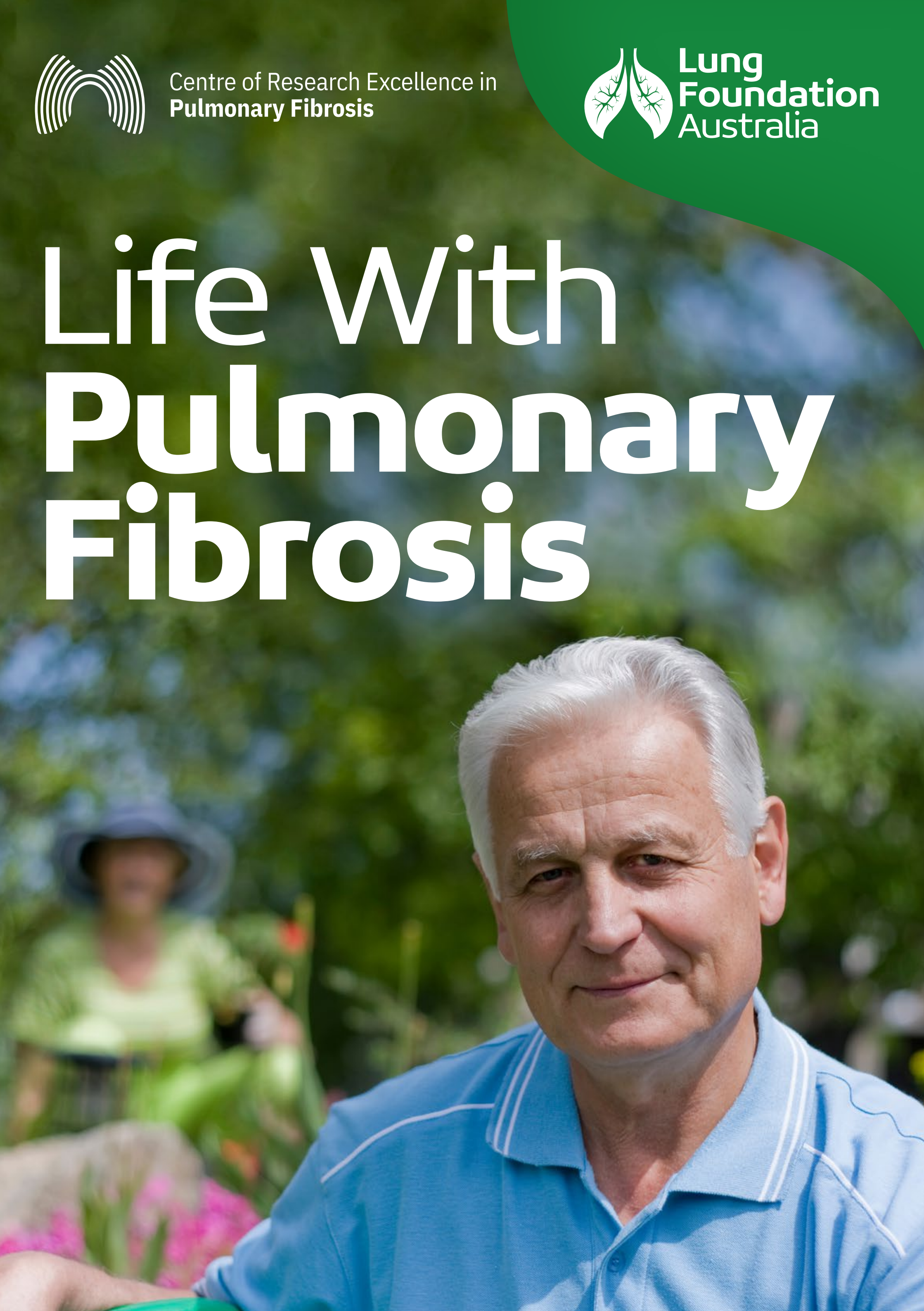
Centre of Research Excellence in
Pulmonary Fibrosis

Lung
Foundation
Australia

Life With
Pulmonary Fibrosis



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Introduction

This booklet contains information for people who have been diagnosed with Pulmonary Fibrosis (PF) and their families and carers. It has been designed to help you understand more about PF, how to manage your condition and help you to live a better life with PF.

The information has been developed by healthcare professionals from the Centre of Research Excellence in Pulmonary Fibrosis and people living with PF. It is a general guide and does not replace the information provided by your healthcare team.

There are many different types of PF. This booklet provides information that is common across the different types. For information that is specific to you and your diagnosis, it is important that you speak with your healthcare team.

What is Pulmonary Fibrosis?

Pulmonary Fibrosis (PF) causes the tissue around the air sacs within the lungs (alveoli) to become thickened and scarred – this is called fibrosis. This scarring causes the lungs to stiffen, making it difficult to transport oxygen and carbon dioxide in and out of the body. With mild PF, you may not notice a difference in how your lungs function. However, in some types of PF, the scarring can get worse over time.

PF can be seen in many types of Interstitial Lung Diseases (ILD). The many causes of PF are summarised in figure 1 below. "Idiopathic" diseases such as Idiopathic Pulmonary Fibrosis refer to specific diseases where the underlying cause is unknown.

Figure 1. Causes of PF

Pulmonary Fibrosis

Pulmonary Fibrosis with known causes	Pulmonary Fibrosis with no known causes (idiopathic)	Other
Examples	**Examples**	**Examples**
Secondary to autoimmune disease (e.g. rheumatoid arthritis, scleroderma) Secondary to non-organic exposures (e.g. heavy asbestos, silica, certain medications) Secondary to organic exposures (e.g. hypersensitivity pneumonitis from mould or birds) Secondary to smoking (e.g. pulmonary Langerhans cell histiocytosis)	Idiopathic Pulmonary Fibrosis (IPF) Idiopathic Non-Specific Interstitial Pneumonia (NSIP) Cryptogenic Organising Pneumonia (COP)	Sarcoidosis Lymphangioleiomyomatosis (LAM)

More information about specific types of PF is available in our other resources:

Idiopathic pulmonary fibrosis (IPF) fact sheet.

