Pulmonary fibrosis

What it is and how it affects your breathing

What is pulmonary fibrosis?

Pulmonary fibrosis is a term that covers many different conditions that cause scar tissue to build up in your lungs. This build-up of scar tissue, which makes your lungs stiff, is called fibrosis.

Pulmonary fibrosis is a type of interstitial lung disease (ILD). 'Interstitial' means the disease affects the interstitium, the lace-like network of tissue that supports the air sacs in your lungs. There are more than 200 different ILDs.

Some types of pulmonary fibrosis have an identifiable cause. But for many types, a definite cause cannot be found.

In ILDs, there can be scarring in your lungs or inflammation in your lungs. Some ILDs mostly cause scarring, some mostly cause inflammation. But often there is a combination of these processes going on. Which of these processes is dominant can determine what kind of treatment you may have.

The treatment and outlook for different types of pulmonary fibrosis vary considerably, so if you're not sure about your diagnosis, check with your doctor or nurse for the exact name of your lung condition.

All types of pulmonary fibrosis are rare. We have specific information about those seen most often in other PDFs:

- **idiopathic pulmonary fibrosis or IPF**
- **hypersensitivity pneumonitis formerly called extrinsic allergic alveolitis**
- **pneumoconiosis, also known as an occupational interstitial lung disease**
- **pulmonary fibrosis associated with connective tissue and autoimmune diseases**
- **drug-induced pulmonary fibrosis**

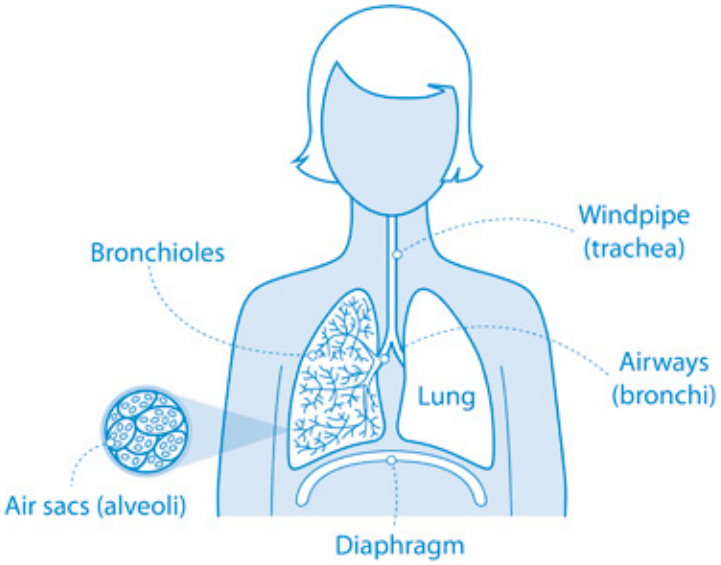
Although we do not always know what causes pulmonary fibrosis, we do know it is *not* a form of cancer or cystic fibrosis, and it is *not* contagious. Cystic fibrosis is not a type of ILD or pulmonary fibrosis.

Sarcoidosis is a relatively common type of disease that usually affects the lungs. It can sometimes cause pulmonary fibrosis but we don't know why. To find out more about this condition, go to **blf.org.uk/sarcoidosis** or call our helpline.

How does pulmonary fibrosis affect your breathing?

