

MEDfacts

An Educational Health Series From National Jewish Health®



Interstitial Lung Disease

Interstitial lung disease (ILD) is a broad category of lung diseases that includes more than 130 disorders which are characterized by scarring (fibrosis) and/or inflammation of the lungs. Some of the disorders included under the heading of ILD are:

- Idiopathic pulmonary fibrosis,
- Connective tissue or autoimmune disease-related pulmonary fibrosis,
- Hypersensitivity pneumonitis,
- Sarcoidosis,
- Eosinophilic granuloma (a.k.a. Langerhan's cell histiocytosis),
- Chronic eosinophilic pneumonia,
- Pulmonary vasculitis,
- Pulmonary hemorrhage syndromes,
- Bronchiolitis obliterans,
- Lymphangioleiomyomatosis and
- Familial pulmonary fibrosis or Familial interstitial pneumonia.

What are Common Symptoms of ILD?

Because ILD disrupts the transfer of oxygen and carbon dioxide in the lungs, its symptoms typically manifest as problems of breathing. The most common symptoms of ILD are shortness of breath with exercise and a non-productive cough. These symptoms are generally slowly progressive, although rapid worsening can also occur. Some people also may have a variety of other symptoms such as fever, weight loss, fatigue, muscle and joint pain, and abnormal chest sounds, depending upon the cause.

What Happens in the Lungs with ILD?

In ILD the tissue in the lungs becomes inflamed and/or scarred. The interstitium of the lung includes the area in and around the small blood vessels and alveoli (air sacs) where the exchange of oxygen and carbon dioxide takes place. Inflammation and scarring of the interstitium disrupts the lung tissue which leads to a decrease in the ability of the lungs to extract oxygen from the air.

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The progression of ILD can vary from disease to disease and person to person. It is important to determine the specific form of ILD in patients because what happens over time and what the most beneficial treatment will differ depending on the cause. Importantly, each person responds differently to treatment so close monitoring during treatment is important.

What are the Causes of ILD?

ILD can develop from a variety of sources, ranging from other diseases to environmental factors. Some of the known causes of ILD include:

Connective Tissue or Autoimmune Disease

- Scleroderma/Progressive systemic sclerosis
- Lupus (Systemic lupus erythematosus)
- Rheumatoid arthritis
- Polymyositis/Dermatomyositis

Occupational and environmental exposure

- Inorganic dust (asbestos, silica, hard metals)
- Organic dust (bacteria, animal proteins)
- Gases, fumes

Drugs and poisons

- Chemotherapy medications
- Antibiotics (rare)
- Radiation therapy

Infections

- Residue of active infection of any type
- Ongoing chronic infections

Some ILDs, while we know a lot about how they affect people, have no known cause. These conditions are termed "idiopathic" (meaning of unknown cause). Some examples of these include:

- Idiopathic pulmonary fibrosis
- Idiopathic nonspecific interstitial pneumonitis
- Acute interstitial pneumonia
- Sarcoidosis

How is ILD Diagnosed?

A multiple step process is necessary to diagnose interstitial lung disease (ILD). Diagnosis begins with an in-depth look at family history, a review of any symptoms suggestive of connective tissue diseases, a listing of all current and former medications, and a review of any environmental and occupational exposures to dusts, gases, chemicals, pets (in particular birds), air conditioners and humidifiers.

Other procedures to help with diagnosis may include:

- A complete physical examination - Findings of crackling sounds in the lungs and changes in the skin, joints and fingernails can help direct further evaluation.
- A high resolution computed tomography (CT) scan - Results are usually abnormal and can often help determine the type of ILD present.
- Pulmonary function tests - These breathing test results are often abnormal with ILD. Your lung function may be checked before and after an inhaled bronchodilator treatment.
- Arterial blood gas - This test measures the amount of oxygen and carbon dioxide in your blood. The results may be normal or show a reduced oxygen level.
- Exercise testing - This test may show a reduction in your oxygen level during activity.
- Bronchoscopy with bronchoalveolar lavage or biopsy – A bronchoscopy may be done to check for inflammatory cells in your lungs or to sample small pieces of the lung to look for evidence of a specific ILD. Bronchoscopy involves inserting a tube through the nose into your trachea (windpipe) to see the airways. In bronchoalveolar lavage a small amount of sterile saline is placed in one area of your lung and then withdrawn. This fluid contains cells that will be analyzed under the microscope. Small biopsy tools may be used through the bronchoscope if indicated.
- Surgical lung biopsy – In many cases of ILD, a surgical lung biopsy is needed to get enough lung tissue to make a specific diagnosis. This may be performed with the use of a thoracoscope which allows the surgeon to biopsy multiple areas of one lung through a few very small incisions.

What is the Treatment for ILD?

Because current therapy is not thought to reverse scarring that has already taken place, it is important to diagnose and treat ILD as early as possible. If your doctor has identified an occupational or environmental exposure, removal from the source of the problem is essential - even if it means giving up your job, hobby, or changing where you live. People who respond well to therapy generally report less shortness of breath with exercise and stable breathing tests.

