IPF
Idiopathic pulmonary fibrosis

What is IPF?

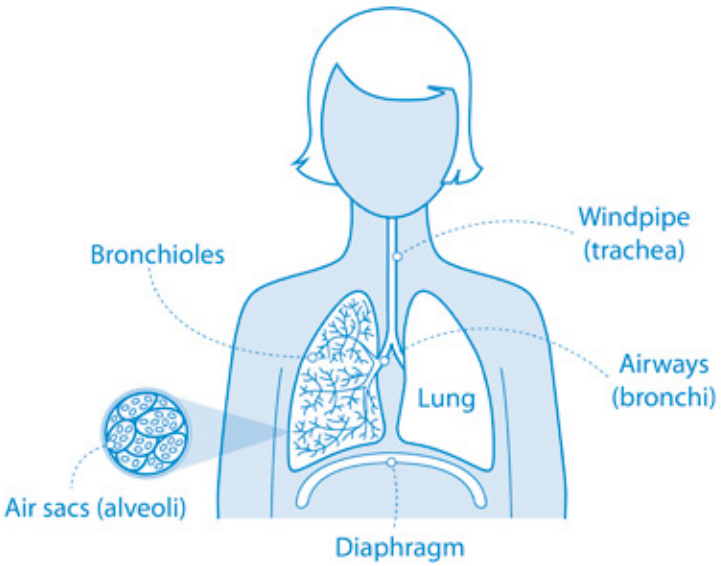
Idiopathic pulmonary fibrosis (IPF) scars your lungs and so reduces the efficiency of your breathing. The build-up of scar tissue is called fibrosis. Fibrosis causes the lungs to become stiffer and lose their elasticity so they're less able to inflate and take oxygen from the air you breathe.

IPF is a progressive condition and usually gets worse over time. In some people the symptoms gradually get worse over several years. For others, the symptoms get worse more quickly.

It's difficult to predict how IPF will progress. Sometimes when the condition has been stable, people can get sudden flare-ups of symptoms, called acute exacerbations. Everyone is different – talk to your specialist doctor about your individual situation.

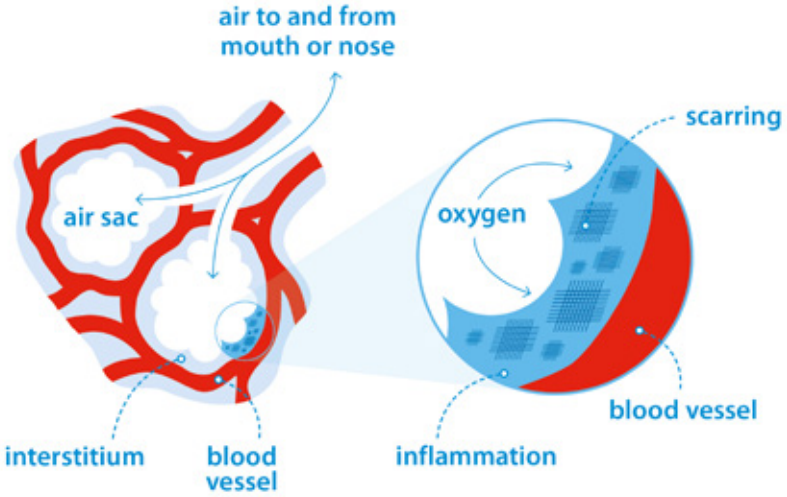
How does IPF affect breathing?

Each time you breathe in, you draw air in, down through your throat and into your windpipe. Your windpipe splits into two smaller tubes, called bronchi, which go to your lungs. The air passes down the bronchi, which divide into thousands of smaller airways called bronchioles.



The bronchioles have many small air sacs. Inside the air sacs, oxygen moves across paper-thin walls to tiny blood vessels and into your blood. The air sacs also pick up the waste gas, carbon dioxide, from your blood ready for you to breathe it out.

If you have IPF, scarring affects the air sacs, limiting the amount of oxygen that gets into the blood. With less oxygen in the blood, you can get breathlessness from everyday activities like walking.



Who's at risk?

Our latest research suggests about 6,000 people are diagnosed with IPF every year in the UK. Men are more likely to have IPF. IPF can affect people of all ages, but around 85% of diagnoses are made in people over 70.

Causes of IPF

The term idiopathic means the cause is not known. Researchers now believe that the body creates fibrosis in response to damage in the lung. The initial damage might be from:

- **acid reflux** from the stomach
- **viruses** – in some studies, IPF has been linked to certain viruses, including the Epstein Barr virus, which causes glandular fever. The herpes virus and hepatitis C have also been suggested as possible causes
- **environmental factors such as breathing in kinds of dusts** – it's more common if you've been exposed at work to dust from wood, metal, textiles or stone, or from cattle or farming

Some people may be genetically predisposed to develop IPF when their lung is damaged.

The scar tissue cannot currently be changed back to healthy tissue, so there is no cure yet for IPF. Current treatments aim to slow the rate of scarring, but they do not stop it.

Before the availability of specific treatments, studies showed that almost half of people with IPF in the UK died within three years of their diagnosis. However, about one in five people lived for more than five years after they were diagnosed. Clinicians believe the treatments now available will mean that people diagnosed today will survive longer.

Symptoms

The first symptom you might notice is getting out of breath when you're exerting yourself, such as climbing stairs. But you might feel constantly short of breath, and not just when you're moving about.