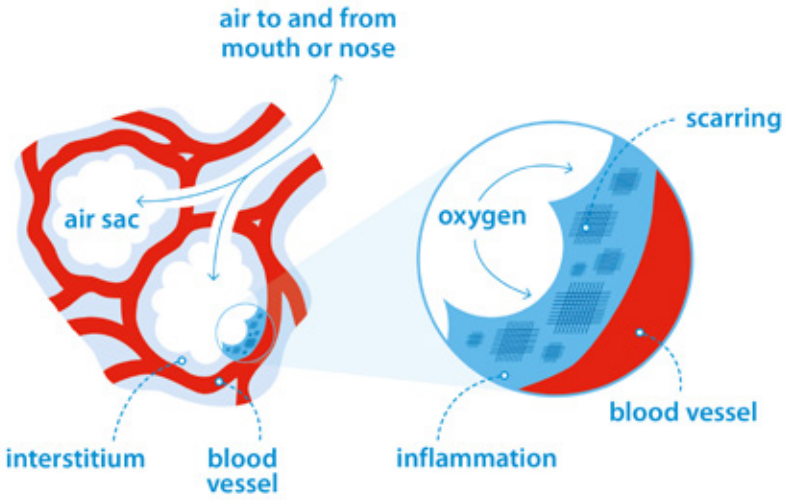
Pulmonary fibrosis scars your lungs and so reduces the efficiency of your breathing. Scarring causes your lungs to become stiffer and less elastic so they are less able to move and take oxygen from the air you breathe.

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



The bronchioles have many small air sacs, called alveoli. Inside the air sacs, oxygen moves across paper-thin walls to the capillaries - 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 pulmonary fibrosis, scarring affects the air sacs in your lungs. The air sacs are supported by the interstitium, a network of tissue, a bit like lace. Scarring fills in the gaps between and around the air sacs and limits the amount of oxygen that gets into the blood.



As scarring increases, your lungs are less able to expand to allow you to take deep breaths and the level of oxygen in your blood can start to drop. Breathing may feel like harder work and you can feel breathless from everyday activities like walking.

What causes pulmonary fibrosis?

In a small number of pulmonary fibrosis cases, it's possible to identify a specific cause. Some causes are:

- being exposed to certain types of dust - including wood or metal dusts or asbestos
- being exposed to allergens - such as bird feathers or mould
- a side effect of a drug

Some types of pulmonary fibrosis occur when you have another condition such as rheumatoid arthritis or scleroderma.

In a very few cases, two or more members of a family may develop pulmonary fibrosis. But current research suggests the genetics of familial interstitial lung disease is complex and there is no clear inherited predisposition to develop pulmonary fibrosis.

In most types of pulmonary fibrosis a specific cause cannot be found. One of the most frequently occurring forms of fibrosis is called idiopathic pulmonary fibrosis (IPF). The word 'idiopathic' means there is no known cause.

Researchers recently set out some common types of interstitial lung disease, grouped by what causes them, in the table below. It is not agreed by all doctors and there's lots of research underway to improve our understanding of the causes.

Classification of common interstitial lung diseases

Idiopathic disorders	• Idiopathic pulmonary fibrosis (IPF) • Acute interstitial pneumonia (AIP) • Idiopathic non-specific interstitial pneumonia (NSIP) • Sarcoidosis
Connective tissue and autoimmune disease	• Scleroderma / progressive systemic sclerosis • Systemic Lupus erythematosus (Lupus) • Rheumatoid arthritis • Polymyositis / dermatomyositis
Occupational and environmental	• Inorganic dust • Organic dust • Gases and fumes • Radiation
Drug-induced	• Chemotherapeutic agent • Radiation therapy • Antiarrhythmics • Antibiotics • Anticonvulsants

Infections	• Viral infections • Bacterial infections
Genetic / inherited	• Familial pulmonary fibrosis • Hermansky-Pudlak syndrome

Adam Wallis and Katherine Spinks: The diagnosis and management of ILDs, British Medical Journal 2015

What are the symptoms?

The different types of pulmonary fibrosis have similar symptoms, which is one of the reasons why you need to take tests and speak to a specialist consultant to find out exactly which type you have.

The first symptom a lot of people notice is **getting out of breath** when they're exerting themselves, such as climbing a hill or stairs. But you might feel constantly short of breath, and not just when you're moving about.

Several forms of pulmonary fibrosis usually occur after the age of 60, so you might think you're getting breathless because you are not as young as you were. This breathlessness will get worse over time, if not treated. Your shortness of breath may also be affected by other long-term conditions, such as chronic obstructive pulmonary disease (COPD), heart disease and being overweight.

