Understanding Lung Disease

Skinny Little Reference Guide™

AlphaNet

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This single-topic brochure is one of a Skinny Little Reference Guide™ series extracted from AlphaNet's Big Fat Reference Guide to Alpha-1 (the BFRG). These educational resources are available on the AlphaNet website (www.alphanet.org).

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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 musculo-skeletal system (bones and muscles), the respiratory system (lungs and air passages), and so on.

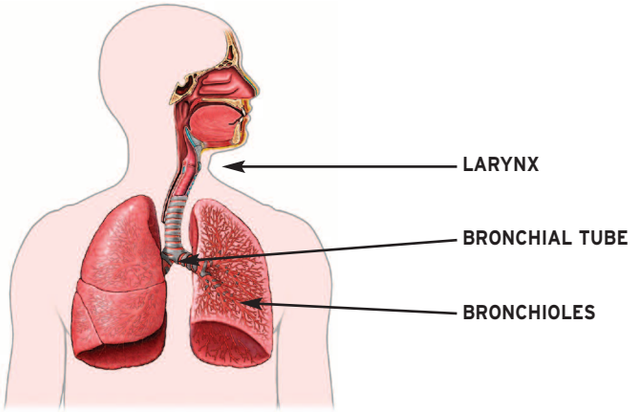
In reality, your body is much more than just the sum of its parts, and it is sometimes difficult to identify the point at which one body system ends and another system begins.

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 health care provider.

YOUR BODY AND YOUR LUNG DISEASE

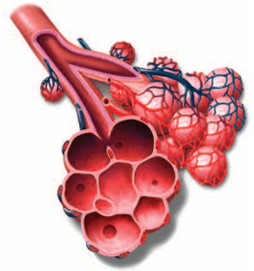
The respiratory system

The lungs are the part of your body where gases are exchanged between the air and your blood. When you breathe in (inspiration), air travels throughout the respiratory passages, exchanging oxygen for carbon dioxide. When you breathe out (expiration), you exhale carbon dioxide back into the air. Contraction and relaxation of the respiratory muscles moves air into and out of the lungs. For the most part, the contraction/relaxation of the respiratory muscles occurs automatically without any conscious effort on your part. When you contract or tighten your respiratory muscles, the diaphragm moves down and the ribs pivot upward allowing air to enter the lungs. When you relax these muscles, the diaphragm moves back up, the ribs pivot down and the lungs contract due to their normal elasticity. Air is pushed out of the lungs and back into the atmosphere.



The **UPPER AIRWAY** is composed of the nose, mouth, pharynx, and larynx. These structures serve as the initial passageway for air to enter the lungs. The upper airway is responsible for warming, humidifying, and filtering the air you breathe, thereby helping to protect the lower airway from foreign matter.

NORMAL BRONCHIOLE AND ALVEOLI



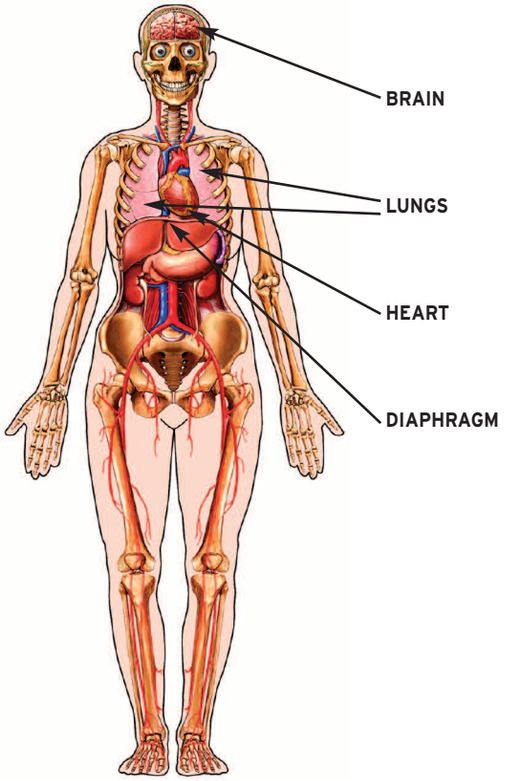
The **LOWER AIRWAY** consists of the trachea, main stem bronchi, secondary bronchi, bronchioles, and terminal bronchioles. The primary purpose for these structures, which are sometimes referred to as "dead spaces," is to provide passage for air to move into and out of the lungs. At the end of each terminal bronchiole, you will find a cluster of structures consisting of respiratory bronchioles, alveolar ducts, and alveolar sacs. The bronchioles and ducts function as conduits for air. The alveoli are the actual functional units of the lungs, and it is within these structures that the exchange of gases finally occurs.

