Interstitial lung disease (ILD) is an umbrella term that covers many different conditions. The most common of these is idiopathic pulmonary fibrosis (IPF). This information sheet tells you the basic facts about IPF and other ILDs, how they are diagnosed and treated and what the symptoms are like. It also talks about different treatments and outlook for IPF and some of the most common types of ILD.

There is not a cure yet for IPF, but the right care can help you feel better and improve your quality of life. If you need to explain the situation to friends and family, this information sheet might be helpful for them to read too.

When someone has IPF or another type of ILD, it affects the whole family. That’s why we’ve included some information for carers, and a reminder about benefits and other help available to you. There’s also a list of places to find more information and support.

About interstitial lung disease and IPF
About 5,000 people are diagnosed with IPF every year in the UK. The term ‘idiopathic’ means the cause is not known. The condition is one of many types of interstitial lung disease (ILD). ‘Interstitial’ means the disease affects the ‘interstitium’, a lace-like network of tissue that supports the alveoli (air sacs) in your lungs.

There are more than 200 different types of ILD. All of them are rare but those seen most often (IPF, collagen vascular disease associated ILD, extrinsic allergic alveolitis and ILD caused by drug side-effects) are covered in this booklet. IPF is the most common type of ILD. Other names for IPF are cryptogenic fibrosing alveolitis or usual interstitial pneumonia.

Sarcoidosis is another common type of ILD. For more information on this condition, go to www.blf.org.uk/Conditions/Detail/Sarcoidosis or call the BLF Helpline.

For most types of ILD, such as IPF and sarcoidosis, the cause is unknown. Some ILDs occur when you have another condition such as rheumatoid arthritis or scleroderma. In a small number of ILD cases it is possible to identify a specific cause such as a drug side effect or exposure to certain types of dust or other allergens. These could include birds, mould, wood or metal dusts or asbestos.

When you have ILD, inflammation or scar tissue builds up in your lungs, making them thick and hard. This build-up of scar tissue is called fibrosis. As your lungs become stiffer and lose their elasticity, they are less able to take oxygen from the air you breathe. People with ILD can feel breathless from...
simple everyday activities like walking. Coughing is another common symptom. Sometimes people have inaccurate ideas about what causes ILD, so it helps to know that it is not a form of cancer or cystic fibrosis, and it is not contagious.

The treatment and outlook for different ILDs vary, so if you’re not sure about your diagnosis, check with your doctor for the exact name of your lung condition.

How the lungs work
To understand how fibrosis affects you, it helps to have a reminder of how the lungs work. Each time you breathe, air is drawn into your nose or mouth, down through your throat and into your windpipe (trachea). The windpipe splits into two smaller air tubes called the bronchi, one of which goes to the left lung and the other to the right lung. The air passes down the bronchi, which divide again and again, into thousands of smaller airways called bronchioles. This system is like branches, and its medical name is the bronchial tree.

Your lungs

The bronchioles have many small air sacs, called alveoli. Inside the alveoli, oxygen moves across paper-thin walls to the capillaries (tiny blood vessels) and into the blood. The alveoli also pick up the waste gas carbon dioxide from the blood, ready to be breathed out.

When your lungs are damaged by the scarring that happens in IPF, or inflammation or scarring caused by another type of ILD, this process becomes less efficient and the oxygen you breathe can’t move easily into your bloodstream.
The diagram below shows how fibrosis due to IPF or other ILDs affects the air sacs in your lungs.

**Normal air sacs (alveoli)**

- Air to and from mouth/nose
- Blood vessel
- Carbon dioxide

**Air sac damaged by IPF**

- Air to and from mouth/nose
- Inflammation
- Scarring
- Blood vessel
- Carbon dioxide

**IPF: Who is at risk?**

About 5,000 people are diagnosed with IPF every year in the UK. Men are more likely to have IPF and they account for six in ten new cases. It is also more likely to affect older people.

The term ‘idiopathic’ means the cause is not known. So it’s important to remember that no one is sure why you have developed this condition. We do know that IPF is more common in people who are smokers, or in people who have smoked in the past.

It is also more common if you have been exposed through your occupation to dust from wood, metal, textile or stone, or from cattle or farming.

Infection might be another cause. 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.

