



# **You, Your Liver, and Alpha-1**



## LIVER FUNCTION AND ALPHA-1

Alpha-1 Antitrypsin Deficiency, or Alpha-1, is a genetic condition that can cause disease of the lungs, liver, skin, and blood vessels. The first descriptions of Alpha-1 pointed to its role in the promotion of lung disease in adults. But, in fact, Alpha-1 is primarily a condition caused by liver problems. Alpha-1 can affect your liver function in many ways and lead to [liver disease](#) in newborns, children, and adults.

To diagnose Alpha-1, first an AAT level test is done. Most individuals with Alpha-1 (Alphas) have a low level of AAT in the blood. If your blood level is low, then genetic tests are needed to find out which abnormality is present in the AAT gene. If you don't have a confirmed diagnosis of Alpha-1, ask your health care practitioner to test you.

## AAT LEVELS AND YOUR LIVER FUNCTION

Your liver makes large quantities of Alpha-1 antitrypsin protein (AAT) and releases it into the blood. People who carry at least one abnormal gene for Alpha-1 produce an abnormal AAT protein. The abnormal genes that can cause liver disease are usually “Z” genes. The “M” gene isn’t abnormal.

Your liver cells don’t release normal amounts of AAT protein (Z). The AAT builds up in the liver, causing lower levels of AAT in the blood. Those low levels may cause lung damage in Alpha-1, and the buildup of Z protein in the liver may cause liver damage.

Because most of the research into Alpha-1 liver disease has been performed on the Z protein, we will use Z as the basis for this guide. There are many other types of AAT gene variants that can cause Alpha-1, but the most common one associated with liver disease is the Z protein.



**KEY LEARNING:** While the lung damage in Alpha 1 appears to be directly related to the low blood level of AAT, the liver is damaged by the unusual internal buildup of the abnormal AAT protein.

To understand how Alpha-1 affects your liver, it helps to understand how your liver functions under normal conditions.

## WHAT DOES NORMAL LIVER FUNCTION LOOK LIKE?

**Synthetic function: It’s your body’s filter and distribution center.**

Your liver makes many proteins and other substances your body needs to function. It also makes many components of the liquid, non-cellular portion of your blood. These substances help with:

- Normal blood clotting
- Getting energy to your body when you exercise or fast
- The distribution of salts, fluid, and nutrients through your bloodstream



**IT’S A FACT:** The liver is the body’s filter and distribution center. Part of its function removes waste while also generating life-sustaining proteins and substances to keep the body working smoothly.

**Digestive Function: It helps you get nutrients from food.**

Blood passes through your liver when it leaves your intestines. Nutrients from food are processed, stored, and distributed to other parts of the body.

Your liver also produces a substance called bile, which is stored in your gallbladder. When you eat, the bile drains into your intestine through the bile duct. It mixes with food to help with fat digestion and nutrient absorption.

Your liver has its very own unique blood supply called the “portal circulation.” This carries blood from the intestines and stomach to the liver and spleen. Portal circulation through the liver slows when the liver is injured.

### **Excretory Function: It’s a waste-removal system.**

Your liver helps remove waste from the body by:

- Cleaning toxins from the food you eat as blood from your intestines passes through it.
- Processing some wastes into useful substances.
- Transferring other wastes into bile, which drains into the intestine and passes out in your stool.
- Clearing some drugs and medicine from your bloodstream, sending them out of your body in bile or urine.

## **SEEING YOUR HEALTHCARE PRACTITIONER ABOUT ALPHA-1 LIVER DISEASE**

When you have Alpha-1, you should see a healthcare practitioner who is willing to learn about Alpha-1 liver disease, as most are unfamiliar with it. Knowing what to expect can help your first visit go smoothly.

Typically, your first visit will include a medical history, a physical exam, and some [lab tests](#). These will help determine if there’s a problem with your liver function.

