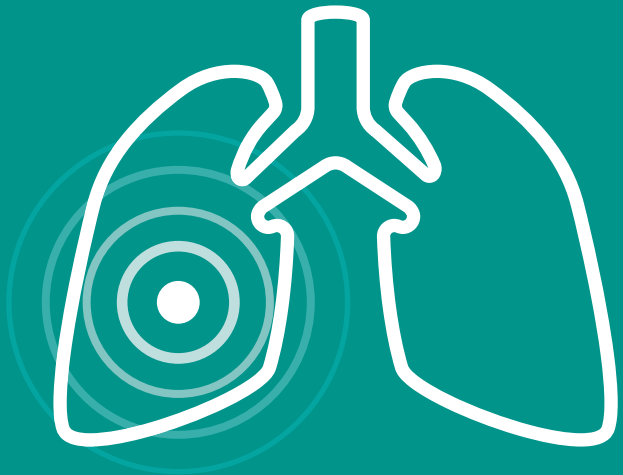
Chest Heart & Stroke Scotland

IDIOPATHIC PULMONARY FIBROSIS



ESSENTIAL GUIDE

This Essential Guide is about idiopathic pulmonary fibrosis.

It explains:

- What idiopathic pulmonary fibrosis (IPF) is
- How IPF is diagnosed
- How to manage and treat IPF
- Where to find support and further information about idiopathic pulmonary fibrosis

What is idiopathic pulmonary fibrosis?

Pulmonary fibrosis is scarring in the lungs. This means the tissue in your lungs is thicker and less flexible, making it harder for oxygen to get from your lungs into your bloodstream.

Pulmonary fibrosis can be caused by many things, like: infections, smoking, medication, asbestos, or a dusty or smoky environment. Fibrosis caused by these things usually gets better once the cause is dealt with.

However, sometimes there is no clear cause, and the condition continues to get worse over time. This is called **idiopathic pulmonary fibrosis.**

Idiopathic	no clear cause
Pulmonary	to do with the lungs
Fibrosis	thickening or scarring

Symptoms of IPF

Symptoms usually develop slowly over time. They may not be noticeable until the disease is well established. The main symptoms are:

Breathlessness
Difficulty breathing after exercise or exertion. This often gets worse as the disease progresses, and you may be breathless even at rest.

Cough
A dry, tickling cough that doesn't go away.

Fatigue
Tiredness and lack of energy.

Clubbing of fingers and toes
The tips of your fingernails and toenails can change shape over time, making them appear more rounded.

What does IPF do?

Pulmonary fibrosis affects your **alveoli**, the sacs in your lungs which transfer oxygen into the blood.

There are millions of alveoli in your lungs, surrounded by tiny blood vessels called **capillaries**. Oxygen passes through the thin walls of the alveoli into the capillaries. Once in the blood, it can be carried to the rest of your body. Carbon dioxide passes the other way, from the blood to the lungs, to be breathed out.

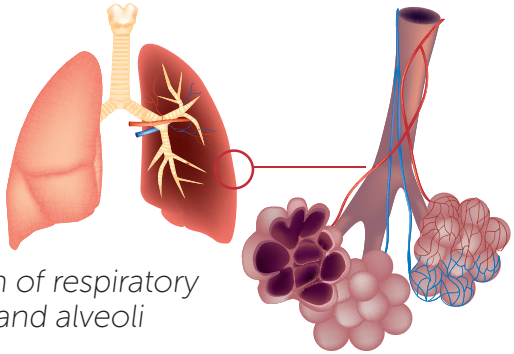


Diagram of respiratory system and alveoli

Who gets IPF?

IPF is increasingly common. Around 32,500 people in the UK have it.

IPF is more common in men than women, and most often occurs in people over 50. It can occur in younger people too, especially if there is a family history of pulmonary fibrosis.

In general, if people in your family have had fibrosis, it is much more likely that you will too. This is called **familial IPF**.

Smoking makes IPF more likely.

People with acid reflux (heartburn) are more likely to develop IPF. Some viral infections can also trigger IPF.

Exposure to some jobs or long-term medications can increase your risk.

In some cases, it is caused by a combination of these factors.

How is IPF diagnosed?

