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IDIOPATHIC PULMONARY FIBROSIS (IPF)

This information is about idiopathic pulmonary fibrosis (IPF). Here we provide support for those who have been diagnosed with IPF, as well as information about the condition.

What is IPF?

Idiopathic pulmonary fibrosis (IPF) is a lung condition that scars your lungs and makes it difficult for you to breathe. It's the most common type of pulmonary fibrosis.

What does IPF stand for?

I **idiopathic** = there is no known cause

P **pulmonary** = it affects your lungs

F **fibrosis** = the name of the scar tissue in your lungs

IPF is a progressive condition. This means it gets gradually worse over time. The scar tissue in the lungs cannot be repaired by the body or any drugs, and there is currently no treatment that stops or reverses the scarring.

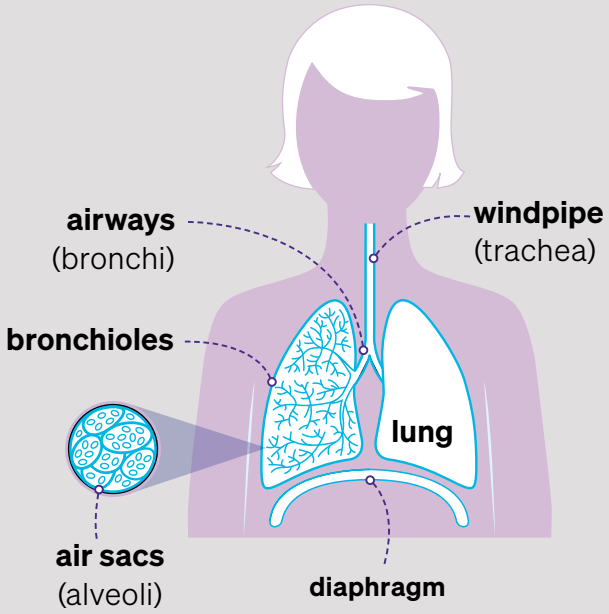
Current treatment focuses on slowing the rate of scarring in the lungs and controlling symptoms. Some people respond well to treatment and find their symptoms remain the same for many years. For others, the symptoms get worse more quickly.

It's difficult to predict how quickly IPF will develop because it varies a lot from person to person. There are no clear stages of IPF, though some people may talk about mild, moderate, or severe disease. Everyone is different – talk to your specialist doctor about your own situation.

How does IPF affect breathing?

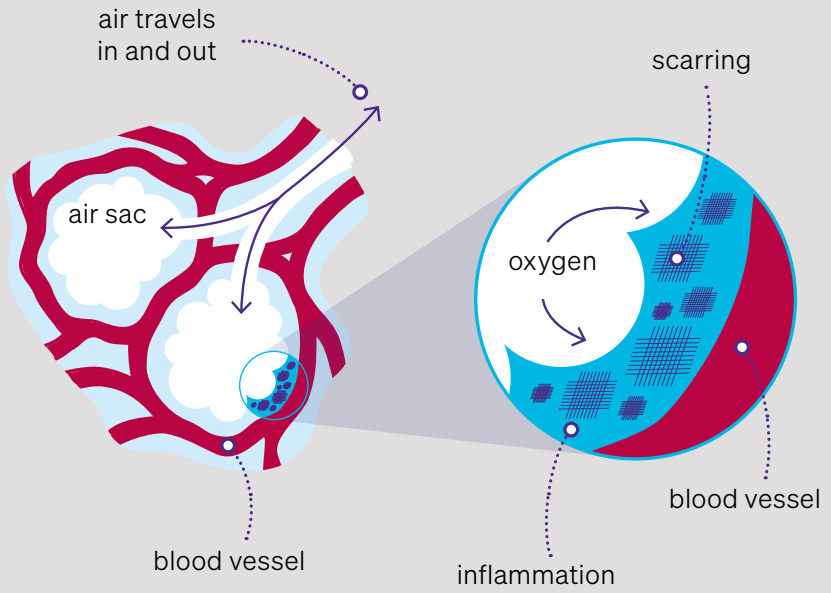
Scar tissue causes the lungs to become stiffer and to lose their elasticity. This stops them working properly: they're less able to inflate and transfer oxygen from the air you breathe into your body.

Each time you breathe in, you take 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 with air.



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 (scarring) in response to an injury to the lung.