A **cough that doesn't go away** and **feeling very tired** all the time are two other symptoms of pulmonary fibrosis. Some people with pulmonary fibrosis can also have a fever, lose weight or experience muscle and joint pain.

You might not think that a lung condition could affect your fingers and toes, but this is often a distinctive sign of pulmonary fibrosis. It's called **clubbing**, and you might notice:

- your nails feel too soft or as if they are coming loose
- the shape of your nails changes
- the tips of your fingers or toes bulge out

Clubbing can also occur with various other lung conditions, and as a result of heart or liver disease.

Get in touch with us to find support near you.

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Monday to Friday, 9am-5pm

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

British Lung Foundation
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Pulmonary fibrosis
Diagnosis and tests

If you have symptoms that might be due to pulmonary fibrosis, your GP will refer you to a consultant who specialises in lung conditions. It's important to identify the cause of your symptoms if possible, and what type of pulmonary fibrosis you have, as it will influence your treatment.

The doctor will ask questions about your medical and work history, listen to your chest, and order some blood tests to rule out possible known causes. Further tests used to diagnose pulmonary fibrosis are described below - you might not need to have all of them. Your consultant will assess this when they see you.

X-rays and scans

You will have a chest X-ray. If your doctor thinks you might have pulmonary fibrosis, this will be followed by a CT (computed tomography) scan, which uses X-rays to produce a very detailed image of your lungs. For some types of pulmonary fibrosis, the results from the scan can be very clear.

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

Breathing tests

These are also known as lung function tests. There are different types, but most involve breathing into a machine through a mouthpiece or tube. The equipment measures how well you can breathe in or out. The results can tell your doctor how much your lungs and breathing ability have been affected. If you are found to have pulmonary fibrosis, these tests will be used to monitor your condition.

You may also take a walking assessment. This usually involves walking for a few minutes, to see what happens when you exercise, including changes to the level of oxygen in your blood. This is measured by pulse oximetry, using a sensor placed on your finger or earlobe.

Looking inside your lungs

To confirm the diagnosis, your doctor may need to look inside your lungs and possibly remove some cells or tissue for testing. The samples are examined under a microscope by a pathologist – a doctor who is expert in laboratory tests.

Different procedures can be used to get some lung tissue:

- **flexible bronchoscopy** involves inserting a narrow tube through your nose or mouth, down into your lungs. You will have a local anaesthetic sprayed inside your nose and throat and often a sedative injection into your vein for your comfort.

  The tube has a camera on the end so the doctor can see inside your lungs. The doctor may flush some water through the tubing to remove and collect cells for analysis. Your doctor may also remove samples of tissue at the same time. These are called biopsies and are painless. This is usually an outpatient or day case procedure.

- **video-assisted thoracoscopy or VATS** involves surgery under a general anaesthetic to get a larger piece of lung tissue. A surgeon makes keyhole incisions in your chest for a video-assisted surgical telescope to enter, and to remove tissue samples from your lungs.

  It has higher risks than a bronchoscopy. It's undertaken only if your specialist thinks more tissue is needed to make a diagnosis and treatment plan. Your doctor will discuss the risks with you. You'll stay in hospital for a few days for this test.

Identifying and diagnosing pulmonary fibrosis is a joint effort by your specialist team, which will include several doctors who are experts in lung conditions, surgery, X-rays and scans, and laboratory tests. The specialist nurse is an important part of this team and a good source of information and support for you. The team will put together the findings from all of your tests to give you a diagnosis.

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Pulmonary fibrosis

Treatment and support

Treatment of your condition

There are treatments for most of the different types of pulmonary fibrosis, and your treatment will depend on the specific form of pulmonary fibrosis you have. You can find out more about the different treatments in our downloads for the specific conditions – *IPF*, *hypersensitivity pneumonitis*, *occupational interstitial lung diseases*, *pulmonary fibrosis associated with connective tissue and autoimmune disease* and *drug-induced pulmonary fibrosis*.

You might be invited to take part in a medical study, also called a clinical trial, to investigate new treatments. It's not an option for everyone – if you want to know more, ask your doctor.

