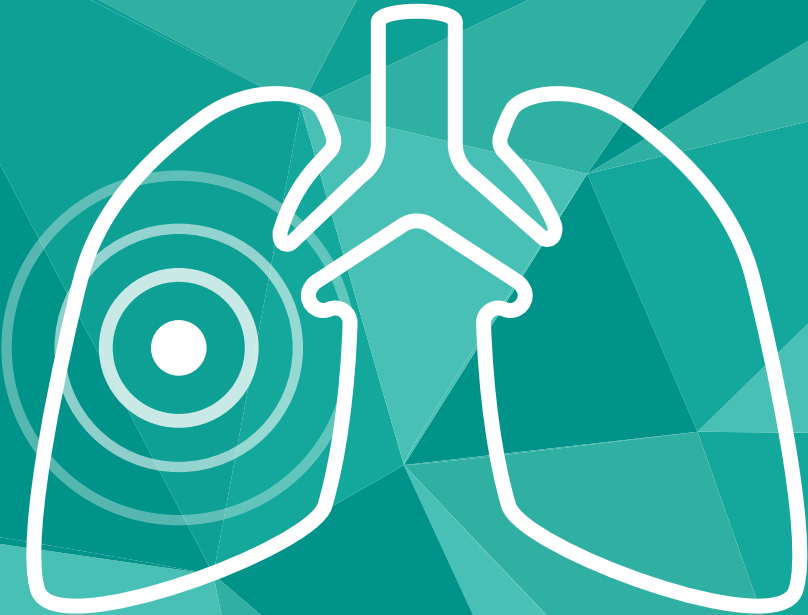


# LIVING WITH IDIOPATHIC PULMONARY FIBROSIS





**NO LIFE HALF LIVED**

The information contained in this booklet is based on guidelines and practice and is correct at time of printing. The content has undergone peer, patient and expert review.

# LIVING WITH IDIOPATHIC PULMONARY FIBROSIS

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# ABOUT THIS BOOKLET

## Who is the booklet for?

The booklet is for people who have idiopathic pulmonary fibrosis (IPF). You may also find it useful if you are a friend, relative or carer of someone who has IPF.

## What is this booklet about?

The booklet explains what IPF is, its causes and symptoms. It explains how IPF is diagnosed and managed and what you can do to help control your symptoms and keep yourself well. The booklet also addresses some of the concerns you may have about living day-to-day with IPF and offers practical advice to help you continue to do the things that you enjoy.

## How to use the booklet

The booklet is divided into different sections. These are outlined in the contents list. You do not have to read all of the sections at once. Some sections may be useful to you now, other sections you may want to read at a later time.

## Further information

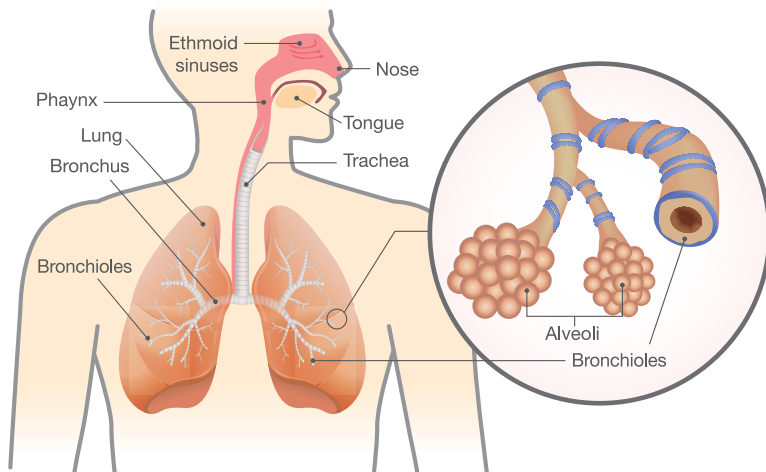
If you would like more information, we have a range of easy-to-read 'essential guides' on a number of different topics discussed in the booklet, for example, breathlessness, tiredness and fatigue and stopping smoking. To see the full list of guides and other resources and to order, view or download them, go to **[www.chss.org.uk/publications](http://www.chss.org.uk/publications)** or call the Advice Line nurses for more information on 0808 801 0899.

# UNDERSTANDING IPF

## How do the lungs work?

In normal healthy airways, air travels into the lungs through the **trachea** (windpipe). The trachea divides into branches to supply your left and right lungs. These branches are called the left bronchus and the right bronchus. These **bronchi** divide further and branch into smaller and smaller air passages called **bronchioles**, which end in millions of tiny air sacs called **alveoli**. Around these air sacs are tiny blood vessels called **capillaries**.

Oxygen from the air that you breathe passes through the thin wall of the alveoli into the capillaries, where it attaches to the red blood cells. The oxygen is then carried through the blood vessels to the rest of the body. At the same time, carbon dioxide (a waste product) passes from the capillaries into the alveoli and is breathed out.



## What is pulmonary fibrosis?

**Pulmonary fibrosis** is scarring and thickening of the cells that line the alveoli in the lungs.

In people with pulmonary fibrosis, the cells that line the alveoli become damaged. It is thought that the cells then try to heal themselves, but the healing process gets out of control causing thickening and damage to the walls of the alveoli and surrounding lung tissue. As the lung tissue becomes scarred and thicker, it becomes more difficult for the oxygen to pass from the alveoli into the bloodstream.

