How is pulmonary fibrosis treated?

Unfortunately there is no cure for IPF. The treatments recommended by your doctor are used to slow down or prevent the disease progressing and to improve your symptoms.

A lung transplant is the only effective treatment for IPF but this is not a suitable option for everyone. It involves a major operation and depends on the severity of the patient’s IPF and general health. If you are being considered for a lung transplant, you will be assessed in the National Lung Transplant Centre in the Mater Misericordiae University Hospital in Dublin.

The medications Perfinidone and Nintedanib can slow down the formation of scar tissue in the lungs and the rate of disease progression. However, they cannot cure IPF or reverse the damage that is already there.

Your doctor may prescribe other medications to help you manage your symptoms and any other health problems you may have.

If your blood oxygen levels are low, your doctor will prescribe medical oxygen. Oxygen can help you manage your breathlessness and improve your quality of life. Your oxygen needs may change over time, so tell your doctor, nurse or physiotherapist if you are more breathless.

Please see our leaflets The treatment of Idiopathic Pulmonary Fibrosis and Oxygen and Idiopathic Pulmonary Fibrosis for more information.

Tips for managing your health and keeping well

IPF can affect you physically and emotionally but you can make positive lifestyle changes to help you cope with the challenges of living with this condition. The most important things you can do are to learn more about your lung condition, take a proactive approach to managing your health and try to stay positive.

Lifestyle tips to help you manage your IPF

- If you smoke, stop. Ask your doctor for help. Ask your family and friends not to smoke around you or in your home to protect you from the effects of second-hand smoke.
- Stay as active as possible. Exercise is very important for people with a lung condition and will help you to stay fit and strong. Exercise can also help improve your mood and emotional wellbeing. Contact ILFA to order your free exercise DVD and walking pack called The 2000 Steps a Day Challenge.
- Join a pulmonary rehabilitation class. These classes are a good way for IPF patients to take part in a supervised exercise programme. Ask your doctor to refer you to a class if there is one in your area.
- Eat healthy foods and maintain a healthy weight. Being overweight can put pressure on your chest and stomach muscles making it harder to breathe and causing you to be more tired. Some patients will lose weight as their IPF progresses. If this happens to you, it is important to get advice to prevent too much weight loss.
- Continue to socialise and enjoy your hobbies. It is important to carry on with these activities as much as you can. It is ok to take things slowly.
- Ask your doctor for help if you are struggling physically or emotionally. If you think you would like to have counselling, ask your doctor to refer you to a counsellor or psychologist.
- Join a lung fibrosis support group to meet others with IPF and learn from their experiences. You can join the ILFA mailing list to receive our free newsletter and keep up to date with developments.

ILFA has a wide range of resources for patients and carers. These include:

- The ILFA 2000 Steps a Day walking pack
- The ILFA Exercise DVD for lung fibrosis patients
- Information leaflets including:
  - The treatment of Idiopathic Pulmonary Fibrosis
  - Oxygen and Idiopathic Pulmonary Fibrosis
  - Getting the most out of your hospital appointments
  - Advice for carers of people with Idiopathic Pulmonary Fibrosis
  - Weight Management and Nutrition for Pulmonary Fibrosis

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What is Pulmonary Fibrosis?
About this leaflet
This leaflet tells you about the causes, symptoms and treatment for pulmonary fibrosis. It also includes tips and advice for managing your health.
The Irish Lung Fibrosis Association (ILFA) hopes you find this leaflet useful.

What is pulmonary fibrosis?
Pulmonary fibrosis is the name for a group of diseases in which scar tissue (fibrosis) develops in the lungs. Pulmonary fibrosis is also known as lung fibrosis.

When we breathe in, our lungs are exposed to many types of dust, particles, chemicals and toxins. Healthy lungs can repair the damage to lung tissue caused by these pollutants. However, if you have pulmonary fibrosis, the repair process goes out of control and too much tissue is produced causing fibrosis.

There are more than 200 different types of pulmonary fibrosis diseases. Doctors call these ‘Interstitial Lung Diseases’ because the tissue damage happens in the interstitial space which is the area around the tiny air sacs in the lungs.

As fibrosis develops, the soft, spongy and elastic tissue in healthy lungs is replaced by hard, thick scar tissue. This makes it harder for your lungs to expand when you breathe in. Fibrosis also prevents the normal movement of oxygen from the air sacs (alveoli) in the lungs into the bloodstream, causing you to be more breathless as your blood oxygen levels fall.

What causes pulmonary fibrosis?
Some pulmonary fibrosis diseases are caused by:
- breathing in dust in the workplace that contains tiny particles of wood, metal, stone or textiles;
- breathing in air or dust that is contaminated with bacteria, viruses or animal material such as bird droppings;
- medical conditions that affect the joints and connective tissue such as arthritis, scleroderma or sarcoidosis;
- medications such as amiodarone (a heart medicine), nitrofurantoin (an antibiotic), and methotrexate, bleomycin and bisulfan (all used for chemotherapy);
- some medical treatments such as radiotherapy.

Your doctor will ask you detailed questions about your job, hobbies, medical history, and previous exposures to chemicals, animals and medications, to help diagnose your lung condition.

However, it is often not possible to find the cause of the pulmonary fibrosis. When the cause is unknown the condition is called idiopathic pulmonary fibrosis.

What is idiopathic pulmonary fibrosis?
Idiopathic pulmonary fibrosis (IPF) is the most common type of interstitial lung disease. The word ‘idiopathic’ means that the cause of the pulmonary fibrosis is unknown.

IPF can be difficult to diagnose and medical experts will need to look at your medical history, symptoms, test results, x-rays and CT scans to reach a diagnosis. IPF is only diagnosed when all other known causes of lung fibrosis have been ruled out.

Who is at risk of developing IPF?
IPF is usually diagnosed in people over the age of 50. It is more common in older adults and rare in younger people. IPF affects more men than women. It is more common in smokers or people who have smoked in the past. However, nearly one in three IPF patients has never smoked.

Sometimes IPF develops in different family members, suggesting a possible genetic link. If more than one member of your family has IPF, it is called ‘familial IPF’. However, the same gene defect is not always present in family members with IPF. This suggests that multiple genetic and environmental factors are involved. At the moment, there is no reliable genetic test available for IPF and genetic testing of family members is not normally recommended.

About 800-1000 people in Ireland are diagnosed with IPF. As people live longer, it is expected that more people will be diagnosed with IPF.

What are the symptoms of IPF?
The first symptom you may notice is shortness of breath especially when you are climbing the stairs, walking, exercising or taking part in an activity. Your breathlessness can worsen as IPF progresses.

Other symptoms can include:
- a dry cough;
- low energy and fatigue causing you to feel exhausted or weak;
- changes in the shape of your fingertips and toes. They may get bigger and more rounded. This is called ‘clubbing’;
- stomach acid reflux (indigestion). This is common in IPF patients and can make your IPF worse if the stomach acid reaches your lungs.

How does IPF progress?
IPF is a progressive lung disease which means it gets worse over time.

The rate of disease progression is different for everyone. Some patients remain stable with a gradual change in their symptoms over many years. Other patients have a steady decline in their health and their symptoms get worse. Some patients may experience a sudden and serious worsening of their symptoms. This is called an acute exacerbation.

Talk to your doctor if you have concerns or questions about your situation and how your IPF is progressing.