BETTER LIVING WITH Pulmonary Arterial Hypertension (PAH)
A PATIENT GUIDE
Lung Foundation Australia developed this booklet to help people diagnosed with Pulmonary Arterial Hypertension (PAH) understand more about this disease, its diagnosis and treatment. You will find information to explain what PAH is, its causes, the symptoms and the different types of treatment options and things you can do to live better with PAH.

PAH forms part of the overarching disease Pulmonary Hypertension (PH). You can find more information about PH by contacting the Lung Foundation.

PAH can be difficult to diagnose due to the vague symptoms. Often, people do not take much notice of these early signs of PAH. They may think they are just over-tired, lacking in fitness or getting old.

As PAH is uncommon and its treatment complex, all individuals with PAH should be managed by an experienced centre. Your doctor/specialist will be able to advise where your local centre is located. Medical research results in new and improved treatments and taking part in a clinical trial can give you access to the latest treatments. Ask your specialist if you are eligible to participate in a clinical trial.

When faced with a diagnosis of PAH there are many decisions to be made. You may want a second medical opinion to help you make decisions about treatment at the various stages of the disease. Your treatment team at an expert centre can help guide with these decisions, and provide you with a multidisciplinary opinion involving several treatment specialists.

Further support and information about living better with PAH is available by contacting Lung Foundation Australia. Support services and patient support groups can help you deal with the range of emotions you’re likely to experience and put you in touch with others who are going through similar experiences.

This booklet may also be helpful for the person who is your main carer.

If you need any further information and support, contact Lung Foundation on freecall 1800 654 301 or www.lungfoundation.com.au
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Is There a Difference between Pulmonary Arterial Hypertension and Pulmonary Hypertension?

There is an important difference between Pulmonary Hypertension and Pulmonary Arterial Hypertension (PAH). Pulmonary Hypertension is a general term and simply means high blood pressure in the lung. The high blood pressure can be caused by many medical conditions such as diseases of the left heart, chronic lung diseases, sleep apnoea, and blood clots in the lungs. PAH is a specific cause of Pulmonary Hypertension when the arteries of the lungs are directly diseased and become narrowed. It is very important that the exact cause of Pulmonary Hypertension is determined since treatment must be directed at the underlying condition.

What is Pulmonary Arterial Hypertension?

Pulmonary Arterial Hypertension (PAH) is the medical term for a progressive disease caused by narrowing or tightening (constriction) of the pulmonary arteries that connect the right side of the heart to the lungs. PAH is a serious condition that affects the blood vessels (pulmonary arteries) that carry blood from the heart to the lungs.

Although PAH is an uncommon condition, it is a disease that can affect males and females of all ages and ethnic backgrounds. Very rarely, PAH occurs in childhood.

PAH gradually restricts the flow of blood through the lungs. As a result, the pressure in the pulmonary arteries increases. The heart must pump against this increased pressure to maintain blood flow in the lungs and to the rest of the body. Over time, this can affect the heart’s ability to work effectively and may eventually lead to heart failure.

To understand more about PAH, it helps to understand how the heart works:

The role of the heart and lungs

The heart is divided into 2 parts – a left and right side. It is responsible for pumping blood to the lungs and around the body.

The blood is pumped by the right side of the heart through the lungs where oxygen moves into the blood; the blood then leaves the lungs and comes back through the left side of the heart, and is pumped out to the body.
This system is called blood circulation. The pressure that the right side of the heart pumps against in order to push blood into the lungs is your pulmonary pressure. Normally, your pulmonary pressure is very low. In PAH, the situation becomes different and very high pulmonary pressures develop due to disease of the blood vessels of the lungs.

**What happens to the blood vessels of the lungs in PAH?**

PAH is a disease of the blood vessels of the lungs. As PAH develops, the pulmonary arteries become narrow. The walls of these vessels also become thicker and less flexible. Over time, this narrowing of these blood vessels restricts the blood flow to the lungs. This means that the heart finds it harder to pump, therefore, the right side of the heart has to generate higher pressure in order to pump blood through the arteries. This results in high blood pressure in the lungs or Pulmonary Hypertension.

