



# Not the Only Alpha-1:

A KID'S GUIDE TO  
ALPHA-1 ANTITRYPSIN DEFICIENCY



FOR AGES 8-11

# A Note to Parents

Welcome to “Not the Only Alpha-1: A Kid’s Guide to Alpha-1 Antitrypsin Deficiency!” The purpose of this book is to teach your child(ren) about Alpha-1. We hope that this book provides useful information and serves as a good start to discussions about Alpha-1. There is no one way to use this book, so feel free to adapt it to the level and needs of your child. Each child and family is different.

It is encouraged that you review this book before you share it with your child as it may include information that you have not yet discussed. You know your child best and can judge when the time is right to discuss some of the information. You may wish to copy pages or cut the book apart so that it can be shared in parts.

Depending on the age of your child, he or she may want to review this book alone or prefer to read it with you, grandparents, an older sibling, or an adult that is a trusted friend. Be sure to provide an opportunity for your child to ask questions and talk about subjects discussed in this book.

The Alpha-1 Association acknowledges the following individuals for their support in the development of this publication:

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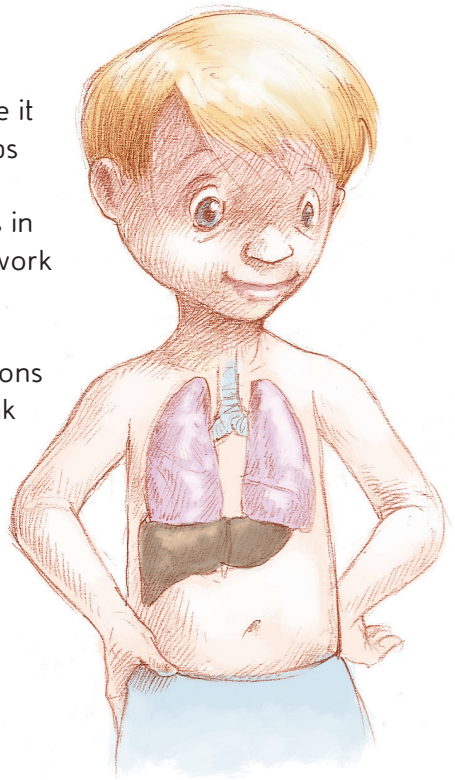
# What Is Alpha-1?

Alpha-1 is a short way to say “alpha-1 antitrypsin deficiency.” Having Alpha-1 changes the way your body works – especially the liver and the lungs.

Your liver is a big, brown organ that sits in your belly near your stomach. The liver is busy, because it does many jobs. Your liver cleans your blood, helps your body break down food, and makes proteins. Proteins can leave the liver and go to other places in your body. These proteins help other body parts work right and stay healthy.

Your lungs help you breathe. They are like balloons that fill up with air when you breathe in, and shrink down when you breathe out. Breathing gets your body oxygen, which your body needs all the time to work right.

This drawing shows your lungs and liver. Can you point to where they are on your own body?

