortable. Breathing may also become more difficult especially when lying down. As ascites advances, there can be swelling of the abdomen that looks like the later stages of pregnancy. This may cause back pain, changes in bowel function, and fatigue. During the day, gravity may carry some of the fluid down into the scrotum and cause worsening leg swelling. Sometimes fluid can fill the lung near the liver.



Ascites is not actually a disease, but a symptom. However, in most patients, ascites is a sign of advanced liver failure or cirrhosis of the liver. The ascitic fluid can also become infected and this can be life threatening.

Treatment of ascites

- **Low salt (sodium) diet**
  Ascites is a problem of the body holding on to too much salt (sodium) rather than a problem of too much water intake. Thus, it is important to cut down on salt intake, not water intake. Dietary sodium intake is usually restricted to less than 2000 mg per day (about 1 teaspoon). Since most salt in a person's diet comes from processed foods and not from a salt shaker, it is important to avoid canned and processed foods.

- **Diuretic therapy ("Water Pills")**
  Diuretics can help the body get rid of extra sodium and water through the kidneys. Common medications include **spironolactone (Aldactone)**, and **furosemide (Lasix)**. These medications can be used alone or in combination. Dosing starts low and is adjusted gradually over a period of weeks to months based on treatment response. Kidney function and blood levels of sodium and potassium can be affected by these medications, so close lab monitoring is recommended.

- **Paracentesis (Tap)**
  This procedure involves draining fluid out of the abdomen with a needle. It is performed by interventional radiologists using a local anesthetic. This procedure is used when the amount of ascites is large and causing symptoms, if you cannot take diuretics, or if samples of fluid need to be taken to evaluate for infection. Paracentesis can rapidly relieve the symptoms of ascites, but it does not correct the underlying cause. The fluid will eventually return and may need repeat paracentesis. Strict sodium (salt) restriction and diuretic therapy must still be used to slow down the accumulation of ascites.

- **Monitor Progress**
  It is important that patients undergo careful monitoring with frequent blood tests and regular measurements of body weight. This is especially true in patients taking diuretics (which may cause reduced kidney function and changes in the blood levels of sodium and potassium). You should check your weight daily. It is also strongly recommended that you use a pill box and have an organized system for taking your water pills (diuretics) and other medications. You should keep a calendar to mark the dates if you need regular paracentesis.

- **Trans jugular intrahepatic portosystemic shunt, (TIPS)**
  This is a procedure which involves placement of a shunt within the liver to improve blood flow. A TIPS procedure is performed through the veins and does not require abdominal surgery. This procedure is performed by an interventional radiologist. It can be performed to control bleeding from varices (if other measures fail) and sometimes to decrease fluid buildup (ascites). About 30% of patients can develop increased mental confusion (encephalopathy) after TIPS, and in some cases the shunt must be closed back down if this occurs. Rarely, progressive jaundice and liver failure develops after a TIPS procedure.

- **Liver transplant**
  Liver transplant should be considered in anyone with decompensated cirrhosis. Unfortunately, not all patients are candidates for a liver transplant. If you are a potential candidate, your gastroenterologist will discuss the eligibility process with you.

Complications of ascites

Spontaneous bacterial peritonitis (SBP)
SBP is when ascitic fluid becomes infected. Symptoms include fever and abdominal pain, but these symptoms may be absent during the early stages. If you have infection of the ascites fluid, you will need to be admitted to the hospital for treatment with antibiotics since this condition can be life threatening. Oral antibiotics are recommended after the first episode of SBP to decrease the chances of recurrence. If you are requiring frequent paracentesis, your doctor may also prescribe antibiotics to prevent this infection from developing.

Hepatorenal syndrome (HRS)
This refers to kidney failure that sometimes develops in patients with advanced liver disease. This may happen suddenly or as a slowly progressive process. Treatment usually involves stopping diuretic therapy, intravenous fluids and albumin infusions, and a search for a reversible cause such as dehydration or infection. Rapid kidney failure in cirrhotic patients with ascites is associated with a 90% chance of death if liver transplant is not performed.