**What happens to the heart in PAH?**

Because the blood vessels are thicker or stiffer, it is harder for the heart to push blood through them. Over time, with the heart working harder, like any overworked muscle (the heart is a very complicated muscle) it gets ‘tired’. As the strain on the heart increases, it starts to function less well, a condition called heart failure. During this process the heart muscle can stretch and the heart can get larger, but this is not a good adaptation. As the heart muscle and pump function weaken, the heart responds less well to strain, which makes you feel tired and dizzy.
Causes of PAH
There are many causes of PAH and your doctor will perform several tests to confirm that you have PAH and to look for a cause. Causes of PAH may include:
- Idiopathic (meaning the cause is unknown) PAH
- Heritable (genetic causes that may be passed to your children)
- Drug and toxin induced
- Associated with other systemic diseases such as:
  - Connective tissue disease such as scleroderma, lupus, rheumatoid arthritis
  - HIV infection
  - Congenital heart disease
  - Liver disease

Some people may develop PAH if there is a family history (heritable) of the disease, while others develop PAH for no known reason (called idiopathic PAH). Because it can be genetically inherited in some cases, once the diagnosis is made it is important for other close relatives to be screened for PAH. You can receive guidance from your expert treatment centre regarding this.
Symptoms of PAH

The first symptoms of PAH may be very slight or difficult to notice, and you may only notice them when you exercise - climbing stairs, running, walking uphill or carrying bags. However, as PAH progresses, the symptoms may become more noticeable with less activity and interfere with daily routine.

Common symptoms of PAH include:

- Shortness of breath – breathlessness, especially with physical activity
- Feeling tired or fatigued
- Dizziness
- Chest discomfort or pain
- A fast and/or irregular heartbeat (also called palpitations)
- Light-headedness or fainting
- Swelling in your legs, and/or the belly (also called fluid retention or oedema)
- Lips and fingers turning purple or blue

Many of these symptoms are nonspecific and can be due to other diseases that affect the heart and lungs. For this reason it is important that the diagnosis of PAH is made by a physician who is an expert in this area.

“I was 35 when I started to experience changes in my health. The first thing I noticed was how much more breathless I was doing normal activities. Then I noticed I was gaining weight without any changes in my diet which was odd. Eventually I also became easily fatigued and needed to rest more.” — Melissa
Diagnosing Pulmonary Arterial Hypertension

How is PAH diagnosed
An accurate diagnosis of PAH is important, so that you can get the correct treatment. It is often difficult to recognise PAH because breathlessness and fatigue are common symptoms that are often overlooked or thought to be caused by many of the other diseases that affect the heart and lungs.

A PAH diagnosis requires a heart catheterisation and many other tests to exclude left heart disease, lung disease and other rare disorders. The diagnosis of a patient with PAH can be investigated by:

- Chest X-ray
- ECG (Recording of the heart's electrical activity using patches on your skin)
- Various blood tests
- Breathing tests (Lung function tests)
- Ultrasound of the heart (Echocardiogram)
- Scan to compare blood and air circulation in the lungs (V/Q scan)
- CT scans (Looks in depth at the tissue in the heart and lungs)

A right heart catheterisation (RHC) is typically undertaken to confirm the diagnosis of PAH.

During the right heart catheterisation, a small tube (catheter) is placed in a blood vessel and threaded through the heart to measure the pressure in different places along the way. This is the most accurate way to measure the pressure in the lung vessels, as well as the amount of blood being pumped through them every minute. In some cases, this test may need to be deferred because of safety or other considerations for some patients.

It is important that the medical work-up for PAH is done by a doctor who is used to diagnosing and managing patients with this condition.
Measuring the severity of PAH

You may have heard your specialist refer to PAH Functional Class or stage of the disease. This is simply a way to measure the effect that PAH is having on your day-to-day living. The Functional Class system has 4 levels and your specialist will use this system to assess how severe your PAH symptoms are. It is also a useful tool for your specialist when considering your treatment plan and comparing how you are before and after treatment. The table below describes the 4 levels.