If you feel breathless, don't ignore it – see your doctor.

A **cough that doesn't go away** and **feeling very tired** all the time are two other symptoms of IPF. Another distinctive sign affects your fingers and toes, and is called **clubbing**.

Diagnosis

There are many different types of pulmonary fibrosis, so a multidisciplinary team will need to rule out these other conditions before they can say if you have IPF. Your doctor will listen to your chest, ask about your medical and work history, and order blood tests to rule out other causes. You might have further tests such as:

- a chest X-ray, which may be followed by a CT scan to produce a very detailed image of your lungs
- breathing tests to measure how well you can breathe in and out
- a bronchoscopy, where a narrow tube with a camera is passed down into your lungs, so the doctor can see inside and also get samples of lung tissue for analysis

Treatment

Your specialist team will try to slow the scarring and treat your symptoms so you feel better and your quality of life improves. You should have hospital appointments every three to six months and your care might include:

- **pulmonary rehabilitation** – a tailored exercise programme, which will help you cope with feeling short of breath
- **oxygen therapy** – if the level of oxygen in your blood falls, you can have a portable oxygen cylinder or an oxygen concentrator at home, to make the air you breathe richer in oxygen
- **medication to help with symptoms**
- **medication to slow the scarring** in your lungs
- **help to stop smoking**, if you smoke

For a very few people, having a **lung transplant** might be an option if the IPF progresses and isn't controlled by treatment. Transplants are rare. Your specialist doctor should discuss lung transplantation with you within six months of being diagnosed, if it's suitable for you. And, if you wish to explore the possibility, your doctor will contact the transplant centre.

Treating symptoms

For coughing, your doctor might treat problems that are making it worse, such as heartburn (acid reflux) or a stuffy nose. Make sure you tell your doctor if you have heart burn – there's some evidence this may make your fibrosis worse as well as your cough.

To help you cope when you **get out of breath**, pulmonary rehabilitation is an important treatment and you'll learn breathing techniques so that you feel more in control.

N-acetyl-cysteine or NAC helps break up mucus in the lungs, so your doctor may recommend it if you find it difficult to cough up phlegm or mucus. There's anecdotal evidence that some people find it helps their cough, but others experience stomach discomfort, trapped wind or nausea.

Medication to slow scarring in your lung

There are currently two drugs which are licensed for use in IPF: pirfenidone and nintedanib. They both slow down the development of scar tissue in the lungs of people with IPF. Several clinical trials are currently looking at possible new treatments, including combinations of existing treatments, so other options may be available in the coming years.

The National Institute for Health and Care Excellence (NICE), which advises the NHS on using new drug treatments, has only recommended the use of pirfenidone and nintedanib for people whose lung function is within a certain range. This means there's a chance that your doctor may not be able to prescribe you either drug – they will explain why.

NICE has also recommended that if your IPF continues to get worse, these drug treatments should be stopped. Again, your consultant will discuss this with you.

Pirfenidone and nintedanib have not been directly compared in clinical trials. Your lung specialist will be able to discuss the pros and cons with you to help you decide which drug is best for you.

Pirfenidone – brand name Esbriet

This treatment comes in the form of capsules: the usual dosage is nine capsules each day (three taken with each meal). Scientists don't know exactly how pirfenidone works yet, but they think it slows down inflammation and the build-up of scar tissue in the lungs. In medical studies, it slowed down the loss of lung function in most people with IPF, decreased the rate at which their symptoms got worse and also improved life expectancy.

There are some common side effects. These are skin reactions to sunlight, feeling sick or nauseous, tiredness and indigestion. Talk to your doctor about possible side effects if you're considering taking pirfenidone.

Nintedanib – brand name Ofev

Nintedanib is a new treatment which has also been shown in trials to slow the rate at which lungs become scarred in IPF. This drug is taken in the form of capsules, usually two a day.

Trials indicated nintedanib slows down the loss of lung function in people with IPF and may also reduce the rate of sudden flare-up of the symptoms. Common side effects include diarrhoea and nausea. If you're taking certain medications such as blood thinners, you may be advised not to take nintedanib.

There's lots of research underway. For the most up-to-date information, visit blf.org.uk/IPF or call our helpline.

Looking after yourself

If you have IPF, there's a lot you can do to look after yourself by leading a healthy lifestyle. Feeling very tired is common and health problems that used to be minor can become serious. Follow these tips to help keep your strength up, reduce your risk of complications and help you feel better generally:

- Have a flu jab each year.
- Avoid being around people with chest infections and colds.
- Ask your nurse for a pneumococcal vaccination to protect against pneumonia and many other infections.
- Stay as fit as you can. It's recommended we exercise 150 minutes each week. Find out more at **blf.org.uk/exercising**
- Eat a healthy, balanced diet. It's a good idea to ask to be referred to a dietician, who can give you tailor-made advice.

Remember to look after your mental health too. When you have a serious condition like IPF, it's common to have emotional struggles. Your doctor or nurse will understand if you are feeling low, and they can help.

It's important to keep enjoying life and we can help with practical advice on staying active. We can also put you in touch with your local support group. Our friendly helpline team are here to help you.

Get in touch with us to find support near you.

Helpline: **03000 030 555**

Monday to Friday, 9am-5pm

Ringing our helpline never costs more than a local call and is usually free, even from a mobile.

helpline@blf.org.uk **blf.org.uk**

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