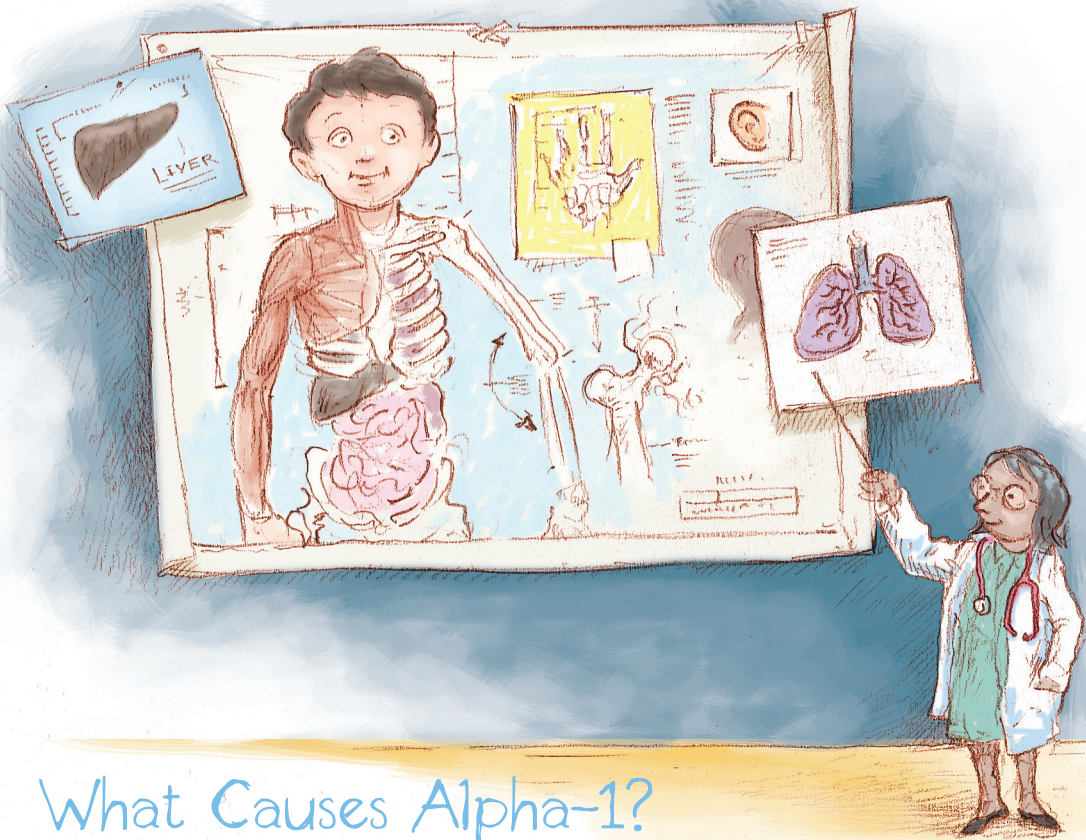


Did you know that  
you breathe about 15  
times every minute?  
Breathing happens  
whether you think  
about it or not!

The liver makes the protein called alpha-1 antitrypsin. This protein protects your lungs and keeps them healthy. When you have Alpha-1, your liver doesn't make the protein in the usual way. Sometimes it can't make the protein at all. Sometimes it can make the protein but can't get it out of the liver to go to the places in your body it needs to go, like the lungs.

Kids with Alpha-1 can get changes in their liver. You can't see the changes happening because they are inside your body, but sometimes you notice signs of them. Jaundice happens when your skin and eyeballs turn yellow. Sometimes your urine (pee) is dark and your stool (poop) is lighter than normal. Liver problems can also cause itching, or make it easier for you to get bruises or bleed when you get hurt. You can also get puffy legs or a swollen belly.

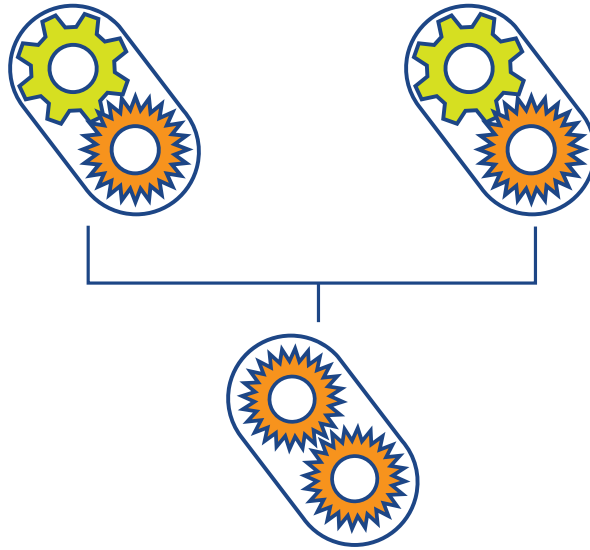
Kids with Alpha-1 don't have all of these changes. Most kids with Alpha-1 don't get serious liver problems, but some do. Although doctors can tell you that you have Alpha-1, they can't tell who will get liver problems and who won't. Kids with Alpha-1 don't usually have changes in their lungs. But, grown-ups with Alpha-1 can have trouble breathing. Breathing problems are worse for people who smoke cigarettes or work in places with dirty air.



