

Life With Pulmonary Fibrosis



Lung
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Australia



Centre of Research Excellence in
Pulmonary Fibrosis



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Introduction

This booklet contains information for people who have been diagnosed with Pulmonary Fibrosis (PF) and their carers. It has been designed to help you understand more about PF, how to manage your condition and help you to live better 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 your diagnosis, it is important that you speak with your healthcare team.



Centre of Research Excellence in
Pulmonary Fibrosis

The role of the Centre of Research Excellence in Pulmonary Fibrosis

Funded in 2017 by the Australian National Health and Medical Research Council, the Centre of Research Excellence in Pulmonary Fibrosis (CRE-PF) aims to build research capacity in the area of Pulmonary Fibrosis. The CRE-PF enables a nationally coordinated, clinically focused research program to address the urgent need for more effective, personalised approaches to identify and better manage fibrotic lung disease, as well as to educate the public and train future generations of researchers in Pulmonary Fibrosis. For more information on CRE-PF, visit cre-pf.org.au



The role of Lung Foundation Australia

As Australia's leading lung health charity, Lung Foundation Australia funds life-changing research and programs to reduce the prevalence of lung disease and improve support and care for all Australians. Lung Foundation Australia collaborates with the Centre of Research Excellence in Pulmonary Fibrosis, working together to drive patient and clinician involvement in Pulmonary Fibrosis research and trials via the Pulmonary Fibrosis Australasian Clinical Trials (PACT) Network and the Australian Idiopathic Pulmonary Fibrosis (IPF) Registry. For more information on PACT, visit pact.lungfoundation.com.au and for more on the IPF Registry, visit lungfoundation.com.au

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. Treatment for this type of PF aims to stop or slow down the scarring with medications, whilst also helping you manage your symptoms to ensure you feel as well as you can.

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 is a relatively rare lung disease and although research is growing, there are still many unanswered questions.

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

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

Diagnosis

Many people with PF do not receive a diagnosis until 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 generally 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.



For a few years prior to this (diagnosis), I noticed I was becoming very short of breath when walking up the stairs or hills. I kept saying to my doctor that it felt like I was getting asthma again.

Lyn, lives with PF, QLD

There are more than two hundred different types of PF. It is important for your healthcare team to identify the underlying type of PF, to help determine the most appropriate treatment options for your condition.

To correctly diagnose PF, detailed examinations and investigations are required. Your specialist doctor will carefully investigate your condition and some of the tests they will use include:



Physical examination

Your specialist doctor 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 early diagnosis.



Blood tests

This is mainly to exclude known causes of lung scarring.



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.



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.

Once the results are available, a specialist team of healthcare professionals (doctors, radiologists, pathologists) may meet together to determine your final diagnosis. This group of healthcare professionals is called a Multidisciplinary Team (MDT).



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

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)

