Lung Disorders
Introduction to Respiratory Care
Obstructive versus Restrictive Lung Disorders

**Obstructive lung disease** -- a decrease in the exhaled air flow caused by a narrowing or blockage of the airways.

**Restrictive lung disease** -- a decrease in the total volume of air that the lungs are able to hold. Often, this is due to a decrease in the elasticity of the lungs themselves or caused by a problem related to the expansion of the chest wall during inhalation. *The ability to inhale is affected.*

Restrictive lung diseases may be divided into the following groups:

- Intrinsic lung diseases (diseases of the lung parenchyma)
- Extrinsic disorders (extra-parenchymal diseases)

Categories of restrictive diseases

- Pulmonary
- Thoracoskeletal
- Neurologic-neuromuscular
- Abdominal
Obstructive Disorders

Asthma

Asthma is a reversible obstructive lung disease, caused by increased reaction of the airways to various stimuli. It is a chronic inflammatory condition with acute exacerbations. Asthma can be a life-threatening disease if not properly managed.

* In 2005, it was estimated that 22.2 million Americans currently have asthma. Of these, 12.2 million Americans (3.8 million children under 18) had an asthma attack.1
* In 2004, asthma accounted for an estimated 14.5 million lost work days in adults.6
* Asthma breathing problems usually happen in "episodes" or "attacks," but the inflammation underlying asthma is continuous. An asthma episode is a series of events that result in narrowed airways. These include: swelling of the lining, tightening of the muscle, and increased secretion of mucus in the airway. The narrowed airway is responsible for the difficulty in breathing with the familiar "wheeze."
* Lung function declines faster than average in people with asthma, particularly in people who smoke and in those with excessive mucus production (an indicator of poor treatment control).
* Asthma medications help reduce underlying inflammation in the airways and relieve or prevent symptomatic airway narrowing. Control of inflammation should lead to reduction in airway sensitivity and help prevent airway obstruction.
* Two classes of medications have been used to treat asthma--anti-inflammatory or controller agents and bronchodilators or relievers. Anti-inflammatory drugs interrupt the development of bronchial inflammation and have a preventive action. They may also modify or terminate ongoing inflammatory reactions in the airways. These agents include corticosteroids, cromolyn sodium, and other anti-inflammatory compounds. A new class of anti-inflammatory medications known as leukotriene modifiers, which work in a different way by blocking the activity of chemicals called leukotrienes that are involved in airway inflammation have recently come on the market.
* Bronchodilators act principally to open the airways by relaxing bronchial muscle. They include beta-adrenergic agonists, methylxanthines, and anticholinergics.
* Asthma is characterized by excessive sensitivity of the lungs to various stimuli. Triggers range from viral infections to allergies, to irritating gases and particles in the air. Each person reacts differently to the factors that may trigger asthma, including:
  - respiratory infections, colds
  - cigarette smoke
  - allergic reactions to such allergens as pollen, mold, animal dander, feather, dust, food, and cockroaches
  - indoor and outdoor air pollutants, including ozone
  - exposure to cold air or sudden temperature change
  - excitement/stress
  - exercise
* Asthma may also be triggered by over the counter drugs. One study found that one adult asthmatic in five can suffer an attack from taking aspirin.

Bronchiectasis

Bronchiectasis is an abnormal stretching and enlarging of the respiratory passages caused by mucus blockage. When the body is unable to get rid of mucus, mucus becomes stuck and accumulates in the airways. The blockage and accompanying infection cause inflammation, leading to the
weakening and widening of the passages. The weakened passages can become scarred and deformed, allowing more mucus and bacteria to accumulate, resulting in a cycle of infection and blocked airways.

Bronchiectasis is one of the chronic obstructive pulmonary diseases COPD (Chronic Bronchitis and Emphysema) and it can be complicated by emphysema and bronchitis. The disease is commonly misdiagnosed as asthma or pneumonia.1

Bronchiectasis can develop at any age, begins most often in childhood, but symptoms may not be apparent until much later. Bronchiectasis can occur as part of a birth defect, such as primary ciliary dyskinesia or cystic fibrosis. About 50% of all cases of bronchiectasis in the U.S. result from cystic fibrosis. It can also develop after birth as a result of injury or other diseases, like tuberculosis, pneumonia and influenza.2

There are three types of bronchiectasis that describe the severity of the condition:

- **Cylindrical** - most common and refers to the slight widening of the respiratory passages. This type can be reversed and may be seen after acute bronchitis.
- **Varicose** - bronchial walls have both extended and collapsed portions.
- **Cystic** - most severe and involves irreversible ballooning of the bronchi.

Symptoms include coughing (worsened when lying down), shortness of breath, abnormal chest sounds, weakness, weight loss, and fatigue. With infections the mucus may be discolored, foul smelling and may contain blood. Symptom severity varies widely from patient to patient and occasionally, a patient is asymptomatic.

Doctors evaluate bronchiectasis with a chest x-ray, breathing tests, sputum culture or computed tomography (CT) scan. Testing for other associated diseases like cystic fibrosis or tuberculosis can also be helpful.

Bronchiectasis patients are often given antibiotics for infection and bronchodilator medicines to open passages. There are also bronchial hygiene techniques to help clear mucus. Lung transplants are also an option for severe cases.

**Chronic Obstructive Pulmonary Disease (COPD)**

*(Chronic Bronchitis and Emphysema)*

Chronic obstructive pulmonary disease (COPD) is a term referring to two lung diseases, chronic bronchitis and emphysema, that are characterized by obstruction to airflow that interferes with normal breathing. Both of these conditions frequently co-exist, hence physicians prefer the term COPD. It does not include other obstructive diseases such as asthma.

- COPD is the fourth leading cause of death in America.
- Smoking is the primary risk factor for COPD. Approximately 80 to 90 percent of COPD deaths are caused by smoking.
- Other risk factors of COPD include air pollution, second-hand smoke, history of childhood respiratory infections and heredity. Occupational exposure to certain industrial pollutants also increases the odds for COPD.
In 2007, the cost to the nation for COPD was approximately $42.6 billion, including $26.7 billion in direct health care expenditures, $8.0 billion in indirect morbidity costs and $7.9 billion in indirect mortality costs.\(^7\)

**Chronic bronchitis** is the inflammation and eventual scarring of the lining of the bronchial tubes. When the bronchi are inflamed and/or infected, less air is able to flow to and from the lungs and a heavy mucus or phlegm is coughed up. The condition is defined by the presence of a mucus-producing cough most days of the month, three months of a year for two successive years without other underlying disease to explain the cough.

- This inflammation eventually leads to scarring of the lining of the bronchial tubes. Once the bronchial tubes have been irritated over a long period of time, excessive mucus is produced constantly, the lining of the bronchial tubes becomes thickened, an irritating cough develops, and air flow may be hampered, the lungs become scarred. The bronchial tubes then make an ideal breeding place for bacterial infections within the airways, which eventually impedes airflow.
- Symptoms of chronic bronchitis include chronic cough, increased mucus, frequent clearing of the throat and shortness of breath.
- Chronic bronchitis doesn’t strike suddenly and is often neglected by individuals until it is in an advanced state, because people mistakenly believe that the disease is not life-threatening. By the time a patient goes to his or her doctor the lungs have frequently been seriously injured. Then the patient may be in danger of developing serious respiratory problems or heart failure.

**Emphysema** begins with the destruction of air sacs (alveoli) in the lungs where oxygen from the air is exchanged for carbon dioxide in the blood. The walls of the air sacs are thin and fragile. Damage to the air sacs is irreversible and results in permanent "holes" in the tissues of the lower lungs. As air sacs are destroyed, the lungs are able to transfer less and less oxygen to the bloodstream, causing shortness of breath. The lungs also lose their elasticity, which is important to keep airways open. The patient experiences great difficulty exhaling.