## What Causes Alpha-1?

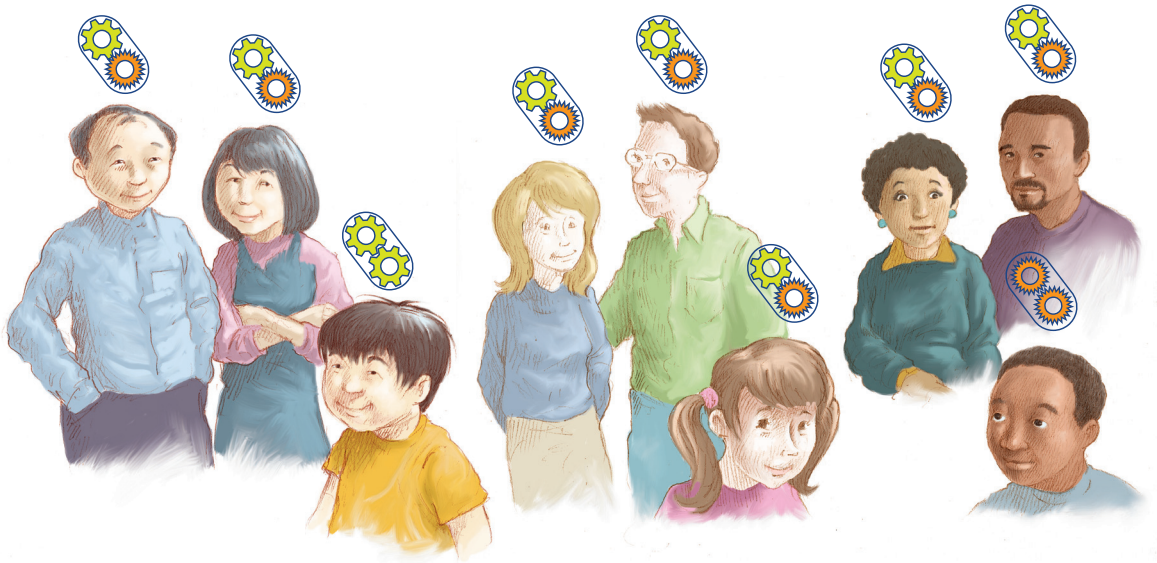
Alpha-1 isn't something you can catch, like a cold. It is something you are born with, because it is caused by changes in genes. Genes are instructions for making a person – they tell the body how to work. Genes control what you look like, as well as whether you get some diseases.

You get your genes from your parents. Half of your genes came from your mom and half came from your dad.



Changes in genes – called mutations – can cause health problems. People with Alpha-1 are born with two changes in the gene that makes alpha-1 antitrypsin. One gene change was passed from each parent.

The parents of a kid with Alpha-1 may have Alpha-1 or they may not. Most of the time, they have one gene with a mutation, but they also have one gene that is working and making protein. This is called being a carrier. Carriers often don't have lung or liver problems from Alpha-1. Carriers who smoke or work in a job with dirty air may get lung problems when they get older. A few carriers do get liver problems when they get older so they should have their livers checked by a liver doctor throughout their lives.



When both mom and dad are carriers, they can pass on different genes to their different children:

If both pass on their working genes, their child doesn't have Alpha-1 and isn't a carrier.

If one passes on a mutation and one passes on a working gene, their child doesn't have Alpha-1 but is a carrier.

If both pass on their mutations, their child has Alpha-1.

Parents can't control which genes they pass on when they have kids. Maybe you've heard that some things about you, like your eyes or the shape of your face, look like one of your parents. Maybe you look like your brothers and sisters, too. That's because they share some of your genes.