Sarcoidosis of the lung fact sheet.

Scleroderma with interstitial lung disease fact sheet.

Connective tissue disease-associated interstitial lung disease (CTD-ILD) fact sheet.

Hypersensitivity pneumonitis fact sheet.

Scan the QR code to access our full range of PF resources.

Diagnosis

Many people with PF receive a diagnosis only when they have had symptoms for some time. Early in the disease, symptoms can be subtle or mild which can make it difficult to detect. Early symptoms of PF may include cough, breathlessness and fatigue.

People diagnosed with PF are often older. It can be easy to put the symptoms down to ageing or being unfit. Older people tend to have other medical conditions, such as joint or heart problems, which can also mask the symptoms. Similar symptoms can also occur in other conditions, such as Chronic Obstructive Pulmonary Disease (COPD), asthma and heart disease. It may take some time before these other conditions are ruled out and a firm diagnosis of PF is made.

> *My initial symptoms had been classed as insignificant and my breathlessness I put down to not being fit enough. I'd had a cough for several years, but it hadn't really led to anything.*
>
> **Marcia, lives with IPF, QLD**

There are more than two hundred different types of PF. To help determine the most appropriate treatment options for your condition, it is important for your healthcare team to try and identify the underlying type of PF. Despite thorough investigation however, approximately 10% of PF remains unclassifiable, with no specific underlying type being able to be identified.

To diagnose PF, detailed examinations and investigations are required. Your healthcare professional will listen to your chest to see if they can hear fine crackles in your chest that sound like Velcro. These can be a valuable sign in diagnosis.

Your specialist doctor will carefully investigate your condition and some of the tests they will use include:

Blood tests

This is mainly to look for known causes of lung scarring such as autoimmune diseases.

Lung function (breathing) tests

To measure how well your lungs are working. This is also an important measure of how your disease is progressing over time.

Chest X-ray

To look for any signs of scarring although this does not always show the disease, especially in the early stages.

High Resolution Computed Tomography (HRCT) chest scan

This is a CT scan that uses specific techniques to provide very detailed pictures of your lungs. The findings on a HRCT scan are critically important when trying to determine which type of PF you have.

Lung biopsy

This might be required in some cases, however in many cases your specialist team can make a confident diagnosis without a lung biopsy.

There are other common signs often found in people with PF. These include:

A reduced oxygen level measured on a finger probe

Fingernail or toenail changes (known as clubbing)

Occasionally blue lips or tongue

Some people with PF may already have features of other associated conditions such as rheumatoid arthritis or scleroderma. In these conditions, changes in the joints, muscles and skin may also be present.

Once the results are available, a specialist team of healthcare professionals (such as: Respiratory Specialists, Radiologists, Pathologists, Rheumatologists, and/or Immunologists, and ILD nurses) may meet together in a Multidisciplinary Meeting (MDM) to determine your final diagnosis.

> *The diagnosis came following complaints to my GP of bouts of breathlessness when doing manual work such as mowing the lawn. I was finally diagnosed after a series of X-rays, CT scans, and a lung biopsy.*
>
> **Jim, lives with IPF, VIC**

Associated medical conditions

Scan to read the Other health conditions and PF fact sheet.

Studies have shown that people with PF are commonly diagnosed with other medical conditions that can impact their quality of life and health outcomes. The most common risk factor for these associated medical conditions is advancing age, but smoking history and being outside the healthy weight range can also play an important role.

Commonly associated lung conditions:

- Chronic Obstructive Pulmonary Disease (COPD), including emphysema – narrowing of the airways in the lungs
- Pulmonary Hypertension – high blood pressure in the lungs
- Obstructive Sleep Apnoea – repeated episodes of partial or complete closure of the throat during sleep.

Less commonly associated lung conditions:

- Lung cancer
- Pulmonary embolism - blood clot/s in the lungs.

Your healthcare team member or specialist doctor will take a detailed medical history and examination and arrange tests to help detect if you also have other medical conditions.

Other medical conditions that may occur along with PF but are not lung related may include:

- Gastro-oesophageal reflux disease (GORD)
- Coronary artery disease
- Depression and anxiety.

Managing these conditions is important for looking after your PF. It can make a big difference to how you feel and function. Speak with your healthcare team to develop a plan that takes into account all of your other conditions and your overall health and wellbeing.

How will my disease behave in the future?

Your future or the “prognosis” of your disease is very important to understand, but it can be difficult to predict.

PF affects each person differently, and the course of the disease is very specific to each individual. It is influenced by factors beyond your diagnosis of PF, including the specific pattern or type of PF, severity of your disease, and other medical conditions. Information about your prognosis is best provided by your specialist doctor, who can take into account your diagnosis, as well as other important factors that may influence your outlook.

PF is a relatively rare lung disease, and although research is growing, many unanswered questions remain. New research is ongoing into blood, breath and CT based biomarkers which can help doctors predict prognosis for an individual.

It is important to seek out sources of information you can trust. When you read information on the internet it can be hard to know if it relates to your specific disease and may make you worry. You should always talk to your specialist doctor for information and advice about your personal circumstances.

Monitoring PF

Regular monitoring of your condition helps your healthcare team to assess whether your disease is progressing, if treatments are working and whether there are other treatments available that could improve how you feel and function. In addition to checking your symptoms and clinical examination, tests commonly used for monitoring include:

Lung function testing

Breathing tests measure how well your lungs are working. They are important indicators of how your disease is progressing over time.

At least two tests will generally be done each time you visit the respiratory laboratory – spirometry and diffusing capacity.