Treatment of your symptoms

You'll also be offered treatment for specific symptoms, such as coughing, alongside disease-specific therapies.

For coughing, your doctor might treat problems that are making it worse, such as heartburn or a stuffy nose.

Do tell your doctor if you have symptoms of gastro-oesophageal reflux, such as **heartburn** or a sour taste at the back of your mouth. There's some evidence this may make any inflammation and fibrosis worse, as well as make a cough worse.

Feeling out of breath can have a serious effect on your everyday life. It can be a frightening experience, too. Pulmonary rehabilitation is an important treatment to help you cope with breathlessness.

Help to stop smoking

If you smoke, stopping is very important. The NHS offers a free stop smoking service – your GP can refer you or visit nhs.uk/smokefree

Pulmonary rehabilitation

Pulmonary rehabilitation, sometimes called PR, includes:

- a physical exercise programme, designed for people with lung conditions and tailored for you
- advice and information on looking after your body and lungs, managing your condition and coping with feeling short of breath

PR is led by a qualified respiratory professional, usually a physiotherapist. Nurses, occupational therapists, psychologists and dieticians may take part too.

PR is designed to support and reassure you, as well as help your condition. You may find you can walk farther, you feel less breathless and you feel generally more positive. Find out more at **blf.org.uk/exercising**

Ways to help your breathing

If you have pulmonary fibrosis, you may tend to breathe very fast and shallowly – a bit like panting. You can use techniques and positions to help you control and slow down your breathing. You can also use them to avoid getting too breathless when you exert yourself, and help yourself to recover when you do get out of breath.

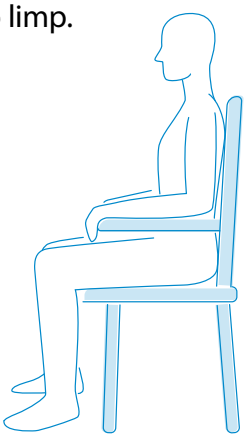
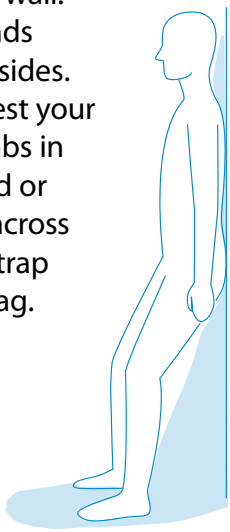
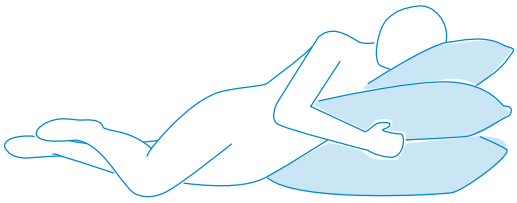
Talk to your respiratory physiotherapist or nurse for help to find out what works for you. Try the different breathing techniques to find what helps you and practise the ones that help. Our suggestions are based on what physiotherapists find works, as little research has been done with people living with pulmonary fibrosis.

tip

Hold a hand-held battery fan near your face if it helps you to feel less breathless.

Breathing positions

Use these positions to practise your breathing control or to recover your breath.

Position 1	Position 2	Position 3
Sit upright in a firm chair If your chair doesn't have arms, rest your arms on your thighs. Let your wrists and hands go limp. 	**Stand leaning backwards or sideways** Have your feet slightly apart, about one foot or 30cms away from the wall. Relax your hands down by your sides. If you prefer, rest your hands or thumbs in your waistband or belt loops, or across the shoulder strap of your handbag. 	**High side lying** Lie on your side with pillows under your head and shoulders. Make sure your top pillow supports your neck. Slightly bend your knees, hips and top leg. 

There's more detail in patient information leaflets developed by the Association of Chartered Physiotherapists in Respiratory Care at **www.acprc.uk/publications**.

Breathing control using your diaphragm

Breathing control means breathing gently, using the least effort. It will help when you're short of breath or feeling anxious. This type of breathing is commonly used in yoga.

