

# LEARN ABOUT **LIVING WITH ALPHA-1**

The small changes  
you can make  
for a big impact



**GRIFOLS**



Many people with **alpha-1 antitrypsin deficiency**, also known as **AATD** or **alpha-1**, live full, productive lives. Here you will find advice about some changes in your lifestyle that will help you to improve your symptoms and stay as healthy as possible.

In addition, you will find information about treatment options for people with alpha-1. Since AATD is a genetic disease and can be inherited, you may want to share this information with your family.

## CHANGES THAT CAN IMPROVE YOUR QUALITY OF LIFE



### • **Quit smoking and avoid second-hand smoke**

If you smoke, it is extremely important that you quit. Smoking significantly increases the risk and severity of emphysema and may decrease your lifespan considerably. If your child has been diagnosed with alpha-1, it is important to protect them from exposure to second-hand smoke and educate them on the dangers of smoking.



### • **Reduce or eliminate alcohol**

Drink alcohol with caution, if at all. Many authorities recommend low, infrequent or no alcohol consumption for ZZ patients, and patients with liver damage should avoid alcohol completely.



### • **Avoid exposure to occupational and environmental pollutants**

These substances include pollen, dust or organic fumes, and second-hand tobacco smoke. They can irritate your lungs, cause or worsen lung problems, and also be absorbed through the skin and thus damage the liver.

Do not use aerosol sprays and try to avoid air pollution as much as possible, both at home and at work.



# EAT HEALTHY TO STAY HEALTHY

Eating well is essential to maintain good health. The nutritional needs of patients with alpha-1 are highly individualised.



## • Develop a nutrition program

If you have lung and/or liver problems, consider working with a nutritionist or a registered dietician to develop an individualised nutrition program. Reduce salt and protein intake because they can cause fluid retention, which is a common complication in alpha-1.

**Nutritional guidance** to keep in mind:

- Reduce the consumption of processed foods, as their high salt content can lead to fluid retention and high blood pressure.
- Vegetable sources of protein are better than animal protein.
- Some vitamins and supplements may stress an already stressed and compromised liver, so your doctor may recommend that you avoid some supplements.

# STAY ACTIVE AND IMPROVE YOUR HEALTH

**Exercise** is essential for alpha-1 patients. Not only does it improve your physical well-being and stamina, but it can also improve your mental outlook.



## • Develop an exercise program and make exercise a core part of your day

A professional can help you to develop an exercise program tailored to your needs. It is important to exercise muscles in the chest and upper body that are necessary for breathing as well as the large muscles of the legs.

You can also add endurance activities to your program, for example walking, strolling, swimming and/or biking. All this may improve your lung function and stamina.

Start exercising slowly and build the intensity of your program over time as your capacity increases.



## • Integrate mental well-being activities in your routine

Learning relaxation techniques can help you to have a more optimistic outlook on life and contribute to your overall well-being. Here are some approaches for you to try and find which one works better for you:

- |                       |                     |
|-----------------------|---------------------|
| · Yoga                | · Biofeedback       |
| · Meditation          | · Visualisation     |
| · Breathing exercises | · Hypnotherapy      |
| · Muscle relaxation   | · Positive thinking |

# WHAT TREATMENT OPTIONS DO YOU HAVE?

In addition to benefiting from lifestyle changes, if you have a lung and/or liver disease, you should seek medical help. You can find different treatment options depending on your condition – your doctor will discuss with you which are the best options for your situation.

Firstly, there are several **preventative measures** you can take to care for your overall health.



## • Vaccinations

- It is very important for you to have annual flu shots.
- A pneumococcal vaccine should be given if you are not yet protected.
- Discuss if you need the hepatitis A or B vaccines with your doctor.
- Make sure you get vaccinated against COVID-19.



## • Hand hygiene

Wash your hands frequently to prevent the passage of viral or bacterial infections.

# TREATMENT OPTIONS FOR LIVER DISORDERS



## • General treatments for liver complications

Once liver disease is identified, the first course of action is to evaluate your lifestyle to discover any substances that may be affecting your liver and aggravating your condition, such as alcohol, certain vitamins or medications.

Some medicines may be harmful to your liver. Compile a list of all the medicines you use and review it with your doctor.