- **Spirometry:** This test measures how much air can be blown out of your lungs and how quickly the lungs empty. This test requires your best effort at blowing out and can be a little uncomfortable but not painful. It may be repeated several times to get your best result.
- **Diffusing capacity (DLCO):** This test reflects the ability of your lungs to transfer oxygen from the air into the bloodstream. You will take in a deep breath of a special gas mixture, hold your breath briefly and then breathe out normally.

Exercise test

The most common exercise test used is the 6-minute walk test which provides important information about your walking capacity and your oxygen levels during exercise. The aim is to walk as far as possible in six minutes whilst your heart rate and oxygen levels are monitored. The test is conducted in a corridor under the supervision of a healthcare professional.

You will sometimes be asked to do a 6-minute walk test at your clinic appointments, so your specialist doctor is aware of any changes in your walking capacity or oxygen levels. For this reason, it is a good idea to wear comfortable shoes to the appointment. More recently, other exercise tests including the 1-minute sit to stand test are also being used at some centres.

High Resolution Computerised Tomography (HRCT) chest scan

This is a CT scan that uses specific techniques to provide very detailed pictures of your lungs. Your doctor will use HRCT scans of your lungs to look for changes, new complications, or worsened fibrosis.

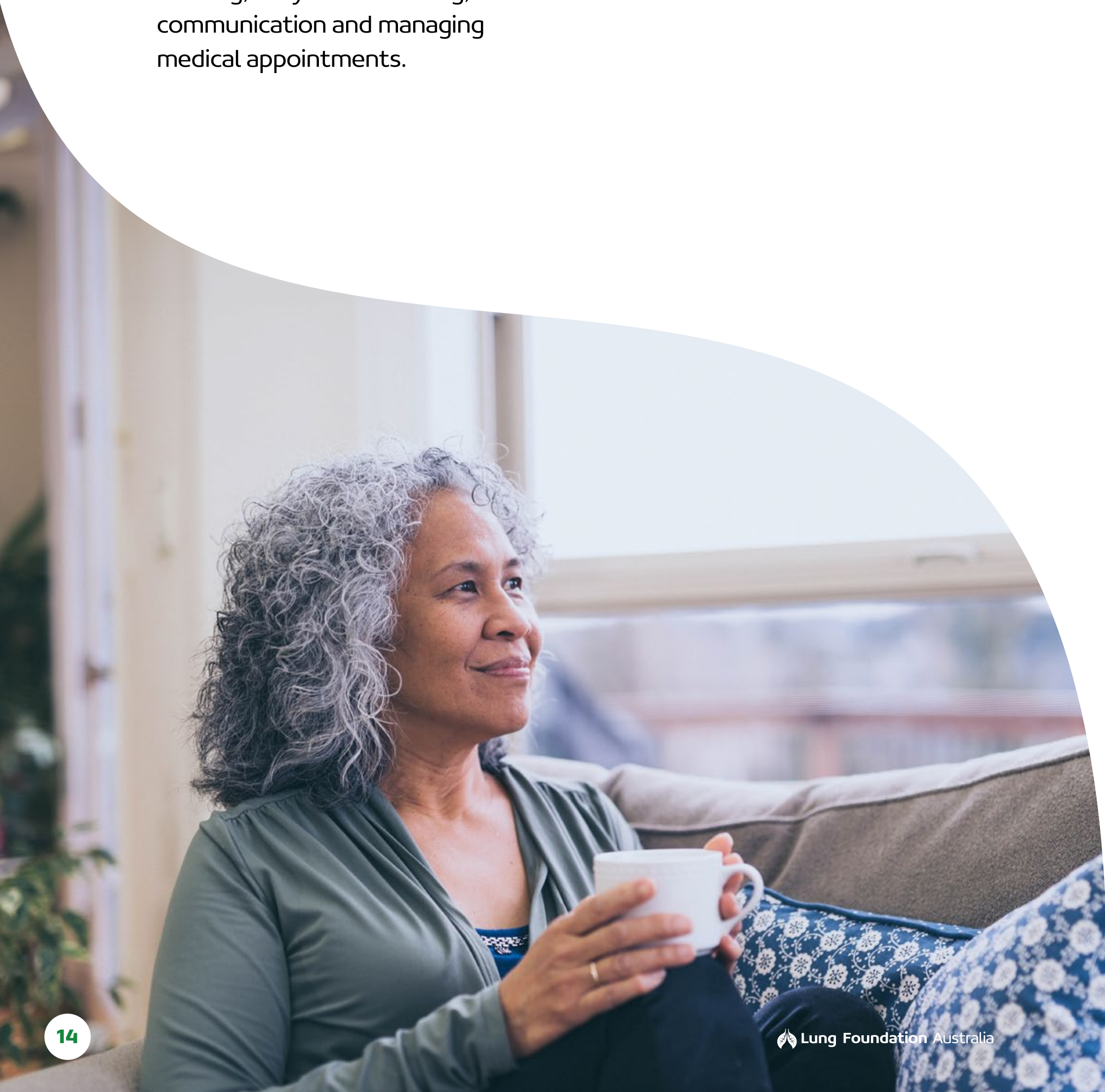
Your treating healthcare team

People with PF may have a Multidisciplinary Team caring for them throughout their disease journey. Not every treating team will be the same, or stay the same over time. Each individual member of your healthcare team will have input depending on the requirements of your disease and your individual needs.

Your healthcare team may include:

- A **Respiratory Specialist** is a doctor who has been specially trained in the diagnosis, treatment and management of PF.
- **Respiratory nurses / ILD nurses** have special training in PF and ILD. They provide evidence-based care, information and guidance, emotional support and referral advice to local services.
- **General Practitioners (GPs)** are qualified in general practice medicine. GPs are often your first point of contact with the healthcare system when feeling unwell. They provide long-term, continuous care, and can refer you to specialists.
- **Physiotherapists** use physical techniques to improve movement, reduce pain and stiffness and increase quality of life.
- **Dieticians** provide advice regarding healthy eating and optimal nutrition.
- **Pharmacists** are responsible for advising you about your medicines, including how to take them, what reactions may occur and answering your questions.
- A **Respiratory Scientist** will conduct lung function tests such as spirometry and DLCO.