### **First, your doctor will take your medical history**

Your doctor usually starts by checking your health history for signs of liver problems that may have gone unnoticed. They’ll start by asking about your health and the health of family members. They’re looking for things such as:

- Jaundice (yellow skin and eyes) as an infant, child, or adult
- Surgeries or hospitalizations
- Gallstones or kidney stones
- Problems with the pancreas gland (pancreatitis)
- Blood transfusions
- Serious injuries
- Significant health events

### ***Risk factors for liver disease***

During your visit, your doctor will review risk factors that can have a big influence on liver health, including:

- Cancer
- Immune diseases
- Alcohol and drug use
- [Exposure to toxic chemicals at work](#)
- Medicines, herbs, and dietary supplements
- Exposure to Hepatitis A, B, or C
- Obesity

**NOTE:** They’ll also ask if you’ve gotten [vaccinations](#) for hepatitis A and B.

## Next, your doctor will ask about symptoms

Your doctor will want to know about specific symptoms that point to liver problems:

- **Diarrhea:** Common in adults and children with abnormal bile flow and fat digestion.
- **Itching:** Common in patients with significant liver disease. When your liver doesn't remove waste products from the blood, they build up within your skin and cause itching.
- **Abdominal pain, indigestion, or vomiting:** Common in patients with liver disease.
- **Unusual bleeding:** May be related to poor vitamin K absorption or poor liver synthetic function.
- **Sleepiness or lack of alertness:** May be caused by toxins in the blood that go to the brain and cause "encephalopathy."
- **Blood in vomit or stool:** May be caused by "portal hypertension," which is a type of high blood pressure in the portal vein. It happens when scar tissue in your liver disrupts blood flow coming from your intestine. Other symptoms of portal hypertension include a swollen, bloated belly from fluid in the peritoneal space called "ascites."

## Other ways Alpha-1 affects your body

If your liver is unhealthy, your body may not get the nutrients it needs. This affects your skin and hair. Cuts and scrapes may take a long time to heal. Your hair may be dry or dull. Fatigue is common.

Your doctor can order lab tests to assess your nutritional status. They can also take key measurements to assess whether

you're getting the nutrients you need, including:

- Height and weight
- Body fat percentage
- Lean muscle mass
- Body Mass Index (BMI)



**IT'S A FACT:** The liver and nutrition are intimately linked.

**Alpha-1, alcohol, and your liver:** Any history of potential liver problems will include questions about how much alcohol you consume. Heavy drinking is the leading cause of liver problems in the world.

## Finally, your doctor will give you a physical exam

During your physical exam, your doctor will look for specific signs of liver disease. These include:

- **Rashes and signs of scratching:** As mentioned earlier, the build-up of waste in the skin often causes itching.
- **Jaundice:** A build-up of a waste product called bilirubin can give your skin and eyes a yellowish color. Newborns often have jaundice that's unrelated to Alpha-1 liver disease. Their livers aren't mature enough to handle the bilirubin caused by the breakdown of red blood cells. If your baby is born with jaundice, there are tests to find out what's causing it.



**IT'S A FACT:** It's important to realize that newborns often have jaundice unrelated to Alpha-1 liver disease.

- **Clusters of veins:** Some patients with liver disease have unusual clusters of veins on their skin, but very few other symptoms. Portal hypertension can also cause large, blood-filled veins near the belly button.
- **Swelling in the feet and hands:** Liver problems may cause abnormal fluid and salt retention. This may lead to swelling of the hands and feet.

### Other signs of liver disease

- **A change in heart and lung function:** Liver-related problems can affect both your heart and lungs.
- **Enlarged or painful liver:** Your doctor can learn a lot about your liver health by examining your abdomen. They can feel if it's gotten bigger. It may be painful to the touch or feel rough through the skin. It may be harder than normal or have an unusual shape. Any of these findings might be a sign of liver disease.
- **A swollen abdomen:** Your abdomen may be swollen for a number of reasons. Abnormal blood flow due to portal hypertension may cause your spleen to swell. Fluid, called ascites, can also build up in your abdomen. Ascites is a sign of severe liver disease.



**KEY LEARNING:** Ascites is a sign of relatively severe liver disease.

- **Hemorrhoids:** Enlarged and painful blood vessels around your anus and rectum may point to portal hypertension.