Doctors have noticed that some people with IPF also have a condition called gastric oesophageal reflux (GORD). This is when the stomach contents leak back up into the oesophagus (gullet), causing the symptoms of heartburn. Reflux is very common in patients with any
type of lung fibrosis, as the gullet becomes stretched by the fibrosis in the lung. Some experts think there could be a link between the two conditions.

In a few cases of IPF, there might be a genetic link.

**IPF: What the name means.** ‘Idiopathic’ means the cause is unknown. It’s pronounced ‘idee-o-path-ic’. ‘Pulmonary’ shows it is related to the lungs and ‘fibrosis’ means scarring.

What IPF is:

- Scarring on the lungs
- Difficulty in breathing
- Unknown cause

What it is not:

- It is NOT cancer
- It is NOT a form of cystic fibrosis
- It is NOT contagious - other people can’t catch it

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**Symptoms of IPF and other forms of ILD**

The different types of ILD, including IPF, 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 most common symptom is feeling short of breath, especially when you’re moving about. This does not mean the normal huffing and puffing you might feel while jogging or climbing a steep hill. It might happen even when you are doing something that would not usually tire you out, such as walking up stairs or even just getting dressed.

IPF occurs in middle age or when people are older, so you might think it’s just that you are not as young as you used to be. But if you feel breathless, you should never ignore it and you should see your doctor.

A cough that doesn’t go away and feeling very tired all the time are two other symptoms that can happen with IPF or another type of ILD. Some people with ILD 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 IPF or other types of ILD. It’s called clubbing, and you might notice that:

- your nails feel too soft as if they are coming loose;
- the shape of your nails changes; and
- the tips of your fingers or toes bulge out.
Clubbing can occur with various lung conditions, and also as a result of heart or liver disease.

**Tests and diagnosis**
If you have symptoms that might be due to IPF or another type of ILD, 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, as that will influence the choice of treatment.

For example, IPF is related to a number of other types of ILD, which your consultant and their team will need to rule out before they can say that you have IPF.

To start with, the doctor will listen to your chest, ask questions about your medical and work history, and order some blood tests to rule out possible known causes. Further tests that can be used to diagnose an ILD are described here, but you might not need to have all of them.

**X-rays and scans**
You will probably have a chest X-ray. If your doctor thinks you might have an ILD, this may 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 ILD, the results from the scan can be very clear. For example, 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.

In some cases, you might need additional tests to confirm your diagnosis.

**Looking inside the lungs**
In some cases, to confirm the diagnosis your doctor may need to look inside your lungs and possibly remove some cells or tissue for testing. This is called a biopsy. The samples are examined under a microscope by a doctor who is expert in laboratory tests (a pathologist).

One test is called flexible bronchoscopy and involves inserting a narrow tube through your nose or mouth, down into your lungs. 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 cells for laboratory tests. You may be sedated for this test and your throat will be numbed first with a local anaesthetic.

Another test is called video-assisted thoracoscopy (VATS). This involves surgery under a general anaesthetic. The doctor uses a tube equipped with a video camera to look at your lungs and remove some tissue samples for laboratory testing. Having this test will mean staying in hospital for a few days.

**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 IPF or another ILD, these tests will be used to monitor your condition.
Identifying IPF and other types of ILD is a joint effort by your specialist team, which might 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.

Treatment

Medication
Options for medication will depend on your specific condition, and you can find more details on medications for IPF and the more common types of ILD later in this booklet. You might also be offered medicines that don’t have an effect on your ILD, but can help with symptoms such as feeling short of breath.

You might be invited to take part in a medical study (clinical trial) to investigate new treatments. It’s not an option for everyone but if you want to know more, ask your doctor. If you decide not to join a clinical trial, you will still receive the best possible care. You can also leave a clinical trial at any time if you change your mind.

Pulmonary rehabilitation
When you have an ILD, breathlessness can have a serious effect on your everyday life. It might be a frightening experience, too. Pulmonary rehabilitation can help you to cope with breathlessness and is an important part of treatment. It includes:

• a physical exercise programme, designed individually for you; and
• advice and information on keeping your lungs healthy and coping with breathlessness.

Sessions are held in hospitals, health centres and in places such as community halls. The groups are led by a qualified professional.