- **Psychologists** explore and talk to you about how you are coping with your circumstances. They can then help you to develop strategies to address problems and worries.
- **Social workers** focus more on “outward” problems around practical and social support like housing, daily tasks of living, communication and managing medical appointments.
- There will also be **Radiologists** and **Pathologists** working behind the scenes that will be actively involved in your initial diagnosis and monitoring your progress.
- Situationally required specialists such as: **Rheumatologists, Immunologists or Palliative Care Specialists**.



Treatment for PF

There is no cure for PF, but there are various treatment options and management strategies to help stop or slow the progression of the condition and manage symptoms. If you also have other conditions, it is important to work with your healthcare team to build a plan for treating PF alongside those conditions.

The following information is a general overview of some of the treatments your specialist doctor can offer for PF. **Please note, this information is not medical advice.** Some treatments may be right for some people, but no one treatment is right for everyone. To ensure you are receiving the best treatment for you, speak with your specialist doctor before starting, changing or stopping any medical treatment.

Anti-fibrotics

Pirfenidone and nintedanib are anti-fibrotic medications that may help reduce the rate of progression of scarring in your lungs. Antifibrotics are used if you have been diagnosed with IPF or another type of PPF.

Scan to read the **Antifibrotics fact sheet.**

While IPF or PPF will still worsen over time, using pirfenidone or nintedanib may help slow or delay this progression. Currently, no medications exist that will improve symptoms or reverse scar tissue in the lungs once it has formed. Pirfenidone and nintedanib are subsidised by the Australian government for people with PF who meet certain eligibility criteria.

Side effects

Like most medications, side effects are relatively common with anti-fibrotics.

- **Nintedanib:** The most common side effect people may experience is diarrhoea. For some people, anti-diarrhoeal medications can be used with good effect. Other gastrointestinal side effects that occur less frequently include nausea, vomiting and loss of appetite. Liver function can be affected by nintedanib and is monitored by regular blood tests.
- **Pirfenidone:** People on pirfenidone may experience skin reactions from exposure to the sun. All patients taking pirfenidone should use SPF 50 sunscreen and other protective measures from the sun. Other common gastrointestinal side effects of this medication are nausea, indigestion, loss of appetite, vomiting and weight loss. Liver function can also be affected by pirfenidone and is monitored by regular blood tests.

Scan to read the **Nintedanib fact sheet.**

Scan to read the **Pirfenidone fact sheet.**

Speak with your healthcare team about ways to best avoid or manage any side effects that you may experience. Your healthcare team may change the dosage of your treatment, suggest trying anti-nausea or anti-diarrhoea medications or dietary changes, and on some occasions switch to the alternate anti-fibrotic. Each person will have individual differences with side effects, so some trial and error may be involved. Work with your healthcare team to develop the best plan for you.

Immunosuppressants

Some forms of PF are caused by inflammation in the lung tissue, often related to autoimmune diseases such as rheumatoid arthritis, scleroderma, Sjögren's syndrome, dermato/polymyositis and hypersensitivity pneumonitis. These inflammatory conditions are often treated with immunosuppressant medications. These medications help control your body's immune system and reduce inflammation in your lungs. By reducing inflammation, immunosuppressants may prevent the formation of more lung fibrosis, and, in some early cases may allow the inflamed lung to return to normal.

Immunosuppressant medications can decrease your ability to fight infection. You need to be alert to the signs of infection and see your doctor immediately if you are concerned.

Before starting immunosuppressant medications, your healthcare team will often perform blood tests to assess for risks of 're-activating' of any dormant infections such as hepatitis and tuberculosis and ensure that you are up to date in your vaccinations.

Commonly used medications include:

- **Prednisolone**
  - **Side effects:** Prednisolone is an effective and powerful drug which is associated with side effects such as increased appetite (and resulting weight gain), fluid retention, gastroesophageal reflux, mood and sleep disturbance, diabetes mellitus, skin thinning and osteoporosis. Side effects are seen especially when used at high doses for long periods of time.
  - **Special precautions:** Prednisolone should not be stopped abruptly. Always check with your specialist doctor before changing the dose.
- **Mycophenolate mofetil (MMF):**
  - **Side effects:** Can cause diarrhoea, abdominal pain, vomiting, muscle aches, abnormal liver function and skin rashes.
  - **Special precautions:** Before taking this medication, you should consult with your doctor if you plan to become pregnant or breastfeed. Regular blood tests are required on this medication for full blood count, kidney and liver tests. Regular skin checks are also required.

Scan to read the **Mycophenolate fact sheet.**

- **Azathioprine (AZA) :**
  - **Side effects:** Can cause diarrhoea, vomiting, abdominal pain, muscle aches, abnormal liver function and an increased sensitivity to sunlight.
  - **Special precautions:** Before taking this medication, you should consult with your doctor if you plan to become pregnant or breastfeed. Regular blood tests are required on this medication for full blood count, kidney and liver. Regular skin checks are also required. You will be checked for a specific enzyme to make sure this medication is appropriate.

Scan to read the **Azathioprine fact sheet.**

Several other immunosuppressant therapies are available and are used to treat different forms of PF. These include methotrexate, cyclophosphamide, rituximab, cyclosporine and tacrolimus. Based on your type of PF, your specialist doctor will tailor their selection of the different medications. Your healthcare team will work with you to manage side effects and maximise benefits from immunosuppressant treatments for PF.