## • Medical and surgical procedures

All these treatment options seek to improve the symptoms of advanced liver disease:

- **Large volume paracentesis:** this procedure removes large volumes of fluid from the abdomen.
- **Banding or sclerotherapy of veins in the oesophagus:** both treatments aim to reduce bleeding from the veins.
- **Portal vein decompression:** this involves major surgery to reduce pressure in the blood vessels that enter the liver from the digestive organs.
- **Liver transplantation:** after transplantation, there will be normal levels of alpha-1 antitrypsin, so the symptoms will improve considerably.

**Please consult your doctor if you have questions about any of these treatments or procedures.**



# TREATMENT OPTIONS FOR LUNG COMPLICATIONS



- **Antibiotics**

One of the main causes of loss of lung tissue may be bacterial infection in the lungs. At the first sign of a lung infection, your physician may prescribe some antibiotics to fight against the infection.

- **Bronchodilators**

Many symptoms of alpha-1 are similar to asthma or chronic obstructive pulmonary disease (COPD). Bronchodilators are usually given as inhalers and improve lung function, allowing better airflow.

- **Corticosteroids**

This treatment (via inhalation, oral or intravenously) reduces inflammation within and around the airways, improving lung function.

- **Supplemental oxygen**

It can be a life-saving therapy for patients with low blood oxygen levels.



- **Surgery options**

- **Lung volume reduction:** this consists of removing part of the damaged tissue. The benefits may last a few years.

- **Lung transplantation:** in this procedure, one or both lungs may be replaced. The benefits and quality of life after transplantation are specific to each person.

- **Augmentation therapy**

This therapy works to increase alpha-1 antitrypsin protein levels in the blood. Augmentation therapy is used to delay the progression of emphysema, reduce the frequency of exacerbations and improve quality of life.

The next page outlines more information about this therapy option.



**Please consult your doctor if you have questions about any of these treatments or procedures.**

## WHAT IS AUGMENTATION THERAPY?



Also called **replacement therapy**, it is a specific treatment for alpha-1 patients with lung disease. It is the process of receiving AAT protein to increase (or augment) the protein levels in your blood. This protein has been purified from the blood of healthy human donors.

AAT protects the lungs from the destructive effects of neutrophil elastase, an enzyme released by our body's white blood cells during inflammation or infection.

## HOW IS AUGMENTATION THERAPY GIVEN?

The therapy is given in a **weekly intravenous infusion**: your doctor calculates the dose you need according to your characteristics. Since the nature of this treatment is ongoing and lifelong, many patients make this part of their routine.

Normally, these infusions are administered by healthcare professionals at home, at a physician's office, at an outpatient infusion centre or other medical facility. You can choose to self-infuse at home after receiving appropriate instruction from a healthcare professional and the approval of your doctor.



**Augmentation therapy is the only specific therapy for alpha-1 lung disease, but it is important to note that this treatment option is not a cure: it will not reverse lung damage that has already occurred nor treat or prevent liver problems related to alpha-1 but it will protect your lungs from future damage.**

## WHAT YOU NEED TO DO BEFORE STARTING AUGMENTATION THERAPY

Ahead of starting treatment, you may be tested to see if you have IgA deficiency. This is done as a precaution to avoid any possible serious allergic reactions to plasma-derived products.

Immunisation against both hepatitis A and B is also recommended in order to reduce the risk of liver injury if you have not had the vaccination already.



## POSSIBLE SIDE EFFECTS



Patients on augmentation therapy have reported various side effects, but the vast majority have never experienced any major complications.

The most common side effect is a sense of feeling tired or having flu-like symptoms (up to 24 hours after an infusion). Other patients have mild allergic-like reactions: rash, itching, tightness of the chest, dyspnoea and/or wheezing.

If this happens, you can continue with your treatment if you take an antihistamine. Adverse events are not usually serious enough to stop the treatment.

**Your doctor will inform you if augmentation therapy can help you and will guide you through the whole process.**



## WHERE TO FIND ADDITIONAL SUPPORT

Here is more helpful information available for you and those in your family who have been diagnosed with alpha-1. The area where you live may have support groups and/or patient organisations that can offer you guidance and support. Patient organisations are the best place to share experiences and to learn more about living with AATD.



You can find out if there is a support organisation in your country here: **[www.alpha-1global.org](http://www.alpha-1global.org)**



Additional information and resources about alpha-1 can be found here: **[www.alpha1.org](http://www.alpha1.org)**