<table>
<thead>
<tr>
<th>Class</th>
<th>How Symptoms Affect You</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>You have no symptoms with normal daily activities.</td>
</tr>
<tr>
<td>2</td>
<td>You have some symptoms with normal daily activities AND some limits when doing physical activity.</td>
</tr>
<tr>
<td>3</td>
<td>You may feel symptoms when doing less than your normal activity. You may also feel more limits when doing physical activity.</td>
</tr>
<tr>
<td>4</td>
<td>You may feel symptoms with any kind of activity, including resting.</td>
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You may have different types of specialists depending on the cause of your PAH and the stage. Speak with your specialist or PAH nurse to find out what resources are available and suitable for you.
You will be cared for by a range of health professionals, each of whom specialises in a different aspect of your treatment. Working with you, the team will develop a treatment plan to ensure you are provided with the best care. You may have different types of specialists depending on the cause of your PAH and the stage. Speak with your specialist or PAH nurse to find out what resources are available and suitable for you.

Your healthcare team is likely to include:

- **PAH Specialist** is a doctor who has been specially trained in diagnosis, treatment and the management of PAH. Most PH specialists are Pulmonologists (lung doctors) or Cardiologists (heart doctors).

- **PAH Nurses** can help with things like information about treatments, managing side effects and providing you with contact details for other support services.

- **Social Workers** can assist with advocacy, referrals to other services (both within and external to the health care service), accessing financial help, education, resourcing and other practical assistance. They can also refer you to counselling and targeted therapeutic services.

- **Occupational Therapists** can assist with equipment that might make things easier at home.

- **Dietitians** can help with advice about healthy diets and strategies for maintaining a healthy body weight as well as being helpful with other dietary problems.

- **Pastoral Care and Spiritual/Religious Care** can assist with psychological and community support, as well as spiritual and religious support.

- **Physiotherapists** can develop an exercise program to fit with your life and your personal needs. It is best to have guidance from those familiar with Pulmonary Arterial Hypertension, and you can ask for recommendations from your treating centre.
PAH is a severe condition and there is no cure at the present time. However, many treatments are now available that can improve your symptoms, quality of life and keep the condition under control. Everyone’s disease is different and taking your treatment as prescribed can help keep you from getting worse. Early treatment is important since PAH can be a progressive disease.

Treatment options will depend on the cause of PAH and the stage at which it is diagnosed. This is not a disease that you should ‘wait and see’ to do something about. It usually gets worse with time, and we know from studies that the sooner we treat it, the better chance you have of improving.

In the past, doctors had little to offer in terms of treatment options. However, this has changed dramatically over the last 10 years with the introduction of several new medications and treatments. Although currently there is no cure for this disease, it can often be well managed for many years with medications.

Types of Treatment
In general, medical treatment of your PAH is divided into PAH medications or medicines and supportive treatment. PAH medicines are given to dilate or open up the blood vessels of the lungs, improve blood flow to the lungs, and reduce the strain on the heart. Your specialist will discuss with you the different PAH medications and those that may be suitable for you.

PAH Medications
There are several medicines that treat PAH specifically. These medications are most commonly taken as tablets by mouth (orally). Other ways include via a needle into the vein (intravenously) or under the skin (subcutaneously) as well as by inhalation (mist). Each medication to treat PAH works in a distinct way and most patients now are given two or more PAH medications for best results.

Prostacyclins help the blood vessels relax. They mimic prostaglandin, a substance in the blood that is often low in PAH patients. Treatments in this group include: epoprostenol (Flolan™ and Veletri™), and iloprost (Ventavis™). Epoprostenol is given as a continuous drip into the vein. Iloprost is taken through a nebuliser and breathed in 5 or 6 times a day. Your PAH nurse will need to teach you how to give yourself these medications.
**Endothelin receptor antagonists (ERA's)** block endothelin, a substance that helps the blood vessels constrict. People with PAH have too much of this in their blood. Medications in this group are: macitentan (Opsumit™), bosentan (Tracleer™) and ambrisentan (Volibris™). These tablets are taken every day.