The initial injury to the lungs might be from:

- **acid reflux** from the stomach
- **viruses** – in some studies, IPF has been linked to certain viruses, including herpes, hepatitis C and the Epstein Barr virus, which causes glandular fever
- **environmental factors** – breathing in certain kinds of dust from wood, metal, textiles or stone, or from cattle or farming
- **cigarette smoke exposure**.

It is not known if these factors directly cause IPF.

Some people may also get IPF if it's in their family history. But this link is only found in a minority of cases. According to a 2021 survey by the **British Thoracic Society**, 5.7% of patients were found to have a known relative with IPF.

Who gets IPF?

IPF is more common in people over the age of 40. It is also more common in men that it is in women.

Symptoms of IPF

The first symptom you might notice is getting out of breath when you're doing everyday activities like walking up the stairs.

The main symptoms of IPF are:

- breathlessness
- a cough that doesn't go away
- feeling tired all the time
- clubbing of the fingertips or toenails.

If you have IPF you may feel short of breath a lot, and not just when you're moving about. If you feel breathless you should visit your GP.

A cough that doesn't go away (usually a dry cough) and feeling tired are other symptoms of IPF.

You may also notice clubbing of your fingertips or toenails. This means the shape of your fingers or toes might appear like a drumstick, with the tips being larger around the nails. The base of the nails may also feel spongy. Having this doesn't mean you have IPF as it can occur in other conditions too. But check with your GP if you notice it developing.

Diagnosing IPF

A team of several experts (called a multi-disciplinary team), often at specialist hospitals, will collect information about you before making a diagnosis. This is because there are many types of pulmonary fibrosis.

IPF is one of many different types of pulmonary fibrosis. This means diagnosing IPF will involve a lot of tests to rule out the other types of pulmonary fibrosis. Your doctor is likely to listen to your chest and ask about your family, medication, work, and medical history. You may also have blood tests to rule out other causes for your symptoms.

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

You may need tests such as:

- **a CT scan** to produce a detailed image of your lungs
- **breathing tests**, such as spirometry, to measure how well your lungs are working
- **a bronchoscopy** to see inside the lung and get samples of the lung tissue – this is done using a small tube with a camera
- **a lung biopsy**, where tissue or cells are taken from the lung for testing.

IPF sometimes shows up as a honeycomb pattern on the lungs in a CT scan. The image shows lots of empty pockets or bubbles appearing where more solid-looking tissue would normally appear.

Being diagnosed with IPF might feel overwhelming at first. It can help to talk to your family and friends about it. You may want to read our information about mental health at **blf.org.uk/support-for-you/looking-after-your-mental-health**

Our Helpline is also here to support you in dealing with your diagnosis. Speak to a member of the team on **0300 222 5800** (Mon-Fri – 9am-5pm).

Treating and managing IPF

Treatment for IPF aims to slow the rate of scarring in the lung and treat the symptoms.

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

Clinical trials and new research

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 at **blf.org.uk/research/our-impact/how-you-can-get-involved-in-research**

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.

New research gives us hope of finding better treatments for IPF. You can read about our research into IPF at **blf.org.uk/research/our-impact/our-ipf-research**

Lung transplant

If the IPF gets worse in spite of treatment, a few people might have the option of having a lung transplant. Your specialist team should discuss lung transplant with you within six months of your diagnosis if it's suitable.

Medication to slow the scarring

There are currently two drugs that can be prescribed to slow down the rate of scar tissue developing in the lungs:

- Pirfenidone
- Nintedanib

These drugs can only be prescribed to people whose lung function is within a set range. If your lung function is outside of that range, you might only be able to access treatment by taking part in clinical trials.

If you're eligible, your specialist team will decide which drug is best for you. Current studies suggest both drugs are equally effective in slowing down the rate of scarring in the lungs.

Pirfenidone – brand name Esbriet

This treatment comes in the form of capsules or tablets taken with meals. Clinical trials found that the drug slowed down the loss of lung function in most people with IPF, slowed the rate at which their symptoms got worse, and improved life expectancy.

Common side effects (up to 1 in 10 people) associated with Pirfenidone are:

- feeling sick
- a rash on the skin
- feeling tired
- diarrhoea.

Some people also might have 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 (up to 1 in 10 people) associated with Nintedanib are:

- diarrhoea
- feeling sick
- stomach pain
- decreased appetite.

If you're taking certain medications, such as blood thinners, you may be told not to take Nintedanib.

Before the availability of specific treatments, studies showed that almost 5 in 10 people with IPF in the UK died within three years of their diagnosis. But some people, about 2 in 10, lived for more than five years after they were diagnosed. Clinicians now believe the treatments available will mean that people diagnosed today will live longer.