IPF can be difficult to diagnose, mainly because the symptoms are similar to other lung diseases like asthma or chronic obstructive pulmonary disease (COPD).

As a result, IPF is sometimes mistaken for another illness. You should always feel able to ask for a second opinion if you are unsatisfied with your diagnosis.

Your healthcare provider may not be familiar with IPF, as it is a rare condition. Some people with fibrosis find that it is helpful to do their own research. Links for this are at the end of this booklet.

If your doctor or nurse practitioner suspects that you may have IPF, you will usually be referred to a chest specialist for a diagnosis.

Your chest specialist may ask you about:

- Your symptoms.
- Past and present jobs.
- Any hobbies or pets you have.
- Whether you smoke, or used to smoke.
- Whether anyone in your family had similar symptoms.
- Prescribed or over-the-counter medicines you regularly take.
- Your general health and wellbeing.

They may also ask you to undergo some medical tests.

Tests for IPF

You may be asked to have some or all of the following tests and investigations:

Listening to your chest

The doctor may use a stethoscope to listen to your breathing. People with IPF often have a crackling sound when they breathe, "like velcro being pulled apart".

Blood and urine tests

These can rule out other conditions that might cause your symptoms.

Spirometry

You will breathe into a machine which measures how much air you can breathe out in one breath.

Gas transfer tests

These measure how well your lungs pass gases in and out of your blood.

Chest X-ray and/or CT scan

Imaging of your lungs may show scarring in your lungs. CT scans will give a more detailed image, and may show "honeycomb" scarring - lots of small bubbles in the lung.

Echocardiogram

This is an ultrasound scan of your heart, which may be used to rule out heart failure and other heart problems that can cause breathlessness.

Bronchoscopy

A thin, flexible tube with a camera at the end is passed into your lungs. This allows a doctor to look at the inside of your lungs.

Lung biopsy

A sample of lung tissue is taken for examination under a microscope. This can show changes to the cells.

Bronchoscopy

Bronchoscopy uses a thin, flexible tube to look at the inside of your lungs and take cell samples for analysis. This tube will be put into your mouth or nose. This is usually done under a local anaesthetic, so you are awake while it happens.

Bronchoscopy can be uncomfortable, but does not last long, and is safe overall.

Lung Biopsy

A lung biopsy is a surgical procedure to take a sample of your lung tissue. This is done under general anaesthetic, so you will be asleep during the procedure.

There are some risks associated with lung biopsy, which your doctor should discuss with you beforehand.

What does having IPF mean for me?

At the moment, there is no cure for IPF, but you can slow its progress. You will need ongoing treatment to manage symptoms.

IPF reduces your life expectancy, you may want to access support with preparing for and coming to terms with death. However, you can still live a full and happy life while managing your IPF.

Symptoms of IPF usually become worse over time. This can happen at different speeds for different people. Some people experience long periods where their symptoms are stable. Others may see a more rapid decline.

IPF can also make you more at risk of a number of other illnesses, including: chest infections, high blood pressure, heart failure, and pulmonary embolism.

Managing IPF

The management of IPF is based on:

- Controlling symptoms and improving quality of life.
- Slowing down the disease's progress.
- Managing any other conditions you have.
- Providing information and support.

Once you are diagnosed, you should be looked after by a multidisiplinary team. This is a range of healthcare professionals assigned to your case who have expertise in lung disease.

You may want to seek out hospice care, counselling, or other end-of-life support. Ask your GP for local options.

Managing IPF symptoms

Currently there is no cure for IPF and the best treatment has not yet been found. The aim of treatment is to manage individual symptoms.

Breathlessness

People with IPF often have reduced oxygen in their blood. You may find that you struggle to breathe, especially when you are active.

As the disease progresses, people often find that less and less activity is needed to make them feel breathless. Periods of breathlessness may also last longer or be worse as the condition goes on.

If you get out of breath during normal daily activities, or while you are resting, your doctor may recommend:

- Oxygen therapy.
- Pulmonary rehabilitation.
- Exercises to clear your chest or control your breathing.
- Lifestyle changes, such as stopping smoking or keeping active.
- Controlling stress and anxiety.