**PDE-5 inhibitors** allow the lungs to make more of their own natural vasodilators. Vasodilators are substances that relax the blood vessels, allowing more blood to flow. These treatments include sildenafil (Revatio™) and tadalafil (Adcirca™) which are taken every day.

**Soluble guanylate cyclase stimulators (sGC stimulators)** are substances in the body which help in chemical reactions that improve blood flow in the lungs. This type of treatment includes a tablet called riociguat (Adempas™).

**Continuous IV treatments for PAH**
There are several continuous IV infusions used to treat Pulmonary Arterial Hypertension. A small pump is commonly used to infuse these medications into the patient. The pump must be carried on the patient at all times. The medication is mixed into a plastic container called a cassette that is attached to the pump and a thin plastic tubing runs from the pump to the patient (a catheter is often inserted through the skin into a large vein). These medications run 24 hours a day, 7 days a week, 52 weeks a year. They must never be interrupted except to change the cassette.

**Transplant Surgery**
Most individuals with PAH do very well with treatments but occasionally and only if these treatments are ineffective or are unable to control the condition, more involved treatments including organ transplant may need to be considered. This is where an organ in your body is replaced with an organ from a donor. Transplant surgery for PAH can involve replacing the heart and lungs, or just the lungs. Transplantation is required less commonly these days because of medications being more effective.

**Chronic Thromboembolic Pulmonary Hypertension (CTEPH)**
PAH can result from blood clots in the lungs known as Pulmonary Emboli (PE). Some patients with PE have incomplete response to therapy and go on to develop CTEPH. In some cases of CTEPH, an operation to potentially cure the condition is performed, called Pulmonary
Thrombendarterectomy, which involves opening the pulmonary arteries and removing the residual clot together with the inner lining of the artery. Alternatively, balloon angioplasty where a balloon catheter is used to stretch the arteries wider is performed. If neither of these is possible, management with medications is used.

**Supportive Treatment**

Other treatments may be prescribed by your specialist to assist in managing your PAH. The treatment options listed below are examples of treatments that may be prescribed for you.

**Oxygen therapy** may be required for some people with PAH. Many people with PAH need to use oxygen at night when they are sleeping or only when they exercise.

If you are planning to travel on a plane or visit a location in a high altitude you may require oxygen and should talk to your specialist when planning your trip. A test can be performed to determine if you would need oxygen and results of this test can take up to three weeks.

Lung Foundation Australia has a Home Oxygen booklet and a Fitness to Fly fact sheet that you may find useful. Please call 1800 654 301 for further information.

**Warfarin** is an anticoagulant as it helps prevent blood clots from forming in the blood vessels. Warfarin is given to some people with PAH who are at a high risk of developing blood clots.

**Diuretics** increase the amount of urine you make. Increasing the amount of urine means less fluid in the blood vessels. This reduces pressure in the blood vessels and makes it easier for the heart to pump blood around the body.

**Exercise rehabilitation** can make you more active and improve your well-being. Patients with PAH have unique heart-lung physiology (make-up) and it is important that exercise rehabilitation is supervised by a therapist with experience in PAH. You should only start an exercise program after discussing with your PAH specialist.

**Accessing New Treatments via Clinical Trials**

Clinical trials are research studies that test new and potentially better ways of improving peoples’ health. They are an important process to assess whether promising approaches to prevention, diagnosis and treatment are possible, safe, and effective in humans. Some trials also look at ways of enhancing a person’s quality of life.
There is ongoing research into the causes and treatments of PAH. There are new medications that are being studied and your specialist may suggest that you participate in a clinical trial. Benefits of taking part in a clinical trial include more personalised care and attention from research nurses and treating doctors. If you would like to take part in a clinical trial, talk to your specialist.

**Treatment Side Effects and Management**

Different drugs and treatments have different side effects. A healthcare team member will discuss expected side effects with you and explain what you can do to manage and reduce your symptoms. Always tell your healthcare team about any side effects that you experience so they can support you and help you manage them. In most cases, they can prescribe drugs to make the side effects less severe.

**Treatment Compliance**

Anything you take or use for therapeutic benefit is a medicine. This includes prescribed medications from your doctor, over-the-counter medicines from the pharmacy, the supermarket or another store. It also extends to herbal remedies, vitamins and other supplements.

