

Support the ILFA



Irish Lung Fibrosis Association

The Irish Lung Fibrosis Association is a voluntary organisation which relies on donations and fundraising activities to achieve its aims of research, education, and support.

If you would like to volunteer your time or if you want to know more about fundraising for ILFA, please contact us by email, telephone, or in writing.

If you would like to make a donation to ILFA, cheques, postal orders, or bank drafts can be made payable to the Irish Lung Fibrosis Association.

ILFA are also registered with www.mycharity.ie for on-line donations.

If you would like to donate on a regular basis, we can help you to set up a Standing Order. Further information is available on the website.

Thank you for your support.

Research Education Support

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What is Pulmonary Fibrosis?



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What is Pulmonary Fibrosis?

Pulmonary fibrosis is a progressive, debilitating disease, which results from the development of scarring or fibrosis of the lung tissue. When this occurs, the lungs stiffen and the ability to breathe normally and transfer oxygen sufficiently into the bloodstream gradually becomes restricted.

How does Pulmonary Fibrosis Progress?

In some instances Pulmonary Fibrosis can remain symptomless or unchanged for many years. Patients can experience mild, shortness of breath, which may have little or no impact upon their quality of life.

As scarring of the lung tissue begins to progress symptoms may worsen. Increased shortness of breath can prevent patients from performing some of their normal daily activities. Progression of disease may be gradual but occasionally it may run a rapid course, which could limit the patient's survival.

Symptoms include:

- Shortness of breath (particularly on exertion)
- Dry, hacking cough
- Fatigue
- Chest discomfort
- Loss of appetite and weight loss
- Clubbing (enlargement of fingertips)

What causes Pulmonary Fibrosis?

Pulmonary Fibrosis comes under the umbrella of many different forms of lung disease that result in scarring of the lung tissue. These are known as Interstitial Lung Diseases (ILD) where the causes and symptoms at times vary.

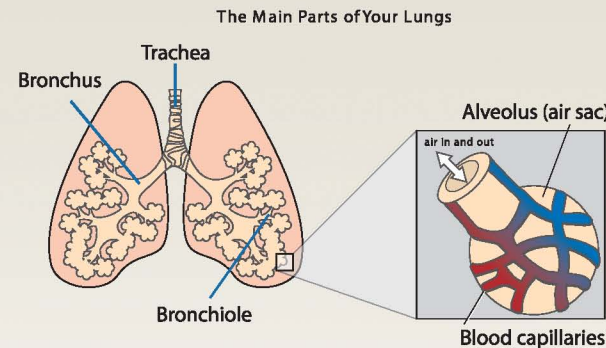


Figure 1. With pulmonary fibrosis, the air sacs become thicker and stiffer, which in turn makes it difficult to breathe

Idiopathic Pulmonary Fibrosis or IPF is a specific form of Interstitial Lung disease where the cause is unknown.

Other known conditions or diseases that may cause pulmonary fibrosis include:

- Inhalation of environmental hazards
- Inhalation of dust contaminated with bacterial, fungal or animal products
- Diseases involving connective tissue disorders such as: Rheumatoid Arthritis, Scleroderma and Sarcoidosis
- Certain medications: e.g. Amiodarone, Methotrexate, Nitrofurantoin, Bleomycin and Busulfan
- Certain therapies; some chemotherapies and radiotherapy

Who is at risk of Pulmonary Fibrosis?

A diagnosis of Idiopathic Pulmonary Fibrosis (IPF) is more commonly seen in men than women, mostly between 50 and 80 years of age.

How is Pulmonary Fibrosis treated?

Certain medications and therapies are used to relieve the symptoms that can occur with Pulmonary Fibrosis, however your treating physician must decide these on an individual basis. Pirfenidone is the only approved drug that specifically helps treat IPF and still remains under development in some countries.

For some patients who are suitable, lung transplantation can be an important and effective treatment. The National Lung Transplant Unit in Ireland is in the Mater Misericordiae University Hospital, Dublin.

Research to find effective medication and treatment remains ongoing.

How is Pulmonary Fibrosis managed?

Home oxygen therapy is very important and is used to help relieve symptoms of breathlessness and fatigue. There are many different ways in which oxygen can be used and it is important to follow guidelines directed by your doctor, nurse or physiotherapist.

The early treatment of respiratory/chest infection is very important and is also best directed by your treating, respiratory physician.

In many instances a pulmonary rehabilitation program may be suitable. Pulmonary rehab is an exercise program that can improve energy, strength and quality of life. The program is often led and monitored by a physiotherapist who can help maximise exercise tolerance and appropriate use of oxygen therapy.

It is important to maintain a healthy diet and continue with recreational activities to the extent that you can manage. This not only can improve outcome but it can often ensure a better quality of life mentally, physically and emotionally.