As well as providing support and reassurance, pulmonary rehabilitation can have a positive impact on your condition. It may help to increase the distance you can walk, help you feel less breathless and improve your sense of well-being. If it has not already been recommended, ask your doctor or nurse about trying pulmonary rehabilitation. The BLF Helpline can also help you find a local group and tell you more about how to get the right kind of exercise.

Oxygen
If your condition gets worse, the level of oxygen in your blood falls and you feel more breathless. If this happens, your doctor might recommend oxygen therapy, which is provided free on the NHS. Most patients with ILD require oxygen for getting about outside when their condition is more advanced. You might 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 are not confined to one room.

You can find out more about oxygen therapy at [www.blf.org.uk/oxygen](http://www.blf.org.uk/oxygen) or by calling the BLF Helpline on 03000 030 555.
**Lung transplant**
For a very few people, having a lung transplant might be an option. This is rare, and only about 120 lung transplants are carried out in the UK every year, including people with other conditions. Whether you can be considered for a lung transplant depends on factors that influence the chance of a successful outcome, such as your age and general health. Then there is the question of whether a donor lung is available.

If you wish to explore lung transplantation and your doctor thinks you might be suitable for this treatment, you could be referred to the transplant unit. You should ask for an initial response from the transplant centre within four weeks.

According to the most recent research, the survival rate (the percentage of people who survive a disease such as IPF for a specified amount of time) for one year after lung transplantation is 79 per cent, and for five years it is 53 per cent. The survival rate for 10 years is 30 per cent. However, the outcome of lung transplantation can be influenced by several factors including the type of procedure you have, your age, and the characteristics of the donor lungs.

**Specific treatments**
This section looks at specific types of ILD and the treatments you might be offered.

**IPF**
This is the most common type of ILD. IPF is a progressive condition and usually gets worse over time. In some people the symptoms gradually get worse over several years. For other people, the symptoms get worse quickly.

Sometimes when the condition has been stable, people can get sudden flare-ups of the symptoms. This is called an acute exacerbation.

The length of time that people live after being diagnosed with IPF varies. Sadly, about half of people with IPF live for only two to three years after their diagnosis. However, many people survive much longer than this and new treatments have become available since these figures were published.

It’s difficult to predict how IPF will progress. Each person is different so you should talk to your consultant about your individual situation. If you or your family need emotional support during this time, many people are ready to help, including the specialist nurses and advisors at the BLF.

Unfortunately, there is no cure yet for IPF. However, there are things your specialist team can do to help you feel better and improve your quality of life. The options can be mixed and matched according to your needs, and your care might include:

- pulmonary rehabilitation (see above)
- oxygen therapy
- medication to help with symptoms
- medication to try to reduce the scarring and inflammation in your lungs.

Ongoing information and support from a specialist nurse should also be a key part of your care. For a very few people, having a lung transplant might be an option, but this is rare (see above).
Medication for symptoms
If you are having trouble breathing, even when you are resting, your doctor might prescribe a sedative or morphine. You might know them as painkillers but these medicines can also help with symptoms of breathlessness when people have a serious lung condition like IPF. For coughing, your doctor might treat problems that are making it worse, such as heartburn or a stuffy nose. Morphine can also help a really bad cough that is becoming hard to live with.

Medication for IPF
For now there are just two main drugs that are used to try and stop the damage to the lungs in IPF:

Pirfenidone (brand name Esbriet)
This is a new treatment for people with mild to moderate IPF, that comes in the form of capsules. Scientists don't know exactly how pirfenidone works yet, but it is thought to slow down inflammation and the build-up of scar tissue in the lungs. In medical studies, it slowed down the loss of lung function in patients with IPF, and researchers hope that this will decrease the rate at which symptoms get worse and also improve life expectancy. The most common side effects are skin reactions to sunlight; feeling sick (nausea); tiredness, diarrhoea and indigestion. Other side effects are possible and your doctor can tell you about these if you are considering pirfenidone.

In April 2013, the National Institute for Health and Care Excellence (NICE), which advises the NHS on using new drug treatments, recommended pirfenidone as a treatment option across the NHS for people who have IPF. However, it has only been recommended for people whose lung function is within a certain range. Therefore there is a chance that your doctor may not be able to prescribe you pirfenidone, but if this is the case then they will explain the reasons to you. NICE has also recommended that if your IPF continues to get worse despite being treated with pirfenidone, then treatment should be stopped after a year.