There are a number of different causes of pulmonary fibrosis including:

- Diseases such as rheumatoid arthritis
- Previous lung infections such as pneumonia and tuberculosis
- Environmental dusts such as asbestos and silica, some gases, birds, moulds and many other irritants
- Exposure to radiation such as that used to treat tumours of the chest
- Some medications, including amiodarone, nitrofurantoin and bleomycin

For some people, however, there is no known or identified cause – this is known as **idiopathic pulmonary fibrosis** or **IPF**.

**IDIOPATHIC** = of unknown cause

**PULMONARY** = of the lungs

**FIBROSIS** = thickening or scarring

Although the exact cause of IPF is not known, there are several possible triggers that may start the initial damage to the cells that line the alveoli. These include:

- Being a smoker or an ex-smoker
- Some viral infections
- Heartburn (acid reflux)
- Family history – IPF seems to run in some families, but most of the time there is no family history

## Who gets IPF?

- ✓ IPF is increasingly common. There are about 3,550 people in Scotland affected by it.
- ✓ IPF is more common in men than in women.
- ✓ IPF usually occurs in people over 50 years, but occasionally it occurs in younger people, especially if there is a family history.
- ✓ IPF is more common in people who smoke.



## What are the symptoms of IPF?

Symptoms usually develop gradually and may not be noticed until the disease is well established.

The main symptoms are:



- **Feeling breathless.** At first you may feel more breathless than usual doing strenuous exercise, such as walking uphill or walking up the stairs. As the disease advances breathlessness often gets worse, making everyday activities such as washing and dressing more difficult.



- **Cough.** Usually a dry, tickly cough that won't go away.



- **Fatigue.** Feeling more tired than usual.

You may also notice 'clubbing' of your fingers and toes. This is a thickening of the tissue at the bottom of your fingernails or toenails. Clubbing occurs in about half of all people with IPF, but it usually occurs later in the course of the disease.



# DIAGNOSIS

Idiopathic pulmonary fibrosis (IPF) can be difficult to diagnose, mainly because the symptoms are similar to other more common long-term lung diseases such as asthma and chronic obstructive pulmonary disease (COPD). These diseases can exist in addition to IPF, making diagnosis even more complicated.

## How is IPF diagnosed?

If your GP suspects you may have IPF, you will usually be referred to a chest (respiratory) specialist in hospital for diagnosis.

You may be asked about the following:

- Your symptoms – in particular cough and breathlessness
- Past and present jobs
- Any hobbies or pets you have
- Whether you currently smoke, or used to smoke
- Whether you have a family history of chest disease
- Any prescribed or over-the-counter medicines that you take
- Your general health

If the doctor suspects that you have IPF you will usually have some or all of the following tests and investigations:

- The doctor may listen to your chest with a stethoscope
  - often crackles can be heard in your lower lungs that suggest you may have IPF.
- Blood and urine tests to rule out other diseases
- Lung function tests:
  - Spirometry measures how much air you can breathe out in one breath.
  - Gas transfer tests measure how well your lungs can pass gases into and out of your blood.
  - To find out more about spirometry and gas transfer tests, visit <http://mylungsmylife.org>.
- Chest x-ray may identify a particular pattern of scarring in your lungs.
- High resolution computerised tomography (CT scan). A CT scan takes a series of x-rays and puts them together using a computer, to create a detailed image of the inside of your body. Classic findings in IPF show scarring of lungs with small bubbles that have a characteristic 'honeycomb' appearance.
- An echocardiogram (ultrasound examination of your heart) may be done if your doctor thinks that you may have developed heart failure.

Often a diagnosis of IPF can be made from your symptoms, history and the results of these tests and investigations. Sometimes, however, more tests may be recommended to confirm the diagnosis. This might be either a **bronchoscopy** or occasionally a **lung biopsy**.

**Bronchoscopy** allows the doctor to look at the inside of your lungs:

- A thin flexible tube is passed through your nose or throat down into your airways.
- The doctor can look through the tube for anything unusual, and can take cell samples to be tested.
- This procedure is usually done using a local anaesthetic, so you are awake throughout. Your nose and throat will be numbed with an anaesthetic spray. It may feel a bit uncomfortable but the procedure does not last long.

A **lung biopsy** allows the doctor to take a sample of lung tissue for examination under a microscope.

- Lung biopsy is performed under general anaesthetic, so you are asleep during the procedure.
- Small cuts are made in the side of your chest and a thin tube with a camera on the end is passed into the area between your chest and your lungs.
- Small samples of lung tissue are taken for examination.
- Once the procedure is finished, the tube is removed and the cuts are closed, usually with stitches.

There are some risks associated with having a lung biopsy. These include:

- A partial collapse of your lung (occurs in about 15 out of every 100 people)
- Severe bleeding (occurs in about 1 out of every 100 people)
- Infection (uncommon, can usually be treated with antibiotics)

Your doctor should discuss with you the benefits of getting a correct diagnosis and the possible risks of having a lung biopsy.



## What does a diagnosis of IPF mean?

The symptoms of IPF will often become worse over time. The rate at which symptoms get worse varies from person to person. Some people with IPF will experience long periods where their symptoms are stable and there is little decline in their condition. For others there may be a more rapid decline and worsening of symptoms.

At the moment there is no cure for IPF. Whilst the average length of survival after diagnosis is between 3 and 5 years, many people survive longer than this.