After taking your medical history and doing a physical exam, your doctor will most likely order [blood tests](#). Doing so will help them get a complete picture of your liver health.

## LAB TESTS FOR LIVER HEALTH AND FUNCTION

Lab tests are an important part of assessing your liver's health. Each test looks at specific functions of your liver.

### General lab tests for liver health

There are many diagnostic tests that provide general information about liver health or point to specific diseases. Let's look at some common blood tests for liver health.

**AST and ALT (formerly called SGOT and SGPT) levels:** If [your healthcare practitioner](#) orders a panel of liver function tests (LFTs), it will include these two tests. AST and ALT are chemicals or enzymes found normally within liver cells. They usually leak into your blood very slowly. But, if your liver cells are irritated or damaged, they'll leak out faster. You'll have more AST and ALT in your blood. The higher the level, the more injury to your liver.

Babies with Alpha-1 may have higher levels of AST and ALT. But, in most cases, the level returns to normal by the time they're two years old. Adults with Alpha-1 commonly have elevated levels of AST and ALT. In general, these tests do not suggest major problems of scarring in the liver unless other tests are also abnormal. If the liver injury happens slowly over time, it may go unnoticed.



**GOOD NEWS:** AST and ALT are commonly elevated in infants with Alpha-1; but in most of these infants, these elevations return to normal during the first two years of life.

### **Alkaline phosphatase (ALP) and gamma-glutamyl transferase (GGT) levels:**

These chemicals are normally found within the cells of the bile ducts. These are tubes that drain the bile from the liver to the intestine. A higher level of these chemicals can be a sign of injured bile duct cells.

**Platelet count:** Low platelet count is often the first abnormal test for Alphas with liver diseases. It's part of the complete blood count (CBC).

Liver disease can affect many other organs of your body. Your healthcare practitioner may order a test to look at other body systems. This might include:

**Alpha-fetoprotein:** Some patients with cancer, including liver cancer, have high levels of alpha-fetoprotein in their blood. Since Alpha-1 is a risk factor for liver cancer, this test may help catch it early.

### **Tests of specific liver functions**

While some tests look at your **general liver function**, the following lab tests look at **specific functions** of the liver.

#### **Lab tests for synthetic function**

These tests usually measure the blood levels of substances that your liver produces.

**Albumin:** This protein helps move salts and nutrients

throughout your body.

**INR:** This is a measure of blood clotting that can uncover liver problems. Some drugs affect INR, so tell your healthcare practitioner about any drugs you take regularly.

#### **Lab tests for digestive function**

Bile from your liver is critical for the normal digestion and absorption of fat-soluble vitamins. Measuring levels of fat-soluble vitamins in your blood is a good test of your liver's digestive function. It can show if you're getting the nutrients you need. Healthcare practitioners sometimes look at blood levels of vitamins A, D, and E, especially in children.



**IT'S A FACT:** Patients with significant liver disease will often be low in vitamin K. This is measured with a high blood test for INR.

Patients with serious liver disease often have low levels of vitamin K, another fat-soluble vitamin. Vitamin K plays a key role in blood clotting. All forms of seriously low levels of vitamin K will show up as a high INR test result.

#### **Lab tests for excretory/detoxification function**

Your liver helps your body rid itself of toxins. So, testing your blood for levels of specific waste products can uncover liver problems.

**Blood bilirubin:** Measuring blood bilirubin levels is a common method used to assess liver function. Your liver eliminates bilirubin through bile.

There are several types of bilirubin in your blood. Levels of certain types will be too high if your liver isn't functioning normally. Other types of blood bilirubin aren't related to liver function.

**Ammonia:** High ammonia levels can be a sign of severe liver disease. When your body digests proteins, it releases ammonia into the blood. Normally, your liver breaks the ammonia into less toxic compounds. But a failing liver will allow ammonia to move into your bloodstream. If your ammonia levels get too high, you may start showing symptoms. These include unusual sleepiness, changes in thinking and personality, and tremors (shaking).

### Other diagnostic tests

Your healthcare practitioner may order other types of tests to get a full picture of your liver health.