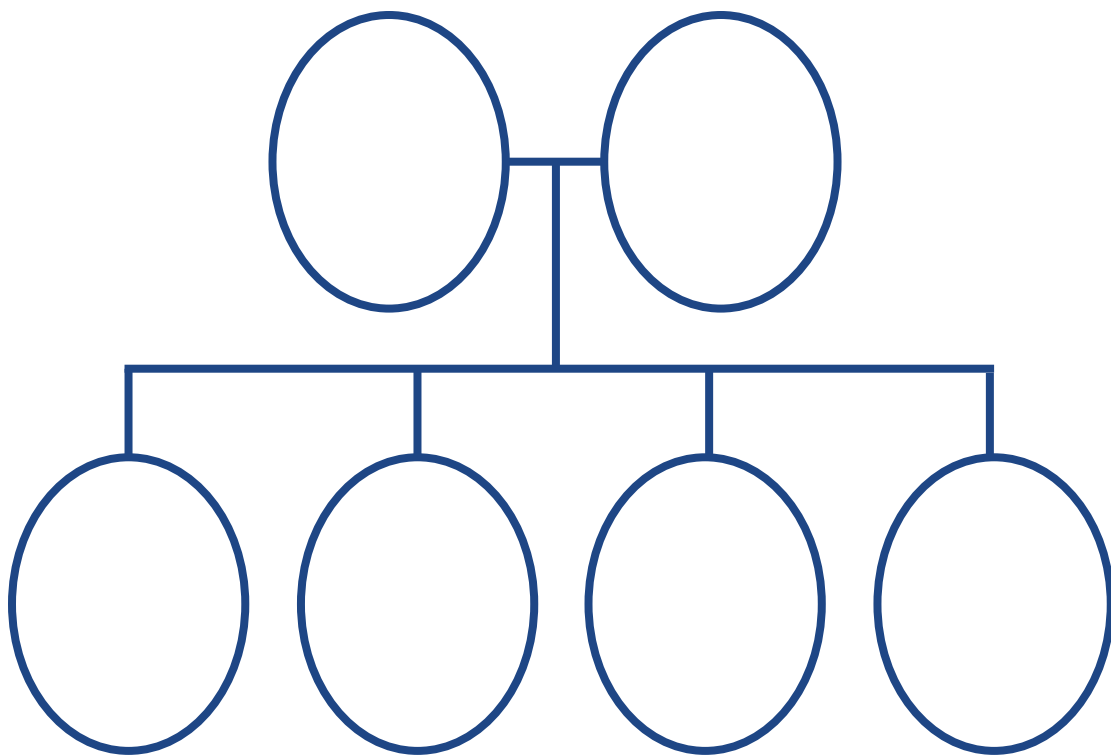
Some silly features are passed in our genes. For example, "hitchhiker thumbs" run in families the same way as Alpha-1. You can have thumbs that are straight, or you can have thumbs that are extra bendy.



Draw your family  
in the circles below.

Does anyone have  
hitchhiker thumbs?

Does anyone have  
Alpha-1?





## How Do I Know If I Have Alpha-1?

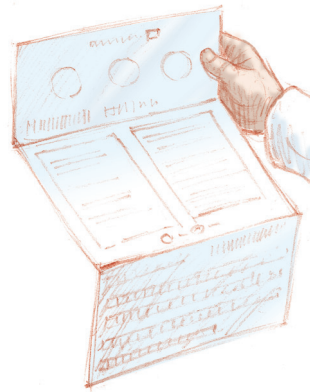


You may already know that you have Alpha-1 because you see a doctor for liver problems. Or, you may know that you have a chance to have Alpha-1 because it is in your family. You can be tested for Alpha-1 to find out for sure whether you have it or not.

The Alpha-1 test checks the type of the gene for Alpha-1 in your blood. To do the test, your doctor will need a small amount of blood from your arm or from a prick on your finger. Being poked with a needle is never fun, but it doesn't hurt too much. It mostly feels like a pinch, and it takes less than a minute.

Your blood will go to a special lab to be tested. There are three main types of the gene for Alpha-1. They are named with the letters: M, S, and Z.

- M is the normal type of the gene.
- S and Z stand for the two most common types of the gene that cause Alpha-1.
  - The S form of the gene makes a protein that works, but not as well as the protein that is made from the M form of the gene.
  - The Z form of the gene makes a protein that is the wrong shape and doesn't work well at all.





The letters can seem a little bit like an alphabet soup. Your test will tell you which two letters you have.

Most kids who have Alpha-1 have ZZ results. If you have ZZ, then you definitely have Alpha-1. It doesn't mean that you're sick. It just means that you will need more check-ups for your liver and lungs.