Having IPF can also make you more at risk of a number of other illnesses. These include:

- Chest infections, such as bronchitis, pneumonia and influenza
- Pulmonary hypertension (high blood pressure in the arteries that carry blood from the heart to the lungs)
- Heart failure
- Pulmonary embolism (a blood clot in your lung)

# MANAGEMENT OF IPF

Currently there is no cure for idiopathic pulmonary fibrosis (IPF). The management of IPF is based on:

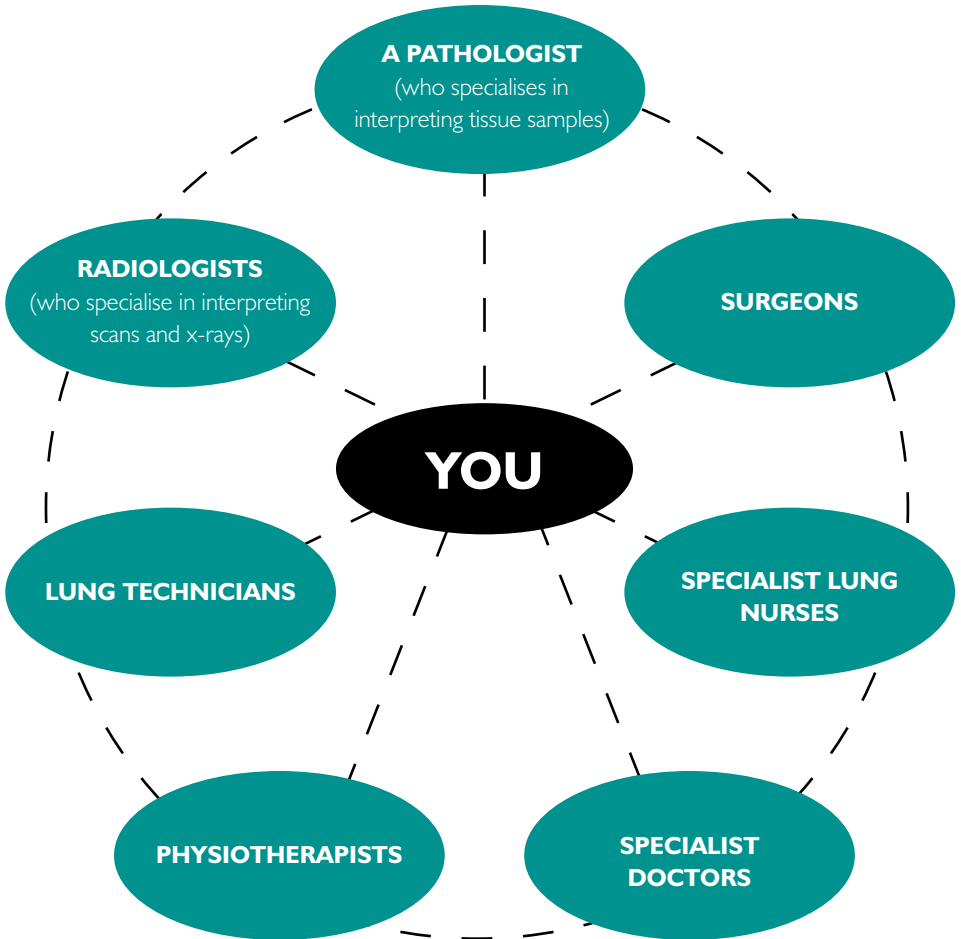
- Controlling symptoms and improving quality of life
- Slowing down the advance of the disease
- Managing other conditions that exist alongside IPF
- Providing information and support

The treatment you receive will depend on the severity of your illness, how quickly it is advancing, and your own personal preferences. Management may also include stopping treatments that are not working or are causing you side effects.



## Who will be involved in my care?

You should be looked after by a multidisciplinary team (a whole range of healthcare professionals) with expertise in lung disease. The team might include:





## How are the symptoms of IPF managed?

### Breathlessness

If you get out of breath just doing normal everyday activities or while you are resting, your doctor may recommend that you are assessed for:

- Pulmonary rehabilitation
- Oxygen therapy

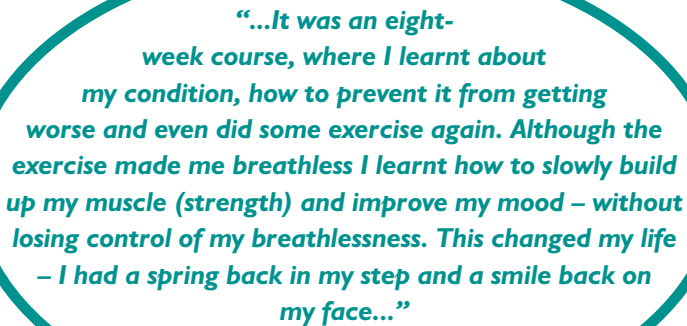
There are also a number of things that you can do yourself to help manage your breathlessness. These include:

- Breathing control techniques
- Chest clearance techniques
- Stopping smoking
- Keeping active
- Learning to control stress and anxiety

## **Pulmonary rehabilitation**

Pulmonary rehabilitation (PR) is recommended for most people with IPF. When you are diagnosed with IPF, your specialist should offer you an assessment to see if you would benefit from PR. If PR could help you, you should be referred to a local programme.

Pulmonary rehabilitation is a structured programme of exercise and education for people with chronic chest conditions. PR is designed to improve your level of fitness, increase your sense of well being and help you understand and cope better with your condition.



***“...It was an eight-week course, where I learnt about my condition, how to prevent it from getting worse and even did some exercise again. Although the exercise made me breathless I learnt how to slowly build up my muscle (strength) and improve my mood – without losing control of my breathlessness. This changed my life – I had a spring back in my step and a smile back on my face...”***

Pulmonary rehab patient

## Oxygen therapy

People with IPF often have lower levels of oxygen in their blood, and this can cause symptoms of breathlessness and fatigue. Oxygen therapy may therefore be recommended to increase the levels of oxygen in the blood, helping to relieve symptoms. Oxygen will only help you if you have low levels of oxygen in your blood.

