

Interstitial Lung Diseases

Definition

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- Interstitial Lung Diseases are:
 - Chronic, nonmalignant, noninfectious diseases of the lower respiratory tract
 - Characterized by inflammation and derangement of the alveolar walls and damage to the lung parenchyma by varying degrees of inflammation and fibrosis

Definition

- Over 200 diseases of known or unknown cause result in interstitial lung disease (ILD)
- Diffuse lung diseases, such as emphysema or chronic obstructive lung disease, bronchiolitis, and pulmonary hypertension are not included in the definition of ILD

Aetiology

Causes

- Interstitial Lung Diseases (ILDs) can be categorized into two major groups:
 - ILDs with known causes
 - ILDs with unknown causes

Known Causes

- Inhalants:
 - Inorganic dusts (silicosis, asbestosis)
 - **Organic dusts (hypersensitivity pneumonitis)**
 - Gases (oxygen toxicity, sulphur dioxide)

Known Causes

- Drugs and toxins:
 - Antibiotics: nitrofurantoin, sulfasalazine
 - Anti-inflammatory agents: gold, penicillamine, aspirin
 - Cardiovascular agents: amiodarone
 - Chemotherapeutic agents: bleomycin, methotroxate
 - Recreational drugs: heroin, methadone, talc-contaminated drugs

Known Causes

- Infections

- Viruses (CMV)
- Bacteria (TB)
- Fungi (pneumocystis carini)

Unknown Causes

- The idiopathic interstitial pneumonias
 - **Idiopathic Pulmonary Fibrosis (IPF)**
 - Desquamative Interstitial Pneumonitis (DIP)
 - Cryptogenic Organizing Pneumonia (COP)
 - Respiratory bronchiolitis-associated interstitial lung disease (RBILD)
 - Nonspecific interstitial pneumonia (NSIP)
 - Lymphocytic interstitial pneumonia (LIP)
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Unknown Causes

- Connective tissue disease associated ILD
- Sarcoidosis
- Eosinophilic granuloma
- Eosinophilic pneumonia
- Pulmonary hemorrhage syndromes
 - Goodpastures syndrome
 - Wegener's granulomatosis
 - Idiopathic hemosiderosis

Clinical Presentation

Clinical Presentation

- Interstitial lung disease (ILD) patients typically present with progressive exertional dyspnea and/or a persistent nonproductive cough, often in patients with pre-existing systemic disease
- There may be clubbing and end-expiratory crackles
- There may also be pulmonary symptoms from an associated disorder

Symptoms

- Cough, usually nonproductive
- Exertional dyspnea, progressive
- Fever especially in acute ILD
- Wheezing is not usual and can occur with ILD associated with asthma (in children or adults), such as Churg-Strauss syndrome

Symptoms

- Hemoptysis is unusual and suggests diffuse alveolar hemorrhage syndromes such as Goodpasture's syndrome or idiopathic pulmonary hemosiderosis
- There may be symptoms from associated collagen vascular diseases, such as rheumatoid arthritis, Crohn's disease, or other tissue disorders

Symptoms

- Recurrent sinusitis occurs in Wegener's granulomatosis and Churg-Strauss syndrome
- Dry eyes and mouth can occur in sarcoidosis
- Pleuritic chest pain can occur with sarcoidosis, systemic lupus erythematosus (SLE), and histiocytosis X

Signs

- End-expiratory crackles that do not shift with coughing (present in 80% cases of ILD)
- Clubbing, especially in idiopathic pulmonary fibrosis

Signs

- Signs of pulmonary hypertension in advanced disease (elevated pulmonary component of the second heart sound (P2), holosystolic murmur along the left lower sternal border, a right-sided S3, and elevated jugular venous pressure)
- Extrapulmonary signs of collagen vascular diseases:
- Skin abnormalities (sarcoidosis, collagen vascular disease, tuberous sclerosis, amyloidosis, neurofibromatosis)

Signs

- Arthritis (collagen vascular disease, sarcoidosis)
- Uveitis/conjunctivitis (sarcoidosis, Behçet's disease, inflammatory bowel disease)
- Neurologic abnormalities (sarcoidosis, tuberous sclerosis)
- Peripheral adenopathy
- Hepatosplenomegaly (sarcoidosis)
- Muscle tenderness (polymyositis)

Diagnosis

Diagnosis

- The diagnosis of ILD relies on the history and examination, results of laboratory investigations, pulmonary physiology, imaging (chest X-ray and high-resolution computed tomography (CT) scan), and histologic examination of open lung biopsies.