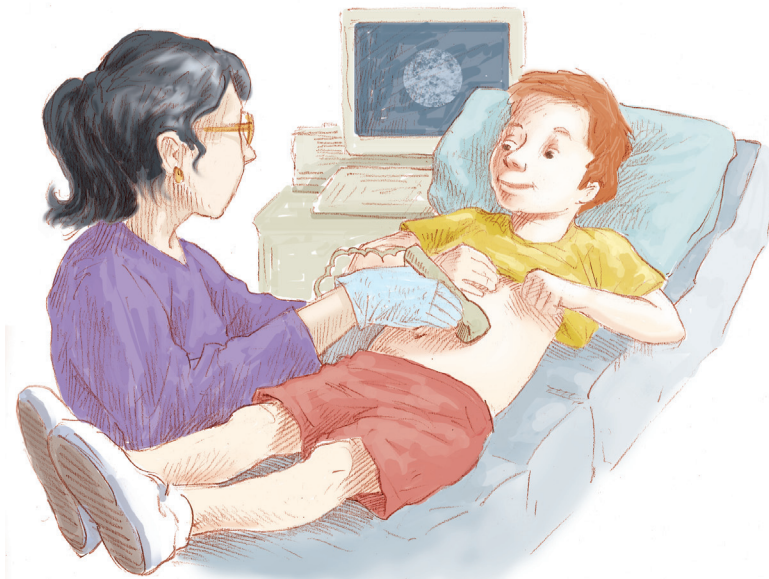
Some kids are SZ. Most of the time, people with SZ don't have any health problems as kids. Some kids that are SZ have liver problems. They also sometimes have lung problems when they grow up.

Kids who definitely don't have Alpha-1 are usually MM. Kids who are carriers may be MZ or MS. If you have MZ or MS, you probably won't have lung or liver problems, even when you get older. But you should still take special care of your liver and lungs and not do anything that could harm them, like smoke or work in areas with dirty air or drink alcohol excessively.

## What If I Have Alpha-1?

If you have Alpha-1, there are special check-ups that your doctors may do to see how your liver and lungs are working.

Did you know that an ultrasound uses sound waves to make a picture? You can't hear it working, though. The waves go so fast people's ears don't notice them!

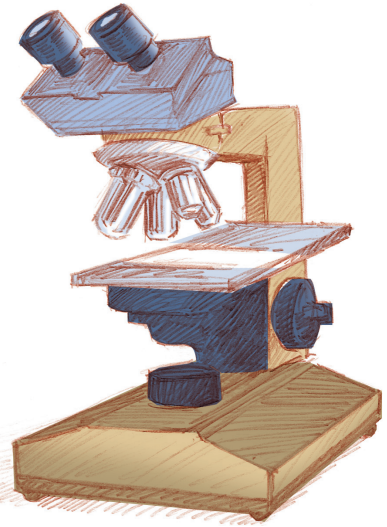


## Checking Your Liver

One way to know whether your liver is working right is to take a picture of it. Ultrasound is a special way of taking pictures of body parts like the liver that are inside of you.

When you get an ultrasound, you lie on a bed. You'll need to raise your shirt, so it's best not to wear a dress or overalls. To do an ultrasound, a doctor will use a special tool to rub some jelly on your belly. The jelly can be cold or feel sticky. You will feel the doctor press on your belly while looking at a TV screen. Having an ultrasound doesn't hurt at all. It usually doesn't take more than a half hour. Your parents can be in the room, and you'll be able to talk to them while you are having your ultrasound. You'll have ultrasounds every so often, depending on whether you have liver problems or not.

Doctors look at liver biopsies under a microscope. Have you ever looked in a microscope? It makes tiny things look really big.