If your doctor thinks that you will benefit from oxygen therapy, he will send you for an oxygen assessment.

In some people oxygen levels only fall when they are physically active; these people will only need to use oxygen when they are active. This is called ambulatory oxygen.

Other people need oxygen even when they are resting. This will be delivered through an oxygen concentrator that you will have at your home.

## Cough

In people with IPF, cough can have a large impact on daily life. Its effects include tiredness, shortness of breath, sleep disturbance, anxiety, dizziness, vomiting, incontinence and social exclusion. However, there are things that can be done to minimise your cough and the effect it has on you.

Sometimes there is an underlying cause of the cough which can be treated. Such causes include acid reflux and asthma. Despite managing these other conditions, it is rare for cough to completely go away in people with IPF. Other treatments that have been shown to reduce cough include:

- Steroids
- Opioids
- Thalidomide

If your cough is troublesome, ask your doctor about available treatments.

Below are some suggestions from other people with IPF for things that have helped to relieve their cough. These are anecdotal reports only and are not supported by research, but you might find that they help you.

- Using throat lozenges containing a local anaesthetic before speaking
- Taking sips of iced water
- Warm water with honey
- Herbal tea
- Breathing techniques such as pursed-lip breathing

When you cough, the pressure on your bladder increases suddenly. For many people this is not a problem, but for some people, the combination of a persistent severe cough and weak bladder muscles can mean that urine leaks out from the bladder.

Although this can be embarrassing to talk about, speak to your GP or a member of your healthcare team, as there are a number of things that can help. These include:

- Losing weight if you are overweight
- Avoiding caffeine-containing drinks (such as tea and coffee) and alcohol
- Pelvic floor exercises
- Bladder training

There are also a number of products you can use such as absorbent pads to give you protection. These are available through your GP or practice nurse.

Practical help and advice can also be given by a continence advisor – ask your GP for more information and a referral if appropriate.

## Fatigue

Many people with IPF experience tiredness to some degree. Sometimes it can feel overwhelming and leave you with little energy for everyday activities. Tiredness can be associated with low oxygen levels in your blood, and your doctor may refer you for an oxygen assessment to see whether you might benefit from oxygen therapy.

Sometimes (although not always) tiredness can be related to disturbed sleep. If you are having trouble sleeping, try the following:

- Establish fixed times for going to bed and waking.
- Try to relax before going to bed – have a warm bath, take a hot milky drink.
- Make your sleeping environment as comfortable as possible: not too hot, cold, noisy, or bright.
- Try not to have drinks containing caffeine or alcohol in the evening.
- Avoid eating a heavy meal late at night.

There are tips and techniques you can adopt to make everyday activities less demanding on your energy, such as:

- Organising your space so that items you need for a particular task (such as washing and dressing) are close at hand.
- Learn to pace yourself; give yourself enough time to do things and build in rests.
- Don't be tempted to overdo it on a good day as you may be overtired the next day.

## Are there any medicines for treating IPF?

Unfortunately, there is not yet a cure for IPF, but there are now medicines available that can slow the advance of the disease. Two medicines, pirfenidone and nintedanib, are common treatments in Scotland for people with mild to moderate IPF. However, they are not suitable for everyone and the benefits of treatment need to be balanced against the possible side effects. Your hospital doctor or nurse can provide you with detailed information about this.

Acid reflux is common in people with IPF. If you have any symptoms of acid reflux, you may be offered treatment with a medicine to reduce the amount of acid in your stomach. Controlling reflux can help limit the damage to the cells in the alveoli and help to improve lung function.

Research into the management of IPF is ongoing. You might want to ask your specialist team if there is a trial in your local area for which you might be suitable to be enrolled. However, there is never any pressure to enrol; participation is entirely voluntary.

***“I volunteered to take part in a 3-year clinical trial for a new drug in 2011. Unfortunately I began to suffer from some side effects which meant that I was withdrawn from the trial and the drug. However, a few weeks later I was prescribed a different new drug, which I am still taking with no significant adverse effects.”***

Patient with IPF



## What about a lung transplant?

A lung transplant may be an option for those who are physically healthy enough to undergo the operation. There is some evidence that a lung transplant may improve survival and quality of life in people with IPF.

However, a lung transplant is not suitable for everyone with IPF and there are many factors that need to be considered before this can be recommended.

If your doctor feels this may be an option for you, you will be referred for a lung transplant assessment. The decision to have a transplant will depend on a number of factors including:

- How bad your IPF is
- How quickly it is worsening
- Your age and general health
- How much your IPF is likely to improve after transplant
- Whether a donor lung is available

## What happens if I have a flare-up of my symptoms?

On top of your usual symptoms, you may experience a **flare-up** (or **exacerbation**) of your IPF. It is important that you receive treatment as soon as possible to minimise further 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 (sputum) you produce

The quicker an exacerbation is managed the better the outcome.

There are other conditions that can also lead to an increase in cough and breathlessness in people with IPF and these will need to be ruled out or treated. These include:

- Pneumonia
- Pulmonary embolism (blood clot in your lung)
- Pneumothorax or 'collapsed lung'
- Heart failure

You may need other investigations such as a chest x-ray, CT scan or echocardiography to rule these out.

If you experience a rapid decline in your symptoms, are very short of breath, or the level of oxygen in your blood drops significantly, you may need to be admitted into hospital for treatment. There is little research to guide treatment of an exacerbation, but you will probably be treated with one or more of the following:

- An antibiotic, in case the exacerbation is due to infection
- A steroid, to reduce inflammation
- An anticoagulant, such a warfarin, if it is possible that your symptoms are due to a blood clot
- Start or increase oxygen therapy
- An urgent lung transplant if appropriate



## What follow-up should I expect?

How often you need to have follow-up appointments will depend on how quickly your condition is declining. If your symptoms are rapidly getting worse, you should be seen at least once every three months. If your symptoms are more stable you may be seen about once every six months.

