Centre of Research Excellence in **Pulmonary Fibrosis**

Lung Foundation Australia

# Idiopathic Pulmonary Fibrosis (IPF): Understanding your diagnosis

## Overview

Idiopathic Pulmonary Fibrosis (IPF) is a lung disease in which the tissue around the air sacs (alveoli) within the lungs become thickened and scarred – this is called fibrosis.

This scarring, or fibrosis, makes the lungs stiff which makes it increasingly difficult to breathe. It also reduces the delivery of oxygen into the bloodstream where it is needed to be transported to the rest of the body.

## Symptoms

Early in the disease, symptoms can be very mild, which can make it difficult to detect. This can often lead to a delayed diagnosis.

**Common symptoms are:**

- Shortness of breath, which is particularly noticeable when walking up hills or climbing stairs.
- A dry, irritating cough that doesn't get better. Some people may produce clear phlegm.
- Reduced exercise capacity or inability to perform usual activities without breathlessness.

**Other symptoms are:**

- Feeling tired
- Gradual unintended weight loss.

## Who gets IPF?

The reason for the scarring in IPF is not known. This is why the disease is called idiopathic, which means "no known cause." However, scientists and healthcare professionals all over the world are working to better understand the disease.

What we do know is that although it affects people of any age, IPF typically affects older people and is more common in men than women. While IPF can be more common in some families, this is unusual, and can be investigated by genetic testing.

### How common is IPF?

**Although IPF is rare, you are not alone. In Australia, the current estimate is that there are approximately 2,170 new cases of IPF each year.**

## Experience

It is important to know that each person experiences IPF differently. Some people can remain stable for many years; others decline rapidly; and others have a series of distinct 'steps' of suddenly feeling worse, followed by a period where their symptoms become stable. As the scarring is progressive and irreversible, most people with IPF do decline over time.

## Treatment

The treatment and management of IPF is based on each person's individual medical needs and lifestyle. It is important to talk to your doctor and healthcare team to understand the range of treatment options. Involving your close family and friends may help you choose the best approach. Once treatment has begun, your respiratory specialist will generally see you several times a year to monitor your disease, symptoms and treatment.

**Current treatment options include:**

**Medicines**

- There are currently two anti-fibrotic medicines - Pirfenidone and Nintedanib – shown to help slow the rate of disease progression. They are subsidised by the government and are available to people that meet certain eligibility criteria. To see if you might be eligible for these medicines, please speak to your specialist.
- Anti-fibrotic medicines may have side effects such as diarrhea, skin rash or reflux. If side effects develop, it is important you discuss them with your treating doctor and develop a management plan.

### Oxygen therapy

- Oxygen therapy may be prescribed to people with low oxygen levels at rest or during exercise. In some this may assist with shortness of breath and staying active. Some people only use oxygen when they feel breathless (e.g., walking or exercising) but others need to use oxygen continuously during the day and night.

### Lung transplantation

- In some cases, lung transplant surgery may need to be considered. This involves a detailed assessment of your lung condition and other medical problems to determine if it is suitable.

### Clinical trials

- There are many new and promising treatments in development for the treatment of IPF. Your specialist may refer you for enrolment in a clinical trial, if appropriate.

### Pulmonary rehabilitation

- Pulmonary rehabilitation is a program of exercise training, education and support, that teaches you the skills you need to manage your breathlessness, to stay well and out of hospital. The program is provided by specially trained health professionals. To find a program near you, please contact Lung Foundation Australia.

## Other management options include:

### Staying active and healthy

- Quitting smoking, being physically active, eating well, getting plenty of rest, enjoying friends, family and hobbies, practising relaxation techniques, joining a support group, and keeping a positive attitude are all things you can do to support managing your IPF.

### Ensuring your vaccinations are up-to-date

- This may include discussing COVID-19 vaccines, seasonal influenza vaccinations and a five-yearly pneumonia vaccine with your doctor in order to help support your immune system. Prompt treatment of respiratory infections is also very important.

### Accessing emotional support

- Anxiety and depression are commonly experienced by people with IPF, and it is important to recognise and treat any symptoms in order to optimise wellbeing. Talk to your doctor or contact Lung Foundation Australia for referral to an appropriate support service.

## FURTHER INFORMATION AND SUPPORT

### Lung Foundation Australia services

- Information and Support Team
- Lung disease information resources
- Education webinars
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- E-newsletter

For more information visit lungfoundation.com.au/PF

### External links

**Centre for Research Excellence in Pulmonary Fibrosis**
www.cre-pf.org.au

**Australasian Interstitial Lung Disease Registry**
www.sydney.edu.au/medicine-health/our-research/research-centres/aildr.html

**Pulmonary fibrosis Australasian Clinical Trials (PACT) Network**
pact.lungfoundation.com.au

**Pulmonary Fibrosis Foundation**
www.pulmonaryfibrosis.org

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