Use this breathing control combined with any of the techniques on the next few pages.

You use your diaphragm – a big umbrella of muscle that's under your lungs, at the bottom of your rib cage. It contracts when you breathe, so pulling the lungs down, stretching and expanding them.

To get used to this, it helps to practise when you're sitting, and are relaxed and not out of breath. You'll then be able to use it when you are breathless, or to reduce your shortness of breath when you're exerting yourself.

Get into a comfortable position, with your arms supported (position 1 on the previous page). Let your shoulders and body be relaxed and loose.

- Put one hand on your chest and the other on your abdomen (tummy)
- Close your eyes to help you relax and focus on your breathing
- Slowly breathe in through your nose, with your mouth closed. If you're relaxed, the air will reach low in your lungs. You'll feel your abdomen rise – it will move out against your hand. The hand on your chest will not move much at all
- Breathe out through your mouth, either like a sigh or through pursed lips. Your abdomen will fall gently. Imagine all the tension in your body leaving as you let the air out
- Try to use as little effort as possible and make your breaths slow, relaxed and smooth. With every breath out, try to feel more relaxed and calm. Gradually try to breathe more slowly

Try the positions on the previous page to help you practise your breathing control, or to help you recover your breath when you get breathless. Your physiotherapist can help you find the positions that work best for you too.

Breathing techniques

Relaxed slow deep breathing is very useful while you're active. Use it from the very start of an activity that makes you out of breath such as walking or making the bed.	**How do I do it?** As you start to exert yourself, slow down your breathing and breathe in more deeply. Breathe in through your nose if you can. Use it with blow-as-you-go or paced breathing and pursed-lips breathing if that helps.

Blow-as-you-go helps make tasks and activities easier. Use it while you're doing something that makes you breathless. You can use it with pursed-lips breathing.	**How do I do it?** Breathe in before you make the effort. Then breathe out while you're making the effort. For example, when going up a step or standing up, breathe in before you stand or step up, and then blow out as you step or stand up. Try using pursed-lips as you blow out.
Paced breathing is useful when you are active, for example, walking or climbing stairs. You pace your steps to your breathing. You can use it at the same time as pursed-lips breathing and blow-as-you-go.	**How do I do it?** Count to yourself as you walk (or move). For example, breathe in for one step and then take either one or two steps as you breathe out. You can take more steps as you breathe in or as you breathe out, if that feels better for you. The right number for you will depend on you. It's worth trying different combinations to find the one that works best for you - for example, one step in, two steps out, or two steps in, three steps out.
Pursed-lips breathing can be used at any time to help you control your breathing.	**How do I do it?** Breathe in gently through your nose, then purse your lips as though you're going to blow out a candle. Blow out with your lips in this pursed position. Imagine blowing out a candle when you breathe out. Blow out only for as long as is comfortable – don't force your lungs to empty.

There's more detail in patient information leaflets developed by the Association of Chartered Physiotherapists in Respiratory Care at **www.acprc.uk/publications**.

Oxygen

If your condition gets worse, the level of oxygen in your blood may fall and this may make you feel more breathless. If this happens, your doctor will refer you for an assessment for oxygen therapy, which the NHS provides for free.

You might be given a portable oxygen cylinder to use when you walk around, or have an oxygen concentrator installed in your home. The concentrator takes normal air from the room and makes it richer in oxygen before you breathe it in. The machine is attached to tubing all around the house, so you're not confined to one room.

Some people with pulmonary fibrosis need supplementary oxygen when they move about, and sometimes later when they're resting too.

Your oxygen prescription will be tailored to meet your individual needs. And you'll be advised how to adjust the flow of oxygen according to how active you are.

You can find out more about oxygen therapy at **blf.org.uk/oxygen** or by calling our helpline.

Lung transplant

For a very few people, having a lung transplant might be an option if the pulmonary fibrosis progresses and isn't controlled by treatment. This is rare - only 175 lung transplants were carried out in the UK in 2011-12. Not all of these were for people with pulmonary fibrosis.