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

- Tests to see how well your lungs are working
- An assessment to see whether you would benefit from oxygen therapy
- A referral to pulmonary rehabilitation (if appropriate)
- Assessment for related illnesses that need to be managed
- Advice to help you stop smoking (if needed)
- Identifying any episodes when your IPF got worse, and trying to find out why
- Discussing the need for palliative care

## WHAT CAN I DO TO KEEP MYSELF WELL?

- ✓ Stop smoking
- ✓ Keep up to date with your vaccinations
- ✓ Maintain a healthy weight
- ✓ Ask for support if you need it
- ✓ Eat a healthy diet
- ✓ Know what to do if you have a flare up
- ✓ Keep as active as possible
- ✓ Attend pulmonary rehab classes
- ✓ Try to avoid people with colds and chest infections

## Stop smoking if you are a smoker

If you smoke, there is no better time to quit than now! Although smoking is not thought to cause IPF, it can increase the damage to your lungs, making your symptoms worse. It can also increase the risk of other lung diseases.

**You are more than four times as likely to stop smoking if you have specialist support.**

Get some help to stop smoking – speak to your GP, nurse or pharmacist for advice and support, or contact Quit Your Way Scotland (Scotland's national stop-smoking helpline): telephone FREE on 0800 84 84 84 or visit [www.canstopsmoking.com](http://www.canstopsmoking.com).



## Keep up to date with your vaccinations

Chest infections such as influenza and pneumonia can become severe if you have lung disease. Try to avoid direct contact with people who have colds, flu or chest infections and make sure that you have the following vaccinations:

- Flu vaccine – you should have this every year. It is usually available from late September.
- Pneumococcal vaccine – most people only need to have this once, although certain people may need it every year.

## Keep as active as possible

As IPF gets worse, it may become more difficult to do daily activities like walking or climbing stairs. However, it is important that you do your best to stay as active as possible. Keeping physically active will increase your general fitness and stamina and give you more energy. It will also help to improve low mood and anxiety.

What form of activity you do will depend on your individual condition. Gradually try to build up the amount of activity you do, but make sure you rest too. Although you may not feel like it, try to keep up your favourite activities, adapting how you go about them if necessary.

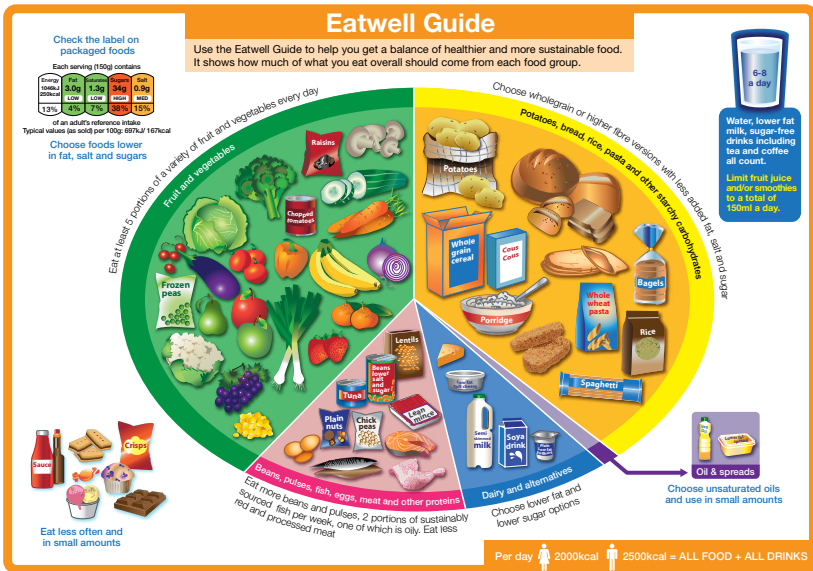
If it is recommended that you attend pulmonary rehabilitation, try your best to do so. You might also want to find out about other exercise groups in your area for people with lung disease. To find out if there is a CHSS affiliated community chest support group in your area:

- visit [www.chss.org.uk](http://www.chss.org.uk) or
- call the Advice Line nurses on 0808 801 0899.

# Eat a healthy balanced diet

As long as you are not experiencing loss of appetite and weight loss, a diet that is low in sodium (salt), fat (particularly saturated fat) and added sugar, but high in whole grains and protein, is good for people with IPF.

- Eat plenty of fruit and vegetables.
- Increase the amount of fibre in your diet.
- Reduce the saturated fat in your diet.
- Cut down on sugar.
- Try to eat less salt.



Source: Public Health England in association with the Welsh Government, Food Standards Scotland and the Food Standards Agency in Northern Ireland

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## Try to maintain a healthy body weight

Many people with IPF experience weight loss and loss of appetite; some people find it hard to eat because of their breathlessness, for others eating itself can be exhausting. As a result, you might not be getting enough energy and you may become underweight. If you notice you are losing weight, or you are having trouble eating, speak to your doctor. You may be referred to a dietician who can advise you on your diet and meeting your nutritional requirements.