N-acetyl cysteine (NAC)
This medicine is used to treat IPF and is taken as tablets, or as fizzy tablets in water. At this time, the evidence of its benefits comes from a single trial, but other studies are going on to confirm the effectiveness of NAC. NAC is an anti-oxidant thought to discourage the creation of new scar tissue in the lungs. Some people experience stomach discomfort, trapped wind or nausea when they take NAC. Many people find that NAC improves their cough but for some people it can make their cough worse.

The use of NAC in combination with steroids or azothiaprine is no longer recommended for IPF, but may be used for other types of interstitial lung disease.

For the most up to date information about treatments for IPF with medicine, visit www.blf.org.uk/ipf or call the BLF Helpline on 03000 030 555.

Clinical trial
Possible new medicines for IPF are being tested in medical studies (clinical trials). If you want to know more about participating in one of these studies, talk to your doctor.
Collagen vascular diseases and ILD (CVD-ILD)
For reasons that we don’t understand fully, sometimes the immune system turns against the body (autoimmune disease). When this affects collagen – the tough protein that forms connective tissue – various organs in the body can be affected, including the lungs. The general term is collagen vascular diseases (CVDs) and a few of these conditions are associated with ILD, including rheumatoid arthritis, Sjögren syndrome and scleroderma. Unfortunately some of the drugs used to treat collagen vascular diseases, such as methotrexate, can also cause interstitial lung disease.

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.

It’s difficult to say what course your ILD will take because this depends including the type of CVD you have, how severe it is and the way it is affecting your lungs. Sadly, 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 the BLF Helpline for confidential one-to-one advice and support from our team of dedicated advisors and nurses.

Extrinsic allergic alveolitis
This condition happens when the lungs have an allergic reaction to something you have inhaled. One example is farmer’s lung, which is caused by breathing 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. Inhaling certain chemicals can be another cause.

The symptoms, including cough and breathlessness, can come on suddenly after you’ve been exposed. This is the acute form of the condition. It often goes away if you can avoid the substance that caused the attack. But it can also develop into a longer-lasting condition that permanently scars the lungs. In this case, you may need treatment with steroids and oxygen therapy.

ILD caused by drug side-effects
Any medication can have side effects and unfortunately, some medicines can damage the lungs and cause ILD. You and your doctor have to weigh up the risks and benefits before you start a medicine and sometimes the choices are difficult, especially when it is a life-saving treatment.

Hundreds of medicines have the potential to cause ILD. Some of the medication types that are known to carry the risk of ILD include certain:

• cancer chemotherapy drugs
• drugs for heart conditions, particularly amiodarone
• anti-inflammatory drugs
• antibiotics, particularly nitrofurantoin
• biological agents, used to treat cancer or immune disorders

Some recreational (illegal) drugs can also cause ILD.

The situation can vary for each individual. Breathing problems from drug-induced ILD can come on suddenly, or develop more slowly over time.

If a drug is causing ILD, people often get better quickly if the medication is stopped before much damage is done. If this does not happen, your doctor might prescribe steroid medication to help calm down your body’s response to the medication. Unfortunately, some people have lasting lung damage from drug-induced ILD.

Sarcoidosis is another common type of ILD, and is covered in a separate BLF leaflet. For more information on this condition, go to www.blf.org.uk/Conditions/Detail/Sarcoidosis or call the BLF Helpline on 03000 030 555.

Looking after yourself

Everyone benefits from a healthy lifestyle and this is especially true when you have IPF or another type of ILD. 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.
• Ask your nurse for a pneumococcal vaccination (pronounced new-mo-coc-al). This is a jab you have just once to protect against pneumonia and many other infections.
• Avoid being around people with chest infections and colds.
• Stay as fit as you can.
• Eat a healthy, balanced diet. 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.
• Make sure you drink enough water each day.
• Try to keep regular bedtime routines to ensure you get enough sleep. If you are having trouble sleeping, mention it to your doctor as they might be able to help.