**CT scans ("CAT" scans) or ultrasounds:** These imaging tests create pictures of your liver. They can reveal birth defects, gallstones, and liver tumors. Unfortunately, these tests are not very good at measuring the amount of liver scarring or fat unless the changes are advanced.

**FibroScan®:** The FibroScan® uses sound waves to determine the density of the liver and has been used to suggest cirrhosis. Because it is a relatively new test, the sensitivity of the test is not clearly known. The value is that it can be repeated over time without X-rays and without the use of needles.

**Liver magnetic resonance imaging (MRI):** This test is very good for detection of liver cancers and new contrast agents are being tried to get a signal for cirrhosis. A new test called the magnetic

resonance elastography (MRE) is offered at some medical centers for this purpose. If you have metal in your body (artificial joints or pacemakers) the MRI technicians will need to know the details about the brand and number before they will let you under the strong magnets of an MRI machine.

**Endoscopy:** An endoscopy can help your healthcare practitioner see abnormal blood vessels associated with portal hypertension. They can also reveal other intestinal problems caused by liver disease. A special endoscopic procedure, called ERCP, can be used to evaluate the bile ducts and even treat gallstones.

During an endoscopy, a technician passes a flexible, lighted tube equipped with a mini-TV through your mouth or rectum. Then, they can directly examine your stomach or intestines.

**Biopsy:** Liver biopsies are used to obtain a sample of liver cells to examine under a microscope. After sedating you, the technician inserts a needle directly into your liver. Then, they remove a sample about the size of a toothpick. Biopsies may be valuable for understanding your liver problems. However, it's an invasive procedure, and there is a small but real risk of complications.

A liver biopsy is the most accurate way to diagnose [Alpha-1 liver disease](#). Researchers are studying other kinds of tests to see if they can take the place of liver biopsy.



**KEY LEARNING:** Liver biopsies are not required for the diagnosis of Alpha-1 or Alpha-1 liver disease. However, liver biopsies are currently the most accurate test to determine the amount of liver scarring and assure that Alpha-1 is the only cause of liver disease.



## OUTPATIENT CARE FOR ALPHA-1 LIVER DISEASE

### Outpatient care: Your first visit

Newborns with severe liver problems are often treated in a hospital setting. But many children and adults with milder liver disease receive outpatient care after their initial diagnosis.

Your initial [outpatient visit](#) after a diagnosis of [Alpha-1-related liver disease](#) should focus on evaluating and treating existing problems and preventing new complications. Your doctor will do a thorough medical history and a physical exam and order some tests.

If your physical exam reveals issues like low weight, poor childhood growth, or signs of abnormal blood flow (portal hypertension), they may order other tests.

If you haven't gotten hepatitis A and B [vaccines](#), you'll start getting these shots at this visit. Your doctor will check for signs of lung disease because Alpha-1 increases your risk for it.



**BURNING ISSUE:** Because all adults with Alpha-1 are at risk for lung disease, your healthcare provider should monitor you for signs of lung disease, even if your presentation was for liver disease.

### Outpatient care: Follow-up visits

Once your doctor has a clear picture of your liver health, they'll make a plan for regular monitoring. If they find problems, they can refer you to a specialist.

Your doctor will do basic [liver blood tests](#) once a year, for:

- Alanine Transaminase (ALT)
- Aspartate Transaminase (AST)
- Bilirubin
- Alkaline phosphatase (ALP)
- Gamma-glutamyl transferase (GGT)
- PT/PTT (INR)
- Albumin