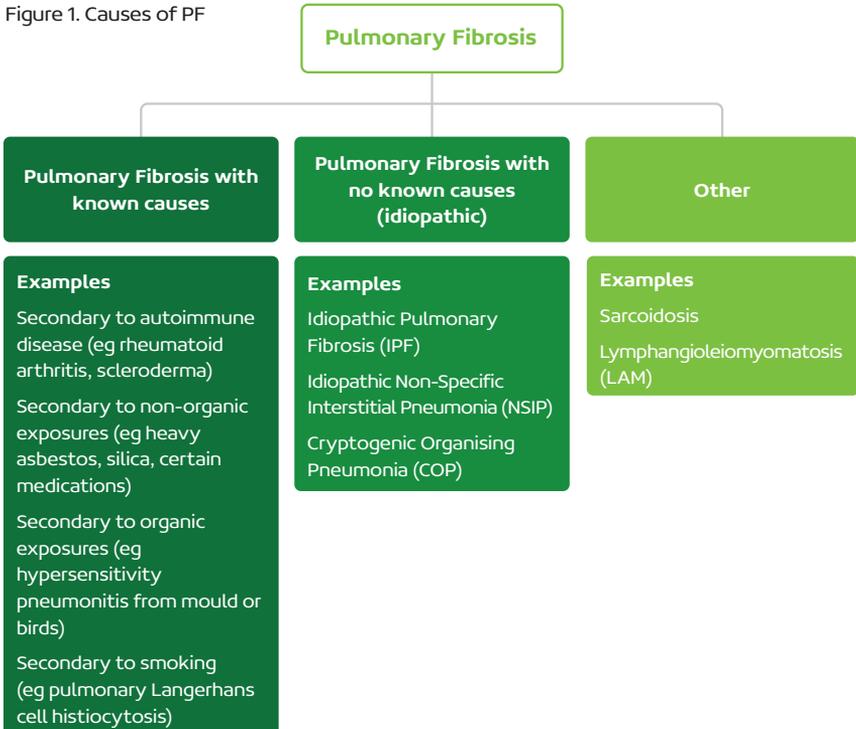


Occasionally blue lips or tongue

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.

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



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. 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. 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.

- **6-Minute Walk Test**

The 6-Minute Walk Test 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 can stop as often as you need.

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.

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 those conditions alongside your PF.

Treatment options

PF can result from a number of different causes. Understanding the cause of your PF will help your specialist doctor to decide which treatment is best.

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 particular treatment is right for everyone. To ensure you are receiving the treatment that best suits your condition speak with your specialist doctor before starting, changing or stopping any medical treatment.

Anti-fibrotics

Idiopathic Pulmonary Fibrosis (IPF) is a specific type of PF for which two anti-fibrotic medications are currently available in Australia. In patients with mild to moderate IPF (as judged by lung function tests), pirfenidone and nintedanib have been shown to slow down the progression of fibrosis within the lungs. While IPF will still worsen over time, using pirfenidone or nintedanib may help slow or delay this progression. Currently, no medications exist that are able to improve symptoms or reverse scar tissue in the lungs once it has formed. Both pirfenidone and nintedanib are subsidised by the Australian government for patients with IPF who meet certain eligibility criteria.

Side effects

Side effects are relatively common with anti-fibrotic medications but are usually mild and almost always completely resolve if treatment is ceased. The frequency of side effects experienced by participants in large clinical trials is described below.

- **Nintedanib:** The most common side effect people may experience is diarrhoea, which occurred in over half of the participants in clinical trials of this medication. Generally, it tends to be mild (less than four motions per day), although an anti-diarrhoea medication is sometimes needed. Other gastrointestinal side effects that occur less frequently include nausea, vomiting and loss of appetite. Liver function can be affected by nintedanib (in about 5% of participants in the clinical trials) and is monitored by regular blood tests.
- **Pirfenidone:** The most common side effect of this medication is nausea, which occurred in about a third of participants in clinical trials of this medication. Other gastrointestinal side effects that occur less frequently include indigestion, loss of appetite, vomiting and weight loss. About a third of people may also experience rash from exposure to the sun. Liver function can also be affected by pirfenidone (in about 4% of participants in the clinical trials) and is monitored by regular blood tests.

Speak with your healthcare team about ways to best avoid or manage any side effects you may experience. Your healthcare team may change the dosage of your treatment, suggest trying anti-nausea or anti-diarrhoea medication or dietary changes. 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.

Immunosuppression

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, dermatomyositis and antisynthetase syndrome. These inflammatory conditions are often treated with immunosuppressive medications. Commonly used medications include:

Prednisolone

- **Action:** A corticosteroid that suppresses the immune system. It works quickly and effectively for most conditions related to inflammation in the lung, however it can increase the risk of infections.
- **Side effects:** Prednisolone is a powerful drug and is associated with side effects such as increased appetite and weight, fluid retention, gastro-oesophageal reflux, mood and sleep disturbance, diabetes mellitus, skin thinning and osteoporosis, especially when used at high doses for long periods of time.
- **Monitoring:** Prednisolone should not be stopped abruptly. Always check with your specialist doctor before changing the dose.

Mycophenolate mofetil (MMF):

- **Action:** Suppresses the immune system. It is often used to reduce the dose of prednisolone. It can be used in combination with other immunosuppressive medications or alone.
- **Side effects:** Can cause diarrhoea, constipation, abdominal pain, vomiting, muscle aches, abnormal liver function and an increased risk of skin cancers.
- **Monitoring:** People on this medication should not plan to become pregnant or breastfeed. Regular blood tests are required on this medication for full blood count, kidney and liver tests as they can be abnormal. Regular skin checks are also required.