If you smoke, stopping is a very important part of your treatment. We know it isn’t easy, but there is help available. Your doctors and nurses will be keen to help you with this too. The NHS offers a free stop smoking service and your GP can refer you. You can also call the NHS Stop Smoking Help Line (see details further down). Taking that first step is a very positive way to look after your lungs.

Feeling breathless can be distressing but there are things you can do to help you cope. Taking a rest
break before or after a strenuous situation, such as going to the shops, can be helpful. There are also certain positions or breathing techniques that can ease the feeling of breathlessness. For more information, ask your doctor, nurse or physiotherapist, call the BLF Helpline or visit www.blf.org.uk

**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 IPF or another type of ILD, it’s common to have emotional struggles too. Your doctor or nurse will understand if you are feeling low, and they can help. You can also call the BLF Helpline, not just for practical advice but also for emotional support.

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 support group.

**Caring for someone with a lung condition**

When someone you love is ill, looking after them just seems like the natural thing to do. But being a carer can become a job in itself, and sometimes a pretty demanding one. It’s important to take good care of yourself so that you can give better quality care to the person living with a lung condition.

You can get more information from our practical booklet, *Looking after someone with a lung condition*. It is packed with useful information about your rights as a carer, how to look after your loved one and how to look after yourself. Read and order the information at www.blf.org.uk/carers or request a copy by calling the helpline on 03000 030 555.

Contact the BLF Helpline for any query related to IPF or any other ILD, as well as practical advice and emotional support for people with IPF, their carers, and their families or friends. Caring can be a tough experience. If you need someone to talk to, don’t hesitate to contact your GP, the nurse or doctor looking after the person you are caring for, or the advisors on our helpline.

**Benefits and family finances**

Having a serious lung condition might leave you struggling with your finances. Help is available and even if you are in work, you might be able to claim financial benefits. For example, if your lung disease is caused by your work you may be able to get Industrial Injuries Disablement Benefit. It is worthwhile to find out what you qualify for. The benefits system is undergoing significant changes at the moment so it’s important to get advice about which benefits to apply for and how to go about it. It’s also important to act right away and not put it off. You can’t backdate most claims and it takes time for any money to come through.

You can find out more information about the benefits you might be entitled to at www.blf.org.uk/your-rights. For the most up to date information about benefits, and for more advice, you can call the BLF Helpline.

**Information and support**

Benefits Enquiry Line - anyone with a disability can get advice from this government helpline.
Telephone: 0800 882 200

Citizens Advice
Information about benefits, money problems, and many other topics including NHS care, housing, and help from social services. You can find your local bureau in the phone book.
Find your local bureau: www.citizensadvice.org.uk
Read information guides: www.adviceguide.org.uk

GOV.UK
Read about benefits and money matters on the government’s main website:
www.gov.uk

Turn2Us
Turn2us helps people in financial need gain access to welfare benefits, charitable grants and other financial help.
http://www.turn2us.org.uk/
Free and confidential helpline: 0808 802 2000

Carers Trust
Find your local carers’ centre, or read information online.
www.carers.org.uk

Carers UK
Call the helpline or visit the website for support and practical information.
www.carersuk.org
Helpline: 0808 808 7777

Carers Direct
This is the NHS resource for carers. You’ll find information and advice on services, as well as a free course to give you skills and confidence as a carer.
www.nhs.uk/carersdirect
Helpline: 0808 802 0202

NHS Stop Smoking Service
Visit the website to find advice, tools and to locate the free NHS stop smoking nearest you. Or phone the helpline for individual advice.
www.smokefree.nhs.uk
NHS Smokefree Helpline: 0800 022 4332

Pulmonary Fibrosis Trust
www.pulmonaryfibrosistrust.org
0333 20 20 991

Action for Pulmonary Fibrosis
www.actionpulmonaryfibrosis.org
info@actionpulmonaryfibrosis.org
The British Lung Foundation has a specialist team of nurses and advisers on our helpline who are dedicated to answering your questions. Whether it’s about a visit to the doctor, concerns about your lungs, coping with symptoms or if you just need a chat, they are here for you.

Ringing the BLF Helpline never costs more than a local call and is usually free, even from a mobile. Lines are open from 9am to 5pm, Monday to Friday.

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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 www.blf.org.uk.