Lung transplant

Lung transplant may be a treatment option for some patients with PF, but not suitable for others. Successful outcomes following transplantation vary according to age, other disease burden and severity of PF. In general, whilst there may be small differences between transplant units in Australia, lung transplant is rarely performed above the age of 70. There are many potential medical complications associated with the procedure including rejection of the organ and infection related to the use of anti-rejection medication.

Over 80% of people will live for a year after transplant, and around 50% of people who have a transplant are alive after approximately six years. Your specialist doctor will provide guidance on whether transplantation is an option and whether you should be referred to your local lung transplant unit.

Scan to read the **Lung transplant fact sheet.**

Clinical trials

A clinical trial is a research study designed to evaluate the success of a new treatment to prevent, stabilise or reduce the effects of a disease. Clinical trials are particularly important when treatment options are limited, as is still the case for PF, and involves the voluntary participation of individuals. Each individual's response to the treatment administered is monitored closely in a clinical trial. This research is then collected and used to further the development of these treatments, in turn, helping those affected by PF now and in the future.

Scan to read the **Clinical trials fact sheet.**

The success of clinical trials has been a major factor in the development of treatment options for people with PF. There are multiple PF clinical trials ongoing in Australia at any one time. Ask your treating specialist about PF clinical trials or visit the Pulmonary Fibrosis Australasian Clinical Trials (PACT) Network for information about current trials.

Scan to access the **PACT website.**

Acute exacerbations in PF

What is an acute exacerbation?

Some people with PF may experience a sudden worsening or intensity of their symptoms which is known as an acute exacerbation or a flare-up. This usually develops over one month or less.

If you experience an acute exacerbation, you may notice:

- Your breathing symptoms are worse than usual
- Difficulty performing tasks that seemed easier a month ago
- Worsening breathlessness or cough.

The cause of an acute exacerbation is not known. It can occur at any stage of your disease, even when you are feeling quite well. Possible events that might trigger an acute exacerbation include a chest infection, specific therapies such as radiation or anti-cancer drugs, and some surgical procedures. Acute exacerbations are more common in the winter season.

How is an acute exacerbation managed?

Your specialist doctor may perform investigations to find out why your symptoms have suddenly worsened. Along with your examination findings, additional tests such as a chest x-ray, CT scan, respiratory swabs and blood tests, may assist in excluding other causes for your worsening symptoms.

Management of an acute exacerbation may include:

- Targeted treatment depending on any underlying causes and your symptoms
- Supportive therapy for feelings of breathlessness
- Time in hospital.

After an acute exacerbation, you may require a long period of recovery and in some cases, you may need to continue using oxygen therapy at home. It may be difficult to return to the same level of breathing function you had before.

How do you reduce your risk of an acute exacerbation?

Whilst it is not always possible to avoid an acute exacerbation, there are ways to minimise the risk:

- Attend all your specialist appointments and follow through with all investigations
- Avoid infections by:
  - Having up-to-date vaccinations
  - Avoiding contact with people, especially children, with respiratory tract infections (i.e. “colds and flu”)
  - Practicing good hand hygiene – wash your hands regularly with soap and water
- Treat chest infections early. Early intervention is crucial. You may need early treatment with antibiotics if you develop a bacterial chest infection
- Discuss any planned surgery with your specialist doctor
- Ensure other medical specialists are aware of your PF
- Contact your specialist doctor if you notice a sudden worsening in your symptoms. This allows for investigations and early treatment
- Maintain a healthy lifestyle for your general wellbeing - see the Living Well with PF section on page 30 for further information.

All people with PF are at risk of having an acute exacerbation. An acute exacerbation will look different for everyone, depending on the type of PF you have. You are encouraged to discuss what to expect during an acute exacerbation with your healthcare team.

Managing your symptoms

Breathlessness

People with PF report that breathlessness is one of their more distressing symptoms. Breathlessness often starts with exertion such as walking, and may progress to breathlessness performing daily activities, such as showering, getting dressed or speaking on the phone and at rest. There are a range of possible causes or contributors to breathlessness in PF, including scarring of the lung tissue itself, other lung or health conditions, weakening of breathing muscles, contaminants in air and anxiety.

Scan to read the **Cough and breathlessness booklet.**

Breathlessness can be frightening. If you are feeling out of breath, it is important to talk with your treating healthcare team. They can suggest ways to help you manage your breathlessness, which may include breathing techniques, pulmonary rehabilitation (see page 24 for further detail) or oxygen therapy (see page 25 for further detail). If you experience any anxiety from your breathlessness, discuss this with your healthcare team as there are trained professionals who can provide help and support in managing this.

If your breathlessness remains serious and persistent, follow up with your specialist doctor as a priority. If you are experiencing shortness of breath which becomes severe, do not hesitate to call an ambulance.

Cough

People with PF often report an irritating cough as an early symptom. The cough is usually dry in nature and may first be noticed as a cough that doesn't get better after a chest infection. Other people with PF may report an intermittent cough or throat irritation as the first symptom they notice. If the condition progresses, the cough may become more severe. Some people with PF experience debilitating coughing fits, often associated with episodes of breathlessness.

Whilst cough can be a symptom of PF, it can also occur for different reasons that can be identified and managed. Check with your healthcare team if other conditions, like heartburn or hay fever could be making your cough worse. Ask your specialist doctor if there are any management strategies, such as cough suppression, or prescription medications to help reduce your cough.