Whether you can be considered for a lung transplant depends on factors that influence the chance of a successful outcome, such as your general health, other medical conditions and your body mass index. There is no age cut-off, but it's unusual to accept people much over 65 years old. Sadly, there are not enough donor lungs available to meet demand.

If you wish to explore this option and your doctor thinks you might be suitable, you'll be referred to a transplant unit.

There are significant risks in having a transplant. According to recent research, the survival rate* at one year after lung transplantation is 79%, and for five years it is 53%. The survival rate at 10 years is 33%.

*Survival rate means the percentage of people who survive after lung transplant surgery.

Palliative care

Your doctor, nurse or physiotherapist should also talk to you about palliative care or symptom control, or refer you to a palliative care specialist. This care is designed to improve the quality of your life and those close to you. It includes controlling pain and other symptoms, which can be interrelated, such as breathlessness, fatigue and anxiety.

Pulmonary fibrosis tends to be progressive and you may become increasingly breathless. If you're having trouble breathing, even when you're resting, your doctor might prescribe a sedative or morphine. You might know them as painkillers or sleeping tablets but these medicines can also help with symptoms of feeling short of breath as pulmonary fibrosis progresses. You may also need oxygen all the time.

Looking after yourself

If you have pulmonary fibrosis, there is also a lot you can do to help yourself by leading a healthy lifestyle. Feeling very tired is a common symptom and health problems that used to be minor - such as catching a cold – can become more serious. Following these tips will help keep your strength up, reduce your risk of complications and help you feel better generally:

- Have a flu jab each year, and avoid being around people with colds.
- Ask your nurse for a pneumococcal vaccination. This is a jab you have just once to protect against pneumonia and many other infections.
- Stay as fit as you can. It's recommended we all do 150 minutes of exercise each week. Find out more at blf.org.uk/exercising
- Eat a healthy, balanced diet and maintain a healthy weight. It's a good idea to ask your doctor or nurse if they can refer you to a dietician, who can give you tailor-made advice.

Emotions and enjoying life

As well as taking good care of your body, it's important to look after your mental health. When you have a serious condition like pulmonary fibrosis, it's common to have emotional struggles too. 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 Breathe Easy patient group or pulmonary fibrosis support group. Have a look at **blf.org.uk/BreatheEasy** or call our helpline.

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IPF
Idiopathic pulmonary fibrosis

What is IPF?

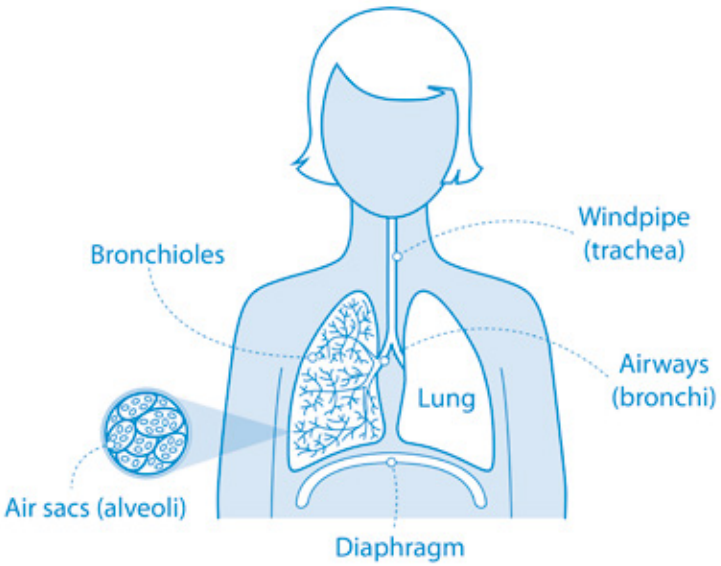
Idiopathic pulmonary fibrosis (IPF) scars your lungs and so reduces the efficiency of your breathing. The build-up of scar tissue is called fibrosis. Fibrosis causes the lungs to become stiffer and lose their elasticity so they're less able to inflate and take oxygen from the air you breathe.

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.