If you have ascites and you suddenly get a fever or new belly pain, you should go to the emergency room immediately. These could be signs of a serious infection that can be life threatening.

Hepatic Encephalopathy (Confusion)

Patients with cirrhosis can develop confusion, disorientation and difficulty thinking. This is called hepatic encephalopathy and is due to a buildup of toxins (ammonia) as a result of an unhealthy liver. These toxins will reach and affect the brain. Hepatic encephalopathy is diagnosed clinically, and an elevated ammonia blood level does not necessarily mean you have this diagnosis.

These toxins may also cause changes in sleep, mood, concentration and memory. Often, the first sign of hepatic encephalopathy is insomnia. Other frequent symptoms include irritability, personality changes and poor work performance. In severe cases, it can cause a coma. If you have encephalopathy, you may have problems driving, writing, calculating and performing other activities of daily living.

Encephalopathy may occur when you have an infection, dehydration or when you have internal bleeding. It may also occur if you are constipated or taking too many medications that affect the brain, like narcotics (morphine-like pain medications) and sedatives or sleeping pills (i.e.: Valium, Librium, Ativan, Xanax, Klonopin). Patients with cirrhosis can be very sensitive to these medications and they should be avoided. If they must be used, the lowest possible dose should be used.

Treating Hepatic Encephalopathy (HE)

A common medication used to treat hepatic encephalopathy is a non-digestible sugar called **Lactulose**. Lactulose is an artificial sugar that cannot be digested by the stomach or intestines. It goes into the large bowel (colon) and traps protein toxins which are then excreted in stool.

Lactulose is routinely prescribed as a liquid. Patients are started on 2 or more tablespoons once or twice a day. The dose can be increased gradually until the patient is having 3 loose stools a day. The dose of lactulose can be adjusted by the patient and/or family**.** For example: you may increase the dose of Lactulose if your stools are firm, if you are constipated, or if you are having fewer than 3 stools per day. You should also increase the dose on days when you are more forgetful or confused. You may decrease the dose if you are having more than 3 loose stools per day.

Another treatment for hepatic encephalopathy is non-absorbable antibiotics that work only in the intestine. These antibiotics include **Rifaximin (Xifaxin)** and

Neomycin. These medications help eliminate the gut bacteria which produce toxins. In severe cases of hepatic encephalopathy, Rifaximin (Xifaxin) can be taken with Lactulose and may be able to decrease the frequency of hospitalizations due to hepatic encephalopathy. Rifaximin can also be used alone if lactulose cannot be tolerated.

- The most effective way to prevent hepatic encephalopathy is to have good nutrition with an early breakfast and very frequent small meals, maintain healthy kidney function, avoid narcotics and sedatives, and to take medications that help decrease accumulation of ammonia.

- You should avoid excessive amounts of protein, but it is not necessary to restrict the amount of protein if eaten in normal amounts. Protein should be divided throughout the day among the several meals you are eating. Patients can eat as much vegetable protein (dried beans and peas, soy products, peanut butter and others) as they like.

- Even mild hepatic encephalopathy may cause problems with safe driving. Patients with confusion or any other signs of encephalopathy are not allowed to drive, due to their slow reaction time and impaired thinking. Patients with encephalopathy should also not operate any heavy machinery.

- Worsening hepatic encephalopathy can be a sign of infection, bleeding, dehydration, and worsening liver function. Cirrhotic patients with a rapid change in their symptoms and with worsening confusion and disorientation should be seen ASAP by a medical professional for further evaluation.

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If you are not acting like yourself, if you are confused, or if you are very sleepy, you should be taken to the emergency room immediately. These symptoms could be a sign of a serious medical problem. You should not drive when you have these symptoms.

Jaundice (yellowing of the eyes and skin)

A liver that is working poorly cannot get rid of bilirubin, a substance that is produced in the liver. Elevated bilirubin causes a yellowing of the eyes and skin, called jaundice. Too much alcohol and some medications can also lead to jaundice. If you have cirrhosis and noticed new onset jaundice, it may be a sign of worsening of your liver function, an infection or other new problems.

If you suddenly develop jaundice with yellowing of the eyes or skin, you should contact your doctor immediately.