Fatigue

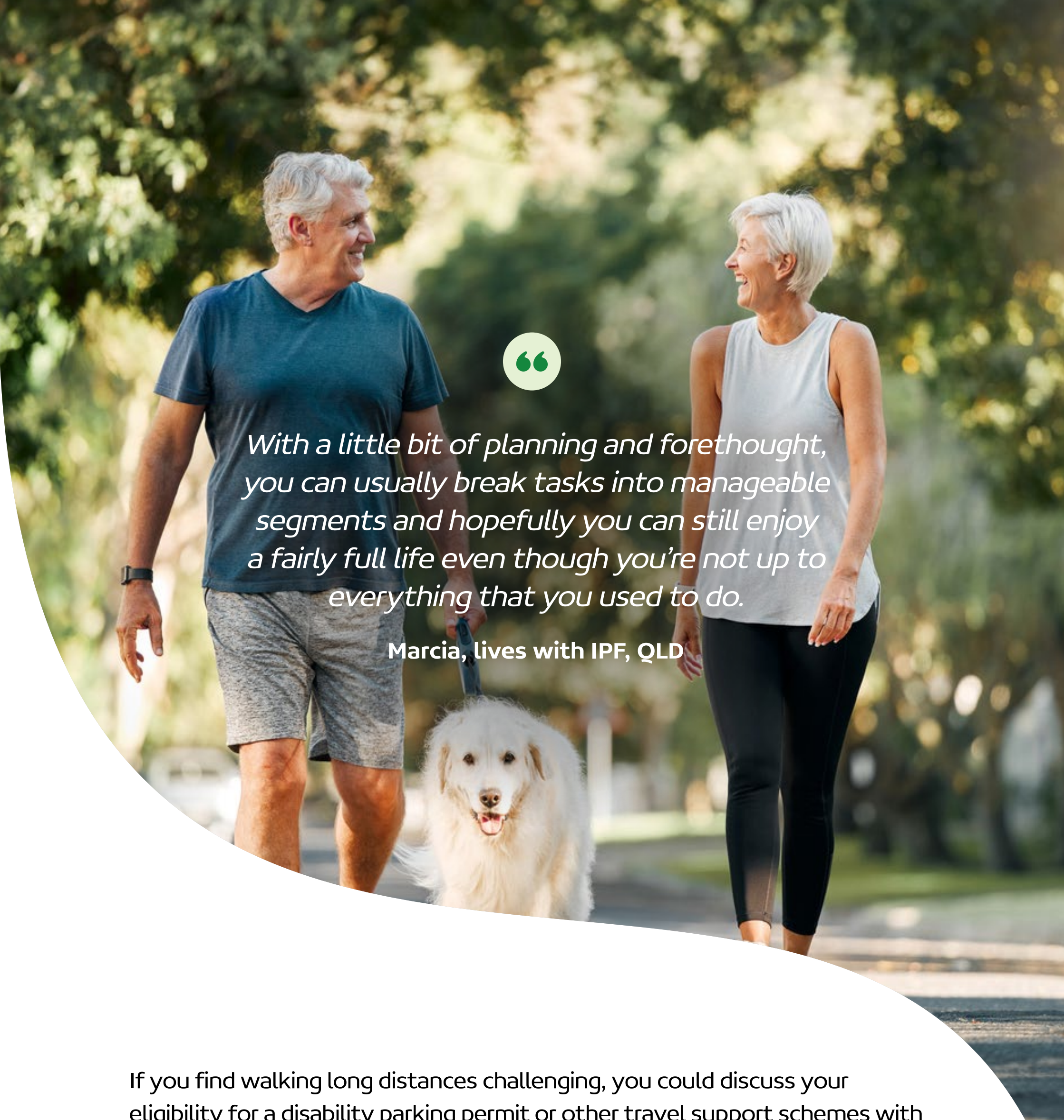
It can be difficult to tell the difference between fatigue (tiredness) and breathlessness on exertion. People living with PF often describe days where they have a total lack of energy and feel completely exhausted. In the early stages, people may find the fatigue is associated with a mild loss of appetite and losing weight without trying. It can affect the way you think and feel, or impact your concentration and memory.

Getting around with PF

Living with PF, you may find it much more difficult to undertake everyday activities such as showering, walking, household chores, shopping or social outings. Rather than avoiding activities, try breaking them into smaller parts, so you can take rests and pace yourself. You may have heard this called “staging” your activities. By resting in between parts of activities, you give your body a chance to “catch up” before continuing on.

Practical tips to help you get around with PF:

- Plan ahead and ensure you have enough time and places to rest
- Split a larger task up into three to four smaller tasks to allow for breaks in between
- Listen to your body and allow yourself to rest when needed
- Let your friends and family know you might need to walk slowly or take breaks. They will appreciate knowing how they can help you
- Talk to your healthcare team about daily living and mobility aids which may be available to you.



> *With a little bit of planning and forethought, you can usually break tasks into manageable segments and hopefully you can still enjoy a fairly full life even though you're not up to everything that you used to do.*
>
> **Marcia, lives with IPF, QLD**

If you find walking long distances challenging, you could discuss your eligibility for a disability parking permit or other travel support schemes with your GP. Walking aids can also provide support and assistance with walking, standing, and balance. Generally, a physiotherapist or occupational therapist will assess your mobility to determine if a walking aid would be beneficial for you, and if so, which type of equipment would be best suited to your needs.

Deconditioning

To avoid the challenges of living with PF, including the unpleasant sensation of breathlessness, it is very common for people with PF to do less activity over time. This is understandable, but in the longer term it results in loss of strength and physical fitness (known as deconditioning). Reduced strength and fitness mean the body has to work harder just to accomplish normal daily activities, which in turn results in more breathlessness and fatigue.

> *Currently I am involved in a good exercise regime with my friends. The main thing that needs discipline is on ascending slopes where I need to slow my pace in order to achieve the target without undue stress.*
>
> **John, lives with IPF, WA**

Staying as fit and active as possible will help to avoid deconditioning - see the Living Well with PF section on page 30 for further information. Your healthcare team may suggest attending your local pulmonary rehabilitation program.

Pulmonary rehabilitation

Scan to read the **Exercise and PF fact sheet.**

Pulmonary rehabilitation is a supervised program of exercise, education and behavioural change designed specifically for people with a chronic lung condition. The aim is to help you to become as fit and strong as possible, and to increase your confidence in managing your breathlessness and your PF.

