



Understanding Lung Disease



INTRODUCTION

To understand how the body works, we often describe how it is made up of various parts or body systems. For example, there is the cardiovascular system (the heart and blood vessels), the musculoskeletal system (bones and muscles), the respiratory system (lungs and air passages), and so on.

In reality, your body is much more than just the sum of its parts, and it is sometimes difficult to identify the point at which one body system ends and another system begins.

Although the space limitations of this brochure only allow us to provide a brief summary of the respiratory system's structure and function, we are hopeful this information will give you a better understanding of your condition and prepare you to have further discussions with your healthcare provider.

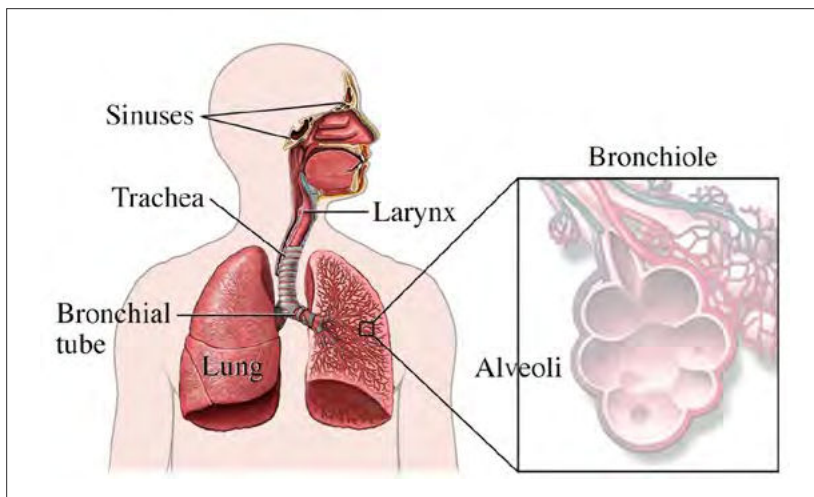
THE RESPIRATORY SYSTEM

How breathing works

When you breathe in (inspiration), air travels through the respiratory passages, exchanging oxygen for carbon dioxide. When you breathe out (expiration), you exhale carbon dioxide back into the air.

You don't have to think about doing this. It just happens. Your respiratory muscles contract and relax, moving air in and out of your lungs. That's where the exchange of gases between the air and your blood takes place.

The parts of your respiratory system



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The **upper airway** is made up of your nose, mouth, pharynx, and larynx. They warm, filter, and moisten the air you breathe. This protects your lower airway from foreign substances.

The **lower airway** is made up of your:

- Trachea (windpipe)
- Mainstem bronchi
- Secondary bronchi
- Bronchioles
- Terminal bronchioles

Air moves into and out of your lungs through these structures.

Each terminal bronchiole ends in clusters of structures made up of respiratory bronchioles, alveolar ducts, and alveolar sacs. The exchange between oxygen and carbon dioxide takes place in the alveoli (air sacs).

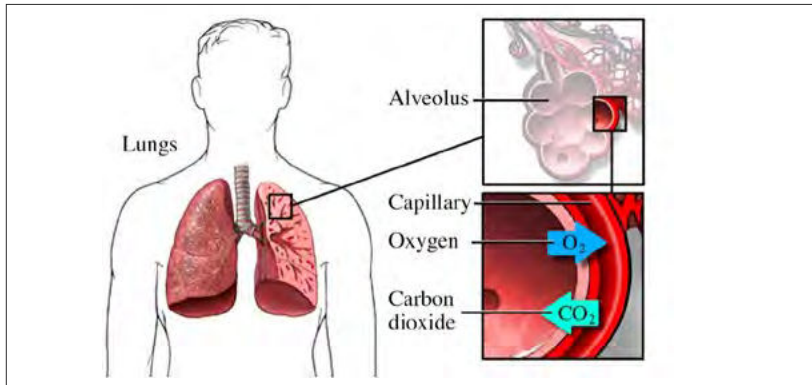
The mucociliary system helps protect your lower airway. It produces mucus, which traps foreign particles. Then, hair-like projections called cilia move them to the upper airway. When you cough or sneeze, you expel them from your body.