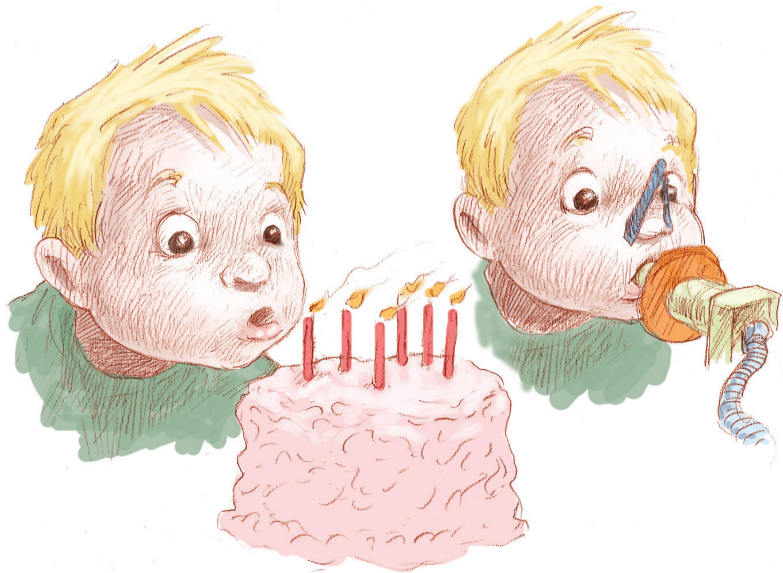


Another way to check your liver is to do special blood tests that find out whether your liver is working right. These blood tests are called liver function tests. To have them done, you need to have some blood drawn from your arm. Results of the test can tell your doctor if your liver is healthy or showing signs of a problem. You'll have to have this test done more than once. Your doctor will let you know when you need it.

Finally, the last way to check your liver is to do a liver biopsy. In a liver biopsy, a doctor takes a small piece of the liver. The liver is sent to the lab to see whether it looks normal or not.

If you need a liver biopsy, it will probably be done at a hospital. You won't be able to eat for a few hours before your test. You'll get some medicine to make you sleepy. Your doctor will use a needle to take a very small amount of your liver. The liver will grow back quickly.

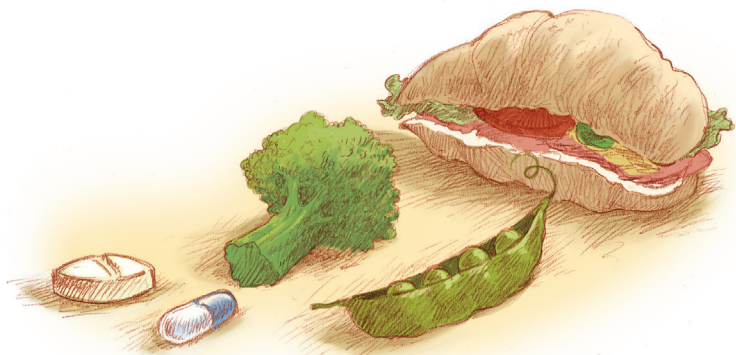
It doesn't take very long for your doctor to do a biopsy, but you may need to stay in the hospital for a few hours to make sure everything is fine. You may be sore afterward or get a bruise where the doctor poked you with the needle. You can ask your parents or your doctor if you can take some medicine so that it hurts less. You'll probably be able to go to school the next day.



## Checking Your Lungs

Most kids with Alpha-1 don't have serious lung problems. Your doctor may want to check your lungs just to be sure they are OK. The way to check how your lungs work is to have a pulmonary function test done.

Pulmonary function tests are easy and don't hurt at all. A doctor will have you take deep breaths while you're hooked up to a special machine. You'll need to blow out hard and fast, for as long as you can. (You can pretend that you're blowing out the candles on a birthday cake!). The machine measures how big your breath was and tells your doctor whether your lungs are OK. Having a pulmonary function test will probably take less than an hour. You won't need to do anything different afterward, so you can do whatever you'd like as soon as you leave the doctor.



## How Can I Try to Stay Healthy with Alpha-1?

A lot of kids with Alpha-1 are healthy. No one would know that they have Alpha-1, even a doctor. They don't look any different than other kids, and they don't have serious health problems.

Some kids with Alpha-1 do have more health problems. They may get sick more often.

For all kids with Alpha-1, it's important to try to stay as healthy as you can. Here are some things you can do:

- Eat your fruits and vegetables and play outside a lot! Eating healthy foods and being active help keep you healthy.
- Get your shots. Vaccines are shots that keep you from getting certain sicknesses. All kids get vaccines, but kids with Alpha-1 may get a couple of extra ones.
- Breathe clean air. Stay away from people who smoke and anything else that is stinky or makes you cough.
- Get your check-ups. The tests that your doctor wants you to have can find problems before they get more serious.
- If you need medicine, take it just like your doctor and parents ask you to.

## How Will I Know If I'm Sick?

There are some signs that can tell you if you are getting sick from Alpha-1. You should look for these signs, and tell your parents if you notice any of them. Having one of these signs doesn't mean that you're sick. They just mean that you may need to see your doctor to get them checked out.

