



Idiopathic pulmonary fibrosis (IPF)

What is IPF?

Idiopathic pulmonary fibrosis (IPF) is a lung condition that scars your lungs and reduces the efficiency of your breathing. It's the most common type of [pulmonary fibrosis](https://www.blf.org.uk/support-for-you/pulmonary-fibrosis) ([blf.org.uk/support-for-you/pulmonary-fibrosis](https://www.blf.org.uk/support-for-you/pulmonary-fibrosis)). What does IPF stand for?

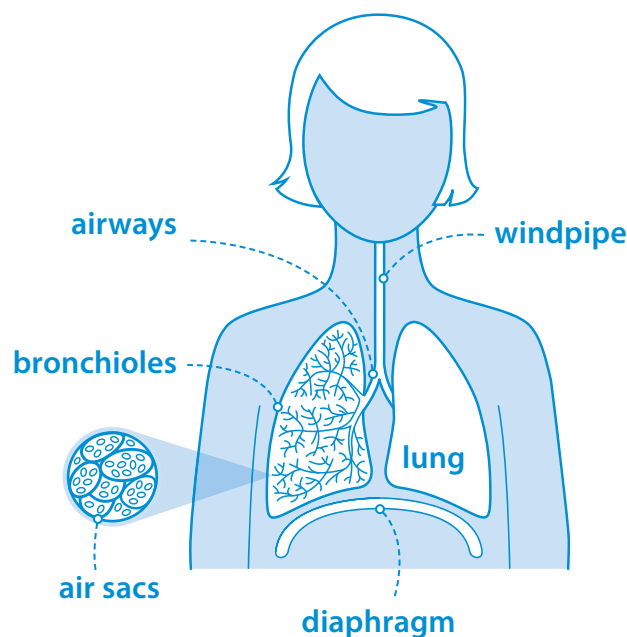
- **idiopathic** because no one knows what causes it
- **pulmonary** because it affects your lungs
- **fibrosis** because that's what the scar tissue in your lungs is called

IPF is a progressive condition and usually gets worse over time. At present, the scar tissue cannot be repaired by the body or any drugs, so there is no cure yet for IPF. Current treatment focuses on trying to slow the rate scar tissue is formed and controlling symptoms. In some people, symptoms get worse gradually over several years. For others, the symptoms get worse more quickly.

It's difficult to predict how rapidly IPF will progress in each person affected. Sometimes when the condition has been stable, people can get sudden flare-ups of symptoms, called exacerbations. There are no defined stages of IPF though some people may talk about mild, moderate or severe disease. Everyone is different - talk to your specialist doctor about your individual situation.

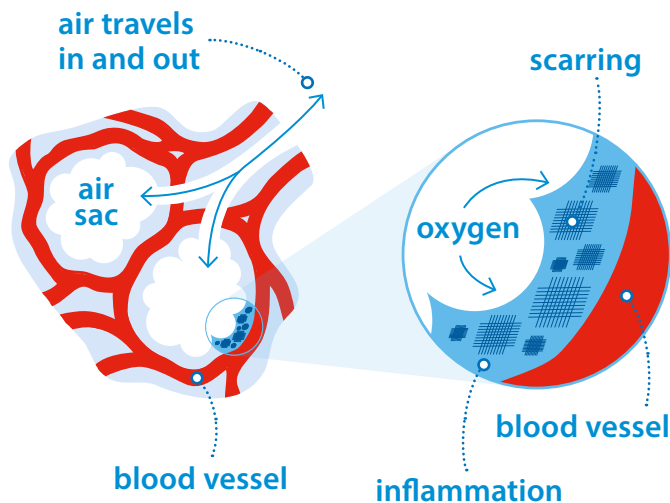
How does IPF affect breathing?

Scar tissue causes the lungs to become stiffer and to lose their elasticity. This stops them functioning properly: they're less able to inflate and transfer oxygen from the air you breathe into your body.



Each time you breathe in, you draw in air through your nose and mouth, down through your throat and into your windpipe (trachea). Your windpipe splits into two main tubes, the right and left bronchi, which supply your lungs. The main bronchi divide into gradually smaller airways called bronchioles. Bronchioles have many small air sacs (alveoli) at their ends. Inside the air sacs, oxygen moves across paper-thin walls to tiny blood vessels and into your blood. The air sacs also exchange waste gas, carbon dioxide, from your blood ready for you to breathe it out.

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



What causes IPF?