- Emphysema does not develop suddenly. It comes on very gradually. Years of exposure to the irritation of cigarette smoke usually precede the development of emphysema.
- Symptoms of emphysema include cough, shortness of breath and a limited exercise tolerance. Diagnosis is made by pulmonary function tests, along with the patient’s history, examination and other tests.
- **Alpha1 antitrypsin deficiency-related (AAT) emphysema** is caused by the inherited deficiency of a protein called alpha1-antitrypsin (AAT) or alpha1-protease inhibitor. AAT, produced by the liver, is a "lung protector." In the absence of AAT, emphysema is almost inevitable. It is responsible for 5% or less of the emphysema in the United States.
- Symptoms of AAT deficiency emphysema usually begin between 32 and 41 years of age and include shortness of breath and decreased exercise capacity. Smoking significantly increases the severity of emphysema in AAT-deficient individuals.
- Blood screening is primarily used to diagnose whether a person is a carrier or AAT-deficient. If children are diagnosed as AAT-deficient through blood screening, they may undergo a liver transplant. In addition, a DNA-based cheek swab test has been recently developed for the diagnosis of AAT-deficiency.
- A recent study suggested that there are at least 116 million carriers among all racial groups, worldwide.

**COPD Treatment**

- Emphysema and chronic bronchitis often co-exist in COPD. The quality of life for a person suffering from COPD diminishes as the disease progresses. At the onset, there is minimal shortness of breath. People with COPD may eventually require supplemental oxygen and may have to rely on mechanical respiratory assistance.
An ALA survey revealed that half of all COPD patients (51%) say their condition limits their ability to work. It also limits them in normal physical exertion (70%), household chores (56%), social activities (53%), sleeping (50%) and family activities (46%).

None of the existing medications for COPD has been shown to modify the long-term decline in lung function that is the hallmark of this disease. Therefore, the goal of pharmacotherapy for COPD is to provide relief of symptoms and prevent complications and/or progression of the disease with a minimum of side effects.

Bronchodilator medications (prescription drugs that relax and open air passages in the lungs) are central to the symptomatic management of COPD. They can be inhaled as aerosol sprays or taken orally.

Additional treatment includes antibiotics, oxygen therapy, and systemic glucocorticosteroids. The efficacy of inhaled glucocorticosteroids in modifying long-term decline in lung function continues to be under study, however short-term benefit has been demonstrated. Corticosteroids have been shown to decrease cardiovascular and lung cancer deaths and the frequency of exacerbations. Chronic treatment with systemic steroids involves the risk of serious side effects; therefore these are used mostly for acute exacerbations.

Pneumonia and influenza vaccines should be given to COPD patients. Those with COPD should also live a healthy lifestyle by exercising, avoiding cigarette smoke and other air pollutants, and eating well.

Pulmonary rehabilitation is a preventive health-care program provided by a team of health professionals to help people cope physically, psychologically, and socially with COPD.

Lung transplantation is being performed in increasing numbers and may be an option for people who suffer from severe emphysema. Additionally, lung volume reduction surgery (LVRS) has shown promise and is being performed with increasing frequency. However, a recent study found that emphysema patients who have severe lung obstruction with either limited ability to exchange gas when breathing or damage that is evenly distributed throughout their lungs are at high risk of death from the procedure.

Treatments for AAT deficiency emphysema including AAT replacement therapy (a lifelong process) and gene therapy are currently being evaluated. It is hoped that a clinical trial on gene therapy will take place within the decade.

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![Emphysema](image)

**Emphysema**

![Chronic Bronchitis](image)

**Chronic Bronchitis**
Restrictive Disorders

*Diseases of the Lung Parenchyma (Intrinsic)*

**Interstitial Lung Disease**

Interstitial Lung Disease (ILD) is a general term that includes a variety of chronic lung disorders. When a person has ILD, the lung is affected in three ways. First, the lung tissue is damaged in some known or unknown way. Second, the walls of the air sacs in the lung become inflamed. Finally, scarring (or fibrosis) begins in the interstitium (or tissue between the air sacs), and the lung becomes stiff.

Breathlessness during exercise can be one of the first symptoms of these diseases. A dry cough also may be present. These are common symptoms that many people ignore. Someone with these symptoms may wait until they feel quite ill before going to the doctor.

People with different types of ILD may have the same kind of symptoms but their symptoms may vary in severity. Their chest X-rays may look alike. Further testing is usually recommended to identify the specific type of ILD a person has. Some ILDs have known causes and some (idiopathic) have unknown causes.

**WHY IS IT CALLED INTERSTITIAL LUNG DISEASE?**

The tissue between the air sacs of the lungs is called the interstitium. Interstitial lung disease is named after this tissue because this is the tissue affected by fibrosis (scarring). Interstitial lung disease is sometimes also known as "interstitial pulmonary fibrosis." The terms interstitial lung disease, pulmonary fibrosis and interstitial pulmonary fibrosis are often used to describe the same condition.

**INTERSTITIAL LUNG DISEASES ARE PUZZLING**

The course of these diseases is unpredictable. If they progress, the lung tissue thickens and becomes stiff. The work of breathing then becomes more difficult and demanding. Some of the diseases improve with medication if treated when inflammation occurs. Some people may need oxygen therapy as part of their treatment.

The diseases may run a gradual course or a rapid course. People with ILD may notice variations in symptoms -- from very mild to moderate to very severe. Their condition may remain the same for long periods of time or it may change quickly.

**COMMON LINK IN INTERSTITIAL LUNG DISEASE**

While the progress and symptoms of these diseases may vary from person to person, there is one common link between the many forms of ILD. They all begin with an inflammation. The inflammation may affect different parts of the lung, as explained below:

1. The walls of the bronchioles (small airways). When inflammation involves the bronchioles, it is called bronchiolitis.
The diagram below shows the changes that happen in lung tissue with interstitial lung disease.

2. The walls and air spaces of the alveoli (air sacs). When inflammation involves the alveoli, it is called alveolitis.

3. The small blood vessels (capillaries) of the lungs. When inflammation involves the small blood vessels, it is called vasculitis.

Inflammation of these parts of the lung may heal or may lead to permanent scarring of the lung tissue. When scarring of the lung tissue takes place, the condition is called pulmonary fibrosis.

Fibrosis, or scarring of the lung tissue, results in permanent loss of that tissue’s ability to transport oxygen. This is because the air sacs, as well as the lung tissue between and surrounding the air sacs, and the lung capillaries, are destroyed by the formation of scar tissue. If this happens, your doctor may prescribe oxygen to help you breathe easier.

**CAUSES OF PULMONARY FIBROSIS**

They include:

1. **Occupational and environmental exposures.** Many jobs - particularly those that involve mining or that expose workers to asbestos or metal dusts -- can cause pulmonary fibrosis. Workers doing these kinds of jobs may inhale small particles (like silica dusts or asbestos fibers) that can damage the lungs, especially the small airways and air sacs, and cause scarring (fibrosis).

   Agricultural workers also can be affected. Some organic substances, such as moldy hay, cause an allergic reaction in the lung. This reaction is called Farmer’s Lung and can cause pulmonary fibrosis. Other fumes found on farms are directly toxic to the lungs.

2. **Sarcoidosis.** A disease characterized by the formation of granulomas (areas of inflammatory cells), which can attack any area of the body but most frequently affects the lungs.

3. **Drugs.** Certain medicines may have the undesirable side effect of causing pulmonary fibrosis. Check with your doctor about the medicines you are taking and ask about any possible side effects.
4. **Radiation.** (treatment for breast cancer)

4. **Connective tissue or collagen diseases** such as rheumatoid arthritis and systemic sclerosis.

5. **Genetic/familial.** This is not as common as the other causes listed.

### IDIOPATHIC PULMONARY FIBROSIS (IPF)

When all known causes of interstitial lung disease have been ruled out, the condition is called "idiopathic" (of unknown origin) pulmonary fibrosis (IPF).

There are several theories as to what may cause IPF; including viral illness and allergic or environmental exposure (including tobacco smoke). These theories are still being researched. Bacteria and other microorganisms are not thought to be the cause of IPF.

There is also a familial form of the disease, known as familial idiopathic pulmonary fibrosis. Additional research is being done to determine whether there is a genetic tendency to develop the disease, as well as to determine other causes of IPF.

### SYMPTOMS OF IPF

Shortness of breath is the main symptom of idiopathic pulmonary fibrosis. Since this is a symptom of many types of lung disease, making the correct diagnosis may be difficult. The shortness of breath may first appear during exercise. The condition then may progress to the point where any exertion is impossible. If the disease progresses, the person with IPF eventually may be short of breath even at rest.

Other symptoms may include a dry cough (without sputum). When the disease is severe and prolonged, heart failure with swelling of the legs may occur.