All medicines have potential benefits and risks, and sometimes interact with each other; which is why it’s important that you discuss all the medicines you’re using with your health professionals.

“Being compliant with medications is both very difficult and simply easy. Being on intravenous epoprostenol and a handful of other tablets can be time consuming, takes careful preparations and you have to always be thinking ahead, on the other hand, without them, I would not be able to do all the things I love to do. This small sacrifice is something I am willing to do each and everyday to feel as good and as healthy as I can!” — Tegan
Life with PAH
In the beginning, learning that you have a chronic (meaning long-term) illness can be worrying. Many people find that the physical symptoms of PAH can be stressful or make them feel sad. Some people feel anxious when they can't breathe properly. Others feel frustrated, left-out and lonely when they can't keep up with others. Ultimately, it may be a relief to finally have a diagnosis and appropriate treatment.

Your specialist, nurse, and PAH healthcare team are there to support you. Living with and treating PAH is a different experience for everyone. While PAH is not curable, there are effective treatments available to help improve your condition and your quality of life.

People living with PAH face many emotional, psychological and practical day-to-day demands on top of the physical impacts of the disease and its treatment. Eating well, exercising and relaxing may help reduce stress and improve wellbeing. Addressing changes in your emotions and relationships early on is also important, and you should speak with your specialist, nurse or GP.

Lung Foundation Australia has developed a brief quality of life checklist to help you or your carer identify whether you might benefit from some extra help in managing your PAH.

In addition, Lung Foundation has developed a set of ten short self-management videos titled Better Living with Your Lung Disease, that aim to increase the knowledge and confidence of people living with PAH to enable them to better manage their condition, its symptoms and their overall wellbeing.

Contact the Lung Foundation if you would like further information about these patient resources – 1800 654 301.
Healthy Eating
Because everyone is different, there is no set diet to help to reduce PAH symptoms. It is important to eat a nutritious diet to keep your energy levels up. You should aim to be a healthy weight. If you notice any significant weight loss or gain, tell your specialist.

Common suggestions for a healthy diet may be:

- Change to a low salt diet (salt causes you to retain water, which may worsen any swelling or oedema you have)
- Avoid packaged and prepared food as these often contain hidden salt (also called sodium), fats and sugar
- You can see a dietician to find out which foods are high in salt
- Eat small, more frequent meals
- Eat fresh fruit, vegetables and whole grains
- Eat fish and lean meat, such as chicken
- Drinking alcohol may be acceptable in moderation
- Some foods and drinks can affect medicines, so you need to check with your specialist or pharmacist if there is anything you should avoid
- Your PAH team may ask you to measure how much fluid you drink each day

Manage Tiredness
A common symptom of PAH can be extreme tiredness. It may feel different to normal tiredness. Many patients have good days and bad days. Getting to know your body and understanding the signs it gives are important. This will enable you to rest when your body needs it. Many people find that pacing themselves – doing one thing, having a rest, doing another, having another rest, means that they can get more done during the day. If you overdo it one day, you may be very, very tired the next day. Take this as a sign that you may have done too much, and wind it back a little the next time. If you find yourself fatiguing more frequently on a regular basis, this can be a sign that your PAH needs reassessment.
Exercise and PAH
There are many benefits to being active. Exercise can boost energy levels, decrease fatigue, increase strength, and relieve stress as well as lower anxiety and depression.

It may be hard to know how much or what type of exercise is possible with PAH. With help from your specialist, you can work out what is right for you. Do not start an exercise program without discussing this with your PAH specialist. Choose activities that you enjoy and that make you feel good.

Feeling down or anxious
Emotions can run high when you have a serious disease that needs ongoing treatment. It’s not uncommon to feel sad, anxious, angry or confused when you’ve been diagnosed with PAH. If you feel sad, you may not have the motivation or energy to go to your medical appointments, take your medications or look after yourself. This can make things worse very quickly.
How can I deal with these emotional problems?