The lower airway also provides additional defense mechanisms that help protect the lungs. The **MUCOCILIARY SYSTEM** and **COUGH REFLEX** are two such mechanisms that help by clearing foreign matter from the lungs.

The mucociliary system produces mucus, which traps foreign particles that are then moved up to the upper airway by specialized, hair-like projections called cilia. These collections of mucus can be expelled by expectoration or coughing. On a cellular level, the lower airway also initiates specialized responses to help fight disease and promote healing.

The molecular exchange of oxygen and carbon dioxide

The average adult lung contains an estimated 300 million alveoli. Each alveolus is supplied by many tiny blood vessels called capillaries. For oxygen and carbon dioxide to be exchanged, they must cross the alveolar capillary membrane. This exchange is promoted by diffusion — the passage of gas molecules through respiratory membranes. In diffusion, oxygen passes to the blood, and carbon dioxide, a by-product of cellular metabolism, passes out of the blood and is channeled away through exhalation.



How does the circulatory system affect oxygenation?

Circulating blood delivers oxygen to all the cells in the body. The amount of oxygen that reaches the cells depends on several factors, including: how much oxygen is in the blood; the concentration of hemoglobin (the principal carrier of oxygen) in the blood; the adequacy of the blood vessels comprising the circulatory system; as well as the ability of the heart to effectively pump blood throughout the body. Clearly, when any of these factors are affected by disease, the levels of oxygen and carbon dioxide in the blood can become out of balance.

How does the brain affect respirations?

Your respiratory rate, or the number of times you take a breath each minute, is controlled by a part of the brain known as the medulla oblongata, located within the brain stem. This part of the brain is sensitive to carbon dioxide levels in the blood, and sends impulses down the nerves to the diaphragm and the intercostal muscles between the ribs to stimulate the periodic contraction of these muscles. As mentioned previously, it is the contraction and relaxation of these muscles that permits the lungs to fill with and expel air. Injuries incurred to this part of the brain stem will impair the ability of these muscles to function on their own. Paradoxically, when oxygen levels in the blood are significantly reduced due to respiratory disease, the brain's ability to stimulate respirations can be negatively affected.

IT'S A FACT: When oxygen levels in the blood are significantly reduced due to respiratory disease, the brain's ability to stimulate respirations can be negatively affected.

COMMON OBSTRUCTIVE DISEASES OF THE LUNGS

CHRONIC OBSTRUCTIVE PULMONARY DISEASE. COPD is a disorder characterized by the presence of airflow obstruction due to chronic bronchitis or emphysema. In COPD, the airflow obstruction generally continues to progress slowly and may be accompanied by airway hyperactivity (asthma). The asthmatic component may be partially reversible.

Symptoms of COPD are:

- Difficulty breathing and shortness of breath
- Coughing
- Wheezing

Most frequently, COPD develops as the result of exposure to environmental toxins, the most common being cigarette smoke. 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). Many Alphas, in fact, were diagnosed with COPD several years before the diagnosis of Alpha-1 itself was made. It is therefore strongly recommended that all individuals diagnosed with COPD be tested for Alpha-1.

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

While COPD may be acquired either environmentally or genetically, most persons diagnosed with the COPD experience the symptoms of the disease in a similar fashion. Regardless of how the disease was acquired, the medical management is similar but has some differences. For those persons diagnosed with Alpha-1 COPD, treatment with Augmentation Therapy is used to supplement significantly reduced levels of the protein alpha-1 antitrypsin.

CROSS REFERENCE: For a comprehensive discussion of the management of Alpha-1 related lung disease, see the section on "Management of Alpha-1 Lung Disease" in the Big Fat Reference Guide™ at www.alphanet.org.

For more detailed information about Augmentation Therapy, see the section "Specific Therapy for Alpha-1 Lung Disease" in the Big Fat Reference Guide™ at www.alphanet.org.

CHRONIC BRONCHITIS literally means persistent inflammation of the bronchus. Bronchitis is said to be "chronic" when an individual has a productive cough that has lasted for at least three months during each of two successive years when other causes for a chronic cough (such as asthma, post-nasal drip, gastroesophageal reflux disease) have been ruled out.