- A yellow color to your skin or eyes
- Any changes in your pee or poop
- Itching
- Feeling tired
- Swelling in your feet or belly
- Getting more bruises than usual

Sometimes it's hard to talk about these signs because you get embarrassed. You may not know for sure whether something is normal or not. You may not know exactly what to say about how you're feeling. You may not want to make your parents worry. You may even want to keep these signs a secret, because you know they mean you could be sick and you don't want to be sick.

It's important that you tell someone even if you are feeling some of these things. Your parents and doctors will try to understand how you feel. Trust them – they want to help you.



## What If I Get Sick?

Knowing that you have Alpha-1 may make you scared about getting sick. It's normal to feel like that. If Alpha-1 makes you sick, your parents and your doctors will do everything they can to help you feel better.

There are medicines that can help with some of the problems in Alpha-1. Medicines can help with the itching or swelling if it happens. Your doctor may also change how you eat, to make sure that you grow and have enough energy.

## What If I Need Surgery?

Sometimes kids with Alpha-1 get pretty sick because their livers aren't working. If this happens to you, you may need a new liver. This is called getting a liver transplant.

Getting a liver transplant is a big deal, but it makes you better. You'll be in the hospital and have surgery. Your doctors open your belly, take out your liver, and put in a different liver. That liver may come from someone you know (like a relative) or someone you don't. You'll need to take medicines after the surgery to help your body get used to having a different liver. Having a liver transplant means being in the hospital and out of school for a little while.

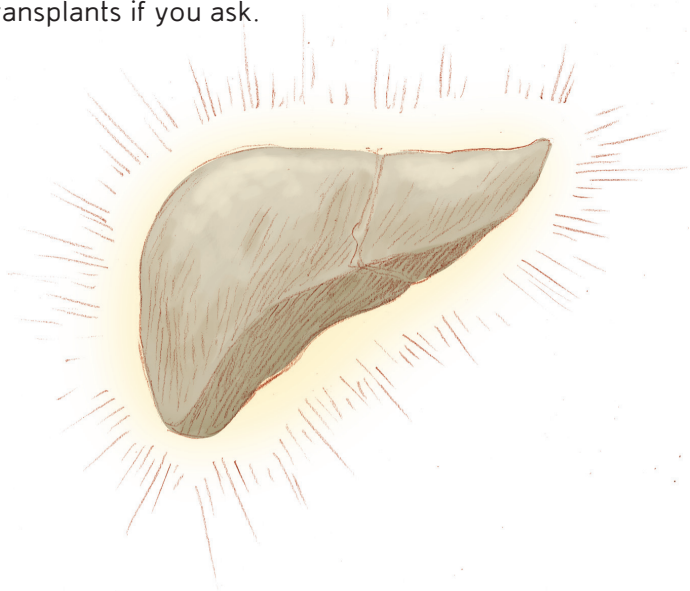
If you have a liver transplant, the new liver will make the Alpha-1 protein normally. So, kids who get liver transplants are cured of Alpha-1.



If you need a liver transplant, ask your doctor what to expect.  
Some questions you might have:

- How long will it take?
- Does it hurt?
- How long will I be in the hospital?
- After my surgery, can I do what I did before the surgery?  
How soon?
- Will I have to take medicine?
- Will anyone notice my new liver?

Your doctor will help you understand what it's like to have a new liver. You may even be able to meet other kids who have had transplants if you ask.



## How Should I Feel About Having Alpha-1?

There are lots of different ways you may feel about having Alpha-1. Some days, you may feel one way, and other days, you may have changed your mind. It may depend on your family, your health, and how your own mind works! No matter what you feel, it is normal for you.

Here are some ways kids might feel about having Alpha-1. Put a mark next to the ones that make the most sense to you. You may want to talk about these with your parents.

- I don't even think that much about having Alpha-1. It's no big deal.
- I hate having Alpha-1. I don't want to be sick.
- I worry about whether Alpha-1 will make me sick.
- I wish my parents didn't worry about me so much.
- I wish I knew someone else with Alpha-1 who understands what it's like.
- I get tired of going to the doctor.
- I don't understand why I have Alpha-1 and other kids don't. It's not fair.
- I don't like thinking about surgery. It makes me really nervous.
- I'm happy my doctors take good care of me.