• Start by talking about your feelings and concerns with your treating team. Don’t keep them to yourself.
• Consider counselling to help deal with your feelings.
• Join a support group to share what you feel with others who may feel the same.
• Look to family and friends for support
• Try relaxation techniques
• Don’t be afraid to take medication if they are prescribed to you. If you are concerned, talk to your pharmacist, specialist or PAH nurse.
• Most of all, try to discover what makes you happy. This is personal – it might be listening to music or reading a good book. Plan to do something you enjoy every day.

There are several organisations that can provide assistance if you are going through a rough time. Don’t be afraid to reach out to one of these groups:

- **Headspace** – the National Youth Mental Health Foundation  
  www.headspace.org.au

- **Beyond Blue** – a national initiative to raise awareness of anxiety and depression  
  www.beyondblue.org.au

“Mum and the many other supports, are the core reason for my current state of health. I have such good supports in my sisters, maternal grand-parents and my best friends.” *Tegan*
Building Your Networks & Support
It is very important to find support to help you manage your condition. Support can be found in many places: your PAH specialist and nurse, allied health services like physiotherapists, social workers or pharmacists, local patient associations for people with PAH or just breathing problems in general, together with the people who are already important in your life.

Family and Friends
As with any long-term condition, family and friends may want to offer support. Don’t be afraid to ask for help when you need it or accept help when it is offered.

Accepting help gives those who care about you a sense of making a contribution at a difficult time. It can also allow you to get on with the things that you can and want to do. While this may not be what you are used to doing, you may find that by accepting help you have more of yourself to give back to others.

Below are some suggestions of ways in which others may help you:

<table>
<thead>
<tr>
<th>Everyday support</th>
<th>Medical support</th>
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<tbody>
<tr>
<td>Running errands such as shopping</td>
<td>Transport to doctor appointments</td>
</tr>
<tr>
<td>Preparing meals</td>
<td>Taking notes at doctor appointments</td>
</tr>
<tr>
<td>Doing household chores</td>
<td>Picking up medicine</td>
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Linking in with Other PAH Patients
Many newly diagnosed patients find it helpful to talk with other people with PAH. After all, no one can understand your experience better than someone with the same problem. Patient Groups and Associations can be a great source of support and comfort in helping you deal with day-to-day living and PAH. Please contact Lung Foundation Australia for information about your nearest support group networks on 1800 654 301 or at www.lungfoundation.com.au
Important Considerations for PAH Patients

Pregnancy and Contraception
Pregnancy can be extremely dangerous for both the mother and baby if the mother has PAH. It is important that you use reliable contraception to avoid unplanned pregnancy. Some PAH medications are also very harmful to an unborn child. This is a very sensitive subject, and as every person is different you should discuss with your PAH specialist team if you are considering trying to become pregnant.

Operations and Procedures
Anesthetics for operations can be harmful for some patients with PAH. Anesthetics are when you are made sleepy for an operation (general anesthetic or sedation), or have numbing in the back (spinal or epidural anesthetic). Let your anesthetist and PAH specialist know if you are going to need an anesthetic, even for a small procedure, so they can make it as safe as possible.

Quitting smoking
If you smoke, stopping when living with PAH is important for your lung and heart health. Smoking is addictive and it can be difficult to quit – you should speak with your specialist or health care team about quitting. If you need help to quit smoking, call Quitline on 13 78 48. You can also visit the Quit Now website at www.quitnow.gov.au.

Dental Work
For those living with PAH your dental hygiene becomes more important. PAH is not caused by dental problems but the resulting increased inflammation may cause your PAH to get worse. You should schedule regular check-ups with your dentist. It is important to inform your dentist of your medical history (including PAH) and any medications that you are taking.

PAH and Other Illnesses
Even minor illnesses or sickness such as common colds can worsen PAH symptoms. Please see your local doctor (GP) promptly if you start to feel unwell.

Vaccinations
It is important that all your routine vaccinations are up to date, and that you and everyone living in your house is vaccinated with the annual flu vaccine. There are other vaccines that may be important when living with a PAH diagnosis – you should discuss these with your specialist.
If you live in regional or remote area within Australia and need to travel to hospital for treatments or tests, you may be eligible for financial assistance for travel and accommodation.
Financial Considerations

Depending on your circumstances you may have a range of medical, personal, travel and accommodation costs associated with your diagnosis and treatment. You don’t need to face these alone.

