

Other medications

Steroids, also called corticosteroids, are used to control inflammation. They may be prescribed if you have a chest infection or if your symptoms get worse. Sometimes high doses may be needed. Do not suddenly stop taking steroids. The dose must be gradually reduced, as advised by your doctor.

Sildenafil, also called Revatio®, is used to treat pulmonary hypertension (high blood pressure in the lungs), a common complication with IPF. Sildenafil lowers the blood pressure in the lungs and improves blood flow and the transfer of oxygen within the lungs helping to relieve breathlessness.

Antibiotics may be needed to treat an infection. You must always take the full amount of antibiotics prescribed by your doctor even if you feel better soon after starting them. Some patients may need to take antibiotics long term.

Proton Pump Inhibitors and **H2 blockers** are medications prescribed to prevent your stomach producing too much acid and to protect against acid reflux (indigestion), which is a common complication of IPF.

Statins are anti-cholesterol medicines that help lower the level of cholesterol in your blood, protect your heart and prevent a stroke.

Cough medications may be prescribed to help control your cough, although they are not always effective. Medicine containing codeine may sometimes be helpful.

Antidepressants may be needed if you have low mood and depression. Depression is common in people with IPF. These medications will help to balance the chemicals in the brain that are needed to ensure your mental wellbeing.

Vaccines are available to protect against influenza (flu) and pneumonia. Ask your doctor if you should get these.

Medication safety tips

It is important to follow your doctor's advice. Here are some useful safety tips:

- Ask your doctor to explain what your medications are for. If you don't understand, ask your doctor to explain it again or in a different way.
- Read the Patient Information Leaflet that comes in the pack with your medication.
- Tell your doctor or pharmacist if you have side effects from medication.
- Do not take over-the-counter medication (non-prescription drugs) without your doctor's advice.
- Never take medication bought over the Internet, herbal drugs or alternative therapies. The safety of these drugs is unknown.
- Use one pharmacy for all of your prescriptions.

Clinical trials and drug development

It takes many years for new drugs to be developed. If there is scientific evidence to show that the drug works, it undergoes more tests to see if it is safe and effective for humans. The next stage of drug development involves a clinical trial where the drug is tested by patients with IPF.

If there is a research study taking place, your doctor may ask you to take part in a clinical trial. Some patients will be treated with the medication being tested and some will receive a placebo (sugar pill) that will have no effect. Neither your doctor nor you will be told which tablet you are being treated with.

If you take part in a clinical trial, you will need to be assessed regularly to make sure that you are able to tolerate the medication, to monitor your progress and to identify any side effects. If there is good evidence to support the use of the drug, it will be approved (licensed) and doctors will be able to prescribe it for patients with IPF.

Evaluation of medication

Medical experts regularly review the treatments for IPF and make recommendations based on the best medical evidence available. Some drug treatments that were used in the past to treat IPF are no longer recommended. These include Interferon, Bosentan, N-acetylcysteine (NAC) and Azathioprine.

Future treatments

The search for better treatments for IPF is ongoing. Current research is focused on:

- drugs that slow down disease progression and prevent further fibrosis;
- treatments to control the complications of IPF and prevent acute exacerbations;
- genetic links and biomarkers (measurements of different biological cell types in the body) that may be involved with the development and progression of IPF.

Need more information?

Contact ILFA if you would like more information about IPF and to request some of our patient resources:

- the ILFA 2000 Steps a Day walking pack and Exercise DVD for lung fibrosis patients
- information leaflets
- the National Patient Charter for Idiopathic Pulmonary Fibrosis

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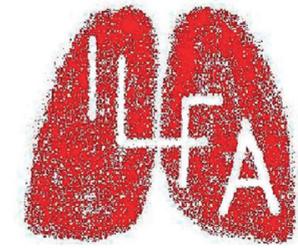
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Irish Lung Fibrosis Association

The treatment of Idiopathic Pulmonary Fibrosis



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About this leaflet

This leaflet explains what Idiopathic Pulmonary Fibrosis (IPF) is and describes three main treatments that your doctor may recommend to help you manage your IPF and your general health.

The Irish Lung Fibrosis Association (ILFA) hopes you find this leaflet useful.

What is IPF?

Idiopathic Pulmonary Fibrosis (IPF) is a progressive disease that causes scarring (fibrosis) and hardening of the lungs. The cause is unknown. Fibrosis prevents the normal movement of oxygen from the lungs into the blood supply and patients develop severe breathlessness, cough, fatigue (tiredness) and low energy.