Azathioprine (AZA):

- **Action:** Suppresses the immune system. Often used to reduce the dose of prednisolone. Can be used in combination with other immune-suppressive medications or alone.
- **Side effects:** Can cause diarrhoea, vomiting, abdominal pain, muscle aches, abnormal liver function and an increased risk of skin cancers.
- **Monitoring:** People on this medication should not plan to become pregnant or breastfeed. Regular blood tests are required on this medication for full blood count, kidney and liver tests as they can be abnormal. Regular skin checks are also required.

Your healthcare team will work with you to manage side effects and maximise benefits from these treatments.

Several other immunosuppressive therapies are available and are used to treat different forms of PF. These include methotrexate, cyclophosphamide, cyclosporine, tacrolimus, rapamycin and rituximab. Based on your type of PF, your specialist doctor will tailor their selection of the different medications to ensure you receive the most benefit.

Lung transplant

Lung transplantation 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 transplantation is rarely performed above the age of 65-70 years. 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. While over 90% of patients will live for a year after transplant, around 50% of people who have a transplant will be alive at five 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.

Managing Your Symptoms



Breathlessness and deconditioning

People with PF report that breathlessness is one of their more distressing symptoms. Breathlessness can occur when resting quietly but is more common when you move around and exert yourself. In some people, it may progress to the point of becoming breathless whilst performing daily activities, such as showering, getting dressed or speaking on the phone.

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

Staying as fit and active as possible will help to avoid deconditioning. Your healthcare team may suggest attending your local pulmonary rehabilitation program. Pulmonary rehabilitation can help to relieve breathlessness by increasing your fitness and teaching you strategies to manage your breathlessness. See the *Pulmonary Rehabilitation* section on page 17 for further information.

Breathlessness can be frightening for some. 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 you start to feel anxious due to breathlessness, try to find ways of calming yourself. Stop what you are doing, breathe quietly and calmly and wait until you feel better before going back to what you were doing. Often, finding a distraction such as listening to music, reading or sitting with a loved one can help your breathing settle.

If your breathlessness remains serious and persistent, speak with your specialist doctor. See the *Acute Exacerbation in PF* section on page 22 for further information. If you are experiencing shortness of breath which becomes severe, do not hesitate to call an ambulance.





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



Cough

People with PF often report a bothersome 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 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 describe debilitating coughing fits, often associated with episodes of breathlessness.

Whilst cough can be a symptom of PF, it can also occur for different reasons. Check with your healthcare team if other conditions, like heartburn or hay fever could be making your cough worse. There are treatment options for both these conditions. Ask your specialist doctor if there are any 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, as well as making you more emotional or impact your concentration and memory.

Tips to manage fatigue:

- 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

See the *Living Well with PF* section on page 26 for further information.



Pulmonary rehabilitation

Pulmonary rehabilitation is a program of exercise, education and support designed specifically for people with a 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 health centre for 6 to 12 weeks, with two outpatient 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 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 well-being.

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



My specialist got me into an eight week pulmonary rehabilitation program which I found very helpful. I have continued with my exercising and fitness program and I believe this has helped greatly in keeping my lungs and body healthy.

Lyn, lives with PF, QLD



Image supplied by Air Liquide Healthcare



Oxygen therapy

You may be prescribed oxygen if you develop low levels of oxygen in your blood. You may only need to use oxygen at night when sleeping or when doing exercise. Alternatively, it may need to be prescribed all the time.

To determine if you would benefit from home oxygen, your specialist doctor may measure your oxygen levels in several 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.

You may be tested:

- While sitting at rest
- While exercising, such as walking in the corridor or on a treadmill
- During sleep.

If you require extra oxygen, your specialist doctor will prescribe the amount as litres per minute (for instance two litres/minute) and for how many hours per day you require oxygen (for instance 24 hours, 18 hours or for sleeping).

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. You may also be given oxygen by mask, however this is less common. The nasal prongs are then attached to a home oxygen concentrator, a portable oxygen concentrator or a cylinder of oxygen.

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 flow-rate (litres/ min) according to the prescription from your specialist doctor
- Requires electricity to operate
- A length of lightweight plastic tubing attached to your nasal prongs will allow you to access all parts of your home including the shower.