### HOW IS IPF DIAGNOSED?

A very careful patient history is an important tool for diagnosis. The history will include environmental and occupational factors, hobbies, legal and illegal drug use, arthritis, and risk factors for diseases that affect the immune system. A physical examination, chest X-ray, pulmonary function tests, and blood tests are important. These tests will help your doctor rule out other lung diseases and determine the extent of disease.

### TREATMENT FOR IPF

Corticosteroids may be administered to treat the inflammation present in some people with IPF. The success of this treatment for many forms of pulmonary fibrosis is variable and is still being researched. Other drugs are occasionally added when it is clear that the steroids are not effective in

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**Diagnostic Tests That May Be Used to Identify Pulmonary Fibrosis or Interstitial Lung Disease**

1. Blood Tests
2. Pulmonary Function Tests
3. Chest X-ray
4. CT Scan
5. Bronchoscopy
6. Bronchoalveolar Lavage
7. Lung Biopsy
8. CT Scan
reversing the disease.

Oxygen therapy may be prescribed for some people with IPF. The need for oxygen will be determined by the doctor and may depend on the severity of disease, as well as your activity level.

Lung transplantation may offer hope for selected people with severe IPF and other lung diseases. Speak with your doctor about eligibility criteria for lung transplant recipients.

Rehabilitation and education programs may help some people with IPF. Local support groups have been of benefit for people with IPF and their family members and friends.

**Occupational lung disease** is the number one work-related illness in the United States based on the frequency, severity, and preventability of diseases. These illnesses are usually caused by extended exposure to irritating or toxic substances that may cause acute or chronic respiratory ailments, although severe single exposures can cause chronic lung disease as well. Smoking can act synergistically to increase the severity of these diseases.

Occupational lung diseases are often not curable, but are always preventable. Improving ventilation, wearing protective equipment, changing work procedures, and educating workers are the key factors for prevention.

Several of the most common occupational lung diseases:

- **Occupational lung cancer**: These occupational exposures account for about 10.3 percent of cancer of the lung, trachea and bronchus, the most frequent occupational cancer

- **Occupational asthma**: the most common form of occupational lung disease. An estimated 15 to 23 percent of new onset adult asthma cases in the U.S. are due to occupational exposures. These exposures within the workplace can also aggravate pre-existing asthma.

- **Asbestosis**: a progressive disease involving scarring of lung tissue as a result of exposure to the microscopic fibers of asbestos. An estimated 1.3 million employees in construction and industry face significant asbestos exposure on the job.

- **Mesothelioma**: an otherwise rare cancer of the chest lining caused by asbestos exposure.

- **Byssinosis (brown lung disease)**: a chronic condition involving obstruction of the small airways, severely impairing lung function. It is caused by dusts from hemp, flax, and cotton processing.

- **Coal workers' pneumoconiosis (black lung disease)**: caused by the inhalation of coal dust that becomes imbedded in the lungs, causing them to harden, making breathing very difficult. An estimated 2.8 percent of coal miners are affected; about 0.2 percent have scarring on the lungs, the most severe form of the disease. Each year, close to 400 people die from black lung disease.

- **Silicosis**: results from exposure to free crystalline silica in mines, foundries, blasting operations, stone, clay and glass manufacturing that cause scar tissue to form in the lungs. Silicosis substantially raises the risk of tuberculosis. Each year 200 people die with silicosis listed as an underlying or nonunderlying cause of death on their death certificates, a rate that has been stable since the early 1990's. About 1 million workers are believed to have been exposed to silica dust.
HYPERSENSITIVITY PNEUMONITIS

Hypersensitivity pneumonitis (also called allergic alveolitis) is a disease in which the air sacs (alveoli) of the lungs become inflamed when certain dusts are inhaled to which the person is sensitized or allergic.

These dusts contain organic substances, such as fungus spores from moldy hay or the droppings of birds.

When a person inhales such dusts the first time, no problem is noticed. But, after repeated exposure to the dust some people may develop symptoms. The tiny air sacs in the lung known as alveoli become inflamed, their walls fill with white blood cells, and sometimes the sacs fill with fluid. If the disease recurs as a result of continued or repeated exposure to the offending dusts, parts of the lung may develop fibrous scar tissue and can no longer function normally in breathing.

WHAT ARE THE SYMPTOMS?

The symptoms of an acute attack are similar to those of the flu and appear some 4-6 hours after the person breathes the offending dust. These symptoms include chills, fever, dry cough, shortness of breath, a tight feeling in the chest, and tiredness. The symptoms may persist for as little as 12 hours or as long as 10 days. Between attacks the person may have no symptoms and feel quite normal.

After repeated exposure to the dust, chronic cough may develop with excessive sputum production containing pus, and eventually there may be chronic shortness of breath. The person may also show loss of appetite and weight loss.

WHO GETS HYPERSENSITIVITY PNEUMONITIS?

Hypersensitivity pneumonitis occurs most often in people exposed to certain organic dusts in their daily work or living, especially dusts which contain fungus spores from mold.

Only a small proportion of people who are exposed, perhaps 5 to 20 percent, develop the disease. When there is frequent exposure to high concentrations of the offending substances, the likelihood of developing hypersensitivity pneumonitis is greatest.

WHAT CAUSES HYPERSENSITIVITY PNEUMONITIS?

The disease is caused by organic material that is inhaled as a fine dust. Sensitization (allergy) develops over a period of several months to a number of years.

Farmer’s lung is the best-known example. It is caused by an organism that grows on hay, straw, or grains, and by other organic materials found on farms. Also known to cause hypersensitivity pneumonitis are dusts from moldy sugar cane and barley, maple bark, cork, animal hair, bird feathers and droppings, mushroom compost, coffee beans, and paprika. Often the disease is named for the occupations: for example mushroom-worker’s lung or paprika-splitter’s lung. The organism of farmer’s lung also grows in the water of both commercial and home humidification systems, particularly if they are not adequately maintained.

HOW IS HYPERSENSITIVITY PNEUMONITIS DIAGNOSED?

A careful, detailed history is necessary so that the occurrence of symptoms can be related to environmental exposure. Laboratory tests of the patient’s blood and identification of specific molds from material at the workplace may help. Chest X-ray films and lung function tests may
show effects of the disease during acute episodes or in the chronic stage of the disease – but they may be normal between episodes when the patient is without symptoms. Also chest X-ray and pulmonary function test results are not specific for hypersensitivity pneumonitis, and positive results may be due to other causes. Inhalation of the materials to which the person is sensitive (challenge test) may be necessary to confirm the diagnosis. The challenge test should be done only under the expert medical supervision of a specialist. Sometimes, a lung biopsy is needed to distinguish it from other lung diseases.

**IS HYPERSENSITIVITY PNEUMONITIS PREVENTABLE?**

Properly dried and stored farm products are unlikely to cause hypersensitivity pneumonitis because the particular organism causing farmer’s lung grows only in moist conditions. Other approaches to prevention are to reduce exposure by proper ventilation and the use of respiratory protection (masks).

If a worker begins to have symptoms of hypersensitivity pneumonitis due to occupational exposure, the process can be stopped if it is identified early enough. Permanent lung damage is prevented by eliminating the exposure. If control measures do not work or are not feasible, it may be necessary to change jobs, even though doing so may be difficult for many reasons. Therefore the medical necessity for a change should be determined only after careful evaluation to establish the diagnosis and specific cause.

**WHAT CAN BE DONE TO TREAT HYPERSENSITIVITY PNEUMONITIS?**

Avoidance of the offending dust is the single most important measure because in the early stages the disease is completely reversible.

Most drugs are of limited value in treatment; antihistamines and bronchodilators are ineffective. Steroids can relieve the symptoms of acute attacks but do not cure the disease. Recovery from acute attacks may take as long as three weeks. Residual lung damage in the form of pulmonary fibrosis is permanent and may occur even after symptoms have disappeared. Avoiding exposure in the first place eliminates any possibility of later permanent damage.

**SARCOIDOSIS**

Sarcoidosis is a disease due to inflammation.

The disease can attack any organ of the body in any location. The disease is characterized by the presence of granulomas, small areas of inflamed cells. They can be either inside the body or on the body’s exterior, appearing as sores on the face or shins. But sarcoidosis is most frequently found in the lungs.