If you are underweight, here are some tips to help you increase the amount of energy you take in:

- Eat small amounts as often as possible.
- Try not to have drinks just before meals, as this can make you feel too full to eat.
- Avoid low fat or diet versions of foods and drinks. Instead have full-fat milk or cream rather than skimmed or semi-skimmed milk, drink sugary drinks instead of diet drinks, try thick, creamy soups instead of watery soups.
- Add extra energy to your food by adding high calorie ingredients. For example:
  - Add cheese to pasta
  - Add skimmed milk powder to full fat milk
  - Add sugar, jam or honey to cereal or porridge
  - Add butter to mashed potatoes or sauces
  - Add cream to fruit

- Snack on high energy and protein foods throughout the day. This includes cheese, creamy yoghurt, nuts and seeds, chips, chocolate.

Note if you have a health condition that meant you previously had to reduce the amount of fat and sugar in your diet, check with a healthcare professional what is a suitable diet for you.

## Seek support

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

- Join a support group. It can be very helpful to hear how other people with similar lung problems cope. To find out if there is a CHSS affiliated community chest support group in your area, visit [www.chss.org.uk](http://www.chss.org.uk) or call the Advice Line nurses on 0808 801 0899.
- Talk to a health professional if you have questions or concerns about your condition or treatment. Some areas have a dedicated interstitial lung disease (ILD) nurse who can provide specific support for people with pulmonary fibrosis.
- Get support from family and friends. Let the people around you know how you are feeling and how they can help. It can also be helpful for your family and friends to know more about what is happening to you.
- Call the Advice Line nurses. CHSS Advice Line nurses can provide support and information for people with IPF. For confidential advice and information, call FREE on 0808 801 0899.

# LIVING WITH IPF

This section of the booklet aims to address some of the concerns that you might have about living day-to-day with idiopathic pulmonary fibrosis (IPF), and to suggest some tips and strategies to help you live as full a life as possible.

## Coming to terms with your diagnosis

Being diagnosed with a serious illness like IPF can turn your life upside down. It is only natural that you will feel a mixture of emotions including:

- Fear and anxiety – it is natural to be scared about what is happening, and what will happen as your disease advances.
- Worry – uncertainty about the future is very worrying, both for you and your family. You may have concerns about work or money, or you may be worried about loss of independence if your health declines.
- Low mood – feeling low is common among people with IPF, and can lead to depression.
- Anger and frustration – you may be angry that this has happened to you, especially if no obvious cause has been identified. Or you may feel frustrated that you can't do some of the things you used to do.

- Isolation – IPF can have a big impact on your social life and on your relationships with families and friends.

These are all natural emotions. What is important is that you recognise these feelings, accept that they are normal, and try to find ways of managing them.

There are things that you can do to help you cope with these feelings.

- Find out as much as you can about IPF. Your main source of **information** will be your healthcare team, but there are also other resources available including written information, help lines, online information and support groups. See the Resources section at the back of this booklet for more details.
- Join a **support group**. Contact with other people who can relate to what you are going through can be very helpful and may help you feel less isolated. You may be able to share tips on coping with the day-to-day challenges of IPF and find out further sources of help and information.
- **Talk** to your family and friends. Let them know how you are feeling and what they can do to help you.
- **Keep active**. As well as keeping you as physically fit as possible, keeping active can also help improve your mood and relieve anxiety.

- Find ways to **relax**. There are many different relaxation techniques. Try a few and choose one that works for you. This may be by learning breathing techniques, meditation, using a relaxation app on your phone, or going to a class such as yoga or tai chi.
- **Mindfulness** can be useful in people with IPF. Find out more about mindfulness at [www.mentalhealth.org.uk/a-to-z/m/mindfulness](http://www.mentalhealth.org.uk/a-to-z/m/mindfulness) or [www.moodjuice.scot.nhs.uk/mildmoderate/Mindfulness.asp](http://www.moodjuice.scot.nhs.uk/mildmoderate/Mindfulness.asp)
- Keep **socially active**. Try not to withdraw from your family and friends. You may find that you want to stop taking part in social activities because it is difficult to get out, you feel embarrassed or you are too tired. Try to maintain an active social life as far as possible, as this will help stop you from feeling isolated.
- Focus on the things you **can do** rather than dwelling on the things you are no longer able to do.
- **Talk** to your doctor about any concerns you have. Although it can be difficult, it is important to address your concerns, ask difficult questions and start talking and planning what will happen when your condition starts to get worse.

Sometimes people become stuck feeling angry, unhappy or anxious most of the time. If this is the case, you have crossed from a normal emotional response to an unhealthy response. It is important that you recognise if your emotional response is unhealthy, and that you seek help from your healthcare team.

Depression is common in people with IPF, with up to half of all people with IPF having depression at some point. Speak to your doctor or one of your healthcare team if you:

- Feel down most of the time
- Feel hopeless or helpless
- Have noticed a change in your sleep patterns
- Have lost interest in doing things you usually enjoy
- Are avoiding social situations or feeling isolated
- Have thoughts of harming yourself

Remember, you are not alone. There are treatments available that can help if you are feeling depressed, including antidepressant medicines and 'talking' treatments such as cognitive behaviour therapy (CBT).

## **Will I be able to continue to work?**

A diagnosis of IPF does not necessarily mean that you will have to stop working. Whether or not you are able to continue to work will depend very much on the severity of your symptoms and on the type of job you do.

In the first instance you should discuss this with your healthcare team and your employer. It may be possible to make changes to your current role (for example working from home or reduced hours, or travelling less) that would enable you to continue working.

## **Where can I get financial help?**

If you are unable to work because of your IPF or your partner has given up work to care for you, you may be left with financial difficulties. However, there are benefits that you may be entitled to.

## How might IPF affect my relationship with my partner?

Having IPF does not mean that you have to give up having an intimate relationship.