You can find more information in Chest Heart and Stroke Scotland's booklets on:

Breathlessness
Oxygen Therapy
Pulmonary Rehabilitation
Stopping Smoking

You can order all of these online at **www.chss.org.uk/resources-hub**

Cough

People with IPF often find that coughing can have a large impact on their daily life. This cough may be dry (no phlegm) or productive (meaning there is a lot of sputum or phlegm).

Coughing can cause:

- tiredness or disturbed sleep.
- shortness of breath.
- anxiety.
- dizziness.
- vomiting or incontinence.

Sometimes, there is an underlying cause which can be treated, like acid reflux or asthma. However, even when these conditions are treated, it is rare for cough to completely go away in people with IPF.

Your cough may be worse in cold or wet weather. It can help to use a humidifier or dehumidifier to control the air quality.

You may be offered medications to reduce coughing or how much sputum you produce, for example: opioids and thalidomide. Or medication for acid reflux or post-nasal drip.

If your cough is causing you trouble, ask your doctor about available treatments.

People with IPF have also reported that the following techniques help to reduce their cough. These are based on people's experience and are not necessarily supported by research, but you may find them helpful.

- Using throat lozenges containing a local anaesthetic before speaking.
- Taking sips of iced water.
- Warm water with honey, or herbal tea.
- Breathing techniques like pursed-lip or circular breathing.

Fatigue

Many people with IPF experience tiredness and fatigue. Sometimes, this can be overwhelming and leave you struggling to do everyday activities.

Tiredness may be associated with low oxygen levels in your blood. Your doctor may refer you for an oxygen assessment to see whether you might benefit from oxygen therapy.

You can manage your fatigue to an extent by pacing yourself, managing your energy and activities carefully, and building in time to rest.

You can find more information in Chest Heart & Stroke Scotland's booklet on **Tiredness and Fatigue**.

You can order copies online at **www.chss.org.uk/resources-hub**

Symptom flare-ups

On top of your usual symptoms, you might have flare-ups (exacerbations) of your fibrosis. These should be treated as soon as possible to minimise damage to your lungs.

Contact your specialist team urgently if you notice an increase in:

- how breathless you are
- how often you cough
- how much phlegm you produce

or if your phlegm changes colour or texture suddenly.

Other conditions can also lead to an increase in cough and breathlessness in people with IPF, and will need to be ruled out or treated.

If you see a rapid decline in your symptoms, are very short of breath, or your blood oxygen level drops sharply, you may need to be admitted into hospital for treatment.

Medicines for IPF

You may be offered medications to manage individual symptoms.

For example, you may be given opioids or thalidomide for your cough, or you may be given medication, like antacids, to prevent acid reflux. This can reduce ongoing damage, including protecting your lungs from stomach acid during a coughing fit.

If you have mild to moderate IPF, you may be prescribed a medicine called an **antifibrotic**. However, these medications are not appropriate for everyone with IPF.

Lung transplant

A lung transplant may be an option if you are healthy enough for the operation. However, it is not appropriate for everyone with IPF.

If a lung transplant is an option for you, your doctor or another member of your specialist team will usually discuss it with you within 3-6 months of your diagnosis. Ask any questions you have about the procedure, and make sure you are comfortable with your understanding of the operation.

Whether a transplant is appropriate for you will depend on many factors, including:

- how bad your IPF is.
- how fast your symptoms are worsening.
- your age and general health.
- how much your IPF is likely to improve after transplant.
- donor lung availability.

Follow-up

How often you need follow-up appointments after your diagnosis will depend on how quickly your condition progresses.

If your symptoms are rapidly worsening, you should be seen at least once every three months.

If your symptoms are only getting worse very slowly, you should be seen once every six months.

If your symptoms don't seem to be getting worse at all, you should have appointments every six months. However, if your symptoms continue to be stable for a year or more, you may be able to reduce to an annual appointment.

If you have a flare-up or exacerbation, you should contact your specialist team as soon as possible.

Your follow-up appointments should include some or all of the following:

- Tests of how well your lungs work.
- Assessment for oxygen therapy.
- A referral to pulmonary rehabilitation if appropriate.
- Identifying any episodes where your symptoms got worse, and what may have triggered them.
- Discussing the need for palliative care.
- Assessment and management of any related illnesses.
- Any changes you can make to your lifestyle to keep your symptoms under control.

Check out **chss.org.uk** for more information.