Pulmonary sarcoidosis can cause loss of lung volume (the amount of air the lungs can hold) and abnormal lung stiffness.

Granulomas can appear on the walls of the alveoli (small air sacs in the lungs) or on the walls of the bronchioles (breathing tubes in the lungs). They also appear in the lymph nodes in the chest, causing them to enlarge.

**WHAT CAUSES SARCOIDOSIS?**

No one yet knows what causes sarcoidosis.
It is thought by most scientists to be a disorder of the immune system, where the body’s natural defense system malfunctions.

**WHAT ARE THE SYMPTOMS OF SARCOIDOSIS?**

In pulmonary sarcoidosis, patients may have a dry cough (without sputum), shortness of breath, or mild chest pain.

In those cases where symptoms do appear outside the lung, they can include a scaly rash, red bumps on the legs, fever, soreness of the eyes, and pain and swelling of the ankles.

There can also be more general symptoms like fatigue, weakness, fever, and weight loss. These symptoms are common in many other lung diseases, so diagnosis may be difficult.

**HOW IS SARCOIDOSIS DIAGNOSED?**

Any of the symptoms listed in the previous section may lead a physician to consider sarcoidosis. Sarcoidosis is initially diagnosed based on a physical examination, laboratory tests, pulmonary function studies, and a chest X-ray. When enlargement of lymph glands in the center of the lungs is seen on X-ray, sarcoidosis may be suspected.

To confirm the diagnosis, a biopsy is usually performed on any of the affected organs or from material in a granuloma on the skin.

**HOW SERIOUS IS SARCOIDOSIS?**

In over half the cases, sarcoidosis appears briefly and heals naturally. Sometimes the patient doesn't even know or do anything about it. From 20 to 30 percent of pulmonary sarcoidosis patients are left with permanent lung damage. And for a small percentage of patients, their sarcoidosis can become chronic, lasting for many years.

**WHO GETS SARCOIDOSIS?**

Sarcoidosis is found throughout the world among almost all races and ages and in both sexes. However, it is most common among African Americans and northern European whites.

Sarcoidosis is mainly a disease of young adults -- patients between the ages of 20 and 40 -- although a few persons past 60 have been known to contract it.

In the United States, a higher percentage of African Americans than whites has sarcoidosis, and the disease is usually more serious in them. The prevalence of sarcoidosis is eight times greater in African Americans than in whites in the U.S.

**WHAT BODY SITES DOES SARCOIDOSIS ATTACK?**

Ninety percent of the cases of sarcoidosis are found in the lungs.

Other commonly affected sites are:

- Skin
- Liver
- Lymph glands
- Spleen
- Eyes
- Nervous system, including the brain
- Musculoskeletal system (the muscles and bones in the body)
- Heart
- Kidneys

**WHAT IS THE COMMON COURSE OF SARCOIDOSIS?**

In most cases of sarcoidosis that have no symptoms, the disease "burns itself out," disappearing with little or no notice to the patient or physician.

If pulmonary sarcoidosis is serious, it can develop into pulmonary fibrosis (the abnormal formation of fiber-like scar tissue in the lung). This actually distorts the structure of the lungs and can interfere with breathing, especially the ability to exchange oxygen in the lungs.

**WHAT IS THE TREATMENT FOR SARCOIDOSIS?**

In a majority of patients, the disease spontaneously disappears, and no treatment is necessary.

When therapy is recommended, the main goal is to keep the lungs and other affected body organs working, and to relieve symptoms. Drugs called corticosteroids are the most common treatment used in fighting sarcoidosis.
Extrinsic Disorders

Myasthenia gravis (Neuromuscular disorder)

Definition

Myasthenia gravis is an autoimmune disease that causes muscle weakness.

Description

Myasthenia gravis (MG) affects the neuromuscular junction, interrupting the communication between nerve and muscle, and thereby causing weakness. A person with MG may have difficulty moving their eyes, walking, speaking clearly, swallowing, and even breathing, depending on the severity and distribution of weakness. Increased weakness with exertion, and improvement with rest, is a characteristic feature of MG.

About 30,000 people in the United States are affected by MG. It can occur at any age, but is most common in women who are in their late teens and early twenties, and in men in their sixties and seventies.

Causes and symptoms

Myasthenia gravis is an autoimmune disease, meaning it is caused by the body's own immune system. In MG, the immune system attacks a receptor on the surface of muscle cells. This prevents the muscle from receiving the nerve impulses that normally make it respond. MG affects "voluntary" muscles, which are those muscles under conscious control responsible for movement. It does not affect heart muscle or the "smooth" muscle found in the digestive system and other internal organs.

A muscle is stimulated to contract when the nerve cell controlling it releases acetylcholine molecules onto its surface. The acetylcholine lands on a muscle protein called the acetylcholine receptor. This leads to rapid chemical changes in the muscle which cause it to contract. Acetylcholine is then broken down by acetylcholinesterase enzyme, to prevent further stimulation.

In MG, immune cells create antibodies against the acetylcholine receptor. Antibodies are proteins normally involved in fighting infection. When these antibodies attach to the receptor, they prevent it from receiving acetylcholine, decreasing the ability of the muscle to respond to stimulation.

The earliest symptoms of MG often result from weakness of the extraocular muscles, which control eye movements. Symptoms involving the eye (ocular symptoms) include double vision (diplopia), especially when not gazing straight ahead, and difficulty raising the eyelids (ptosis). A person with ptosis may need to tilt their head back to see. Eye-related symptoms remain the only symptoms for about 15% of MG patients. Another common early symptom is difficulty chewing and swallowing, due to weakness in the bulbar muscles, which are in the mouth and throat. Choking becomes more likely, especially with food that requires extensive chewing.

Weakness usually becomes more widespread within several months of the first symptoms, reaching their maximum within a year in two-thirds of patients. Weakness may involve muscles of the arms, legs, neck, trunk, and face, and affect the ability to lift objects, walk, hold the head up, and speak.
Symptoms of MG become worse upon exertion, and better with rest. Heat, including heat from the sun, hot showers, and hot drinks, may increase weakness. Infection and stress may worsen symptoms. Symptoms may vary from day to day and month to month, with intervals of no weakness interspersed with a progressive decline in strength.

"Myasthenic crisis" may occur, in which the breathing muscles become too weak to provide adequate respiration. Symptoms include weak and shallow breathing, shortness of breath, pale or bluish skin color, and a racing heart. Myasthenic crisis is an emergency condition requiring immediate treatment. In patients treated with anticholinesterase agents, myasthenic crisis must be differentiated from cholinergic crisis related to overmedication.

**Diagnosis**

Myasthenia gravis is often diagnosed accurately by a careful medical history and a neuromuscular exam, but several tests are used to confirm the diagnosis. Other conditions causing worsening of bulbar and skeletal muscles must be considered, including drug-induced myasthenia, thyroid disease, Lambert-Eaton myasthenic syndrome, botulism, and inherited muscular dystrophies.

MG causes characteristic changes in the electrical responses of muscles that may be observed with an electromyogram, which measures muscular response to electrical stimulation.

Blood tests may confirm the presence of the antibody to the acetylcholine receptor, though up to a quarter of MG patients will not have detectable levels. A chest x ray or chest computed tomography scan (CT scan) may be performed to look for thymoma.

**Treatment**

While there is no cure for myasthenia gravis, there are a number of treatments that effectively control symptoms in most people.

Edrophonium (Tensilon) blocks the action of acetylcholinesterase, prolonging the effect of acetylcholine and increasing strength. An injection of edrophonium rapidly leads to a marked improvement in most people with MG. An alternate drug, neostigmine, may also be used.

Pyridostigmine (Mestinon) is usually the first drug tried. Like edrophonium, pyridostigmine blocks acetylcholinesterase. It is longer-acting, taken by mouth, and well-tolerated. Loss of responsiveness and disease progression combine to eventually make pyridostigmine ineffective in tolerable doses in many patients.

Immune-suppressing drugs are used to treat MG if response to pyridostigmine and thymectomy are not adequate. Drugs include corticosteroids such as prednisone, and the non-steroids azathioprine (Imuran) and cyclosporine (Sandimmune).

