Idiopathic pulmonary fibrosis (fibrosing alveolitis)
What is idiopathic pulmonary fibrosis (IPF)?

IPF used to be known as fibrosing alveolitis

Idiopathic pulmonary fibrosis (IPF) is caused by repeated injury to small areas of your lung. This results in inflammation of your lungs and then scarring. If you have IPF your lung function will get gradually worse.

Scars serve a good purpose in the skin - they heal injured areas. But in the lung, scar tissue stops the lung doing its job - taking oxygen from the air and passing it into the blood. So you may have breathing difficulties or get breathless from simple activities such as walking and talking.

If it’s not known what has caused the condition, it is called ‘idiopathic pulmonary fibrosis’ - IPF (or ‘cryptogenic fibrosing alveolitis’ - CFA, these are different names for the same thing).

It is important to note that IPF is not cancer. It is not infectious either.
Who can get IPF?

The number of IPF cases seem to be rising, but it’s not clear why. It generally affects people in middle age (usually aged 50+) and it’s more common in men. It is also more common among smokers.

We don’t know what causes IPF. But we do know that:

- It is not an infection
- It can not be caught from other people
- It is not a form of cancer
- It is not a form of cystic fibrosis.

What causes IPF?

Although we don’t know exactly what causes IPF, a number of factors are thought to trigger it. These factors include:

- Viral infections: Epstein-Barr virus and Hepatitis C
- Gastro-oesophageal reflux: this is where the contents of your stomach come back up into your throat. If you inhale it, this can cause damage to your lungs
- You may be more likely to get IPF if you are exposed to a variety of occupational dusts or if you smoke
- There is evidence that you can inherit a tendency to develop some types of IPF - it can run in families.

What are the symptoms?

The most common symptom you may have is breathlessness, especially when taking exercise, such as walking up hills or stairs. The disease may come on in your mid-50s, but you shouldn’t think that breathlessness is just part of middle age. If you feel breathless, you should see your doctor.
We do have less energy as we get older and this can have an effect on breathing. However, your breathing will be affected more severely if you have IPF.

Less common symptoms you may have are a dry cough and a change in the shape of your fingernails and toenails.

If you feel you have these symptoms, you should ask your doctor for a diagnosis as quickly as possible.

**How is IPF diagnosed?**

As IPF is one of the less common lung diseases your GP will probably have seen very few, if any, people who have it. So to find out if you have IPF, you will need to be referred to a chest specialist.

The investigations will usually include a chest X-ray and a set of breathing tests. These involve breathing in and out of a lung function machine, which shows how well your lungs are working. You will usually also have a blood test. You should also have a 'high resolution CT scan.' This produces a detailed three-dimensional image of your lungs. The consultant may also need samples of lung cells to examine. To do this, they pass a small, flexible telescope (a bronchoscope) down your breathing tubes – you will have a mild sedative while they do this. This process allows the consultant to collect samples of the cells that line the lungs. This helps diagnosis.

Because other diseases can look like IPF, the consultant may need to remove a small piece of your lung to confirm the diagnosis. This ‘keyhole’ procedure always involves a general anaesthetic. The modern technique is called a ‘video assisted thoracoscopic sample’ - or VATS.

These investigations allow your consultant to have as clear a picture as possible of the precise nature of the problem.

**What treatment is available?**

IPF gets worse over time, but there may be periods when it remains stable. Whatever treatment you receive, if you smoke it is important that you stop.

Unfortunately there are currently no fully licensed medications that can cure IPF or slow down its progression. However, several new drugs are being tested in clinical trials. If you are willing to take part in an appropriate trial you should speak to your doctor about being referred to a specialist centre.

In the past it was thought that inflammation in the lungs was a big part of IPF. Therefore many treatments that have been used, and in many cases are still being used, focus on reducing this inflammation. It was thought that by reducing the inflammation any further scarring would be prevented.

It is now thought that inflammation does not play a big role in IPF and that medications used to reduce inflammation are not helpful for most people. However, in some cases doctors might still suggest trying these medications, such as steroids, for a trial period to see if they help to improve your symptoms. IPF is a part of a group of conditions called interstitial lung disease (ILD) and steroids may be effective for other types of ILD.

One thing to remember about these medications is that they can have side-effects – your doctor will take this into account when prescribing them.
These are some types of treatment that might be suggested to you.

- **Steroids** - a low dose of prednisolone is often used to help reduce inflammation in the lungs.
  