Additional Recommendations for Patients with Cirrhosis

Diet

Cirrhotic patients have limited capability to store nutrients in the liver. For that reason, you need to eat very frequently in order to preventing use of your own muscle mass as a source of nutrition for vital organs

- Eat at least three meals a day and three to four snacks between meals
- Patients who are overweight should be careful to eat low-calorie meals and low-calorie snacks
- Patients who are under-nourished should have high-calorie meals and snacks. Bedtime snacks are especially important.
- Patients with malnutrition and/or loss of muscular mass can improve their nutrition and muscular mass by drinking two cans of Ensure Plus, or Boost-Plus at bedtime, or in case of overweight or diabetes, 2 cans of Glucerna, or Boost Glucose-Control at bedtime, or 1 can of Boost VHC at bedtime.
- Cirrhotic patients are at increased risk of acquiring food-borne infections; we recommend that all foods that you eat from animal organs should be fully cooked. This is especially true for seafood and poultry.
- Raw seafood, especially shellfish, should not be eaten. Only pasteurized milk and juices should be consumed.
- Patients with cirrhosis require normal amounts of protein but may poorly tolerate excessive amounts. We recommend that your protein intake be around 1.2 grams per kilogram of body weight divided into meals throughout the day and not eaten in a single sitting. Patients should strive to eat at least 60 gm of protein per day. Protein can be taken in the form of animal sources (beef, pork, fish/seafood, poultry, eggs, and milk) and from vegetable sources, such as beans and lentils.
- Patients with cirrhosis tend to retain sodium (salt) and water which causes formation of swelling (edema) and free fluid in the abdomen (ascites). Patients with edema and ascites are at very high risk of developing infections that, in turn, carry a very high risk of death. To avoid formation

of edema and ascites, the most important factor is to avoid sodium in your diet. Salt comes with different names, including “Salt, Sea Salt and Light Salt”; is also in high amounts in Soy Sauce, pickles, and olives.

- Patients with cirrhosis and edema or ascites should not exceed 2,000 mg (2 grams) of sodium (salt) per day. Sodium is present in small or moderate amounts in fresh or frozen foods but in large amounts in processed foods and canned foods and soups, including cakes and candy bars that use salt or baking soda to enhance flavor or cause a cake mix to rise. Avoid all processed foods and cured meats, such as ham, baloney, bacon, pickles, potato chips, corn chips, and other salty snacks. Milk and cheese should be used only in moderation because they have large amounts of sodium. Tomato juice also contains a lot of salt. Well water with a water softener, may also contain a lot of salt.

- A diet with more than 2,000mg of sodium per day will make it extremely difficult for you to control ascites and edema and may force us to use higher doses of water pills that, in the end, may cause kidney failure and death.

- Salt substitutes, like “No-Salt or Light-Salt”, that contain potassium should not be used if you are taking diuretics Spironolactone (Aldactone), Triamterene (Dyrenium), or Amiloride (Midamor) because it can cause severe elevations of potassium and subsequent heart problems.

- The safest commercial seasoning you can use is “Mrs. Dash.”

- Using multivitamins and vitamin supplements, overall, is a good idea, except for large amounts of vitamin A which can be very toxic to the liver. If you have excessive iron in your body, taking a large amount of vitamin C may be dangerous. Excessive amounts of iron can also cause worsening of liver disease. You may have to avoid vitamins that contain iron, unless you have an iron deficiency. Use of moderate amounts of vitamin E, which is an antioxidant that helps the liver, is a good idea, however, high doses may place you at increased risk. Only use herbals or supplements after your doctor has approved their use.

- Sugar and fat will not harm the liver when eaten in moderation. However, you may need to watch these if you have diabetes, heart disease, or high cholesterol.

Leg Cramps

- Leg cramps are very common in patients with cirrhosis. They are frequently due to decreased amounts of minerals in the body, such as calcium, magnesium and zinc
- If you have leg cramps, we would prefer that you take these kinds of mineral supplements and avoid taking medications like quinine. Quinine can cause a further drop in your blood platelet count and place you at higher risk of bleeding

Increased Risk of Complications from Infections

- Patients with cirrhosis do not tolerate additional liver damage (hepatitis) or other infections.
- If you are not already immune, you should receive vaccinations against the hepatitis A and hepatitis B viruses.
- You should get the influenza or "flu" vaccine every year. You should not get the nasal formulation of the flu vaccine because it is a live vaccine.
- You should get a pneumonia vaccine (Pneumovax) every five to 10 years.