Programs are generally conducted in a hospital or community health centre for six to eight weeks, with two sessions each week. Before you start you will be thoroughly assessed by the pulmonary rehabilitation team, which usually includes a 6-minute walk test and questionnaires. Then the physiotherapist or exercise physiologist will design an exercise program that suits you. It will start slowly and progress as you feel able. You will be supervised at all times. Research shows that for people with PF, pulmonary rehabilitation is very effective in improving exercise capacity, reducing breathlessness and increasing wellbeing.

Ask your healthcare team about the possibility of a referral to your nearest pulmonary rehabilitation program. You can also find a directory of pulmonary rehabilitation programs on the Lung Foundation Australia website. If a pulmonary rehabilitation program is not available in your area, telerehabilitation options may be available and/or a physiotherapist or exercise physiologist can develop an exercise program that is right for you. Speak with your GP about your eligibility for a chronic disease management plan to access subsidised visits to a physiotherapist or exercise physiologist.

Oxygen therapy

Your specialist doctor may prescribe you oxygen therapy if you develop low levels of oxygen in your blood. You may only need to use oxygen during the day with exertion/ exercise. However, as the disease progresses, you may also need to use oxygen at night when sleeping. Oxygen may be needed all the time at the later stages of the disease.

To determine if you would benefit from home oxygen, your specialist doctor may measure your oxygen levels in a few different ways:

- **Oximetry** - A small, clip-on probe which is placed on your finger or ear lobe
- **Arterial blood gas** - Blood sample taken from an artery, usually at your wrist.

Oxygen equipment

Oxygen is normally delivered through nasal prongs which are two small, soft curved plastic tubes that go into your nostrils and are attached to a lightweight plastic tubing. Nasal prongs allow you to eat and talk while receiving oxygen therapy. The nasal prongs are then attached via tubing to a home oxygen concentrator, a portable oxygen concentrator or a cylinder of oxygen. Rarely, a mask may be used instead of nasal prongs.

Not all oxygen machines may be compatible with your prescription. Discuss the suitability of different oxygen equipment with your healthcare team and oxygen supplier.

Home oxygen concentrator

- A medium-sized machine that draws in air and concentrates that air to give you greater than 90% oxygen. You can adjust the flowrate (litres/min) according to the prescription from your specialist doctor.
- Requires electricity to operate.
- A length of plastic tubing attached to your nasal prongs will allow you access to all parts of your home including the shower.

Portable oxygen concentrator

- Smaller versions of the home concentrator that are designed to be taken out of the house.
- Powered by batteries that require recharging.
- There are different types of machines with various sizes and weights that deliver different amount of oxygen flow.
- Smaller, compact machines may not be compatible with your prescription. Discuss the suitability of a portable oxygen concentrator with your healthcare team and oxygen supplier.

Oxygen Cylinders

- Compressed pure oxygen in metal cylinders that can be refilled or replaced once used.
- Depending on the size of the cylinder and the flow rate (litres/min) a cylinder may last two to four hours before requiring a refill.
- To help carry the cylinder, you can buy or rent a backpack or trolley or perhaps place it in the basket of a walker.

Practical considerations

- Smoking is not permitted around people using oxygen therapy
- Oxygen is highly flammable so avoid gas cooking flames, candles or any other source of open flame
- Oxygen tubing poses a significant trip hazard. Be aware of risks to yourself and others
- Consult your oxygen provider for further safety information
- Develop an emergency management plan for power outages with your treating healthcare team
- Some states and territories have government-assisted concessions to support with energy costs to run essential medical equipment. Speak with your treating healthcare team to see if you qualify.

Starting oxygen therapy can be overwhelming and concerning for some people. Over time you will get used to making it part of your life. You may find you feel less breathless, less tired and more able to do some of the things you enjoy the most.

Scan to read the **Oxygen therapy booklet.**

Palliative care

Palliative care is about living as well as possible with your condition, not only end-of-life care. The goal of palliative care is to reduce your symptoms at any stage of living with PF and improve your quality of life. Some people with PF may benefit from palliative care because it focuses not just on your lung health but on you as a whole person, including physical, mental, emotional and social aspects. Palliative care can be provided to people of any age and at any stage in their disease, including right from the time of diagnosis. Research shows that people who are linked with a palliative care team early on in their condition have better symptom management throughout their journey. Palliative care teams are multidisciplinary in nature and will often change their level of involvement depending on your need at the time. They may provide support in the community, including at your home, if required.

You can discuss the role of palliative care with your treating healthcare team at any time, you don't have to wait for them to start the conversation. Taking an active role in how your condition is managed may help you to feel more empowered in decisions about your care and treatment.

Scan to read the **Palliative care fact sheet.**

Planning for the future

Scan to read the **Managing progression fact sheet.**

Although not an easy topic to think about or discuss, it is a good idea to make arrangements for your future so that your wishes are known. This will ensure you get the care you want when you need it. Such arrangements are known as advance care planning.

Regardless of your age and health, everyone should consider making an advance care plan. It outlines your values, and the future medical care you would or would not like to receive if you can't decide for yourself at the time. It means your opinion and choices are heard during end-of-life care, and helps to relieve some of the pressure and stress from your loved ones to make decisions on your behalf.

If you find it difficult to talk about these topics with your family, don't be afraid to ask for help from your specialist doctor or treating healthcare team.

Living well with PF

Living with PF may significantly change life for you and your loved ones, even in simple ways such as needing to attend medical appointments more often. As the symptoms of PF increase, many people discover that their roles in life are also affected. Working together with your treating healthcare team, wider support team (friends and family) as well as taking positive actions to live a healthy lifestyle will help you stay prepared for some of the challenges you may face living with PF, allowing you to live your life to its fullest.