Idiopathic means the cause is not known but researchers now believe that the body creates fibrosis in response to injury to the lung. The initial injury 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 certain 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. But this link is only found in a minority of cases - about 10% in the UK in 2018. Current drug treatments can 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 3 years of their diagnosis. But some people, about 1 in 5, lived for more than 5 years after they were diagnosed. Clinicians now believe the treatments available will mean that people diagnosed today will survive for longer.

Who gets IPF?

Latest research suggests about 6,000 people are diagnosed with IPF every year in the UK. Nearly 80% are men, and on average, about three quarters are over 73.

Symptoms of IPF

The first symptom you might notice is getting out of breath when you're exerting yourself. The main symptoms of IPF are:

- breathlessness
- a cough that doesn't go away
- feeling tired all the time
- clubbing

If you have IPF, 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 change is clubbing, which that can affect some people's finger and toe nails. You may notice your nails changing shape to become like a drumstick: the tips of the fingers get bigger and the nails curve around the finger tips, and the base of the nail feels spongy. Having this doesn't mean you have IPF, it can occur in other conditions too. But check with your doctor if you notice it developing.

Diagnosing IPF

There are many different types of pulmonary fibrosis, not just IPF, so a multidisciplinary team of experts, often at a specialist hospital, will gather a lot of information about you before a definite diagnosis of IPF can be made. Your doctor will listen to your chest, ask about your medical, family, medication and work history, and order blood tests to rule out other causes. You may need tests such as:

- [a CT scan \(blf.org.uk/support-for-you/breathing-tests/other-tests#imaging\)](https://www.blf.org.uk/support-for-you/breathing-tests/other-tests#imaging) to produce a very detailed image of your lungs

On a CT scan, IPF often shows up as a distinctive pattern on the lungs. You might hear your doctor call this honeycomb lung. The image shows lots of empty pockets or bubbles appearing where more solid-looking lung tissue would normally appear.

- [breathing tests \(blf.org.uk/support-for-you/breathing-tests/tests-to-measure-breathing\)](https://www.blf.org.uk/support-for-you/breathing-tests/tests-to-measure-breathing) to measure how well your lungs are working
- [a bronchoscopy \(blf.org.uk/support-for-you/breathing-tests/other-tests#look\)](https://www.blf.org.uk/support-for-you/breathing-tests/other-tests#look), 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 or cells for analysis
- a lung biopsy. A chest surgeon may be asked to do a keyhole operation under anaesthetic to obtain samples of lung tissue for examination under a microscope. This information can be very helpful in identifying the cause of the fibrosis if other investigations haven't been conclusive.

If you have IPF, doctors listening to your chest can often hear crackles in your lungs that sound like opening velcro.

Treatment for IPF

You should be treated at a specialist centre, or by a team that comes from the specialist centre to your local hospital, and have hospital appointments every 3-6 months.

Your specialist team will discuss treatments:

- to try to slow the rate of scarring, if you're eligible
- to treat your symptoms

Clinical trials

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. You may want to discuss with your specialist team if you can take part in trials studying new treatments or therapies.

You may also be asked if data about your lungs can be collected for a national database, the British Thoracic Society IPF Registry, to help improve care for people with pulmonary fibrosis.

Medication to slow lung scarring

Currently 2 anti-fibrotic drugs can be prescribed

- pirfenidone
- nintedanib

Both slow down the development of scar tissue in the lungs of people with IPF. But, at the moment, they can only be prescribed to people whose lung function is within a set range. If your lung function is outside this range, you might be able to access this treatment by taking part in clinical trials. Pirfenidone and nintedanib have not been directly compared in clinical trials. Your lung specialist will be able to discuss the pros and cons of each with you to help you decide which drug is best for you. If you take either drug and your lung function gets worse and outside the recommended range, these treatments may be stopped. Your specialist will discuss this with you.

Pirfenidone - brand name Esbriet

This treatment comes in the form of capsules or tablets taken with meals. 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 clinical trials 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 feeling sick or nauseous, tiredness, indigestion and sometimes skin reactions to sunlight. Talk to your doctor about possible side effects if you're considering taking pirfenidone and ask about using sunscreen to prevent skin problems.

Nintedanib – brand name Ofev

Nintedanib has also been shown in trials to slow the rate at which lungs become scarred in IPF. This drug is also taken in the form of capsules. Common side effects include diarrhoea, abdominal pain and nausea. If you're taking certain medications such as blood thinners, you may be advised not to take nintedanib.

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 – sometimes because your lung function has not yet declined sufficiently.

