

Idiopathic Pulmonary Fibrosis (IPF)



Lung
Foundation
Australia

when you can't breathe... nothing else matters®

Understanding your diagnosis: The way forward ...

Overview

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

This scarring makes the lungs stiff which makes it increasingly difficult to breathe. This slows the efficient 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, hacking 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.

Experience

It is important to know that each person experiences IPF differently. Some people can remain stable for many years; others may 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, all people with IPF do decline over time.



I got to the point in my IPF diagnosis where the only option was to accept it, and then seek all the information I could to develop a plan to tackle this imposition on my life.

- Bill Van Nierop, IPF patient Brisbane



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 middle aged to older people and is more common in men than women.

How common is IPF?



Although IPF is rare, you are not alone. In Australia, there is a conservative estimate of approximately 1,250 people being diagnosed with IPF each year, however currently it is not known exactly how many people are affected.

Treatment

The treatment and management of IPF is based on each person's individual medical and social situation. It is important to talk to your doctor and health care 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.

Please see an overview of treatment and management options in the next section.



Current treatment options include:

- **Medicines**

There are currently two anti-fibrotic medicines - Pirfenidone and Nintedanib - which have been shown to help slow the rate of disease progression. They are subsidised by the government and are available to patients that meet certain eligibility criteria. To see if you might be eligible for these medicines, please speak to your specialist.

- **Oxygen therapy**

Oxygen therapy may be prescribed to assist with shortness of breath and to help you stay active. Some patients only use oxygen when they feel breathless (e.g. walking or exercising) but other patients need to use oxygen continuously during the day and night.

- **Lung transplantation**

In some cases, complicated treatments like lung transplant surgery may need to be considered depending on other health conditions.

Self-management options include:

- **Pulmonary rehabilitation**

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

- **Staying active and healthy**

Quitting smoking, being physically active, eating well, getting plenty of rest, enjoying friends, family and hobbies, practicing 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 seasonal influenza vaccinations and a 5-yearly pneumonia vaccine with your doctor in order to help support your immune system.

- **Accessing emotional support**

Anxiety and depression is not uncommon in IPF and it is important to access support. Talk to your doctor or contact Lung Foundation for referral to an appropriate support service.



FURTHER INFORMATION AND SUPPORT

Contact Lung Foundation Australia for more information, to access our support services and join our mailing list for regular updates and latest news.

Lung Foundation Australia Services

- Information and Support Line
- Lung disease information resources
- Education seminars and webinars
- Lung Cancer Support Nurse
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- Newsletter

External Links

- **Pulmonary Fibrosis Foundation**

www.pulmonaryfibrosis.org

- **Canadian Pulmonary Fibrosis Foundation**

www.cpff.ca

Produced in collaboration with:



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