The most obvious challenge to having sex for someone with IPF is that, like all other physical activity, sexual activity can make you out of breath and exhausted. You may face other challenges too. You may think that your partner does not find you attractive anymore because you have IPF or you need to use oxygen. Your partner may be worried about making your symptoms worse.

Whatever your concerns, try and talk with your partner about your worries. It is not always the easiest conversation to start, but you may well find that it is something that worries your partner too.

You may need to plan sexual activity a bit more than you previously have, for example:

- Propose sexual activity at a time of day that is least stressful for you, or when you feel most rested.
- If you normally increase your oxygen rate during exercise, plan to increase it by the same amount during sexual activity.
- Plan your environment – make sure that you are cool and comfortable.

Remember, if you get out of breath, like any other physical activity, it is perfectly acceptable to have a rest.



Some people who need to use oxygen during sexual activity can feel awkward about this. They may be worried that the tubing will get caught or the nasal cannula will fall out. There are ways of working around these worries; for example, use tape to keep the tubing in place, or use more tubing to allow greater flexibility.

If you find that breathlessness is restricting your sexual activity, you might want to experiment with different positions. Use positions that don't require you to support too much of your own or your partner's weight. Also, try to avoid positions that put extra weight on your chest or stomach.

Remember, there may be other ways of expressing physical intimacy, such as touching, holding hands, kissing and hugging. The common goal is satisfaction and pleasure for both partners.

## **Can I continue to drive?**

There are no restrictions on driving if you have IPF, and there is no need to notify the DVLA unless you experience giddiness, or you faint or lose consciousness.

If you use oxygen, you should inform your car insurance provider that you are transporting oxygen cylinders or a concentrator. This should not affect insurance premiums, but it will ensure you are fully covered in the event of a claim.

Remember, when you are travelling with oxygen in a private car:

- Make sure that the person using oxygen and the driver both know how to use the oxygen equipment properly.
- Use the equipment with the car ventilation system set to draw in fresh air from outside the car, or with a window partly wound down.
- Do not allow anyone to smoke in the car.
- Do not use the oxygen equipment whilst the car is being refueled.

If your mobility is limited

you may be entitled to apply for a disabled parking permit (Blue Badge). The Blue Badge scheme allows severely disabled people (travelling as a driver or a passenger) to park in certain restricted areas, allowing you to get closer

to where you need to go. To find out more or to apply for a Blue Badge if you live in Scotland, go to the government website [www.mygov.scot/apply-blue-badge/](http://www.mygov.scot/apply-blue-badge/) or contact your local authority.



You may also be eligible for a National Entitlement Card (bus pass). This will allow you to travel free on most local bus services and on many longer journeys between Scottish cities. To find out more about this, and help with other forms of transport, go to [www.mygov.scot/transport-help/](http://www.mygov.scot/transport-help/).

Remember, when you are travelling on public transport with oxygen:

- Only carry enough oxygen for the journey.
- Make sure you know how to use the oxygen equipment.
- Make sure you know what to do in the event of an incident involving the oxygen equipment.
- Turn off the equipment when not in use.
- Make sure that the equipment is secure, not free to roll about, and where required, is supported upright.

## **Can I still go on holiday?**

With proper preparation you should still be able to travel and enjoy going on holiday.

Most people with mild IPF should be able to enjoy going on holiday and fly safely with relatively few additional arrangements. Make sure your vaccinations are up to date, speak to your doctor about taking a course of antibiotics and/or steroids with you in case you experience an increase in respiratory symptoms, and make sure that you have adequate insurance cover.

For people with more severe symptoms, more careful planning may be needed. Plan a trip that is not going to be too strenuous, as you are likely to be feeling fatigued. If you use oxygen, make arrangements in advance. If you are planning to fly, contact the airline to find out about provision of oxygen during your flight. Again, make sure that you have adequate insurance cover. It is probably sensible to avoid destinations at higher altitudes, as there is less available oxygen in the air. Plan a day or two of rest into your holiday, especially after a long journey.

For people with advanced IPF, flying may not be a suitable option. However, you can still travel safely by car or rail. Make sure that you make arrangements with your oxygen provider in advance, and that you have all the equipment you need.

# THINKING ABOUT YOUR CARE AS YOUR IPF GETS WORSE

## Palliative care

Palliative care is a central part of the management of people with idiopathic pulmonary fibrosis (IPF). Palliative care is about managing your symptoms and relieving physical and emotional suffering.

## End-of-life care

Thinking about and talking about the end of your life, or the life of someone close to you, can be very difficult. Often just knowing where to start is the hardest part. Many people find talking about these difficult issues reassuring, as it can take away some of the fears of the unknown. It can also be helpful for those caring for you, as they can feel reassured that they are making the right decisions on your behalf.

It is important to start talking about and planning your care early on in your illness, whilst you are still stable. These conversations should be ongoing, as your needs and wishes will change over time. Issues you might want to discuss with your specialist team and your family include:

- Managing your symptoms and relieving suffering
- Whether you want to be admitted to hospital if you have a bad flare-up

- Where you would like to be cared for towards the end of your life
- What life-support measures you want to be taken
- Getting your affairs in order (for example, making a will or a Power of Attorney)

## **Advanced care planning**

It is important that your wishes are documented. This can be done using an Advanced Care Plan. An advanced care plan is the documentation (or writing down) of the discussions between an individual and their care provider (including their family or a friend if they so wish) regarding what would happen if their condition were to deteriorate. An advanced care plan expresses your wishes but it is not legally-binding. However, it can also provide an opportunity to discuss other legally-binding issues such as an advance decision to refuse treatment or power of attorney for financial, health and social care.

