

Interstitial Lung Disease

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Interstitial lung disease is a general category that includes many different lung conditions. All interstitial **lung diseases** affect the interstitium, a part of the **lungs'** anatomic structure.

The interstitium is a lace-like network of tissue that extends throughout both lungs. The interstitium provides support to the lungs' microscopic air sacs (alveoli). Tiny **blood** vessels travel through the interstitium, allowing gas exchange between **blood** and the air in the lungs. Normally, the interstitium is so thin it can't be seen on chest X-rays or CT scans

Definition

- **Interstitial lung** disease is a group of **lung** disorders in which the deep **lung** tissues become inflamed and then damaged. The **lungs** contain tiny air sacs (alveoli), which is where oxygen is absorbed. These air sacs expand with each breath. The tissue around these air sacs is called the **interstitium**.

Types of Interstitial Lung Disease

- All forms of interstitial lung disease cause thickening of the interstitium. The thickening can be due to inflammation, scarring, or extra fluid (**edema**). Some forms of interstitial lung disease are short-lived; others are chronic and irreversible.

Causes of Interstitial Lung Disease

- Bacteria, viruses, and fungi are known to cause interstitial pneumonias. Regular exposures to inhaled irritants at work or during hobbies can also cause some interstitial lung disease. These irritants include:
- Asbestos
- Silica dust
- Talc
- Coal dust, or various other metal dusts from working in mining
- Grain dust from farming
- Bird proteins (such as from exotic birds, chickens, or pigeons)

Diffuse Interstitial Pulmonary Fibrosis

This disease is known as diffuse interstitial pulmonary fibrosis (IPF) in the United States and cryptogenic fibrosing alveolitis in Europe. Initially, some type of injury to the pulmonary parenchyma causes an influx of inflammatory and immune cells, resulting in a diffuse inflammatory process distal to the terminal bronchiole (alveolitis)..

The etiologic factors of this condition are uncertain. Similar conditions can be produced by certain drugs or poisons and are found in patients with rheumatoid arthritis and systemic sclerosis. There appears to be a genetic factor, since twins, siblings, and other members of the same family have been reported with diffuse interstitial pulmonary fibrosis. This condition has also been reported in a few individuals with Raynaud's phenomenon, ulcerative colitis, and other diseases, but their exact relationship remains unclear.

Symptoms

- fatigue,
- dyspnea on exertion,
- A chronic unproductive cough. As the disease progresses, the patient becomes steadily more dyspneic and cyanotic.

DIAGNOSIS

On **auscultation**, one notes sharply crackling rales. Chest expansion is reduced and clubbing of the digits is often present.

- The chest x-ray usually indicates diffuse reticular markings, most prominent in the lower lung fields.
- Pulmonary function tests show a reduced vital capacity and total lung capacity with no impaired flow rates.

Compliance is markedly reduced to less than half of the predicted value. A reduced diffusing capacity is the earliest and most consistent change.

At first, the arterial Pao₂ may be normal at rest but may drop significantly with exercise. Later the Pao₂ is markedly reduced because of the thickened alveolar membrane and ventilation-perfusion mismatching.

The Paco₂ is reduced as a result of hyperventilation,

Bronchoalveolar lavage (BAL) is often used to assess the amount of inflammation and the accumulation of immune effectors cells and proteins in the alveoli. This is a **medical procedure** in which a **bronchoscope** is passed through the **mouth** or nose into the **lungs** and fluid is squirted into a small part of the lung and then collected for examination. BAL is typically performed to **diagnose lung disease**. In particular, BAL is commonly used to diagnose **infections** in people with **immune system problems**, **pneumonia** in people on **ventilators**,

Broncho alveolar lavage (BAL) is often used to assess the amount of inflammation and the accumulation of immune effectors cells and proteins in the alveoli. The technique consists of wedging a fiber optic bronchoscope in a sub lobar airway, then infusing 20 to 50 ml aliquots of saline into the peripheral airway, which are immediately aspirated by syringe. A total of 150 to 300 ml is instilled and recovered. The fluid and cells are analyzed. High-intensity alveolitis is defined by **10%** or more polymorphonuclear granulocytes (PMNs) in BAL cell differential counts; low-intensity alveolitis consists of **10%** PMNs or less (Reynolds, 1986).

Medical management

- Corticosteroids are the mainstay of treatment for IPF
- Medications are usually continued for a year and then a decision is made about continuing or tapering the dosage.
- Individuals who are not responsive to corticosteroids may be placed on other immunosuppressive drugs such as cyclophosphamide, azathioprine, or penicillamine . Penicillamine is more effective in patients with connective tissue diseases and interstitial fibrosis other than those with IPF.

Patients should stop smoking at once. Supplemental oxygen is important with exercise as there is a characteristic significant fall in arterial oxygen tension (PaO_2) . Individuals that require more than 4 L flow/min by nasal prongs may prefer direct administration of oxygen into the trachea.

In addition to supplying higher concentrations of oxygen to the lungs, many patients prefer the cosmetic effective of not wearing obvious nasal prongs. Patients that have refractory disease limited to the lungs may be candidates for single or double lung transplantation.