Plasma exchange may be performed to treat myasthenic crisis or to improve very weak patients before thymectomy. In this procedure, blood plasma is removed and replaced with purified plasma free of autoantibodies. It can produce a temporary improvement in symptoms, but is too expensive for long-term treatment. Another blood treatment, intravenous immunoglobulin therapy, is also used for myasthenic crisis. In this procedure, large quantities of purified immune proteins (immunoglobulins) are injected. For unknown reasons, this leads to symptomatic improvement in up to 85% of patients. It is also too expensive for long-term treatment.
**Prognosis**

Most people with MG can be treated successfully enough to prevent their condition from becoming debilitating. In some cases, however, symptoms may worsen even with vigorous treatment, leading to generalized weakness and disability. MG rarely causes early death except from myasthenic crisis.

**Prevention**

- There is no known way to prevent myasthenia gravis.

**Pleural effusion (Pleural Disease)**

**Definition**

Pleural effusion occurs when too much fluid collects in the pleural space (the space between the two layers of the pleura). It is commonly known as "water on the lungs." It is characterized by shortness of breath, chest pain, and cough.

**Description**

There are two thin membranes in the chest, one (the visceral pleura) lining the lungs, and the other (the parietal pleura) covering the inside of the chest wall. Normally, small blood vessels in the pleural linings produce a small amount of fluid that lubricates the opposed pleural membranes so that they can glide smoothly against one another during breathing movements. Any extra fluid is taken up by blood and lymph vessels, maintaining a balance. When either too much fluid forms or something prevents its removal, the result is an excess of pleural fluid—an effusion. The most common causes are disease of the heart or lungs, and inflammation or infection of the pleura.

Pleural effusion itself is not a disease as much as a result of many different diseases. For this reason, there is no "typical" patient in terms of age, sex, or other characteristics. Instead, anyone who develops one of the many conditions that can produce an effusion may be affected.

There are two types of pleural effusion: the transudate and the exudate. This is a very important point because the two types of fluid are very different, and which type is present points to what sort of disease is likely to have produced the effusion. It also can suggest the best approach to treatment.
**Transudates**

A transudate is a clear fluid, similar to blood serum, that forms not because the pleural surfaces themselves are diseased, but because the forces that normally produce and remove pleural fluid at the same rate are out of balance. When the heart fails, pressure in the small blood vessels that remove pleural fluid is increased and fluid “backs up” in the pleural space, forming an effusion. Or, if too little protein is present in the blood, the vessels are less able to hold the fluid part of blood within them and it leaks out into the pleural space. This can result from disease of the liver or kidneys, or from malnutrition.

**Exudates**

An exudate—which often is a cloudy fluid, containing cells and much protein—results from disease of the pleura itself. The causes are many and varied. Among the most common are infections such as bacterial pneumonia and tuberculosis; blood clots in the lungs; and connective tissue diseases, such as rheumatoid arthritis. Cancer and disease in organs such as the pancreas also may give rise to an exudative pleural effusion.

**Special types of pleural effusion**

Some of the pleural disorders that produce an exudate also cause bleeding into the pleural space. If the effusion contains half or more of the number of red blood cells present in the blood itself, it is called hemothorax. When a pleural effusion has a milky appearance and contains a large amount of fat, it is called chylothorax. Lymph fluid that drains from tissues throughout the body into small lymph vessels finally collects in a large duct (the thoracic duct) running through the chest to empty into a major vein. When this fluid, or chyle, leaks out of the duct into the pleural space, chylothorax is the result. Cancer in the chest is a common cause.

**Causes and symptoms**

Causes of transudative pleural effusion

Among the most important specific causes of a transudative pleural effusion are:

* Congestive heart failure. This causes pleural effusions in about 40% of patients and is often present on both sides of the chest. Heart failure is the most common cause of bilateral (two-sided) effusion. When only one side is affected it usually is the right (because patients usually lie on their right side).

* Pericarditis. This is an inflammation of the pericardium, the membrane covering the heart.

* Too much fluid in the body tissues, which spills over into the pleural space. This is seen in some forms of kidney disease; when patients have bowel disease and absorb too little of what they eat; and when an excessive amount of fluid is given intravenously.

* Liver disease. About 5% of patients with a chronic scarring disease of the liver called cirrhosis develop pleural effusion.

Causes of exudative pleural effusions

A wide range of conditions may be the cause of an exudative pleural effusion:

* Pleural tumors account for up to 40% of one-sided pleural effusions. They may arise in the pleura itself (mesothelioma), or from other sites, notably the lung.

* Tuberculosis in the lungs may produce a long-lasting exudative pleural effusion.
* Pneumonia affects about three million persons each year, and four of every ten patients will develop pleural effusion. If effective treatment is not provided, an extensive effusion can form that is very difficult to treat.

* Patients with any of a wide range of infections by a virus, fungus, or parasite that involve the lungs may have pleural effusion.

* Up to half of all patients who develop blood clots in their lungs (pulmonary embolism) will have pleural effusion, and this sometimes is the only sign of embolism.

* Connective tissue diseases, including rheumatoid arthritis, lupus may be complicated by pleural effusion.

* Patients with disease of the liver or pancreas may have an exudative effusion, and the same is true for any patient who undergoes extensive abdominal surgery. About 30% of patients who undergo heart surgery will develop an effusion.

* Injury to the chest may produce pleural effusion in the form of either hemothorax or chylothorax.

**Symptoms**

The key symptom of a pleural effusion is shortness of breath. Fluid filling the pleural space makes it hard for the lungs to fully expand, causing the patient to take many breaths so as to get enough oxygen. When the parietal pleura is irritated, the patient may have mild pain that quickly passes or, sometimes, a sharp, stabbing pleuritic type of pain. Some patients will have a dry cough. Occasionally a patient will have no symptoms at all. This is more likely when the effusion results from recent abdominal surgery, cancer, or tuberculosis. Tapping on the chest will show that the usual crisp sounds have become dull, and on listening with a stethoscope the normal breath sounds are muted. If the pleura is inflamed, there may be a scratchy sound called a "pleural friction rub."

**Diagnosis**

When pleural effusion is suspected, the best way to confirm it is to take chest x rays, both straight-on and from the side. The fluid itself can be seen at the bottom of the lung or lungs, hiding the normal lung structure. If heart failure is present, the x-ray shadow of the heart will be enlarged. An ultrasound scan may disclose a small effusion that caused no abnormal findings during chest examination. A computed tomography scan is very helpful if the lungs themselves are diseased.

In order to learn what has caused the effusion, a needle or catheter is often used to obtain a fluid sample, which is examined for cells and its chemical make-up. This procedure, called a thoracentesis, is the way to determine whether an effusion is a transudate or exudate, giving a clue as to the underlying cause. In some cases--for instance when cancer or bacterial infection is present--the specific cause can be determined and the correct treatment planned. Culturing a fluid sample can identify the bacteria that cause tuberculosis or other forms of pleural infection.

**Treatment**

The best way to clear up a pleural effusion is to direct treatment at what is causing it, rather than treating the effusion itself. If heart failure is reversed or a lung infection is cured by antibiotics, the effusion will usually resolve. However, if the cause is not known, even after extensive tests, or no effective treatment is at hand, the fluid can be drained away by placing a large-bore needle or catheter into the pleural space, just as in diagnostic thoracentesis. If necessary, this can be
repeated as often as is needed to control the amount of fluid in the pleural space. If large effusions continue to recur, a drug or material that irritates the pleural membranes can be injected to deliberately inflame them and cause them to adhere close together—a process called sclerosis.

**Pneumothorax (Pleural Disease)**

* Primary Spontaneous Pneumothorax
* Secondary Spontaneous Pneumothorax

Spontaneous pneumothorax is a sudden collection of air or gas in the chest that causes the lung to collapse in the absence of a traumatic injury to the chest or lung. Usually, a distinction is made between a spontaneous (primary) pneumothorax, and secondary (complicated) pneumothorax.

**SPONTANEOUS (PRIMARY) PNEUMOTHORAX**

Spontaneous pneumothorax occurs in individuals with no known lung disease. It affects close to 9,000 persons in the United States each year—most often among tall, thin men between 20 and 40 years old. The cause of this type of pneumothorax is the rupture of a bleb or cyst in the lung.

Symptoms include:

* Chest pain on affected side
* Dyspnea (shortness of breath)
* Cough
* Abnormal breathing movement
* Rapid respiratory rate

Spontaneous pneumothorax is diagnosed by chest radiographs.