The goals of treatment are to:

- Identify the specific disease, start therapy early to decrease inflammation and prevent further lung scarring.
- Remove the source of the problem, when possible.
- Minimize and manage potential complications of ILD.
- Improve or prevent deterioration in a patient's quality of life.

The type of medication and length of therapy depends on the specific type of ILD. With some types of ILD, removing the source of exposure - including cigarette smoke, some medications, and environmental irritants - may be all that is needed. With other types of ILD, medications and even surgery may be required.

What are Some of the Medications?

Many medications are used to treat ILD and its various symptoms. The medications

seek to make breathing more productive, reduce inflammation, and suppress overactive immune systems. Medications to treat ILD include:

- **Oral Corticosteroids** — Prednisone or some other form of corticosteroid, is frequently the first medication used. For some people, corticosteroids alone may decrease lung inflammation and cause an improvement in symptoms. Other people may have to use corticosteroids in combinations with other therapies. It may take as long as 3 months to see results. Corticosteroids can have significant side effects. Some of the side effects include: increased appetite, weight gain, high blood pressure, salt and fluid retention, tendency to bruise easily, depression, psychosis, or hyperexcitability, tendency to develop diabetes, peptic ulcer, infections, cataracts and osteoporosis (a tendency to break bones). Talk with your health care provider about preventing and watching for these side effects.
- **Cytotoxic Agents or Immunosuppressive Medications** - Cyclophosphamide (Cytoxan®) may be used if steroid therapy has failed to be effective or if corticosteroid treatment is not possible. In some cases, a combination of a corticosteroid and cyclophosphamide is used with good results. This medication reduces inflammation by killing some inflammatory cells and suppressing their function. Response to therapy may be slow and require up to 6 months or longer. Cyclophosphamide can have significant side effects. Some potential side effects of cyclophosphamide include: gastrointestinal irritation, bladder inflammation, bone marrow suppression, infection, irregular menstruation and blood disorders.
- **Azathioprine (Imuran®)** – Azathioprine is another medication often used in combination with corticosteroids for the treatment of ILD. It is used if the side effects from other medications are not tolerable. Though early studies have shown this medication may not be as effective the side effects may be more manageable. Azathioprine's side effects include fever, skin rash, gastrointestinal irritation and blood disorders.
- **Mycophenolate (CellCept®)** - Mycophenolate can be used to help reduce the amount of steroids required. It works to prevent the immune system from attacking cells in the body that result in fibrosis. Myophenolate may produce side effects. The most common side effects include abdominal distress, sleepiness, muscle or joint pain.

Because of the potential side effects of the above medications, your doctor will carefully monitor you while on therapy. The decision to treat patients with ILD involves a careful weighing of the potential risks and benefits of therapy. The potential benefits from the treatment usually outweigh the risk from the medications side effects.

What are Some Other Therapies?

There are many investigational therapies currently being studied for the treatment of certain types of ILD. Some people with ILD may qualify for involvement in clinical trials of potential therapies; you should discuss this with your doctor.

- **Oxygen Therapy** - Oxygen therapy is required for some people with ILD because of a low level of oxygen in the blood. Decreased lung function and/or pulmonary hypertension may cause blood oxygen levels to be too low. Some may need oxygen therapy all of the time while others may need it only during sleep and exercise. There are many systems used to deliver oxygen. They allow patients to be active and travel while using oxygen.
- **Pulmonary Rehabilitation** - A pulmonary rehabilitation program is often recommended to help you achieve your highest level of functioning. This program includes education, exercise conditioning, breathing techniques, energy saving techniques, respiratory therapy evaluation, nutritional counseling and psychosocial support.
- **Lung Transplant** - If the above therapies fail to adequately treat ILD, lung transplant is an option for some advanced cases. With improved surgical techniques and post-transplant treatment, transplantation may offer improved quality of life and prolonged survival to selected patients.

Response to therapy varies widely. Some types of ILD may respond quickly and others may not respond at all. Treatment is considered successful if symptoms, physiological and x-ray findings are stabilized. Even with treatment, many types of ILD progress naturally with a worsening of symptoms, x-ray and physiologic findings. Sometimes worsening is due to a complication of the disease or therapy. This may include conditions such as pulmonary hypertension or right heart failure. Some therapies may result in infection, muscle weakness and osteoporosis.

What is the Role of National Jewish Health?

As a center specializing in the care of patients with ILD, our health care providers have vast experience in treating people with these rare and complex conditions. In conjunction with your local provider, doctors at National Jewish develop and implement a detailed plan of diagnosis and care based on the latest information available regarding ILD.

There is a tremendous amount of interest in understanding the mechanisms of inflammation and scarring in ILD, and extensive research in this area is being conducted at National Jewish. The National Institutes of Health has designated and funded National Jewish as a Specialized Center of Research for ILD. Together with our basic scientists, the doctors and staff at National Jewish work to broaden our understanding of the causes of ILD and develop new treatment approaches.

Note: This information is provided to you as an educational service of LUNG LINE® (1-800-222-LUNG). It is not meant to be a substitute for consulting with your own physician.

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