Diffuse Interstitial Lung Disease



Diffuse interstitial (in-tur-STISH-ul) lung disease refers to a large group of lung disorders that affect the interstitium, which is the connective tissue that forms the support structure of the alveoli (air sacs) of the lungs. Normally when you inhale, the air sacs fill with air and oxygen passes into the blood stream. When you exhale, carbon dioxide that passed from the blood into the air sacs is expelled from the body. When interstitial lung disease is present, the lung is inflamed and stiff, preventing the air sacs from fully expanding. This limits both the delivery of oxygen to the blood stream and the removal of carbon dioxide from the body. As the disease progresses, the interstitium thickens, which further impedes lung function.

Blood tests, pulmonary function tests (spirometry), pulse oximetry, chest x-ray, chest CT, bronchoscopy with biopsy, surgical biopsy, or a combination of the above may be performed to help diagnose your condition. Treatment may depend on the underlying cause of the disease and your health status. Medication, respiratory therapy or surgery may be prescribed to help improve lung function.

What is diffuse interstitial lung disease?

Diffuse interstitial lung disease (ILD) is a group of complex disorders that affect the connective tissue (interstitium) that forms the support structure of the alveoli (http://www.radiologyinfo.org) (air sacs) of the lungs.

In a normal lung, the air sacs fill with air during inhalation. Oxygen within the air passes through the walls of the air sacs into the blood stream and carbon dioxide passes from the blood into the air sacs. When affected by interstitial lung disease, the tissue supporting the air sacs becomes inflamed and stiff, which makes it difficult for the air sacs to fully expand. The abnormal interstitium (http://www.radiologyinfo.org) limits the delivery of oxygen to the body and the removal of carbon dioxide from the body. As interstitial lung disease progresses, the interstitium scars and thickens the alveolar walls, further decreasing lung function. In some cases, inflammation may be confined to one or more areas of the lung. However, it is frequently diffuse, meaning it occurs throughout both lungs.

Interstitial lung diseases can be grouped based on cause, association with other diseases, or pathology. Interstitial lung disease may be associated with:

- rheumatoid arthritis
- scleroderma (systemic sclerosis)
- polymyositis and dermatomyositis (an inflammatory disease that causes muscle weakness)
- inflammatory bowel disease

It may also be caused by exposure to drugs, such as:

- chemotherapy agents
- antiarrythmics (used to treat irregular heart rhythm)
- statins (used to lower cholesterol levels)

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- antibiotics

It may also stem from the inhalation of substances, such as

- asbestos
- silica
- chemicals
- animal proteins
- mold
- smoke

Interstitial lung disease related to the inhalation of cigarette smoke can occur as a spectrum of injury that includes respiratory bronchiolitis-interstitial lung disease and desquamative interstitial pneumonia.

In some cases, interstitial lung disease may be associated with a collection of inflammatory cells (granuloma), as is seen in sarcoidosis. Other interstitial lung diseases associated with inflammation include idiopathic pulmonary fibrosis (IPF), usual interstitial pneumonia (UIP), acute interstitial pneumonia (AIP), cryptogenic organizing pneumonia (COP), and non-specific interstitial pneumonia (NSIP). Occasionally, interstitial lung disease is associated with a familial cause or specific genetic disease. Although there are many causes of interstitial lung disease, in some cases, the cause cannot be determined.

The most common symptoms of diffuse interstitial lung disease are shortness of breath and dry cough. As the disease progresses, a patient may experience weight loss, muscle and joint pain, and fatigue. At a more advanced stage, an individual may develop an enlarged heart, enlargement of the fingertips clubbing (http://www.radiologyinfo.org)), and cyanosis (http://www.radiologyinfo.org) (blue coloration in the lips, skin and fingernails as a result of reduced oxygen levels in the blood).

How is diffuse interstitial lung disease diagnosed and evaluated?

To determine the cause of interstitial lung disease, a physician may perform a physical examination and order diagnostic tests, including:

- Blood tests: These tests may help identify autoimmune diseases, such as scleroderma and rheumatoid arthritis, which can be associated with interstitial lung disease.
- Spirometry (http://www.radiologyinfo.org) : This is a test of lung function, in which the patient exhales quickly and forcefully through a tube to measure how much air the lungs can hold and how quickly the air moves in and out of the lungs. Spirometry can help determine if there is a problem getting air into (restriction, such as fibrosis) or out of the lungs (obstruction, such as asthma).
- Pulse oximetry (http://www.radiologyinfo.org) : This test uses a small device placed on a fingertip. By shining a specific wavelength of light through the end of the finger it painlessly measures the amount of oxygen in the blood.
- Chest x-ray (https://www.radiologyinfo.org/en/info/chestrad) : The patterns of lung damage associated with various interstitial lung diseases are often identifiable on chest x-rays. Chest x-rays may also be used to track the progression of interstitial lung disease. *See Radiation Dose in X-ray and CT Exams (https://www.radiologyinfo.org/en/info/safety-xray) for more information about x-rays.*
- CT imaging of the chest (https://www.radiologyinfo.org/en/info/chestct) : Computed tomography (CT), including a specific technique known as high resolution CT, is used to see fine detail of the interstitium that may not be visible on a chest x-ray. In some cases, a specific diagnosis (such as idiopathic pulmonary fibrosis) can be confirmed based on the CT appearance, potentially avoiding the need for lung biopsy. A CT scan can also often help determine the extent of damage to the lungs, guide biopsy when needed, and help determine appropriate treatment(s). *See Radiation Dose in X-ray and CT Exams (https://www.radiologyinfo.org/en/info/safety-xray) for more information about x-rays.*

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- Bronchoscopy (http://www.radiologyinfo.org) and bronchoscopic biopsy: In this procedure, a very small sample of tissue is removed from the lung using a small, flexible tube called a bronchoscope (http://www.radiologyinfo.org) that is passed through the mouth or nose, down the trachea (windpipe) and into the lungs.
- Surgical biopsy: A surgical biopsy may be needed to obtain a larger piece of lung than is possible with bronchoscopy. During this procedure, surgical instruments and a small camera are inserted through two or three small incisions between the ribs, allowing a surgeon to obtain a small piece of lung tissue.

How is diffuse interstitial lung disease treated?

Treatment depends on the cause of the interstitial lung disease and the overall health of the patient. Various drugs may be prescribed to reduce inflammation in the lungs and/or to suppress the immune system.

Oxygen or respiratory therapy (http://www.radiologyinfo.org) may be offered to help improve lung function. Some patients with advanced lung disease may require a lung transplant (http://www.radiologyinfo.org) .

Which test, procedure, or treatment is best for me?

- *Chronic Dyspnea-Noncardiovascular Origin (https://www.radiologyinfo.org/en/info/acs-chronic-dyspnea-noncardiovascular)*

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