The way the condition is treated is dependant on its size and course. The objective of treatment is to remove the air from the pleural space, allowing the lung to reexpand. A small pneumothorax will resolve on its own in 1 to 2 weeks. Larger pneumothoraxes require either needle aspiration or a chest tube. Hospitalization is required for chest tube management as the reexpansion of the lung may take several days with the chest tube left in place. Surgery may be performed for a repeated episode to prevent recurrence.

**SECONDARY PNEUMOTHORAX**

Secondary pneumothorax occurs in the setting of known lung disease, most often chronic obstructive pulmonary disease (COPD). Other lung diseases commonly associated with spontaneous pneumothorax include tuberculosis, pneumonia, asthma, cystic fibrosis, lung cancer, and certain forms of interstitial lung disease. This condition is generally severe and often life threatening.

Symptoms and diagnostic procedures of secondary pneumothorax are identical to that of primary spontaneous pneumothorax.

The therapeutic options for this condition are also the same as those for primary spontaneous pneumothorax, but the circumstances are much more urgent. A small pneumothorax can be life threatening and virtually all patients are treated with chest tubes. Sudden death may occur before chest tubes can be placed and respiratory failure can occur within hours after the tubes are inserted. The mortality rate associated with secondary pneumothorax is high (15%).
The recurrence rate for both primary and secondary spontaneous pneumothorax is about 40% and occurs in intervals of 1.5 to 2 years. Patients suffering from this condition should be advised to discontinue smoking and avoid high altitudes, scuba diving, or flying in unpressurized aircrafts to prevent the recurrence of pneumothorax.

Pickwickian syndrome (Abdominal Restrictive)

Definition
A group of symptoms that generally accompany massive obesity.

Description
Pickwickian syndrome is a complex of symptoms that primarily affect patients with extreme obesity. The major health problem that occurs in patients with this disease is sleep apnea. This is caused in part by the excess amounts of fatty tissue surrounding the chest muscles. This excess fat places a strain on the heart, lungs, and diaphragm of the patient, making it difficult to breathe.

Causes and symptoms
The major cause of Pickwickian syndrome is extreme obesity. This obesity places an excessive load on the pulmonary system. The role of genetics is also being studied. Symptoms of Pickwickian syndrome include excessive daytime sleepiness, shortness of breath due to elevated blood carbon dioxide pressure, disturbed sleep at night, and flushed face. The skin can also have a bluish tint, and the patient may have high blood pressure, an enlarged liver, and an abnormally high red blood cell count.

Diagnosis
Some tests that can be used to diagnose this condition include echocardiography to determine heart enlargement or pulmonary hypertension. Giving the patient multiple sleep latency tests can help give an objective measurement of daytime sleepiness. Magnetic resonance imaging (MRI), computed tomography (CT) scans, or fiberoptic evaluation of the upper airway may also be used.
Treatment

The primary treatment for Pickwickian syndrome is focused on weight loss and increased physical activity. Also, medroxyprogesterone may help improve the condition.

Prognosis

Pickwickian syndrome is entirely reversible if it is diagnosed and treated properly. If the problem goes undiagnosed, the outcome can be fatal.

Prevention

Prevention of Pickwickian syndrome can be achieved by maintaining a healthy body weight and getting the proper amount of exercise. For prevention of the sleep apnea that generally accompanies Pickwickian syndrome, there are several possible treatments. If the sleep apnea is only present when the patient is flat on their back, a tennis ball can be sewn into the sleep clothes to remind the patient not to sleep on their back. For more severe cases of sleep apnea, a tonsillectomy or the use of dental appliances may be recommended.

SLEEP APNEA

This condition, sometimes referred to as Sleep-Disordered Breathing (SDB), involves repeated arousals from sleep--not necessarily to full awakening because breathing has ceased. In the most important form (obstructive sleep apnea) it is because the upper airway for breathing has momentarily been obstructed.

Sleep Apnea may or may not be evident, to the patient or to others. It may be manifested by marked daytime drowsiness--or noted by a spouse who is frequently awakened by the snoring, which almost always accompanies obstructive sleep apnea.

Sleep Apnea is considered a health problem because so many individuals lose so much sleep due to this condition that their lack of alertness poses a serious hazard. It is also associated with hypertension and heart disease.

Sleep Apnea may also cause impaired mental functioning, delayed reaction times, and difficulty maintaining vigilance and concentration. In its severe form it is accompanied by high pressures in the lung arteries leading to heart failure.

PREVENTION: RECOGNIZING AND REMEDYING A PROBLEM

The focus is on managing risks associated with the condition and on effective treatment. Try this advice:

Talk to your doctor. The first step to doing something about SDB is admitting that it's a problem and discussing daytime sleepiness or excessive snoring with your physician so that he can help you deal with the condition.

Slimming down may help. Sleep Apnea is more common among people who are moderately overweight to obese, so a diet and exercise program may eliminate or dramatically reduce occurrences.

Get technical help. If necessary, your doctor will advise a visit to a sleep lab to evaluate your condition. A technique called Nasal CPAP (continuous positive airway pressure) delivers air
through a mask while a patient sleeps, which is effective in helping people get a good night's rest and avoid daytime accidents. In some cases, anatomical surgery may be recommended. In mild cases, special dental appliances may help.

Children. Sleep apnea occurs in children, usually due to enlarged tonsils and adenoids. A child who is sleepy during the day and snores at night should be medically evaluated.

### Atelectasis

**Definition**

Atelectasis is a collapse of lung tissue affecting part or all of one lung. This condition prevents normal oxygen absorption to healthy tissues.

**Description**

Atelectasis can result from an obstruction (blockage) of the airways that affects tiny air sacs called alveoli. Alveoli are very thin-walled and contain a rich blood supply. They are important for lung function, since their purpose is the exchange of oxygen and carbon dioxide. When the airways are blocked by a mucous "plug," foreign object, or tumor, the alveoli are unable to fill with air and collapse of lung tissue can occur in the affected area. Atelectasis is a potential complication following surgery, especially in individuals who have undergone chest or abdominal operations resulting in associated abdominal or chest pain during breathing. Congenital atelectasis can result from a failure of the lungs to expand at birth. This congenital condition may be localized or may affect all of both lungs.

**Causes and symptoms**

Causes of atelectasis include insufficient attempts at respiration by the newborn, bronchial obstruction, or absence of surfactant (a substance secreted by alveoli that maintains the stability of lung tissue by reducing the surface tension of fluids that coat the lung). This lack of surfactant reduces the surface area available for effective gas exchange causing it to collapse if severe. Pressure on the lung from fluid or air can cause atelectasis as well as obstruction of lung air passages by thick mucus resulting from various infections and lung diseases. Tumors and inhaled objects can also cause obstruction of the airway, leading to atelectasis.

Anyone undergoing chest or abdominal surgery using general anesthesia is at risk to develop atelectasis, since breathing is often shallow after surgery to avoid pain from the surgical incision. Any significant decrease in airflow to the alveoli contributes to pooling of secretions, which in turn can cause infection. Chest injuries causing shallow breathing, including fractured ribs, can cause atelectasis. Common symptoms of atelectasis include shortness of breath and decreased chest wall expansion. If atelectasis only affects a small area of the lung, symptoms are usually minimal. If the condition affects a large area of the lung and develops quickly, the individual may turn blue (cyanotic) or pale, have extreme shortness of breath, and feel a stabbing pain on the affected side. Fever and increased heart rate may be present if infection accompanies atelectasis.

**Diagnosis**
To diagnose atelectasis, a doctor starts by recording the patient's symptoms and performing a thorough physical examination. When the doctor listens to the lungs through a stethoscope (auscultation), diminished or bronchial breath sounds may be heard. By tapping on the chest (percussion) while listening through the stethoscope, the doctor can often tell if the lung is collapsed. A chest x-ray that shows an airless area in the lung confirms the diagnosis of atelectasis. If an obstruction of the airways is suspected, a computed tomography scan (CT) or bronchoscopy may be performed to locate the cause of the blockage.

Treatment

If atelectasis is due to obstruction of the airway, the first step in treatment is to remove the cause of the blockage. This may be done by coughing, suctioning, or bronchoscopy. If a tumor is the cause of atelectasis, surgery may be necessary to remove it. Antibiotics are commonly used to fight the infection that often accompanies atelectasis. In cases where recurrent or long-lasting infection is disabling or where significant bleeding occurs, the affected section of the lung may be surgically removed.