Symptoms of chronic bronchitis are:

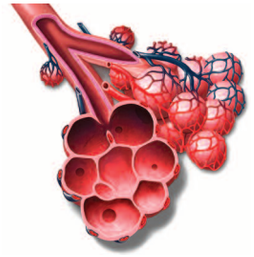
- Cough
- Sputum production
- Shortness of breath

When you have bronchitis, the bronchial tubes become inflamed, mucus glands multiply, and mucus production is increased — all of which leads to coughing and shortness of breath. In chronic bronchitis, the lining of the bronchial tubes lose the hair-like projections (cilia) that normally help propel the mucus up the bronchial tubes so it can be coughed up. When this happens, it becomes harder to cough up mucus, which in turn causes more coughing, more irritation, and more mucus production. This cycle results in the airways becoming swollen and clogged, which causes obstruction and increased shortness of breath.

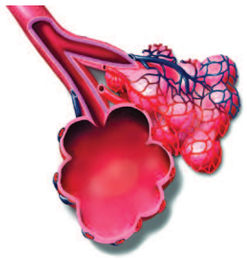
EMPHYSEMA is characterized by destruction of the air sacs, or alveoli, and loss of the lungs' elasticity. This loss of the walls of the air sacs causes these small structures to combine into larger units. These larger air sacs are less functional than the smaller ones, because they reduce the area where oxygen and carbon dioxide can be exchanged between the inhaled air and the blood. Consequently, less oxygen can be absorbed into the blood and, in the most severe cases; the amount of carbon dioxide getting exhaled is decreased.

Following the destruction of the small alveoli, the lungs become stretched out, less flexible, and are no longer able to recoil as they once did to expel air during exhalation. The supporting tissue of the bronchial tubes can be lost, causing "flabby" airways that collapse when air is exhaled. Due to the amount of air that is trapped in the lungs, the diaphragm, the broad muscle at the bottom of the lungs that acts like a bellows during breathing, can become flattened and unable to assist in respiration.

NORMAL BRONCHIOLE AND ALVEOLI



EMPHYSEMA



Thus, the major problems caused by emphysema are poor exchange of oxygen with the blood and difficulty exhaling air out of the lungs.

- In mild or moderate emphysema, you may have shortness of breath with strenuous activity. This can progress slowly and not be noticed.
- In more severe emphysema, you may have shortness of breath with mild activity and even at rest.

ASTHMA is defined as reversible airflow obstruction with increased reactivity or "twitchiness" of the muscles surrounding the bronchial tubes. The key word here is **REVERSIBLE**. As opposed to other lung diseases where the obstruction is not reversible, a key distinction of asthma is that the airway obstruction is reversible.

Asthma is characterized by three elements: constriction of the muscles surrounding the bronchial tubes, inflammation of the bronchial tubes, and overproduction of mucus. These lead to the obstruction of airflow in and out of the lungs. Reversible obstruction means that medication can cause this condition to improve or normalize.

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.

In COPD, airflow obstruction also may include a significant reversible component but, in general, this obstruction can never be entirely reversed, even with the use of medication. Individuals whose airflow obstruction is completely reversible are not generally considered to have COPD.

Some people with asthma may develop irreversible airflow obstruction that appears to be indistinguishable from COPD when it is measured by a simple spirometer.

Symptoms of asthma include:

- Wheezing
- Chest tightness
- Coughing

BRONCHIECTASIS is a condition that is fairly common in individuals with Alpha-1 lung disease. It is defined as chronically enlarged and severely damaged bronchial tubes. With the increased use of lung computerized tomography, or CT scanning, there has been a growing appreciation that individuals may have significant bronchiectasis without symptoms. This is especially true in Alpha-1.

Bronchiectasis can be worsened by recurrent airway infections or by a single episode of severe infection, such as bronchial pneumonia, tuberculosis, or whooping cough. Symptoms of bronchiectasis are due to the pooling of secretions in these damaged airways, which provide a good breeding ground for bacteria of various types.

When symptoms occur, they can include:

- Coughing
- Coughing up copious mucus that may be foul smelling, discolored, and/or bloody
- Shortness of breath
- Fatigue

WHAT CAN YOU DO TO MANAGE YOUR LUNG DISEASE?

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

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 www.alphanet.org

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This brochure is produced by AlphaNet as part of its Alpha-1 Disease Management and Prevention (ADMAP) program.

AlphaNet is a not-for-profit organization providing disease management services and support to individuals affected by Alpha-1 through a staff of medical professionals and specially trained AlphaNet Patient Services Coordinators, available 24 hours a day, 7 days a week. To learn more about ADMAP or to find the AlphaNet Coordinator nearest you, visit our website (www.alphanet.org).