Ask lots of questions.  
Asking questions is how  
you learn. Tell people  
about your feelings.  
Listening to you is how  
they learn.

## Should I Talk About Having Alpha-1?



Talking to your parents and your doctor about Alpha-1 is going to be important. It can be hard to know what to say or which questions to ask. If you are worried about something, or if you don't understand something, tell someone. Be brave and let them know how you're feeling.

Talking about anything that makes you different from your friends can be tough. If you just found out you have Alpha-1, it might take some time for you to be ready to tell anyone else about it. If you are sick from Alpha-1, you may or may not feel like telling your friends about why.

If you want to tell your friends about Alpha-1, talk to your parents first. They can help you think about whom to tell. They can even help you practice what to say. Once you tell a few people, you'll see that it isn't as hard as you think.

# Are There Other Kids With Alpha-1?

You may be the only person you know with Alpha-1. That can feel lonely. But there are a lot of other kids with Alpha-1. With your parents' help, you can learn more about Alpha-1 and connect to other kids through these organizations:

## **Alpha-1 Association**

(800) 521-3025 | [www.alpha1.org](http://www.alpha1.org)

## **Alpha-1 Kids**

(877) 346-3212 | [www.alpha1kids.org](http://www.alpha1kids.org)

## **Alpha-1 Association Genetic Counseling Program**

(800) 785-3177 | [www.alpha1.org/support/gencounselingcenter.php](http://www.alpha1.org/support/gencounselingcenter.php)

## **Alpha-1 Association Patient Information Hotline**

(800) 245-6809

## **Alpha-1 Foundation**

(877) 228-7321 | [www.alpha-1foundation.org](http://www.alpha-1foundation.org)

## **Clinical Resource Centers**

[www.alpha-1foundation.org/healthcare/?c=05-Clinical-Resource-Centers-CRCs-Healthcare](http://www.alpha-1foundation.org/healthcare/?c=05-Clinical-Resource-Centers-CRCs-Healthcare)

## **Alpha-1 Coded Testing Study**

(877) 886-2383 | [www.alphaoneregistry.org/](http://www.alphaoneregistry.org/)

## Word List

**Alpha-1 antitrypsin deficiency** – a condition that can cause lung and liver problems

**Carrier** – having one normal gene and one gene mutation

**Genes** – instructions for making a person

**Liver** – the organ in your belly near your stomach that cleans your blood and makes proteins

**Liver biopsy** – a test that takes a small piece of the liver to check at the lab

**Liver function test** – a blood test that checks to see if your liver is having problems

**Lungs** – the big balloons in your chest that fill with air when you breathe

**Mutation** – a change in a gene that can cause a disease

**Oxygen** – the part of air that we breathe and need to keep our bodies working

**Pulmonary function test** – a breathing test

**Transplant** – a surgery to take out a sick body part and put in a new one

**Ultrasound** – a test that takes a picture of your organs, like your liver

**Vaccine** – a shot that you get so that you don't get certain sicknesses

**ZZ** – the test result for most kids with Alpha-1

# Word Search

N A U D G O W U E T U T R J P  
P O P B X E S F S R E N E H M  
S Y I Y B G L J I A T P V Z L  
V B G T N B A N C N R E I U X  
K E L U A V P Y R S O N L M B  
N L L Q Y T R T E P K T Q F I  
X B R T A E U S X L R Z H W O  
V A C C I N E M E A Z I T E P  
D Y D R V B W P S N S F P F S  
U V R K C O L O R T E F Z H Y  
L A X S T I U R F O B G W O M  
C D L O K N Q K S F T U N W H  
E O G Z D S E L B A T E G E V  
Q S J Y B W B R T O Z I I Z N  
D V T M W H S W Y J O H A N H

BIOPSY  
CARRIER  
EXERCISE  
FRUITS  
GENES

LIVER  
LUNGS  
MUTATION  
OXYGEN  
PROTEIN

TRANSPLANT  
ULTRASOUND  
VACCINE  
VEGETABLES



**Alpha-1** Kids

The Alpha-1 Kids program is dedicated to providing support, education and information for parents and families of children affected by Alpha-1.

For more information about the Alpha-1 Kids program, please contact us at:  
Telephone: (877) 346-3212 • E-mail: [info@alpha1.org](mailto:info@alpha1.org)



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