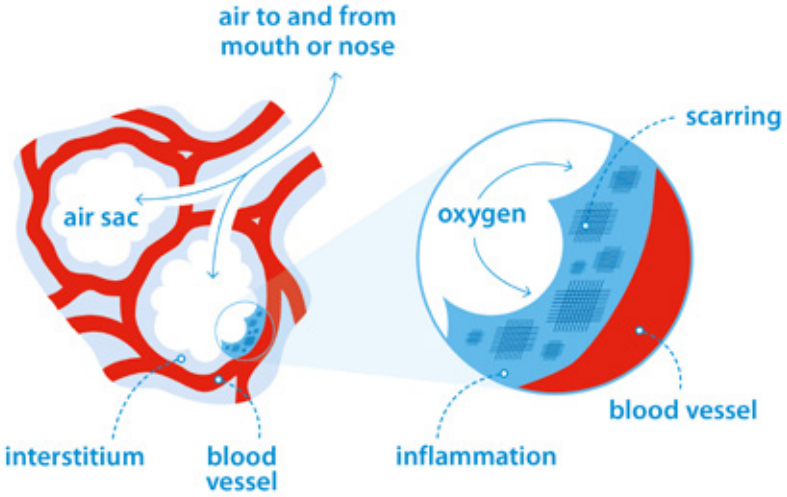
How does IPF affect breathing?

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**

British Lung Foundation
73-75 Goswell Road
London EC1V 7ER

Registered charity in England and Wales (326730), Scotland (038415) and the Isle of Man (1177)

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The Information Standard Certified Member

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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**

Hypersensitivity pneumonitis

formerly called extrinsic allergic alveolitis

This condition happens when your lungs develop an immune response – hypersensitivity – to something you breathe in which results in inflammation of the lung tissue – pneumonitis.

One example is **farmer's lung**. This is caused by breathing in mould that grows on hay, straw and grain. Another is **bird fancier's lung**, which is caused by breathing in particles from feathers or bird droppings. There are many other substances which can cause similar disease patterns. It can be very difficult to find the exact cause.

Symptoms

The symptoms, including cough, shortness of breath and sometimes fever and joint pains, can come on suddenly after you've been exposed. This is the acute form of the condition. It goes away without leading to fibrosis of the lung if you can permanently avoid the substance that caused the attack.

In other cases, symptoms of breathlessness and cough may only appear more gradually, perhaps over many years, as a result of permanent scarring of the lungs. This form of the condition is called chronic, or long-term, hypersensitivity pneumonitis and often a specific cause cannot be identified.

Treatment

If a specific cause is identified, it is really important to completely avoid exposure to it.

You may need to take anti-inflammatory medication called steroids for a few weeks or months.

If you need steroids to control the condition for longer, your doctor may recommend additional drugs to reduce the risk of steroid associated side effects.

Further information

Hypersensitivity pneumonitis is a type of pulmonary fibrosis. You can find more information about pulmonary fibrosis in the following downloads:

- *Pulmonary fibrosis – What it is and how it affects your breathing*
- *Pulmonary fibrosis – Diagnosis and tests*
- *Pulmonary fibrosis – Treatment and support*

"I may never know what's causing my condition"

Jane, 61, was first diagnosed with hypersensitivity pneumonitis 10 years ago

"When I moved to London, I developed a cough. It got so bad I ended up in hospital and was diagnosed with hypersensitivity pneumonitis. I took high dose steroids – and my symptoms disappeared! For seven years I had no symptoms at all.

But when the symptoms came back they didn't go away. So now my hypersensitivity pneumonitis is long-term. Tests showed I have a hypersensitivity to pigeon and budgie droppings, but my doctor says there are thousands of other things that I could be reacting to. I may never know what's causing my condition.

I get unpleasant bouts of coughing. And a small things makes me tired – like carrying shopping home.

I found my work as a lawyer more and more difficult. My employers suggested I claim on their permanent health insurance policy. My claim was accepted and I'm now on long-term sick leave.

I take steroids and immune-suppressing drugs every day. I'll probably take drugs for the rest of my life."