IPF affects each patient differently and the symptoms vary from person to person. Often patients can have other health problems or develop new complications related to their IPF.

The rate of IPF progression is different for each person. Some patients can be stable and have a slow decline in their lung health, some can have a steady worsening and some have sudden flare ups and a rapid worsening of symptoms called acute exacerbations.

Treatments for IPF

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. There are three types of treatments:

1. **non-pharmacological** – no medication.
2. **lung transplant surgery** (if patients are suitable).
3. **pharmacological treatments** – medications are used to manage your symptoms.

Your doctor will carefully look at the risks and benefits of each treatment and consider your stage of fibrosis (early or advanced disease), symptoms, breathing test results and general health when deciding on the best treatment for you. You may need a combination of treatments at different times. Treatments may need to be stopped if your health gets worse.

1. Non-pharmacological treatments

Oxygen

Oxygen is one of the most important treatments for IPF patients. Your doctor will prescribe oxygen for you if your blood levels are low. Taking the correct amount of oxygen will help your body and vital organs to cope better with the stress of IPF and help ease your breathlessness and fatigue.

If you use oxygen correctly, it won't harm you. You should always follow your doctor's advice. Speak with your doctor, nurse or physiotherapist if you have any questions about oxygen.

Pulmonary rehabilitation

This is a special exercise programme for people with lung conditions including IPF. It teaches you about exercise, breathing methods and how to manage anxiety and stress. The classes are given by healthcare professionals and usually last for 6-10 weeks. The benefits of the programme to IPF patients are:

- improved lung function,
- increased exercise tolerance (ability to do more exercise before you become breathless and tired),
- improved quality of life, and
- other health benefits.

Ask your doctor to refer you to a pulmonary rehabilitation class in your area.

Exercise is as important as medicine

If you cannot attend a pulmonary rehabilitation programme, why not try the ILFA 2000 Steps a Day exercise challenge and the ILFA Home Exercise DVD?

The exercise DVD and 2000 Steps walking challenge were developed especially for lung fibrosis patients and take into account your limitations and oxygen requirements.

Contact ILFA for more information and to order your ILFA exercise resources.

Tel: 086 871 5264 or email info@ilfa.ie

Palliative care

This focuses on relieving your symptoms, particularly breathlessness and anxiety. It also maximises your wellbeing and improves your quality of life as IPF progresses.

Ask your doctor if your health is getting worse and you would like to know more about palliative care and planning for the future.

2. Lung transplant surgery

Some IPF patients **may** be suitable for a lung transplant operation which can lead to improved quality of life and increased survival time. The operation removes one or both lungs from the IPF patient and replaces the lung or lungs with a healthy lung (or lungs) from an organ donor.

Early medical assessment is important for patients who may benefit from a lung transplant. Your doctor needs to refer you as soon as possible for an assessment. You will be referred to The National Lung Transplant Unit at the Mater Misericordiae University Hospital in Dublin. You will need to undergo a lot of medical tests to determine if you meet the strict conditions necessary to be considered for a lung transplant.

It is important to exercise and keep as fit as possible to help with your recovery after the operation.

3. Pharmacological treatments

Oral anti-fibrotic medications

Pirfenidone (pronounced "pir-fen-id-own") also called Esbriet®, and **Nintedanib** (pronounced "nin-ted-a-nib") also called Ofev® are anti-fibrotic (anti-scarring) drugs used to treat patients with mild to moderate IPF. These medicines work in different ways to slow down the rate of IPF progression by reducing inflammation and scarring in the lungs but do not reverse damage that is already there.

As with all medicines, Pirfenidone and Nintedanib can cause side effects in some patients. Your hospital consultant will decide which is the most suitable medication for you and will take into account the stage of your fibrosis, the results of your blood tests and breathing tests, and the risk and benefits of the treatments.

Common side effects of Pirfenidone include nausea (feeling sick), vomiting, diarrhoea, tiredness, weight loss, stomach pain, changes to your liver function tests and a rash following sun exposure (photosensitivity). You will have to wear factor 50 sun-screen if you take Pirfenidone.

Common side effects of Nintedanib include diarrhoea, nausea, vomiting, stomach pain, loss of appetite, weight loss and changes to your liver function tests. You may have to take anti-diarrhoea medication if you take Nintedanib.

It is important to tell your doctor if you develop any side-effects from the medicine.

Your doctor may decide to change the dose of your medicine to help control symptoms. Once your symptoms improve, it may be possible to increase your dose over time.