

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 (i.e. “fibrosis”) and/or inflammation of the lungs. ILD accounts for 15 percent of the cases seen by pulmonologists (lung specialists).

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

There are different types of interstitial lung disease that fall under the category of ILD. Some of the common ones are listed below:

- Idiopathic (unknown) Pulmonary Fibrosis
- Connective tissue or autoimmune disease-related ILD
- Hypersensitivity Pneumonitis
- Wegener’s Granulomatosis
- Churg Strauss (vasculitis)
- Chronic Eosinophilic Pneumonia
- Eosinophilic granuloma (Langerhan’s cell histiocytosis)
- Drug Induced Lung Disease
- Sarcoidosis
- Bronchiolitis Obliterans
- Lymphangioleiomyomatosis

The progression of ILD varies from disease to disease and from person to person. It is important to determine the specific form of ILD in each person because what happens over time and the treatment may differ depending on the cause.. Each person responds differently to treatment, so it is important for your doctor to monitor your treatment.

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What are Common Symptoms of ILD?

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. They may include: fever, weight loss, fatigue, muscle and joint pain and abnormal chest sounds, depending upon the cause.

What are the Causes of ILD?

Some of the known causes of ILD include:

Connective Tissue or Autoimmune Disease such as:

- Scleroderma
- Vasculitis
- Systemic Lupus Erythematosus
- Rheumatoid Arthritis
- Polymyositis/Dermatomyositis

Occupational and Environmental Exposure such as:

- Inorganic dust (silica, hard metal dusts)
- Organic dust (bacteria, animal proteins)
- Gases, fumes

Drugs and poisons

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 Pneumonia
- Acute Interstitial Pneumonia
- Sarcoidosis

How is ILD Diagnosed?

When the diagnosis of ILD is suspected, your doctor considers the following:

- **Your medical history** – This includes: a review of all the person's symptoms, a listing of current and former medications and a review of any environmental and occupational exposures to dust, gases, chemicals, pets (in particular birds) and humidifiers.
- **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 chest X-ray and high resolution computed tomography (CT) scan** - Results are often abnormal with ILD. This 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 decrease in your oxygen level during activity. Testing may include a six minute walk, oxygen titration and exercise tolerance test.
- **Laboratory data** – Blood work may be done to test for autoimmune disease and other disorders that can cause ILD.
- **Bronchoscopy with bronchoalveolar lavage** – 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. This allows the surgeon to biopsy multiple areas of one lung through a few very small incisions.

What is the Treatment for ILD?

Treatment for ILD is based upon the diagnosis and may differ depending on the diagnosis. Many forms of ILD can be treated successfully with medications. ILD that is primarily the result of inflammation in the lung can often be treated with immunosuppressive medications. Idiopathic pulmonary fibrosis (IPF) does not respond to immunosuppressive medications, but two medications have recently been approved by the FDA for the treatment of IPF. They are pirfenidone and nintedanib.

However, our current available therapies are not thought to reverse scarring or fibrosis that has already taken place. We are constantly researching new medications that may work to halt or reverse scarring. In some cases where medication therapy is not felt to be useful, a lung transplant is recommended. Because ILD can result in respiratory failure, it is important to diagnose and treat it 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 hobby or pet, or changing what you do in your job or where you live.

Regardless of the cause of ILD, the goals of treatment are:

- To decrease inflammation and prevent further lung scarring
- To remove the source of the problem, when possible
- To minimize and manage potential complications of ILD

- To improve or prevent deterioration in a person's quality of life

What are some of the Medications?

- **Oral Corticosteroids** — Prednisone or some other form of corticosteroid, is often the first medication used for many forms of ILD. For some people, corticosteroids alone may decrease lung inflammation and cause an improvement in symptoms. 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 hyper excitability, difficulty sleeping, tendency to develop diabetes, peptic ulcer, infections, cataracts and osteoporosis (weakening of the bones)
 - Talk with your health care provider about preventing and watching for these side effects.
- **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. Mycophenolate may produce side effects. Some of the side effects include:
 - Abdominal distress (diarrhea, cramping and nausea), sleepiness, fatigue and muscle or joint pain
- **Azathioprine (Imuran®)** – Azathioprine is another medication that may be used to help reduce the amount of steroids required. It is used if the side effects from other medications are not tolerable. Azathioprine is not typically recommended for treatment of people with Idiopathic Pulmonary Fibrosis. Some of the side effects include:
 - Fever, skin rash, abdominal distress (diarrhea, cramping and nausea) and blood disorders
- **Cyclophosphamide (Cytoxan®)** Cyclophosphamide 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 six months or longer. Some side effects include:
 - Abdominal distress (diarrhea, cramping and nausea), bladder inflammation, bone marrow suppression, infection, irregular menstruation and blood disorders.
- **Pirfenidone (Esbriet®)** - Pirfenidone is an oral medication taken three times daily. The specific mechanism is unknown, but it seems to have anti-fibrotic and anti-inflammatory properties. Pirfenidone slows the progression of disease for some people with IPF. Some side effects include:
 - Nausea, loss of appetite, stomach upset and photosensitivity (a tendency to easily develop sunburn, this may be severe).
- **Nintedanib (Ofev®)** – Nintedanib is an oral medication taken twice daily. It is a

triple kinase inhibitor that blocks several pathways that lead to the development of scars. Like pirfenidone, nintedanib slows the progression of disease for some people with IPF. Some side effects include:

- Diarrhea and less often nausea and vomiting.

Because of the potential side effects of the above medications, your doctor will carefully monitor you while on therapy. This will include routine blood work. The decision to treat people 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 medication side effects.

What are Some Other Therapies?

Oxygen Therapy- Oxygen is required for some people with ILD because of low level of oxygen in the blood. Some may need oxygen therapy all of the time while others may need it only during sleep and exercise. Improving the level of oxygen in the blood through the use of supplemental oxygen can help relieve the strain on the heart and lungs and improve symptoms of shortness of breath and fatigue. There is a stigma often associated with oxygen. Many people are embarrassed and are concerned with how it will look and worry about how it will change their lifestyle. There are different options for oxygen systems and people are still able to get out their homes and even travel. Most people find once they are on oxygen they are able to be more active as they are not as short of breath. We can discuss what oxygen system will best fit your lifestyle.

Pulmonary Rehabilitation-This is a program that is often recommended to help you achieve your highest level of functioning. This program includes education, exercise conditioning, breathing techniques and energy saving techniques, respiratory therapy evaluation, nutritional counseling and psychosocial support.

The specific goals of pulmonary rehabilitation are to improve quality of life by: decreasing respiratory symptoms and complications, encouraging self management and control over daily functioning, improving physical conditioning and exercise performance, improving emotional well-being and reducing hospitalizations.

Lung Transplant- If other therapies fail to adequately treat ILD, lung transplant is an option for some advanced cases. With improved surgical techniques and post-transplant care, this 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. For transplant centers nearest to you go to Organ Procurement and Transplantation Network at www.opt.transplant.hrsa.gov.

Clinical Trials- There are areas of active investigation into new treatment options for people with idiopathic pulmonary fibrosis. To find out more information you can go to our website at NJHealth.org or to www.clinicaltrials.gov for general information on clinical trials.

Support Group- The **Joe Walsh Memorial Pulmonary Fibrosis Support Group** at National Jewish Health meets the second Tuesday of every month. For questions regarding this group call Carol Bair at 303.398.1912.

What is the Role of National Jewish Health?

As a center specializing in the care of people 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 Health 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 Health. 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 Health 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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