

Understanding Pulmonary Fibrosis (ILD)

What is pulmonary fibrosis?

“Pulmonary” refers to lungs, and “fibrosis” is a term used to refer to scarring, so pulmonary fibrosis means scarring throughout the lungs. In pulmonary fibrosis, the air sacs in the lungs gradually become replaced by scar tissue. When scar tissue develops in the lungs, oxygen cannot be delivered to the body.

What causes it?

Pulmonary fibrosis can be caused by many conditions, including chronic inflammatory processes, infections, environmental agents (asbestos, silica, exposure to certain gases), exposure to ionizing radiation (such as radiation therapy to treat tumors of the chest), chronic conditions (lupus, rheumatoid arthritis), and certain medications.

In some people, the disease may develop without an identifiable cause. This is called idiopathic pulmonary fibrosis (IPF). Most people with IPF do not respond to medical therapy, while some of the other types of fibrosis, such as nonspecific interstitial pneumonitis (NSIP), may respond to immune suppressive therapy.

While there is no known cause of pulmonary fibrosis, there are certain risk factors that contribute to its development. These include:

- Cigarette smoking
- Inhaled environmental and occupational pollutants
- Diseases such as scleroderma, rheumatoid arthritis, lupus and sarcoidosis
- Infections (Epstein Barr virus, hepatitis C)
- Certain medications
- Therapeutic radiation
- Family history
- Rheumatologic disorders

Who gets it?

Worldwide, more than 500,000 people are diagnosed with pulmonary fibrosis each year. The disease most commonly affects males between the ages of 40 to 70. Most people are diagnosed in their 60s. However, people of all sexes and ages have been diagnosed with the disease.

What are its symptoms?

The signs and symptoms of pulmonary fibrosis can be very vague and often are initially attributed to other things such as a bad cold or the flu. As a result, the disease is frequently not diagnosed until it is very advanced. Symptoms usually progress over several months or years and may include:

- Shortness of breath (dyspnea)
- Chronic dry, hacking cough
- Chest discomfort
- Loss of appetite and weight loss

- Fatigue and weakness

The severity of symptoms and the worsening of symptoms over time can vary and are at least partially dependent upon the cause of the fibrosis.

How is pulmonary fibrosis diagnosed?

There is no single test that can diagnose pulmonary fibrosis. A pulmonologist usually relies on the results from several tests. A diagnosis of pulmonary fibrosis can be confirmed by:

- History and physical examination
- Computerized tomography (CT) scan of the lungs
- Removal of lung tissue (biopsy) for examination under a microscope
- Pulmonary function tests

How serious is it?

Pulmonary fibrosis is a very complex disease and the survival rate after diagnosis varies greatly. In general, the disease has a poor prognosis. Survival rates are highest when the disease is diagnosed at a young age, in an early stage and in those who have had a good response to medications. More than 40,000 people die from this disease annually.

There is new research, however, that offers hope both in understanding the causes of the disease and in developing effective treatments.

How is pulmonary fibrosis treated?

There is no cure for pulmonary fibrosis. Once scar tissue has formed in the lung, it cannot be removed surgically or repaired with medication. Treatment is directed at slowing the progression of the disease and minimizing symptoms. Some therapies a doctor may recommend include:

- Supplemental oxygen
- Medications that affect the immune system
- Pulmonary rehabilitation and exercise
- Lung transplantation

The toxicity and side effects of treatments can be serious. Therefore, patients with pulmonary fibrosis should be followed by a lung specialist experienced in this condition. The lung specialist will determine the need for treatment, the duration of treatment, and will monitor the response to therapy along with any side effects.

More information about pulmonary fibrosis

A patient's own lung specialist is the best source for information about pulmonary fibrosis.

Other sources include:

The American Lung Association

www.lungusa.org

National Institutes of Health, National Heart, Lung and Blood Institute

www.nhlbi.nih.gov

Contact Us

For more information/question or comment about Pneumonia

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