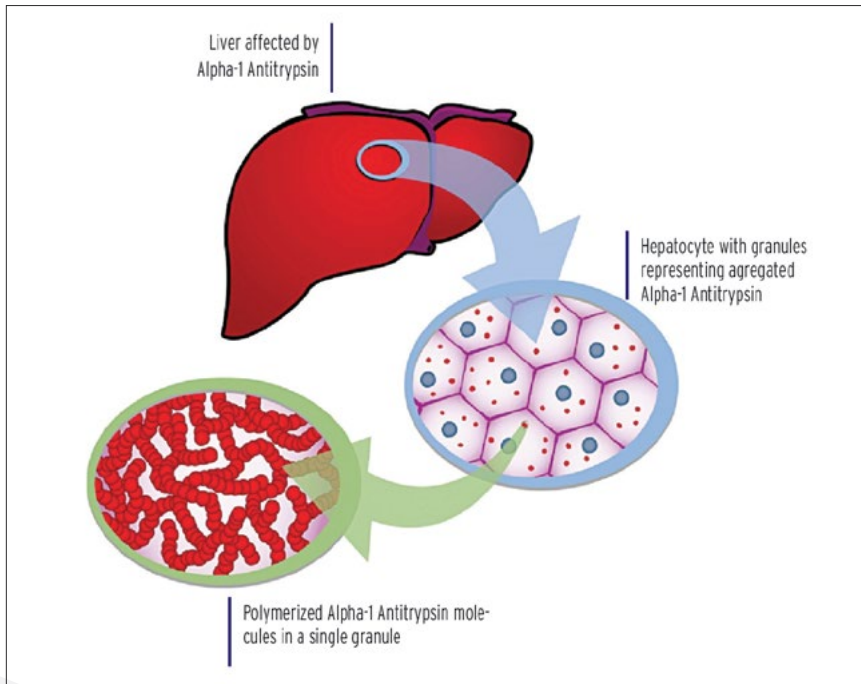
Every few years, you'll need specialized tests for liver cancer. Your risk is small but elevated, and regular testing may protect you in the long run.

### What if you have more severe liver disease?

If you show signs of more severe liver disease, you should see a liver specialist with specific knowledge of Alpha-1. They'll monitor you closely for signs of liver disease.

## ALPHA-1 LIVER DISEASE: CAUSES, TREATMENTS, AND MORE

Alpha-1 liver disease occurs when abnormal Z-type AAT proteins build up within liver cells. Some of the AAT proteins link to other AAT proteins in a process called polymerization. These polymers can form large or small clusters that damage the liver cell.



Silverman et al. NEJM

### Do all Alphas develop liver disease?

Some Alphas never develop liver disease. Sometimes these people have an unusual form of Alpha-1 — the Null deficiency. We assume they don't get a buildup of abnormal AAT protein because they don't make AAT in their livers. Other individuals seem to handle the AAT polymers without getting scarring in the liver. And sometimes early stages of liver disease produce no symptoms.



**IT'S A FACT:** Some Alphas never develop liver disease.

About 2% of children born with Z-type Alpha-1 develop liver failure and need a liver transplant before age two. Research suggests their liver's inability to handle protein build-up leads to damage.

Most infants born with two Z genes have abnormal liver function tests during this same time period. But, if they don't develop severe disease, their tests usually return to normal as they get older.

### Adults and Alpha-1 liver disease

Most adults with Alpha-1 won't develop significant liver disease. If liver disease does occur, it is often very mild and chronic. It may remain stable for years. However, some adults develop liver disease suddenly and quickly need a liver transplant. So far, there's no way to predict whether you'll have mild or severe liver disease.

Studies have shown that almost all Alphas over age 65 have liver scarring that can be seen under the microscope. This is true even when their liver function tests are normal. This suggests that all or most people with Alpha-1 have ongoing, low levels of liver injury throughout their lives.

Fortunately, your liver can tolerate this type of injury and still function normally.

We don't know all the reasons why some people have higher levels of liver damage. But we know that environmental and genetic factors play a role.

### **Treatments for AAT associated liver disease**

There is no specific treatment for Alpha-1 liver disease. But there are many treatments that help the outcome of all liver diseases that can be used. They focus on preventing complications from chronic liver disease. And, if liver problems arise, they treat them quickly.

### ***Lifestyle risk factors***

Obesity is the #1 lifestyle risk factor for liver disease. The "metabolic syndrome" occurs when obesity causes diabetes, high blood pressure, and increased cholesterol. The result is a fatty liver that is not tolerated well in Alpha-1.



**IT'S A FACT:** The metabolic syndrome associated with obesity is the most common cause of liver injury in our society and in Alphas.