Treatment

Treatment

- Even if expert consensus guidelines are followed, therapy is effective in only about 20% of ILD cases
- Given the variety of ILDs and diverse nature of their pathogenic mechanisms, treatment must be individualized and consultation with or referral to a

Treatment

- Therapy should be targeted at the underlying disorder, e.g. if the etiology is infectious, appropriate antibiotics should be administered; if the etiology is drug-related, the drug should be stopped; if it is collagen vascular disease-related, the underlying diseases should be treated appropriately, with corticosteroids; if the etiology is malignant, appropriate chemotherapy may be indicated
- Patients should avoid inhalation of any possible causative or contributory agents

Treatment

- Smoking cessation is the recommended treatment for respiratory bronchiolitis-associated interstitial lung disease (RB-ILD) and histiocytosis X
- All patients should be given influenza vaccine and pneumococcal pneumonia vaccine; all patients with chronic lung disease are recommended to receive these vaccines
- Supportive measures, such as oxygen therapy, may be required
- Rehabilitation and education programs may help to improve quality of life

Treatment

- Anyway, in some cases, further treatment may be recommended by the pulmonologist according to the special type of the disease.

Prognosis

Prognosis

- The overall prognosis for interstitial lung disease (ILD) is poor, with a 75% mortality rate in the 8 years after presentation.
- However, the prognosis depends on the underlying cause.

Idiopathic Pulmonary Fibrosis

Idiopathic Pulmonary Fibrosis

- Idiopathic pulmonary fibrosis (IPF) is a chronic progressive interstitial lung disease of unknown aetiology, characterized by inflammation and fibrosis of the lung parenchyma. No specific pathognomonic clinical or pathologic findings are associated with IPF, and diagnosis is made after excluding other causes of interstitial lung disease.

Idiopathic Pulmonary Fibrosis

- Clinical features consist of progressive dyspnea upon exertion, interstitial infiltrates on chest radiographs, and a restrictive ventilatory defect found on pulmonary function test results.

Idiopathic Pulmonary Fibrosis

- The chest radiology shows patchy, predominantly peripheral, subpleural, reticular opacities in the lower lung zones.
- There may also be a ground-glass opacity usually associated with traction bronchiectasis and bronchiolectasis or subpleural honeycombing.
- Pulmonary function tests often reveal a restrictive pattern, a reduced DLco, and arterial hypoxemia that is exaggerated or elicited by exercise.

Idiopathic Pulmonary Fibrosis

- Histologic examination is essential to confirm the diagnosis.
- The histologic hallmark and chief diagnostic criterion of IPF is a heterogeneous appearance at low magnification with alternating areas of normal lung, interstitial inflammation, fibrosis, and honeycomb changes.

Idiopathic Pulmonary Fibrosis

- Treatment with systemic corticosteroids, other immunosuppressants, or both may benefit patients with lung histopathology patterns of high cellular inflammation and less fibrosis.

Take-Home Points

Take-Home Points

- Interstitial Lung Diseases:
 - Chronic, nonmalignant, noninfectious diseases of the lower respiratory tract
 - Associated with a wide variety of causative agents
 - Suspect possible interstitial lung disease in a patient who presents with dyspnea, exercise intolerance, dry cough, or diffuse parenchymal abnormalities on chest X-ray
 - There are characteristic changes on the chest X-ray and high-resolution computed tomography (CT) scans of the chest

Take-Home Points

- Lung function tests show a restrictive pattern with decreased gas transfer
- Many require open lung biopsy for definitive diagnosis
- Patients with ILD or suspected ILD should be under the direct or joint care of a respiratory physician
- Treatment is aimed at reducing or minimizing the inflammation in the lung parenchyma
- Prognosis is generally poor