You may wish to keep a Preferred Priorities of Care with you. This is where an individual writes down (and keeps with them) what they would like to happen, in the event that their illness deteriorates. Again, it is not legally binding, but it will be taken into account when planning care and will help both the healthcare team and those caring for you to make decisions about your care.

## SUPPORT FOR CARERS

It is very common for someone to put aside their own needs while they are busy caring for a person with a long-term illness such as IPF. However, this can be a difficult time for both of you, and as a caregiver it is important that you feel supported and able to cope.

There are various sources of help available from your local council as well as private and voluntary organisations.

- Chest Heart & Stroke Scotland (CHSS) provides access to Advice Line nurses, affiliated chest support groups and health information resources.
- Your local social work department can provide for assistance at home and a carer's assessment.
- Carers' organisations can provide advice, information and support.
- Local organisations sometimes provide a 'sitting service' to give carers a break for a few hours a week.
- Shared Care Scotland can provide help with respite care.

## USEFUL RESOURCES

### **Chest Heart & Stroke Scotland**

Rosebery House, 9 Haymarket Terrace

Edinburgh EH1 2 5EZ

Tel: 0131 225 6963

Advice Line Nurses: 0800 801 0899 (free number) (Monday to Friday 9.30am - 4pm)

Email: [adviceline@chss.org.uk](mailto:adviceline@chss.org.uk)

Website: [www.chss.org.uk](http://www.chss.org.uk)

*Chest Heart & Stroke Scotland improves the quality of life for people in Scotland affected by chest, heart and stroke illness, through medical research, influencing public policy, advice and information and support in the community.*

### **Action for Pulmonary Fibrosis**

Tel: 01543 442152

Email: [info@actionpulmonaryfibrosis.org](mailto:info@actionpulmonaryfibrosis.org)

Website: [www.actionpulmonaryfibrosis.org](http://www.actionpulmonaryfibrosis.org)

*Action for Pulmonary Fibrosis aims to promote the relief of sickness and the preservation of health by advancing education and raising awareness of pulmonary fibrosis, the symptoms associated with it and encouraging earlier diagnosis of the condition; promoting research into the understanding, diagnosis, causes and treatments of pulmonary fibrosis; and providing support to patients and families suffering from pulmonary fibrosis.*



## **Breathing Space**

Tel (free): 0800 83 85 87

Website: [www.breathingspace.scot](http://www.breathingspace.scot)

*Breathing Space is a free, confidential phone and web based service for people in Scotland experiencing low mood, depression or anxiety.*

## **Carers Scotland**

Helpline: 0808 808 7777 (Mon-Fri 10-4pm)

Email: [advice@carersuk.org](mailto:advice@carersuk.org)

Website: [www.carerscotland.org](http://www.carerscotland.org)

*Carers Scotland (part of Carers UK) offers information to carers throughout Scotland as well as details of local support.*

## **Carers Trust Scotland**

Tel: 0300 123 2008

Email: [scotland@carers.org](mailto:scotland@carers.org)

Website: [www.carers.org/scotland](http://www.carers.org/scotland)

*Carers Trust Scotland provides comprehensive carers' support services throughout the UK including independently-run carers' centres.*

## **NHS 24**

Telephone (free): 111

Website: [www.nhs24.scot](http://www.nhs24.scot)

*This phone service is designed to help you get the right help from the right people at the right time and GP out of hours advice. The website provides comprehensive up-to-date health information and self-care advice for people in Scotland.*

## **Quit Your Way Scotland**

Tel: 0800 84 84 84 (8am-10pm, daily)

Website: [www.canstopsmoking.com](http://www.canstopsmoking.com)

*Quit Your Way Scotland offers initial and ongoing telephone support and encouragement to callers wishing to stop smoking or who have recently stopped and want to stay stopped. Quit Your Way Scotland can also give you information on your nearest free stop-smoking service, give you access to specialist counsellors and send you further information.*

## **Shared Care Scotland**

Tel: 01383 622462

Email: [office@sharedcarescotland.com](mailto:office@sharedcarescotland.com)

Website: [www.sharedcarescotland.org.uk](http://www.sharedcarescotland.org.uk)

*Shared Care Scotland work to improve the quality and provision of short breaks in Scotland. They also operate a 'short breaks fund' on behalf of the Scottish Government. The fund provides grants to organisations that support unpaid carers to take a break.*

# OUR PUBLICATIONS

We hope this information has been useful to you.

Our publications are free to anyone in Scotland who needs them.

To view, download or order any of our resources, visit **[www.chss.org.uk/publications](http://www.chss.org.uk/publications)**

If you'd like more information about our publications, please contact our Health Information team:



[www.chss.org.uk](http://www.chss.org.uk)



[healthinformation@chss.org.uk](mailto:healthinformation@chss.org.uk)



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**Chest Heart & Stroke Scotland** is a Scottish charity. Our ambition is to make sure that there is no life half lived in Scotland.

After a diagnosis of a chest or heart condition or a stroke, many people experience fear and isolation and struggle with the impact on their lives. **Chest Heart & Stroke Scotland** won't stand for that. The care and support we deliver every day ensures everyone can live the life they want to.

We offer our resources free of charge to anyone in Scotland who needs them. To help us to continue to do this, and to help provide vital services to people in Scotland affected by chest, heart and stroke conditions, you can donate at:

[www.chss.org.uk/supportus/donations](http://www.chss.org.uk/supportus/donations)

## CONTACT US

For confidential advice, support and information call the CHSS Advice Line nurses on:

Freephone 0808 801 0899 or email [advice@chss.org.uk](mailto:advice@chss.org.uk)

### Head Office

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[www.chss.org.uk](http://www.chss.org.uk)

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