  **Side-effects** - Long-term use of steroids results in weight gain. They may also trigger diabetes and raised blood pressure if you have the tendency to get them. Other side-effects include:
  
  - Osteoporosis (thinning of the bones) particularly in women past the menopause
  - Cataracts
  - Glaucoma
  - Stomach problems.

  **A balance is needed between the potential benefits of treatment and the risk of side-effects.**

Steroids can also be prescribed with:

- **Immunosuppressants** - azathioprine or cyclophosphamide. These act to suppress the body’s immune system, which is thought to be partly responsible for IPF.
  
  **Side-effects** - This treatment can increase the likelihood of infections and may affect your blood in other ways. For example it can produce anaemia or clotting problems. Occasionally methotrexate can cause lung inflammation.

  **Once again, a balance is needed between the potential benefits of treatment and the risk of side-effects.**

- **Oxygen** - when IPF is very bad, the level of oxygen in your blood falls and you feel more breathless. Your respiratory specialist will prescribe an oxygen concentrator for you, which will help your breathing. You should also ask for portable oxygen to help you get out and about. The level of oxygen in your blood will be monitored so that you are getting the correct flow of oxygen from your concentrator.

- Tablets that help to reduce inflammation, called n-acetylcysteine, may also help.

The more inflammation you have, the better you will respond to treatment. Scarring is less likely to respond well to treatment than inflammation.

Changes in your symptoms and lung function tests show how you are responding to treatment. When, and if, the most improvement is achieved, the aim is to maintain that while giving you less medication. The long-term aim is to keep IPF suppressed on the smallest possible dosage of medication.

People may need treatment for the rest of their lives. This is because IPF is suppressed rather than cured. Your blood will be checked regularly to make sure you are not getting too much treatment. If you do not respond to treatment, your drugs will be stopped.

If your IPF continues to get worse, your consultant may think about a lung transplant. This decision is based on:

- The severity of your condition
- How quickly your condition is getting worse
- Whether you will be more likely to do better with or without a transplant
- Whether a donor lung is available.

Few people are candidates for a lung transplant.
How can I help myself?

You can help yourself stay as healthy as possible if you:

- Get a flu jab each year
- Get immunised against infection with the pneumococcus bacterium (you only have this once)
- Keep away from people with chest infections and colds
- Stay as fit as you can
- Eat a good, balanced diet. Speak to your doctor/nurse for a referral to a dietician. He/she can suggest the most effective diet for you.

The right diet will help your muscles keep their strength - including your breathing muscles.

If you are overweight because you have become less active, then a weight-reducing diet may help.

Pulmonary rehabilitation

If you have IPF, you might well benefit from pulmonary rehabilitation courses. These courses mix physical exercise, education about how to live with your condition with psychological and social support. Many people living with chronic (long-term) lung diseases, such as IPF, feel they lack this help.

Each course is run by a team that may include doctors, nurses, physiotherapists, psychologists and dieticians. We know that pulmonary rehabilitation can improve your physical function, enhance your quality of life and increase your sense of independence.

First, talk with your consultant about whether you will benefit from pulmonary rehabilitation. 160 hospitals or chest clinics in the UK currently run courses - you can check with your consultant or call the BLF Helpline on 03000 030 555 to find your nearest course.

Welfare benefits

You may be eligible for a number of benefits. If you need help with personal care or getting about, you may be entitled to Disability Living Allowance or Attendance Allowance. If you care for someone living with IPF, you may also be entitled to Carer’s Allowance.

We’re here to help

Call the British Lung Foundation Helpline

Whether it’s about the benefits you can claim, coping with symptoms or if you just need a chat, the specialist nurses and advisers on our helpline are here for you. Ringing the helpline never costs more than a local call and is usually free, even from a mobile. Lines are open 10am to 6pm, Monday to Friday.

Find out about more than 40 conditions

We provide clear and trustworthy information about lung disease. All of our information is available in print and online. To order, call the helpline or visit our website.

Get support when you need it most

There is all sorts of support available for you, from becoming a member of the BLF to join one of our local Breathe Easy support groups. To find out about all of these options and more, call the helpline or visit our website.
One person in five in the UK is affected by lung disease. Millions more are at risk.

We are the UK’s lung charity and we are here for every one of them, whatever their condition.

Lung disease can be frightening and debilitating. We offer hope and support at every step so that no one has to face it alone.

We promote greater understanding of lung disease and we campaign for positive change in the nation’s lung health.

We fund vital research, so that new treatments and cures can help save lives.

We are the British Lung Foundation. Leading the fight against lung disease.

The British Lung Foundation has offices across the UK. Get in touch to find support near you.

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