Image supplied by Philips Australia

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
- Many PF patients need machines that deliver continuous flow, requiring the larger portable concentrators which are on wheels
- 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.



Image supplied by Philips Australia

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. This applies both at home and while you are out with your portable oxygen.
- Oxygen is highly flammable so do not expose yourself to gas cooking flames, home fireplaces, candles or any other source of open flame when using oxygen.
- Oxygen tubing poses a significant trip and fall risk. Ensure you hold the tubing up and to the side when walking and warn others of the risk, particularly the elderly and young children.
- Consult your oxygen provider for further safety information.

Starting oxygen therapy can be overwhelming and concerning for some people. You are not alone. Many others feel this way while they are adjusting to using oxygen. Be assured that 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.



Acute Exacerbation in PF

What is an acute exacerbation?

Some patients with PF may experience a sudden worsening of their condition (usually develops over one month or less) which is known as an acute exacerbation (pronounced *ex-zass-cer-bay-shun*) or a flare-up of symptoms.

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 and some surgical procedures. Acute exacerbations are more common in the winter season.

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.

How is an acute exacerbation managed?

Your specialist doctor may perform investigations to find out why you are more breathless than usual. Changes on your CT scan, along with your symptoms, may confirm you are experiencing an acute exacerbation.

Management of an acute exacerbation:

- Treatment will depend on your symptoms and possible causes
- Supportive therapy for feelings of breathlessness
- Possibly time in hospital

After an acute exacerbation, you may require a long period of recovery and in some cases, you may need long-term oxygen therapy. 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 of your specialist appointments and follow through with all investigations.
- Avoid infections:
 - Have up-to-date pneumonia and seasonal influenza vaccinations
 - Avoid contact with people with active chest infections
 - Practice 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.
- 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 well-being - see the *Living Well with PF* section on page 26 for further information.

Acute exacerbations can be difficult to understand. What happens during an acute exacerbation will be different for everyone, depending on the type of PF you have. You are encouraged to discuss what you may experience during an acute exacerbation with your healthcare team.

Associated Medical Conditions

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

Common associated lung conditions (*occurring in more than 30% of people with PF*):

- Chronic Obstructive Pulmonary Disease (COPD), including emphysema – a chronic lung condition, often related to smoking, causing breathlessness and frequent chest infections
- Pulmonary Hypertension – high blood pressure in the lungs
- Obstructive Sleep Apnoea – repeated episodes of partial or complete closure of the throat during sleep.

Less common associated lung conditions (*occurring in less than 10% of people with PF*):

- Lung cancer
- Pulmonary embolism – blood clot in lungs.

Your healthcare team or specialist doctor will take a detailed medical history to help detect if you have associated medical conditions. The investigations which aid in making the diagnosis of PF may also highlight the presence of other lung conditions.

Additional tests for the diagnosis and screening of associated medical conditions may include:

- **Echocardiography** – a non-invasive test that uses sound waves to produce images of your heart. It is used to screen for pulmonary hypertension.
- **Right heart catheter** – a catheter is passed into your neck or groin, to measure the blood pressure in your lungs which can sometimes be high in patients with PF.
- **A sleep study** – a non-invasive overnight study which measures your brain waves and breathing for diagnosis of obstructive sleep apnoea.
- **Coronary angiogram** – a catheter is passed into your neck or groin, to look at the blood vessels around your heart, for diagnosis of coronary artery disease.
- **A contrast CT scan or a nuclear medical ventilation / perfusion scan** – these scans are able to detect any blockages of the arteries within your lungs, for diagnosis of pulmonary emboli.

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
- Diabetes mellitus.

Managing these conditions is just as important as 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 well-being.

Living Well with PF

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.

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.

There may be times when you feel quite good. There may be other times when you feel flat and low, especially if you do experience an acute exacerbation. Feeling flat and low during these times is part of the normal emotional reaction to an acute exacerbation. However, if these feelings last or increase in strength, then seeking professional help may be very useful. Your GP, respiratory clinic or Lung Foundation Australia have resources and staff who can help connect you to relevant services.

It is important to develop strategies that suit your situation including how you will maintain your relationships and activities with friends and family. Research shows us that people with strong social connections have greater feelings of well-being. It can be a challenge, but generally it is well worth the effort.

