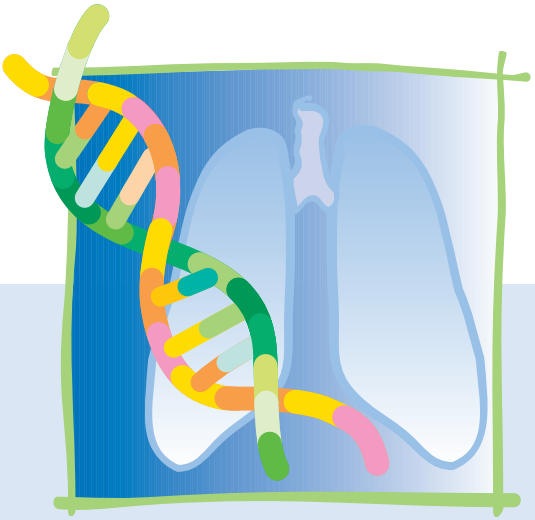
Breathless

The Whys and Wherefores



Living with Alpha-1-Antitrypsin Deficiency

Patient Information Program

Breathlessness

Short of breath after running the marathon? Short of breath after climbing a flight of stairs? Short of breath after gentle exercise? Short of breath even at rest? There are many reasons why someone may feel short of breath. When we consider shortness of breath we think mainly about the lungs and disorders that affect them.

No doubt you've met or observed people who clearly have problems with their lungs.

Mr. M., for example, whom you may encounter at the bakery shop in the morning, coughs into his handkerchief. You get the impression that he always has a cold. When he enters the bakery shop he's completely out of breath, yet he lives just around the corner.

Sometimes a lady can be seen sitting on a bench in the park. She's usually reading a book. When you look closely you may notice a thin tube leading to her nose. Next to her on the park bench is an oxygen cylinder that supplies her lungs and her body with vital oxygen. Without an outside oxygen supply Mrs. B. would be unable to walk without assistance. Because of hyperinflation (emphysema), her lungs are no longer able to provide her body with sufficient oxygen.

Then there's the neighbor's little girl. She suffers from frequent asthma attacks when she plays or exercises vigorously. She always moves cautiously and has to be careful not to catch a cold.

If you see yourself in any of these people or if you've had the feeling that you can't get enough air in other situations, you should read this brochure.

As shown by the examples, there are many causes of shortness of breath: There's asthma with sometimes severe attacks that in some circumstances can only be treated in hospital. You're probably familiar with chronic bronchitis, which often affects people who have smoked for years. Unlike asthma and chronic bronchitis, emphysema is a disease of the lung tissue accompanied by irreversible hyperinflation.

Emphysema

Hyperinflation of the lungs is caused by destruction of the tiny air sacs. The air sacs are responsible for the exchange of gases, including the uptake of oxygen from the air. Destroyed air sacs are no longer able to supply sufficient oxygen to the body. Sufferers have a feeling of shortness of breath and find it increasingly difficult to carry out normal activities of daily life.

Alpha-1-Antitrypsin Deficiency

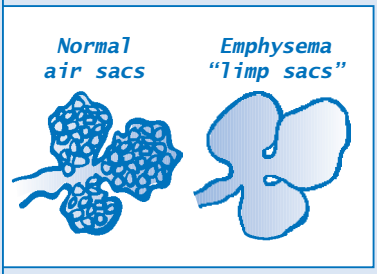
There are other causes of emphysema besides those just mentioned. They include the hereditary disease Alpha-1-Antitrypsin deficiency, or AAT deficiency for short.

It is assumed that far more than 100,000 people carry the Alpha-1-Antitrypsin deficient genotype in Europe. So far very few of these people have been identified and are receiving appropriate medical treatment. This means that most of those affected are not even aware that they have the condition (high number of unreported cases) and are therefore unable to take preventive measures.

The following provides brief information about several important aspects of this disorder that should help you judge whether you have it or perhaps you know someone who might find this information useful.

What does AAT deficiency mean?

AAT (Alpha-1-Antitrypsin) is a protein produced in the liver. It prevents another protein (neutrophil elastase), which is produced mainly by white blood cells and some types of cells in the lungs, from damaging the walls of the air sacs in the lungs. In healthy individuals the two opposing substances are balanced, i.e. AAT is able to neutralise neutrophil elastase before any damage to lung tissue occurs.



If AAT is absent or is not produced in sufficient quantities, the walls of the air sacs are destroyed by neutrophil elastase, leaving only "limp sacs". Because the air sacs are no longer able to engage in vital gas exchange (carbon dioxide out of the body / oxygen into the body), the body does not receive an adequate supply of oxygen.

The first symptoms of emphysema, such as shortness of breath on exertion, are usually occurring between the ages of 30 and 40 and varying in severity.