Maintaining a healthy mind

Being diagnosed with PF can be an emotional time, for both the person with PF and their loved ones. Whilst everyone processes their feelings differently, it is normal to feel a mixture of emotions after the initial diagnosis. If you have experienced a long period of investigation, you may experience relief that you have a diagnosis, mixed with feeling uncertain about the future. You may be getting used to new medications, regular doctor visits and lifestyle changes.

There may be times when you feel quite good. There may be other times when you feel flat and low, especially if you are experiencing worsening symptoms. Feeling flat and low during these times is part of the normal emotional reaction to living with a chronic condition. One of the best ways to cope with strong emotional reactions is to talk to someone you trust about how you are feeling and the thoughts that accompany those feelings. This might be family, friends or a healthcare professional, including your GP.

It's important to seek help from a mental health professional when these negative feelings cause significant distress and begin disrupting your life. Your GP is typically a good place to start, and you may be able to get a referral to see a mental health professional such as a psychologist or social worker.

Maintaining a healthy mind will help you to better maintain your physical health as well. Talking to people about your feelings, maintaining social connections and treating yourself kindly will all help to improve wellbeing.

Maintaining engagement with your life

Research shows that people with strong social connections have greater feelings of wellbeing. Staying engaged with things and people in your life that are important to you will also help you feel more optimistic and resilient. It can be helpful to talk with your loved ones, your GP or other healthcare team members about developing strategies that suit your situation, including how you will maintain your relationships or activities with friends and family.

> *An important message to remember is that you are more than just your Pulmonary Fibrosis diagnosis, that is just one part of who you are.*
>
> **Debra, Clinical Psychologist, SA**

Some people find that as their symptoms persist or worsen, it can become more challenging to get out and about and feel like being social. Generally, with some planning and maybe shorter, less strenuous outings, you will find it is worth the effort. Support groups complement formal support services and can significantly contribute to people coping with the challenges of living with, or caring for someone living with, a lung condition.

Peer support offers a platform for people to come together to share experiences, provide mutual support, and help encourage and guide each other. To learn more, and express your interest in joining a support group, contact our Information and Support Centre on free call 1800 654 301.

Scan to find out more about peer support.

> *It is incredibly important to be in touch with other patients who have PF because they have a better understanding of what you are going through perhaps than anybody else.*
>
> **John, lives with IPF, WA**

Maintaining a healthy body

There are a range of things you can do to help keep your body as healthy as possible while living with PF and be an active participant in your own healthcare.

- **Stay up-to-date with vaccinations**
  - Annual seasonal influenza vaccination
  - COVID-19 vaccinations
  - Pneumococcal vaccination
  - Other vaccinations may be recommended by your healthcare team, including RSV, Shingles, Pertussis.

Scan to read the **Non-pharmacological treatments booklet.**

People with PF have greater difficulty recovering from respiratory and other illnesses, so every attempt should be made to protect you with the available vaccinations.

- **Maintain a healthy diet**
  - Your general health is greatly impacted by your diet. It is best if your weight is within a healthy range. Enjoy a wide variety of healthy foods including fruit and vegetables, lean protein and calcium-rich foods such as dairy products according to your specific dietary requirements and situation. If you need further support, you can ask your GP or healthcare team to refer you to a dietitian.
  - You may find with some medications that your appetite is greater than usual (prednisolone) or less than usual (anti-fibrotic medication). Be mindful of trying to eat regular meals and drinking enough fluids. Speak with your specialist doctor if you are not sure how much fluid you should be drinking.
- **Stay active**
  - Pulmonary rehabilitation programs can be a great way to start incorporating exercise into your everyday life. Check out the pulmonary rehabilitation section in this booklet on page 24 and talk to your treating healthcare team about getting started.
  - A physiotherapist or exercise physiologist can also provide advice on an exercise program that is right for you.
  - Find physical activities that you enjoy to help you stay motivated.
- **Treatment of chest infections**
  - Work with your specialist doctor to develop a plan to follow in the event you get a chest infection. Ensure your GP is aware of this plan.
  - Report any changes in your breathing symptoms to your specialist doctor or GP as soon as possible. This will help them manage any chest infections or acute exacerbations in a timely manner.

Travel with PF

Many people with PF are still able to enjoy travelling, either locally or overseas. However, it is very important to discuss this with your specialist doctor because not all forms of travel are suitable for all people. You may need to do things a bit differently and plan ahead more, but it is important to maintain the things you love in your life.

Scan to read the **Preparing for travel booklet.**

Before travelling, you should ensure your vaccinations (for example, seasonal influenza, pneumococcal, tetanus, hepatitis, COVID-19) are up-to-date. While you're travelling it is essential you stay as healthy as possible. Pack all your medications in your hand luggage, and a written plan to follow if you experience an acute exacerbation. Make sure to wash your hands frequently, stay away from other travellers with respiratory illnesses and most importantly, schedule rest days to make sure you are not over-exerting yourself.

If you use oxygen therapy, your plans must accommodate this. Travelling by plane can be challenging because the available oxygen in the cabin is reduced. You may need to have a high-altitude simulation test to see how your body will cope with changes in altitude.

Some people with PF require oxygen while on a plane even if they don't usually require oxygen therapy. It is important to start the process of organising this as soon as possible and work closely with your airline or travel agent to complete the necessary paperwork. As with any travel, you should always take out travel insurance and list your PF as a pre-existing condition. You will need to pay extra for this policy and may need to shop around to find an insurer.

A diagnosis of pulmonary fibrosis can feel isolating, but you're not alone. Lung Foundation Australia, in partnership with the Centre of Research Excellence in Pulmonary Fibrosis, are here to provide you with the tools and information you need to live well with your condition. Scan the QR code below to access our full range of PF resources.

Scan to access PF resources and information.

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

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Centre of Research Excellence in
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