Oxygen and your circulatory system

Circulating blood delivers oxygen to all the cells in your body. The amount of oxygen that reaches the cells depends on several factors, including:

- How much oxygen you have in your blood
- How much hemoglobin (the principal oxygen carrier) you have in your blood
- The condition of your blood vessels
- How well your heart pumps blood through the body

When any of these factors are affected by disease, the levels of oxygen and carbon dioxide may become unbalanced.



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Breathing, your brain, and your respiratory system

Located in your brain stem, the medulla oblongata controls the number of breaths you take per minute (respiration). This part of the brain senses carbon dioxide levels in the blood. It sends impulses to the diaphragm and the intercostal muscles. This makes them contract and relax.

If you injure your medulla oblongata, the diaphragm and intercostal muscles can't function on their own. This may happen if you have a stroke.

In summary, your respiratory system has many moving parts that must work together to keep you healthy.



IT'S A FACT: When oxygen levels in the blood are significantly reduced due to respiratory disease, the brain stimulates extra respirations to compensate. Breathing quickly after exercise is a sign that oxygen levels should be checked.

COMMON LUNG DISEASES

Let's review some of the common lung diseases that can cause respiratory problems. Understanding them may help you take a more active role in managing your lung disease.

Chronic Obstructive Pulmonary Disease (COPD)

Chronic obstructive pulmonary disease (COPD) is a chronic inflammatory lung disease that causes airflow blockage and breathing problems. Emphysema and chronic bronchitis are the most common types of COPD.

In COPD, the airflow blockage progresses slowly and may be accompanied by airway hyperactivity (twitchiness or spasm) similar to what occurs in asthma.

Common COPD symptoms include:

- Difficulty breathing and shortness of breath
- Coughing
- Wheezing

What causes COPD?

Cigarette smoke causes about 3 out of 4 cases of COPD. But the other 1 out of 4 cases can be caused by exposure to wood smoke, air pollution, fuel, dust, and/or chemicals.

COPD can also develop as the result of the inherited disorder Alpha-1 Antitrypsin Deficiency. This genetically acquired COPD is referred to as Alpha-1 Antitrypsin Deficiency-related Chronic Obstructive Pulmonary Disease (COPD). We often abbreviate this as Alpha-1 lung disease.

In fact, many Alphas are diagnosed with COPD several years before they get their Alpha-1 diagnosis. We strongly recommend Alpha-1 testing for anyone with COPD.



KEY LEARNING: It is strongly recommended that all individuals diagnosed with COPD be tested for Alpha-1.

What are my treatment options for COPD?

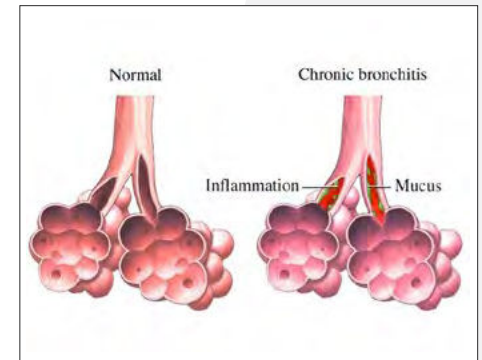
COPD symptoms are the same, whether you get it from your environment or genetics. How you manage the disease is also similar, but with some differences. Some people with Alpha-1 COPD get [Augmentation Therapy](#) to boost their alpha-1 antitrypsin levels.

However, each individual with COPD has differences in the types and severity of symptoms. Symptoms can change from day to day, so COPD requires treatment tailored to each individual person.

Chronic Bronchitis

The word bronchitis means inflammation of the air passages in your lungs. When you have bronchitis, your bronchial tubes swell, mucus glands multiply, and mucus production increases. This leads to coughing and shortness of breath.

In chronic bronchitis, the lining of your bronchial tubes loses its hair-like projections (cilia). When this happens, it's harder to cough up mucus. That causes an endless cycle of coughing, irritation, and more mucus production. Much of the shortness of breath comes from mucus obstructing the airways.



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What makes it “chronic”?