Lifestyle management

There are some things you can do to help keep yourself well and reduce the impact of your symptoms:

Keep up to date with vaccines
Flu and pneumococcal vaccines reduce your chance of getting dangerous chest infections.

Healthy diet and weight
People with IPF often lose weight, so it is important to keep your weight up and eat high-energy foods. A high-protein, low-sodium, low-sugar diet is best for people with IPF.

Stop smoking
Smoking can cause more damage to your lungs, making your symptoms worse. Help quitting is available through your GP or pharmacy.

Keep active

Exercise can become harder as IPF gets worse, but safe exercise helps increase your fitness, stamina, and energy.

Know what to do in a flare-up

Discuss with your support team what you should do if your symptoms get worse quickly. For example: emergency contacts, medications you might need, and warning signs to call for help.

Attend pulmonary rehabilitation classes

Pulmonary rehabilitation is a course of exercises and education which your doctor may refer you to. Regularly attending pulmonary rehabilitation and doing the exercises will strengthen your lungs and may improve your symptoms. For more information, check out the CHSS booklet "**Pulmonary Rehabilitation**".

Palliative care

IPF is a life-limiting condition. As early as you are comfortable with, it can be worth looking into end-of-life care, pain management, and hospice planning.

Getting a plan in place before your symptoms become too severe to handle can reduce stress and make the course of the disease easier on both you and the people around you.

Mental health support

Living with IPF can be stressful and upsetting. Seeing a counsellor, psychotherapist, or support group can help to make your symptoms easier to manage.

Getting involved in research

Many people with IPF are interested in research into the condition. As it is a rare condition, clinical trials are often recruiting people with IPF to take part in studies.

If you feel that this is something you would like to do, you can find clinical trials which are recruiting patients at:
www.eu-ipff.org

You can also find regular research updates through:

ActionPF
www.actionpf.org/news-category/research

Asthma & Lung UK
www.asthmaandlung.org.uk/research-lung-health

Finding information and support

There is help and support available for people with IPF and their families.

Join a support group

To find out whether Chest Heart & Stroke Scotland has a chest support group near you, call the **Advice Line on 0808 801 0899** or visit **www.chss.org.uk**

The **Chest Heart & Stroke Scotland Advice Line** (**0808 801 0899**, or email **adviceline@chss.org.uk**) can offer personalised support and information.

Chest Heart & Stroke Scotland offers a range of booklets on lifestyle changes, symptoms, and treatments. Find these booklets online at: **www.chss.org.uk/resources-hub**

Talk to a health professional if you have questions or concerns about your conditions or treatment.

Some areas have a dedicated **interstitial lung disease (ILD) nurse** who can offer specific support for your IPF.

Friends and family

Be honest with the people around you about your feelings and what you are experiencing. They can often be a valuable source of support.

Hospices

Depending on where you live, hospice charities like Marie Curie, Sue Ryder, or local hospices can offer support to you and your family when dealing with end-of-life care and bereavement.

Action for Pulmonary Fibrosis
A charity which promotes research into pulmonary fibrosis and provides support for people with IPF or related conditions.
www.actionpf.org
01733 475642
Email: info@actionpf.org

Asthma + Lung UK
A UK-wide charity offering information and support aroung lung and respiratory disease.
www.asthmaandlung.org.uk

NHS Inform
The central NHS Scotland website, which includes detailed information on IPF.
www.nhsinform.scot/illnesses-and-conditions/lungs-and-airways/idiopathic-pulmonary-fibrosis

Breathing Matters
A research network focusing on lung disease, including IPF. **www.breathingmatters.co.uk**

My support team's details

Use this page to jot down any contact details for your healthcare team.

Our publications are free to everyone in Scotland, in PDF and in print. See them all at **www.chss.org.uk/resources-hub**

For free, confidential advice and support from our Advice Line Team, contact:

0808 801 0899 (Mon-Fri 9am-4pm)
text ADVICE to 66777
adviceline@chss.org.uk

One in five people in Scotland are affected by chest, heart and stroke conditions or Long Covid. Go to **www.chss.org.uk/supportus** to find out how you can help us support more people in Scotland.

To give feedback or request alternative formats, email: **health.information@chss.org.uk**

Chest Heart & Stroke Scotland

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