EDITORIAL

Interstitial Lung Disease

A general term called Interstitial Lung Disease is one that includes a variety of chronic lung disorders. It affects lung in three ways, damaging it in a subtle way, with inflammation of the air sacs and scarring or fibrosis in the interstitium (or tissue between the air sacs) that makes the lung stiff.

Breathlessness during exercise is usually the first symptoms of these diseases along with dry cough something that mostly commonly people would ignore.

People with different types of Interstitial Lung Disease may have the same kind of symptoms but it may vary in severity. Chest X-rays may look alike. Further testing is usually recommended to identify the specific type. Some Interstitial Lung Disease have known causes and some (idiopathic) have unknown causes.

Interstitium is the tissue between the air sacs of the lungs. It is named after this tissue because this is the tissue affected by fibrosis. It’s also sometimes known as “interstitial pulmonary fibrosis.” The terms interstitial lung disease, pulmonary fibrosis and interstitial pulmonary fibrosis are often used to describe the same condition.

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. Patients with Interstitial Lung Disease may notice variations in symptoms — from very mild to moderate to very severe. Condition may remain the same for long periods of time or it may change quickly. It’s important for a patient to stay in touch with his or her doctor and report any changes in symptoms. The disease has to be managed with mutual cooperation.

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 such as bronchioles causing bronchiolitis, alveoli causing alveolitis or the small blood vessels (capillaries) of the lungs causing 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. The level of disability that a person experiences depends on the amount of scarring of the tissue. 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.
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.

Sarcoidosis is another condition characterized by the formation of granulomas (areas of inflammatory cells), and it may attack any area of the body but most frequently affects the lungs. Certain medicines, radiation therapy for lung cancer and connective tissue or collagen diseases such as rheumatoid arthritis and systemic sclerosis may have the undesirable side effect of causing pulmonary fibrosis.

When all known causes of interstitial lung disease have been ruled out, the condition is called "idiopathic" 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.

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. When the disease is severe and prolonged, heart failure with swelling of the legs may occur.

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.

Bronchoalveolar lavage (BAL) and Lung biopsy may be used to diagnose 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 reversing the disease.

Some physicians may use corticosteroids in combination with other drugs when the diagnosis is
first established. Which drug treatment plan is effective and how long to use the drugs is the focus of current research.

Oxygen therapy may be prescribed for some people with IPF. Severity of disease, as well as the activity level of a patient determines the need for oxygen.

Influenza vaccine and pneumococcal pneumonia vaccine are both recommended for people with IPF or any lung disease. These two shots may help prevent infection and keep you healthy. Lung transplantation may offer hope for selected people with severe IPF and other lung diseases.

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.

Dr. I.S. Burki
Editor in Chief