Prevention

When recovering from surgery, frequent repositioning in bed along with coughing and deep breathing are important. Coughing and breathing deeply every one to two hours after any surgical operation with general anesthesia is recommended. Breathing exercises and the use of breathing devices, such as an incentive spirometer, may also help prevent atelectasis. Although smokers have a higher risk of developing atelectasis following surgery, stopping smoking six to eight weeks before surgery can help reduce the risk. Increasing fluid intake during respiratory illness or after surgery (by mouth or intravenously) helps lung secretions to remain loose. Increasing humidity may also be beneficial.

Postural drainage techniques can be learned from a respiratory therapist or physical therapist and are a useful tool for anyone affected with a respiratory illness that could cause atelectasis. Because foreign objects blocking the airway can cause atelectasis, it is very important to keep small objects that might be inhaled away from young children.

Vascular Lung Disease

Vascular lung disease refers to conditions which affect the pulmonary capillary vasculature. Alterations in the vasculature manifest in a general inability to exchange blood gases such as oxygen and carbon dioxide, in the vicinity of the vascular damage (other areas of the lung may be unaffected).

Pulmonary embolism

Definition

Pulmonary embolism is an obstruction of a blood vessel in the lungs, usually due to a blood clot, which blocks a coronary artery.

Description

Pulmonary embolism is a fairly common condition that can be fatal. According to the American
Heart Association, an estimated 600,000 Americans develop pulmonary embolism annually; 60,000 die from it. As many as 25,000 Americans are hospitalized each year for pulmonary embolism, which is a relatively common complication in hospitalized patients. Even without warning symptoms, pulmonary embolism can cause sudden death. Treatment is not always successful.

Pulmonary embolism is difficult to diagnose. It occurs when emboli block a pulmonary artery, usually due to a blood clot that breaks off from a large vein and travels to the lungs. More than 90% of cases of pulmonary embolism are complications of deep vein thrombosis, blood clots from the leg or pelvic veins. Emboli can also be comprised of fat, air, or tumor tissue. When emboli block the main pulmonary artery, pulmonary embolism can quickly become fatal.

Causes and symptoms

Pulmonary embolism is caused by emboli that travel through the blood stream to the lungs and block a pulmonary artery. When this occurs, circulation and oxygenation of blood is compromised. The emboli are usually formed from blood clots but are occasionally comprised of air, fat, or tumor tissue.

Risk factors include: prolonged bed rest, surgery, childbirth, heart attack, stroke, congestive heart failure, cancer, obesity, a broken hip or leg, oral contraceptives, sickle cell anemia, congenital coagulation disorders, chest trauma, certain congenital heart defects, and old age.

Common symptoms of pulmonary embolism include:

- labored breathing, sometimes accompanied by chest pain.
- a rapid pulse.
- a cough that produces bloody sputum.
- a low fever.
- fluid build-up in the lungs.

In some cases there are no symptoms.

Diagnosis

Pulmonary embolism can be diagnosed through the patient's history, a physical exam, and diagnostic tests including chest x ray, lung scan, pulmonary angiography, electrocardiography, arterial blood gas measurements, and leg vein ultrasonography or venography.
A chest x-ray can be normal or show fluid or other signs and rule out other diseases. The lung scan shows poor flow of blood in areas beyond blocked arteries. The patient inhales a small amount of radiopharmaceutical and pictures of airflow into the lungs are taken with a gamma camera. Then a different radiopharmaceutical is injected into an arm vein and lung blood flow is scanned. A normal result essentially rules out pulmonary embolism. A lung scan can be performed in a hospital or an outpatient facility and takes about 45 minutes.

Pulmonary angiography is the most reliable test for diagnosing pulmonary embolism but it is not used often, because it carries some risk and is expensive, invasive, and not readily available in many hospitals. Pulmonary angiography is a radiographic test which involves injection of a pharmaceutical “contrast agent” to show up the pulmonary arteries. A cinematic camera records the blood flow through the lungs of the patient, who lies on a table. Pulmonary angiography is usually performed in a hospital's radiology department and takes 30 minutes to one hour.

Treatment

Patients with pulmonary embolism are hospitalized and generally treated with clot-dissolving and clot-preventing drugs. Oxygen therapy is often needed to maintain normal oxygen concentrations. For people who can't take anticoagulants and in some other cases, surgery may be needed to insert a device that filters blood returning to the heart and lungs. The goal of treatment is to maintain the patient's cardiovascular and respiratory functions while the blockage resolves, which takes 10-14 days, and to prevent the formation of other emboli.

Thrombolytic therapy to dissolve blood clots is the aggressive treatment for very severe pulmonary embolism. Streptokinase, urokinase, and recombinant tissue plasminogen activator (TPA) are thrombolytic agents. Heparin is the injectable anticoagulant (clot-preventing) drug of choice for preventing formation of blood clots. Warfarin, an oral anticoagulant, is usually continued when the patient leaves the hospital and doesn't need heparin any longer.

Prognosis

About 10% of patients with pulmonary embolism die suddenly within the first hour of onset of the condition. The outcome for all other patients is generally good; only 3% of patients who are properly diagnosed and treated die. In cases of undiagnosed pulmonary embolism, about 30% of patients die.

Prevention

Pulmonary embolism risk can be reduced in certain patients through judicious use of antithrombotic drugs such as heparin, venous interruption, gradient elastic stockings and/or intermittent pneumatic compression of the legs.

Pulmonary edema

Definition

Pulmonary edema is a condition in which fluid accumulates in the lungs, usually because the heart's left ventricle does not pump adequately.

Description

The build-up of fluid in the spaces outside the blood vessels of the lungs is called pulmonary edema. Pulmonary edema is a common complication of heart disorders, and most cases of the
condition are associated with heart failure. Pulmonary edema can be a chronic condition, or it can develop suddenly and quickly become life threatening. The life-threatening type of pulmonary edema occurs when a large amount of fluid suddenly shifts from the pulmonary blood vessels into the lung, due to lung problems, heart attack, trauma, or toxic chemicals. It can also be the first sign of coronary heart disease.

In heart-related pulmonary edema, the heart's main chamber, the left ventricle, is weakened and does not function properly. The ventricle does not completely eject its contents, causing blood to back up and cardiac output to drop. The body responds by increasing blood pressure and fluid volume to compensate for the reduced cardiac output. This, in turn, increases the force against which the ventricle must expel blood. Blood backs up, forming a pool in the pulmonary blood vessels. Fluid leaks into the spaces between the tissues of the lungs and begins to accumulate. This process makes it more difficult for the lungs to expand. It also impedes the exchange of air and gases between the lungs and blood moving through lung blood vessels.

**Causes and symptoms**

Most cases of pulmonary edema are caused by failure of the heart's main chamber, the left ventricle. It can be brought on by an acute heart attack, severe ischemia, volume overload of the heart's left ventricle, and mitral stenosis. Non-heart-related pulmonary edema is caused by lung problems like pneumonia, an excess of intravenous fluids, some types of kidney disease, bad burns, liver disease, nutritional problems, and Hodgkin's disease. Non-heart-related pulmonary edema can also be caused by other conditions where the lungs do not drain properly, and conditions where the respiratory veins are blocked.

**Early symptoms of pulmonary edema include:**

- shortness of breath upon exertion
- sudden respiratory distress after sleep
- difficulty breathing, except when sitting upright
- coughing

In cases of severe pulmonary edema, these symptoms will worsen to:

- labored and rapid breathing
- frothy, bloody fluid containing pus coughed from the lungs (sputum)
- a fast pulse and possibly serious disturbances in the heart's rhythm (atrial fibrillation, for example)
- cold, clammy, sweaty, and bluish skin
- a drop in blood pressure resulting in a thready pulse

**Diagnosis**

A doctor can usually diagnose pulmonary edema based on the patient's symptoms and a physical exam. Patients with pulmonary edema will have a rapid pulse, rapid breathing, abnormal breath and heart sounds, and enlarged neck veins. A chest x ray is often used to confirm the diagnosis. Arterial blood gas testing may be done. Sometimes pulmonary artery catheterization is performed to confirm that the patient has pulmonary edema and not a disease with similar symptoms (called adult respiratory distress syndrome or "noncardiogenic pulmonary edema").
**Treatment**

Pulmonary edema requires immediate emergency treatment. Treatment includes: placing the patient in a sitting position, oxygen, assisted or mechanical ventilation (in some cases), and drug therapy. The goal of treatment is to reduce the amount of fluid in the lungs, improve gas exchange and heart function, and, where possible, to correct the underlying disease.