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Occupational interstitial lung diseases

Pneumoconiosis is a term for a group of lung diseases caused by breathing in specific dusts in your workplace. They get lodged inside your lungs and cause scarring.

The most common type is coal worker's pneumoconiosis, caused by breathing in coal dust. Other forms are silicosis, caused by exposure to silica dust and asbestosis, caused by breathing in asbestos.

There is often a long delay (20 years or more) between breathing in the dust and showing symptoms, so new cases usually reflect past working conditions. You'll often be retired before you're diagnosed.

Symptoms

Symptoms may include:

- shortness of breath
- persistent cough
- tiredness
- difficulty breathing
- chest pain
- coughing up black phlegm (coal worker's pneumoconiosis only)

Treatment

The main treatment is avoiding the dust or fumes causing the condition. There are no specific drug treatments. Oxygen therapy and pulmonary rehabilitation may help with your symptoms.

If you have been exposed to certain substances in the course of your work, you may be entitled to compensation or benefits. Call our helpline to find out more.

Further information

Pneumoconiosis is a type of pulmonary fibrosis. You can find more information about pulmonary fibrosis in the following downloads:

- *Pulmonary fibrosis – What it is and how it affects your breathing*
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- *Pulmonary fibrosis – Treatment and support*

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Pulmonary fibrosis associated with connective tissue and autoimmune diseases

For reasons that we don't understand fully, sometimes the immune system turns against the body. This is known as **autoimmune disease**. When your immune system attacks your body's own connective tissues, they scar.

Connective tissues lie under the surface of your skin and around your internal organs and blood vessels. If autoimmune diseases, including rheumatoid arthritis, Sjögrens syndrome and scleroderma, affect your lungs, it can cause pulmonary fibrosis. Unfortunately some of the drugs used to treat these diseases can also cause interstitial lung disease as a side effect.

Symptoms

It's difficult to say what course your lung disease will take because this depends on many factors, including the particular form of autoimmune disease you have, how severe it is and the way it is affecting your lungs. Some people live just a few years after their diagnosis. But other people survive much longer. Talk to your doctor about your individual situation.

If you or your family are facing an emotional time, call our helpline for confidential one-to-one advice and support from our dedicated team.

Treatment

In addition to treatment of your lung symptoms, the best possible management of your underlying condition is essential to protect your lungs from more damage. You'll usually be treated with immunosuppressant drugs tailored to your specific condition.

Further information

You can find more information about pulmonary fibrosis in the following downloads:

- *Pulmonary fibrosis – What it is and how it affects your breathing*
- *Pulmonary fibrosis – Diagnosis and tests*
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Drug-induced pulmonary fibrosis

Any medication can have side effects; some medicines can damage the lungs and cause pulmonary fibrosis. You and your doctor have to weigh up the risks and benefits before you start a medicine. Sometimes the choices are difficult, especially when it's a life-saving treatment.

Some of the commoner medication types that are known to carry the risk of pulmonary fibrosis include certain:

- cancer chemotherapy drugs
- drugs for heart conditions, particularly amiodarone
- immunosuppressant drugs such as methotrexate
- antibiotics, particularly nitrofurantoin
- biological agents, used to treat cancer or immune disorders

Some recreational drugs can also cause pulmonary fibrosis.

The situation varies for each individual and for each drug. Breathing problems from drug-induced pulmonary fibrosis can come on suddenly, or develop more slowly over time.

If a drug has caused the fibrosis, people often get better quickly if the medication is stopped before much damage is done. So identifying this problem, and stopping the drug is the key intervention. Unfortunately, some people have lasting lung damage.

Treatment

Your doctor will stop you taking the drug causing fibrosis. Steroid medication can help calm down your body's response to the medication.

Further information

You can find more information about pulmonary fibrosis in the following downloads:

- *Pulmonary fibrosis – What it is and how it affects your breathing*
- *Pulmonary fibrosis – Diagnosis and tests*
- *Pulmonary fibrosis – Treatment and support*

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