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

The BLF and the Taskforce for Lung Health are campaigning to change the prescribing rules so people in the early stages of the disease and more advanced stages are also eligible for these drugs. The UK is the only country worldwide to impose the current restrictions.

Supportive treatment

Treating just the symptoms rather than the cause of a disease is often called best supportive care. Your lung specialist may suggest asking other specialists, such as palliative care specialists, to help you with specific problems. They are experts at symptom control and can also give psychological support to you and your family to improve quality of life. They can help with troublesome symptoms at any stage of your illness, not just at the very end of life.

Your team should discuss if you'd benefit from a course of [pulmonary rehabilitation](#) to improve your breathlessness and your ability to do your day-to-day activities. Your care might include:

- medication to help with symptoms
- [oxygen therapy](#) – (blf.org.uk/support-for-you/oxygen) if the level of oxygen in your blood falls when you're active or when you're resting
- [help to stop smoking](#), (blf.org.uk/support-for-you/smoking) if you smoke

For a very few people, having a lung transplant might be an option if the IPF progresses and isn't stabilised by treatment. Your specialist team should discuss lung transplant with you within six months of your diagnosis, if it's suitable for you (blf.org.uk/support-for-you/pulmonary-fibrosis/treatment)

Treating specific symptoms

Coughing

For coughing, your doctor will look for and treat problems that could be making it worse, such as heartburn (acid reflux) or a stuffy nose. Your doctor may also be able to refer you to a physiotherapist who can suggest ways to manage your cough, for example sipping water or swallowing.

Severe coughing can often be suppressed with low doses of liquid morphine. It is usually given by syringe into the mouth and works quite quickly. There are some potential side effects, so your doctor will discuss these with you before you decide if you want to take it.

If you find it difficult to cough mucus up, your doctor may recommend a medicine such as carbocisteine or N-acetyl-cysteine (NAC) to help loosen and thin mucus in the lungs. Some people find it helps their cough, but others may experience side effects such as stomach discomfort, trapped wind or nausea.

Reflux

Tell your doctor if you have symptoms of gastro-oesophageal reflux, such as heartburn, indigestion or a sour taste at the back of your mouth. There's evidence this may make fibrosis worse and also make a cough worse. You can have treatments that reduce the amount of acid your stomach makes, such as omeprazole. Liquid antacids such as Gaviscon can also be helpful in preventing reflux at night.

Breathlessness

To help you cope when you get out of breath, [pulmonary rehabilitation](https://www.blf.org.uk/support-for-you/keep-active/pulmonary-rehabilitation) ([blf.org.uk/support-for-you/keep-active/pulmonary-rehabilitation](https://www.blf.org.uk/support-for-you/keep-active/pulmonary-rehabilitation)) is an important treatment and you'll learn breathing techniques so that you feel more in control.

For more severe symptoms of [breathlessness](https://www.blf.org.uk/support-for-you/pulmonary-fibrosis/help-your-breathing) ([blf.org.uk/support-for-you/pulmonary-fibrosis/help-your-breathing](https://www.blf.org.uk/support-for-you/pulmonary-fibrosis/help-your-breathing)), which can sometimes make you very anxious, your doctor might recommend low doses of a short-acting medication such as lorazepam which can help take away sensations of severe breathlessness and panic. These tablets dissolve under your tongue.

Anxiety

You might feel very anxious or even depressed when living with a long term, progressive condition and your symptoms. Talking about your concerns with your medical team, friends and family can be helpful. But some people may also need medication or other therapies to help improve their mood. Don't be afraid to ask if you or your family member needs help to improve their mental wellbeing ([blf.org.uk/support-for-you/dealing-with-your-mental-health](https://www.blf.org.uk/support-for-you/dealing-with-your-mental-health))

Support for IPF

Pulmonary fibrosis support groups

Being diagnosed and living with a lung condition can be challenging. Our support groups can help you to make new friends who know what you're going through, and get lots of information about pulmonary fibrosis.

IPF support service

We have a free service for people living with IPF, their family and carers. Our IPF support service runs for 6 months when you'll talk to our IPF specialist advisor about topics important to you.

Looking after yourself

If you have idiopathic pulmonary fibrosis (IPF), there's a lot you can do to look after yourself by leading a healthy lifestyle. Why not have a look at our tips about looking after yourself (blf.org.uk/support-for-you/pulmonary-fibrosis/support)

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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We value feedback on our information. To let us know your views, and for the most up to date version of this information and references, call the helpline or visit blf.org.uk

British Lung Foundation

73-75 Goswell Road

London EC1V 7ER

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