Overweight

- To have clinical obesity increases the stress on your liver and accelerates liver damage.
- We recommend that patients with obesity should follow a diet for weight reduction with moderate decreases in caloric intake, following the American Diabetic Association diet guidelines. This situation is particularly important for patients who have diabetes mellitus or who have abnormal lipids (cholesterol or triglycerides) in their blood.
- Excellent control of glucose level, as well as cholesterol and triglycerides, will help slow down progression of liver damage.

Fatigue

- Fatigue is very common in advanced liver disease. There is evidence that decreased physical activity will worsen the fatigue.
- Patients with cirrhosis get some regular exercise, such as having a brisk walk. The exercise should ideally be from 30-45 minutes per day.
- If you have varices in the esophagus or stomach, we do NOT recommend exercises that make you lift heavy weights because that may increase the pressure on your vessels which can cause them to burst.

Tobacco, Alcohol and Drugs Use

- There is evidence that smoking accelerates scarring of the liver. In addition, the use of tobacco products increases the risk of lung cancer, cancer of the mouth, and cancer of the swallowing tube (esophagus). Smokers who have surgery are at increased risk of death.
- We strongly recommend that you not smoke and that you avoid all tobacco products.
- Alcohol accelerates liver damage and often makes it impossible to eliminate viral infections of the liver. In addition, alcohol tremendously increases the risk of liver cancer.
- Patients with liver disease should not drink alcohol
- Use of other substances of abuse, such as cocaine, crack, heroin, ecstasy, marijuana, methamphetamine, etc., increase the risk of further liver damage and sometimes cause severe hepatitis. In a cirrhotic patient, this is likely to cause death. Substances of abuse should not be used.
- Active use of substances of abuse, tobacco, and alcohol may prevent you from being a candidate for liver transplantation.

Pain Medications

- Patients with advanced liver disease are at very high risk of serious complications from the use of pain medications.
- Non-steroidal anti-inflammatory (NSAIDS**)** medications such as aspirin, ibuprofen, Motrin, Midol, Aleve, Advil, Naproxen, sulindac, ketoprofen, Indocin, Celebrex, Alka-Seltzer, Diclofenac (Cataflam), Oxaprozin (Daypro), Feldene (piroxicam), Mobic (meloxicam), Nabumetone (relafen), Bextra (valdecoxib), and others, can cause severe kidney failure in patients with cirrhosis.

- NSAIDS can also increase the retention of sodium and water and should be avoided.
- Narcotics such as morphine, codeine, hydrocodone, fentanyl, demerol, etc., can cause severe confusion/hepatic encephalopathy in patients with advanced liver disease. These medications should be avoided.
- If there is no other choice, these medications should be used at the lowest possible dose.
- Larger amounts of acetaminophen (Tylenol) can cause severe liver damage. This is particularly true if the patient has not been eating regularly. In patients who have been eating regularly, a moderate amount of acetaminophen (Tylenol) is the best choice for pain control.
- For severe pain, we recommend that you not exceed 2 grams (Four 500 mg tablets) of Tylenol (acetaminophen) per day for no more than two days.

Screening for liver cancer

- Patients with cirrhosis are at increased risk for developing liver cancer called hepatocellular carcinoma or HCC
- The risk of developing HCC is about 1 in 100 per year (each year, out of 100 patients with cirrhosis, one will develop liver cancer).
- All patients with cirrhosis should have an ultrasound and blood test called an alfa-fetoprotein (AFP) every 6 months
- Although these tests are not perfect, they can often detect the tumor when it is small, before symptoms develop. If the ultrasound or AFP is abnormal, the next step is MRI or CT scan. Often MRI or CT scans are good enough to make a definitive diagnosis of liver cancer, so biopsies are not required.