Not enough Alpha-1-Antitrypsin

Some diseases are "acquired" in the course of life, e.g. as a result of an unhealthy lifestyle. They include, for example, chronic bronchitis and narrowing of the coronary vessels in the heart. Other diseases are present from birth. Referred to as hereditary diseases, they include AAT deficiency.

Every person is a "miracle of nature" and is unique as an individual (even "identical" twins). For this reason several variants of one and the same disease may exist. This is the case with AAT deficiency. Not everyone who has the predisposition will develop emphysema in the course of his or her life. For example, you may have healthy parents yet still develop the disease.

What about me?

Whether or not a person has a predisposition for AAT deficiency can be determined today with the help of various tests.

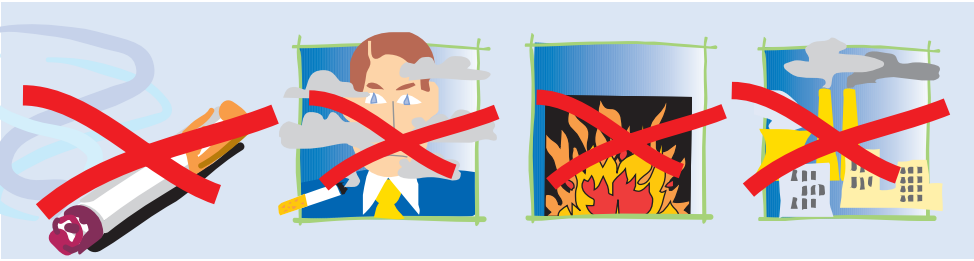
You should have a word with your doctor if

- you have a close relative who has developed emphysema or is known to have AAT deficiency.
- you yourself have a chronic lung disease that does not improve despite treatment.

A test known as the AlphaKit® now permits rapid, reliable diagnosis of AAT deficiency. Further information can be found on the internet at www.alphakit.de.

What can I do?

If it turns out that you yourself have (or your child has) AAT deficiency, there are a few important things you can do.



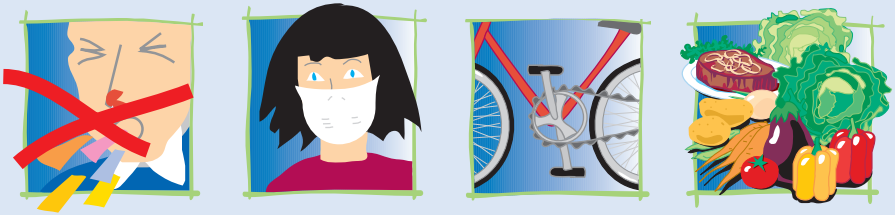
If no symptoms are present

- Protect
- Prevent

Because emphysema may not necessarily have already developed at the time AAT deficiency is diagnosed, affected individuals can help delay progression of the disease as long as possible and minimize damage to their lungs by:

- Not smoking
- Avoiding irritants at the workplace
- Avoiding open wood fires
- Getting regular (pulmonary) exercise
- Ensuring a healthy balanced diet
- Avoiding respiratory infections

Your doctor can also provide assistance. For example, he or she can help you stop smoking (patches, smoking cessation programs) and avoid respiratory infections (vaccinations against flu and pneumococcus, a common cause of pneumonia).



If symptoms are present

- ***Slow down progression of the disease***
- ***Relieve the symptoms***

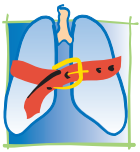
If you have been found to have AAT deficiency and you already have emphysema, it is extremely important that you help halt or slow the progression of the disease and minimize the symptoms:

- The first commandment is do not smoke (including passive smoking).
- According to the latest studies on pulmonary rehabilitation, exercise improves lung function and stamina.
- A balanced protein and vitamin-rich diet will provide your body with energy reserves for longer and more strenuous activities.
- To prevent colds you should avoid large crowds.
- You should avoid any irritants that could affect your lungs; these include irritants at your place of work (dust, vapors), open wood fires and high ozone concentrations.

Needless to say, your doctor will answer any medical questions you may have. In particular, he or she can vaccinate you against flu and pneumococcus and give you moral and practical support if you still smoke. Last but not least, effective drugs are available today that reduce symptoms and are able to slow the progression of the disease.

Want to find out more?

The brochures illustrated below can be ordered free of charge on the internet at www.alpha-1-info.com.



What happens in Alpha-1-Antitrypsin Deficiency?

How can people with Alpha-1-Antitrypsin Deficiency protect their lungs?

Why is nutrition so important for people with Alpha-1-Antitrypsin Deficiency?

Is exercise important for people with Alpha-1-Antitrypsin Deficiency?

How can people with Alpha-1-Antitrypsin Deficiency manage stress?

For further information and useful links see ***www.alpha-1-info.com.***

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