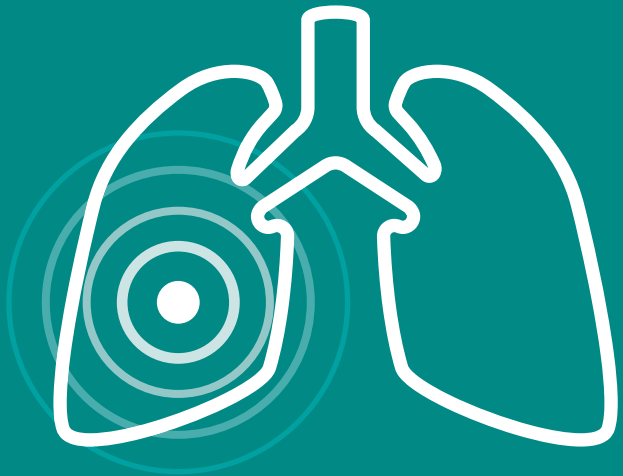
Chest Heart & Stroke Scotland

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 a problem where the tissue in your lungs is thickened and scarred. This makes your lungs less flexible and makes it more difficult for oxygen to get from your lungs into the bloodstream.

Pulmonary fibrosis can be caused by a lot of things - dust, infections, smoking, medication - but sometimes it isn't clear what causes the damage. When this is the case, it is called **idiopathic** pulmonary fibrosis.

Idiopathic	no obvious 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 of IPF are:

Breathlessness
Difficulty breathing after exercise or exertion, which often gets worse as the disease progresses.

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

Fatigue
Tiredness and lack of energy.

Clubbing of fingers and toes
Thickened tissue at the bottoms of your fingernails and toenails. This usually comes later in the disease.

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

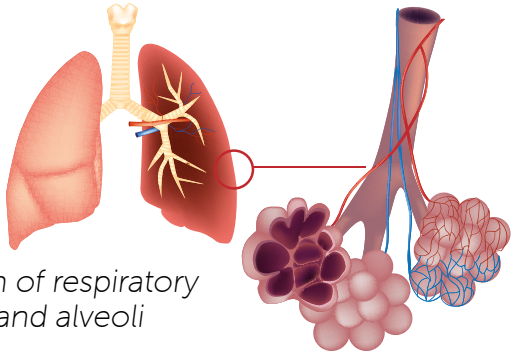


Diagram of respiratory system and alveoli

Who gets IPF?

IPF is increasingly common. Around 32,500 people in the UK have it. Around 2,500 people in Scotland are living with IPF.

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 misdiagnosed as something else. 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 identify patterns of scarring in your lungs. CT scans will give a more detailed image, and may show classic "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 forexamination under a microscope.

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 throughout.

Bronchoscopy can be uncomfortable or painful, 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, although it can be managed.

People with IPF have a reduced life expectancy and usually need ongoing treatment. However, you can still live a full life while managing IPF.

Symptoms of IPF often become worse over time, although this can happen at different speeds for different people. Some people will experience long periods where their symptoms are stable and there is little decline in their conditions. 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 rate at which symptoms get worse
- Managing any other conditions you may have
- Providing information and support

Once you are diagnosed, you should be looked after by a multidisciplinary team - a range of healthcare professionals assigned to your case - with expertise in lung disease.

This may include pathologists, surgeons, specialist nurses and doctors, lung technicians, radiologists, and/or physiotherapists.

Managing IPF symptoms

Although IPF does not currently have a cure, there are steps you can take to reduce your 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/publications**

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 (producing 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, such as acid reflux or asthma. However, even when these conditions are treated, it is rare for cough to completely go away in people with IPF.

You may also find that your cough is worse in cold or wet weather.

You may be offered medications to reduce coughing or how much sputum you produce, particularly:

- Steroids
- Opioids
- Thalidomide

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

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 **Fatigue**.

You can order copies online at **www.chss.org.uk/publications**

Symptom flare-ups

On top of your usual symptoms, you might experience 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 treating IPF

Although there is not yet a cure for IPF, and there are no medications which are used to treat severe IPF directly, you may still be offered medications to manage individual symptoms.

For example, you may be given steroids, opioids, or thalidomides for your cough, or you may be given medication to prevent acid reflux. This can reduce ongoing damage.

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. Your doctor will be able to tell you if this is suitable for you.

Lung transplant

A lung transplant may be an option if you are physically 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 other 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 to have follow-up appointments after your diagnosis will depend on how quickly your condition is worsening.

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.

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 chest infections which can be dangerous when you have IPF.

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
If you smoke, this 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 when you have 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 ensuring you do 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.blf.org.uk/research

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 which you may be experiencing.

You can order these online: **www.chss.org.uk**

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 and 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 available for free to anyone in Scotland who needs them. Go to **www.chss.org.uk/publications** for all our resources, including other Essential Guides in this series.

For free, confidential advice and support from our **Advice Line nurses**, call: 0808 801 0899 (Mon-Fri 9.30am-4pm), text: NURSE to 66777 or email: adviceline@chss.org.uk.

Across Scotland, over one million people – that's one in five of us – are living with the effects of a chest, heart or stroke condition. We are here to help everyone who needs us. But we need your support to do this. Go to **www.chss.org.uk/supportus** to find out how you can help more people in Scotland.

If you would like this resource in an alternative format, please contact our Advice Line nurses.

Chest Heart & Stroke Scotland

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