To help the patient breathe better, he/she is placed in a sitting position. High concentrations of oxygen are administered. In cases where respiratory distress is severe, a mechanical ventilator and a tube down the throat (tracheal intubation) will be used to improve the delivery of oxygen. Non-invasive pressure support ventilation is a new treatment for pulmonary edema in which the patient breathes against a continuous flow of positive airway pressure, delivered through a face or nasal mask. Non-invasive pressure support ventilation decreases the effort required to breath, enhances oxygen and carbon dioxide exchange, and increases cardiac output.

Drug therapy could include morphine, nitroglycerin, diuretics, angiotensin-converting enzyme (ACE) inhibitors, and vasodilators. Vasopressors are used for cardiogenic shock. Morphine is very effective in reducing the patient's anxiety, easing breathing, and improving blood flow. Nitroglycerin reduces pulmonary blood flow and decreases the volume of fluid entering the overloaded blood vessels. Diuretics, like furosemide (Lasix), promote the elimination of fluids through urination, helping to reduce pressure and fluids in the blood vessels. ACE inhibitors reduce the pressure against which the left ventricle must expel blood. In patients who have severe hypertension, a vasodilator such as nitroprusside sodium (Nipride) may be used. For cardiogenic shock, an adrenergic agent (like dopamine hydrochloride [Intropin], dobutamine hydrochloride [Dobutrex], or epinephrine) or a bipyridine (like amrinone lactate [Inocor] or milrinone lactate [Primacor]) are given.

**Prognosis**

Most patients with pulmonary edema who seek immediate treatment can be treated quickly and effectively.

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**Infectious Respiratory Disease**

**Infectious Respiratory Diseases** are, as the name suggests, typically caused by one of many infectious agents able to infect the respiratory system (for example the bacterium Streptococcus pneumoniae).

**Pneumonia**

Pneumonia is a serious infection and/or inflammation of your lungs. The air sacs in the lungs fill with pus and other liquid. Oxygen has trouble reaching your blood. If there is too little oxygen in your blood, your body cells can't work properly. Because of this and spreading infection through the body pneumonia can cause death.

Pneumonia can have over 30 different causes.

Pneumonia affects your lungs in two ways. Lobar pneumonia affects a section (lobe) of a lung. Bronchial pneumonia (or bronchopneumonia) affects patches throughout both lungs.

**CAUSES OF PNEUMONIA**
Pneumonia is not a single disease. It can have over 30 different causes. There are five main causes of pneumonia:

- Bacteria, Virus, Mycoplasmas, Other agents, such as fungi, Various Chemicals

**BACTERIAL PNEUMONIA**

Bacterial pneumonia can attack anyone from infants through the elderly. Alcoholics, the debilitated, post-operative patients, people with respiratory diseases or viral infections and people who have weakened immune systems are at greater risk.

Pneumonia bacteria are present in some healthy throats. When body defenses are weakened in some way, by illness, old age, malnutrition, general debility or impaired immunity, the bacteria can multiply and cause serious damage. Usually, when a person's resistance is lowered, bacteria work their way into the lungs and inflame the air sacs.

The tissue of part of a lobe of the lung, an entire lobe, or even most of the lung's five lobes becomes completely filled with liquid (this is called "consolidation"). The infection quickly spreads through the bloodstream and the whole body is invaded.

The organism streptococcus pneumoniae is the most common cause of bacterial pneumonia. It is one form of pneumonia for which a vaccine is available.

Symptoms: The onset of bacterial pneumonia can vary from gradual to sudden. In the most severe cases, the patient may experience shaking chills, chattering teeth, severe chest pain, and a cough that produces rust-colored or greenish mucus.

A person's temperature may rise as high as 105 degrees F. The patient sweats profusely, and breathing and pulse rate increase rapidly. Lips and nailbeds may have a bluish color due to lack of oxygen in the blood. A patient's mental state may be confused or delirious.

**VIRAL PNEUMONIA**

Half of all pneumonias are believed to be caused by viruses. More and more viruses are being identified as the cause of respiratory infection, and though most attack the upper respiratory tract, some produce pneumonia, especially in children. Most of these pneumonias are not serious and last a short time but some may be.

Infection with the influenza virus may be severe and occasionally fatal. The virus invades the lungs and multiplies, but there are almost no physical signs of lung tissue becoming filled with fluid. It finds many of its victims among those who have pre-existing heart or lung disease or are pregnant.

Symptoms: The initial symptoms of viral pneumonia are the same as influenza symptoms: fever, a dry cough, headache, muscle pain, and weakness. Within 12 to 36 hours, there is increasing breathlessness; the cough becomes worse and produces a small amount of mucus. There is a high fever and there may be blueness of the lips.

In extreme cases, the patient has a desperate need for air and extreme breathlessness. Viral pneumonias may be complicated by an invasion of bacteria, with all the typical symptoms of bacterial pneumonia.
MYCOPLASMA PNEUMONIA

Because of its somewhat different symptoms and physical signs, and because the course of the illness differed from classical pneumococcal pneumonia, mycoplasma pneumonia was once believed to be caused by one or more undiscovered viruses and was called "primary atypical pneumonia."

Identified during World War II, mycoplasmas are the smallest free-living agents of disease in humankind, unclassified as to whether bacteria or viruses, but having characteristics of both. They generally cause a mild and widespread pneumonia. They affect all age groups, occurring most frequently in older children and young adults. The death rate is low, even in untreated cases.

Symptoms: The most prominent symptom of mycoplasma pneumonia is a cough that tends to come in violent attacks, but produces only sparse whitish mucus. Chills and fever are early symptoms, and some patients experience nausea or vomiting. Patients may experience profound weakness that lasts for a long time.

OTHER KINDS OF PNEUMONIA

Pneumocystis carinii pneumonia (PCP) is caused by an organism believed to be a fungus. PCP may be the first sign of illness in many persons with AIDS.

PCP can be successfully treated in many cases. It may recur a few months later, but treatment can help to prevent or delay its recurrence.

Other less common pneumonias may be quite serious and are occurring more often. Various special pneumonias are caused by the inhalation of food, liquid, gases or dust, and by fungi. Foreign bodies or a bronchial obstruction such as a tumor may promote the occurrence of pneumonia, although they are not causes of pneumonia.

Rickettsia (also considered an organism somewhere between viruses and bacteria) cause Rocky Mountain spotted fever, Q fever, typhus and psittacosis, diseases that may have mild or severe effects on the lungs. Tuberculosis pneumonia is a very serious lung infection and extremely dangerous unless treated early.

TREATING PNEUMONIA

If you develop pneumonia, your chances of a fast recovery are greatest under certain conditions: if you’re young, if your pneumonia is caught early, if your defenses against disease are working well, if the infection hasn't spread, and if you’re not suffering from other illnesses.

In the young and healthy, early treatment with antibiotics can cure bacterial pneumonia, speed recovery from mycoplasma pneumonia, and a certain percentage of rickettsia cases. There is not yet a general treatment for viral pneumonia, although antiviral drugs are used for certain kinds. Most people can be treated at home.

The drugs used to fight pneumonia are determined by the germ causing the pneumonia and the judgment of the doctor. After a patient's temperature returns to normal, medication must be continued according to the doctor's instructions, otherwise the pneumonia may recur. Relapses can be far more serious than the first attack.

Besides antibiotics, patients are given supportive treatment: proper diet and oxygen to increase oxygen in the blood when needed. In some patients, medication to ease chest pain and to provide relief from violent cough may be necessary.
The vigorous young person may lead a normal life within a week of recovery from pneumonia. For the middle-aged, however, weeks may elapse before they regain their accustomed strength, vigor, and feeling of well-being. A person recovering from mycoplasma pneumonia may be weak for an extended period of time.

Adequate rest is important to maintain progress toward full recovery and to avoid relapse. Remember, don't rush recovery!

THANK YOU TO THE FOLLOWING SOURCES:
www.lungusa.org
www.healthatoz.com