### ***What does the future hold?***

This is an exciting time for new drug therapies that target Alpha-1 liver disease. Drugs are in clinical trials that stop AAT production in the liver. Other drugs help to get AAT out of the liver cell. At the present time, these treatments are only available through clinical trials.

Researchers are also working on [diagnostic tests](#) to help detect liver damage earlier.

## **TARGETED TREATMENT FOR ALPHA-1 RELATED LIVER PROBLEMS**

Your healthcare practitioner will suggest targeted treatment based on how Alpha-1 affects your liver. Let's take a look at some of these treatments.

### **Targeted treatment for poor digestive function**

#### ***Ensure proper nutrition***

Treatments for babies and children focus on ensuring normal growth and nutrition. If oral feeding is an issue, they may need a feeding tube. Poor growth and problems with oral feedings in infants with Alpha-1 are common. They even occur in infants without serious liver problems.

### **Boost fat digestion**

If you can't digest fat, you may lack critical vitamins. This can lead to major health problems.

VITAMIN	HEALTH IMPACT OF VITAMIN DEFICIENCY
Vitamin K	Life-threatening bleeding
Vitamin D	Rickets (bone damage in children)
Vitamin A	Brain and nervous system problems, like blindness and confusion
Vitamin E	Nerve damage in the hands and feet (Peripheral neuropathy)

### **Targeted treatment for poor excretory function**

#### **Stop severe itching**

Low bile flow makes life unpleasant because it causes severe itching. That can have a big impact on sleep, school and work, and quality of life.

Treatments include:

- Antihistamines to block itching
- Oral medicines to remove excess bile
- Drugs to increase bile flow

#### **Reduce ammonia buildup**

Severe liver damage causes waste products to build up in your blood. This may make you feel sleepy and confused. But, you can take medicine to flush waste products from your intestines.

### **Targeted treatment for poor synthetic function**

#### **Reduce fluid buildup**

Low blood albumin levels may lead to fluid buildup and swelling. When it happens in your belly, it's called ascites.

Your healthcare practitioner may recommend a range of treatments to reduce fluid buildup:

- A low-salt diet (not recommended for children)
- Drugs to remove excess fluid from your body (diuretics)
- Draining the fluid with a needle (paracentesis)
- IV albumin (injecting albumin into the blood)



**KEY LEARNING:** Limiting salt intake can help control swelling in the body.

#### **Boost blood clotting**

Your liver makes many proteins, including some clotting proteins. When you have liver disease, you may have low levels of these clotting proteins. If your clotting issues stem from poor digestion, taking vitamin K may help.

### **Targeted treatment for cirrhosis & portal hypertension**

Some Alphas with cirrhosis and portal hypertension live normally for decades without other health problems. However, others may have swelling and fluid buildup (ascites). We discussed treatment for those symptoms earlier.

### ***Avoid or limit certain drugs***

Patients with portal hypertension should avoid aspirin, acetaminophen, ibuprofen, and other non-steroidal anti-inflammatory drugs (NSAIDs). These drugs may lead to serious bleeding in liver disease patients. If you have mild to moderate liver disease, talk to your healthcare practitioner. You may be able to take small doses.



**IT'S A FACT:** Acetaminophen (such as Tylenol) overdose can cause liver damage.

### ***Reduce severe bleeding***

Portal hypertension (high blood pressure in the portal vein) may lead to severe bleeding from the esophagus (the tube connecting your throat and stomach), stomach, intestines, or rectum.

Treatments include:

- Endoscopy (inserting a flexible tube with a camera)
- Medicines (oral and IV)
- Blood transfusions
- Shunt surgery
- Transjugular intrahepatic portosystemic shunt (TIPS)

### **Targeted treatment for infections**

Chronic liver disease can weaken your immune system. Therefore, your healthcare practitioner should watch for signs of [infection](#). Call your healthcare practitioner if you have a fever with any of these other symptoms:

- abdominal pain
- vomiting
- diarrhea
- bleeding
- jaundice

Liver disease patients can develop severe infections. They may occur in the blood, liver, or built-up fluid in the belly. Your healthcare practitioner may treat these infections with [oral or IV antibiotics](#).