Supportive treatment

Treating the symptoms rather than the cause of a disease is called **best supportive care**. Your doctors may discuss pulmonary rehabilitation to help with your symptoms of breathlessness and oxygen therapy to help maintain oxygen levels to other organs.

Your doctors may also discuss palliative care – this is a plan to help you through any stage of a life-limiting illness and focuses on improving symptoms of the condition.

Coughing

Your doctor will look for and treat problems that could be making your coughing worse, such as heartburn (acid reflux) or a blocked nose. Your doctor may also be able to refer you to a physiotherapist

who can suggest ways to manage your cough, such as 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 potential side effects of morphine, so your doctor will discuss these with you before you decide if you want to take it.

If you find it difficult to cough up mucus, you may be prescribed medicine such as **carbocisteine** 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 feeling sick.

Breathlessness

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.

IPF can cause the level of oxygen in your blood to fall. This can make you feel more breathless. If this happens, you may be prescribed home oxygen treatment to allow you to be more active.

For more severe symptoms of breathlessness, 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.

Acid 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 IPF worse and 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.

Sometimes acid reflux is caused by a Hiatus Hernia, and this can be more common in those with IPF.

Anxiety

You might feel anxious or even depressed when living with a long term, progressive condition. 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.

The importance of self-care

It's important to look after yourself to stay as well as possible if you have IPF.

- If you smoke, the best thing you can do for your health is to quit.
- Exercise as often as you can to stay fit. This can also help to improve your mood.
- Eat a healthy, well-balanced diet.
- Look after your mental health. The NHS and Mind also has more advice on how to improve your mental health and wellbeing.
- Try to get enough sleep.
- Make sure you get your annual flu jab, coronavirus vaccines, and the one-off pneumonia vaccine.
- Try to keep away from people who are ill with coughs or colds.
- Let your doctor know if your symptoms are getting worse. If you are producing more mucus than normal, this could be a sign of a chest infection.

Enjoying life

We know that living with a condition like IPF can be difficult – but with the support of friends, family and your doctors, you can continue to enjoy life. We have some advice and ideas to support your emotional wellbeing:

- **Stay connected** – If you're getting out less, you might feel isolated. It's important to stay in touch with family and friends, whether that's meeting in person or chatting on the phone. You could also join our Health Unlocked forum to connect with other people who live with a lung condition at **healthunlocked.com/asthmalunguk-lung**
- **Take a break** – With some forward-planning, you can have a day out, visit family, or ask people to come and visit you. You may even be able to go on a holiday.
- **Start a hobby** – You may already have a hobby, such as a sport, but if you feel like you need something less physical, you could try something new. You could try something like art or music – which can be very therapeutic and can benefit your mental health. You can even try singing to improve your lung health.

Support for IPF

We have lots of support available for people living with IPF, from support groups to our helpline, and our pulmonary fibrosis passports.

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 with people who know what you're going through. You can also get lots of useful information about pulmonary fibrosis.

Find your local support group at **blf.org.uk/support-for-you/pulmonary-fibrosis/support-groups**

Join one of our support groups

Our support groups are about making new friends and connecting with others who know what you're going through. They are run by volunteers who organise information and support about how to manage and live with a lung condition.

Find your local support group at **blf.org.uk/support-for-you/breathe-easy**

Pulmonary fibrosis passports

The pulmonary fibrosis patient passport can help you to find out whether you're receiving the care you're entitled to, and what to do if not.

Download your free passport at **shop.asthmaandlung.org.uk/products/pulmonary-fibrosis-patient-passport**

Get help

Call our helpline on **0300 222 5800**

We are here to help if you want:

- answers to your questions – whether it's about coping with symptoms, your rights or finding equipment
- clear and trustworthy information about breathing problems and living with a lung condition
- to get in touch with your local support group
- Our friendly team are here Monday to Friday 9am to 5pm. Ringing will cost the same as a local call. It's usually free, depending on your call package, even from a mobile.

Or visit **AsthmaAndLung.org.uk** to find support and information or to join our web community

- get support and information **blf.org.uk/support-for-you**
- sign up to our newsletter **blf.org.uk/signup**
- find your local Breathe Easy group **blf.org.uk/breathe-easy**
- join our web community **healthunlocked.com/asthmalunguk-lung**

Together we fight for lung health

Helpline: **0300 222 5800**

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Asthma + Lung UK 2022

FL38 Version 4
Last reviewed: February 2022
Due for review: February 2025