Maintaining a healthy mind will help you to maintain your health as well. Talking to people about your feelings, maintaining social connections and treating yourself kindly and gently will all help to contribute to a greater sense of well-being.



Maintaining a healthy body

There are many opportunities for you to participate in caring for yourself and to become a member of your own healthcare team.

- **Maintain up-to-date vaccinations**
 - Annual seasonal influenza vaccination and
 - Pneumonia vaccination (every 5 years)
 - COVID-19 vaccination

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



As a grandparent, I pick up every germ or illness that my grandchildren have, so it's especially important for me to protect against vaccine-preventable infections.

Carole, lives with IPF, QLD



- **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 foods across the full spectrum of food groups including fruit and vegetables, lean protein and calcium-rich foods such as dairy products all according to your specific dietary requirements. If you need further support, ask your GP or healthcare team to refer you to see 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**

- Ask about a referral to a pulmonary rehabilitation program.
- 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**

- Discuss with your specialist doctor what the plan should be if you get a chest infection. Ensure your GP is aware of this plan.
- Reporting any changes in your breathing symptoms to your specialist doctor or GP as soon as possible will help them manage any chest infections or acute exacerbations in a timely manner.

Getting Around with PF

You may find that you become breathless undertaking activities that previously didn't affect you such as showering, shopping or gardening. Try not to avoid these activities. Instead you can break the activity into smaller parts, take rests in between or pace yourself. You may have heard this called "staging" your activities. By resting in between activities, you give your body a chance to "catch up" before continuing on.

If you find walking long distances challenging, you could discuss your eligibility for disability parking with your GP.

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 patients. Travelling by plane can be challenging because the available oxygen in the cabin is reduced. You may need to have a 'fitness to fly' test to see how your body will cope with altitude.

Some people with PF are required to have oxygen while on a plane. 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.

Before travelling, you should ensure your vaccinations (for example, seasonal influenza, pneumococcal pneumonia, tetanus) are up-to-date. While you're travelling it is essential you stay as healthy as possible. Remembering to pack all your medications including a written plan to follow if you experience an acute exacerbation, washing your hands frequently, staying away from other travellers with respiratory illnesses and most importantly, scheduling rest days to make sure you are not over-exerting yourself.



Preparing for Life with PF

Being diagnosed with PF may result in life changing significantly 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. There are some things you and your loved ones can do to make adjusting easier.

Maintaining engagement with your life

Discuss with your loved ones, your specialist doctor, healthcare team and other supportive people around you, about ways to keep engaged with things and people in your life that are important to you. This will help to keep you optimistic, resilient and engaged.



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 become more bothersome, 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.



I embarked on significant research, and set about planning my own journey, rather than how IPF would determine my journey.”

Bill, lives with IPF, QLD

Your Multidisciplinary Team

People with PF may have a Multidisciplinary Team (MDT) caring for them throughout their disease journey. Some of the team you will get to know well, like your specialist doctor, GP, lung function testing team, respiratory nurses and physiotherapists.

You may also find consulting with a psychologist helpful. Psychologists can help you manage the emotional aspect of living with PF. Your pulmonary rehabilitation team will usually be supervised by a physiotherapist and have other allied health specialists like dietitians, occupational therapists, rheumatologists and others involved to help you live the best life possible.

There will also be radiologists and pathologists working behind the scenes that will be actively involved in your initial diagnosis and monitoring your progress.

Palliative care

Many people believe that we only need to access palliative care at the end stage of our lives, this is actually incorrect. Palliative care is about symptom management and helping you to live well. An expert palliative care team may be helpful at all stages of your PF journey, if your symptoms are bothersome. Research shows that people who are linked with a palliative care team early on in their condition have better symptom management throughout their journey. Often you will be introduced to palliative care in the earlier stages of your diagnosis and it continues alongside other treatments.

It is okay to ask your specialist doctor or GP about palliative care. Taking an active role in how your condition is managed will help you to feel more in control.

Working together with your treating healthcare team, wider support team (friends and family) and taking positive actions to live a healthy lifestyle can help you stay prepared for some of the challenges you may face with PF and live your life to its fullest.



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