**KEY LEARNING:** Any person with significant liver disease should call their physician if they have a fever.

## LIVER CANCER

There is an increased risk of liver cancer (hepatocellular carcinoma) in Alpha-1. This rare form of liver cancer happens in all diseases that cause liver scarring. This is rare even in individuals with Alpha-1; it's just not as rare in Alpha-1 as it is in the general population. Cancers also are seen more frequently if the body is immune suppressed with transplant medications. The fastest growing hepatocellular carcinomas still do not grow very fast and they rarely metastasize out of the liver. The primary cure for this cancer is a surgery to remove the cancer nodule from the liver.



**KEY LEARNING:** Alphas are at an increased risk for liver cancer. Regular ultrasounds are recommended for early detection.

This surgery can be done if the cancer is small and does not involve critical structures inside the liver. For this reason, some experts recommend a liver ultrasound every 6 months to screen for a small cancer that can be removed for a cure, if found. Other liver experts would suggest that the right interval is yearly, and only if significant scarring is present in individuals over age 50. Importantly, there is no study that has been done to define the best interval and each center may have different suggestions.

## LIVER TRANSPLANT: WHO SHOULD GET ONE AND WHY

When your [liver disease](#) becomes life-threatening, it's time to consider a liver transplant. This procedure involves removing your diseased liver and replacing it with a healthy one.

You may have heard about "living-related donor" transplants. In these procedures, relatives donate a piece of their liver. This type of transplant doesn't work well for people with Alpha-1 for a number of reasons:

1. Alpha-1 is a genetic disease, so close relatives may share the condition.
2. Most centers won't transplant a liver from an Alpha-1 related donor. They're concerned that the liver will be at increased risk of injury in the future. The donor might also be at increased risk of liver disease.

Most people with Alpha-1 get their new livers from an unrelated organ donor, once the donor has been declared brain-dead.

### Preparing for a liver transplant

Your body's immune system sees the transplanted organ as a foreign "invader" and may attack it. There are a number of things that must be done to help your new liver survive and thrive.

First, tests must be performed to ensure that your cells and blood type are a good match to the donor's. Next, you'll have to take powerful medicines to suppress your immune system. This is intended to prevent your body from rejecting your new liver.

Of course, a weak immune system can put you at risk of serious [infections](#). You'll have to be careful for the rest of your life.

### Liver transplants save lives

Successful liver transplantation can provide decades of healthy life. Plus, your new, healthy liver makes normal quantities of normal AAT. Essentially, liver transplantation cures Alpha-1! However, you'll still have abnormal AAT genes. That means you can pass them to any future children.

So, if liver transplants are so great, why doesn't everyone with Alpha-1 have one? There are several reasons.

1. There aren't enough donor livers available.
2. Liver transplants are complicated surgeries and carry a high risk of death.



**GOOD NEWS:** Liver transplantation has become a highly successful and long-term solution to life threatening end-stage liver disease.

### Waiting for a liver transplant

As mentioned above, needing a liver transplant and getting one are two different things. To start the process, you must be evaluated at a liver transplant center.

At the center, they'll [test your bilirubin, PT, and creatinine](#) to assess the severity of your liver disease. Based on the results, they'll assign you a Model for End-State Liver Disease (MELD) score. Your score will fall between 6 and 40, with 6 being less ill and 40 being gravely ill. Children get Pediatric End-stage Liver Disease (PELD) scores.

When a donor liver becomes available, centers look at a range of criteria to decide who gets it. The patient with the highest MELD or PELD score and a good tissue match usually receives the liver. However, "Status 1 patients" whose livers have suddenly failed may be moved to the top of the transplant list.

### What might prevent a transplant?

There are several things that can prevent you from receiving a liver transplant, even if your liver disease is severe. You can't have a transplant if you have:

- Cancer in another part of your body
- Serious heart, lung, or nerve disease
- Active alcohol or illegal drug abuse
- An active, severe infection
- Problems following your treatment plan

Because people with Alpha-1 liver disease may also have [lung disease](#), this may be a roadblock to getting a liver transplant.







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