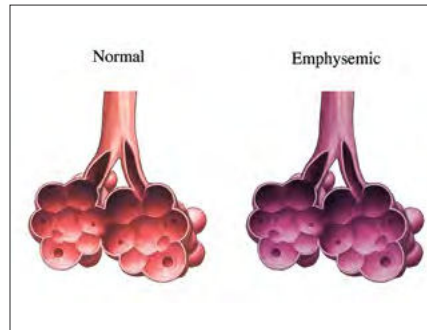
- You have a productive cough — one that brings up mucus.
- The cough lasts at least three months.
- You get this cough two years in a row.
- There are no other causes for this cough, like asthma, post-nasal drip, or reflux disease.

Common symptoms of chronic bronchitis include:

- Cough
- Mucus production
- Shortness of breath

Emphysema

This is a condition in which your lung's air sacs (alveoli) are damaged and enlarged. There's less surface area for the exchange of oxygen and carbon dioxide in these larger air sacs. So, less oxygen is absorbed into your blood. In severe cases, you exhale less carbon dioxide.



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As your small alveoli are destroyed, your lungs stretch out and become larger. They can't expel air when you exhale. Your bronchial tubes (airways) become "flabby" and collapse when you exhale. When air builds up in your lungs from the trapped air, your diaphragm becomes flattened and can't help you breathe.

Emphysema causes two major problems:

- Poor exchange of oxygen and carbon dioxide in your blood
- Difficulty exhaling air out of your lungs

Common emphysema symptoms include:

- **In mild or moderate emphysema:** Shortness of breath with strenuous activity. This can progress slowly. You may not notice it.
- **In more severe emphysema:** Shortness of breath with mild activity and even at rest.

Asthma

Asthma is a chronic condition that makes it hard to move air in and out of your lungs. It is one of the most common lung diseases.

When you have asthma:

- The muscles surrounding your airways (bronchial tubes) narrow.
- Your airways swell.
- You have extra mucus in your airways.

This blocks the airflow in and out of your lungs.

Unlike other lung diseases, if you have asthma, you can take medicine to help unblock your airways and make it easier to breathe. It is a "reversible" condition.



KEY LEARNING: As opposed to other lung diseases where the obstruction is not reversible, a key distinction of asthma is that the airway obstruction is reversible with appropriate medications.

Asthma & COPD

You may also be able to reverse some blocked airflow if you have COPD. But you won't be able to unblock it entirely — even with medicine.

If your blocked airflow is reversible, you don't have COPD. However, some people with asthma may develop irreversible airflow obstruction that looks just like COPD when it's measured by a simple spirometer.

Common symptoms of asthma include:

- Wheezing
- Chest tightness
- Coughing

Bronchiectasis

People with Alpha-1 lung disease often have bronchiectasis or severely damaged and chronically enlarged airways. Thanks to the increased use of CT scanning (lung computerized tomography) we now understand that many people can have severe airway enlargement without symptoms. This is especially true for Alphas.

Airway infections make bronchiectasis worse, whether you get many infections or just one bad one. These infections can come from viruses, bacteria, or atypical mycobacteria that are similar to tuberculosis.

Symptoms occur when secretions from your damaged airways form pools. This creates a breeding ground for all kinds of germs.

When bronchiectasis symptoms occur, they can include:

- Coughing
- Coughing up a lot of mucus that may be smelly, discolored, and/or bloody
- Shortness of breath
- Fatigue

As you can see, many lung diseases may affect your experience as an Alpha. You'll need breathing tests and a computerized tomography (CT) scan to sort out if you have one or more of them.

WHAT CAN YOU DO TO MANAGE YOUR LUNG DISEASE?

Now that you have learned about how the respiratory system normally works and some of the diseases that can cause respiratory problems, you are in a better position to discuss Alpha-1 and COPD with your healthcare providers. This knowledge will allow you to participate in the assessment and management of your healthcare more actively.



CROSS REFERENCE: For many more details about living with Alpha-1, including its diagnosis and management, review the comprehensive information found in the *Big Fat Reference Guide* at BFRG.alphanet.org

NOTES





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This single topic brochure is one of a series extracted from AlphaNet's Big Fat Reference Guide to Alpha-1 (the BFRG), which is available on the AlphaNet website (www.alphanet.org).

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