To find out about financial assistance you should start by talking to your PAH nurse, hospital social worker or the legal service attached to the hospital.

Below is a list of places you can contact to see whether you can get some financial assistance. Most schemes are means-tested, so the assistance you get may depend on your income and other assets.

**Centrelink**
You may be eligible for a benefit or pension, such as the Sickness Allowance.

**The Health Department in your State or Territory**
There is a patient’s travel assistance scheme for people who have to travel over a certain distance for treatment.

**Department of Veterans Affairs**
There are a number of benefits and pensions for veterans or partners of veterans.

**Local councils**
Some local councils help with practical matters, such as child care, meals and general home help.

**Medical**
It is recommended that you talk about the likely costs of medical tests, treatment and support services with your specialist and/or PAH nurse. In general, PAH is managed in the public hospital system (regardless of whether you have private health insurance or not). You will be treated by specialists nominated by the hospital and you will not be charged for most treatments and care costs.

**Travel and accommodation**
If you live in regional or remote area within Australia and need to travel to hospital for treatments or tests, you may be eligible for financial assistance for travel and accommodation. Speak with your PAH nurse to find out whether you are eligible for such assistance.
You may be reading this booklet because you are caring for someone with PAH.

Being a carer can be very stressful. Try to look after yourself. Give yourself some time out and share your worries and concerns with somebody neutral, such as a counsellor or your doctor.

Caring can be rewarding but many carers also find it demanding, both physically and emotionally. If you have been caring for someone for some time, you may feel exhausted. You may feel guilty about making time for yourself. However, looking after yourself can help relieve the stress and exhaustion of caring, and can reduce feelings of frustration and isolation.

Supportive services such as Meals on Wheels or visiting nurses can help you in your role as a carer. Many organisations and groups can provide you with information and support. To learn more, contact Carers Australia on www.carersaustralia.com.au or (02) 6122 9900.

To find out more about different support services, call Lung Foundation Australia on 1800 654 301.

“My daughter and I attend the PAH support group every month and I attend a carers group meeting once a month. Being with people who understand makes you feel you are not alone or fighting the illness by yourself.” Robyn
Helpful Tips

There are some small things you can do to help manage life with PAH. You may find the following tips useful:

Preparing for your appointment
Your specialist will want to talk to you about your care, provide you with choices and answer your questions. If there are things about your PAH symptoms or about your treatment that you do not understand, ask your specialist to explain them to you. Remember to write down questions that you have and take them to your appointment – many people have questions but cannot remember them when they see their doctor.

It may also help to have a partner or family member with you at your specialist appointments. Many patients ask their family member to take notes while they are speaking with their specialist. This may also assist in them gaining a better understanding of PAH and how you and your specialist are managing it.

Keep a diary
Keeping a diary, perhaps for a month before your next appointment, can be very helpful. You may want to record how you are feeling on a day-to-day basis for a few weeks especially when things like medications are changed.

Write down any questions to ask your specialist or nurse. Remember to include questions you have on working, financial issues, housing and equipment concerns. You can also discuss any emotional, physical or sexual concerns you may have. You can use the space on pages 25-27 to get started.

Update your medical information
Keeping your medical information organised is important. It can be helpful to you and to those important people in your life. You can make a list of the names of your different doctors; include their phone numbers and addresses, to keep by the phone.

It is also important to keep a list of your current medications, vitamins, supplements, and anything else you are currently taking. Be sure to note the dose and how often you take each. Consider making this information available to the important people in your life. Sharing this information with your specialist and nurse allows them to better understand your PAH symptoms and your treatment.
**Interpreters**

If English isn’t your first language an interpreter may be useful. Most hospitals have access to translation and interpretation services. If you need an interpreter, speak with the PAH nurse before your appointment. He or she can then arrange for an interpreter to be present.

If you need any further information and support, contact Lung Foundation on freecall 1800 654 301 or www.lungfoundation.com.au
Your Notes
Looking for support and/or further information on PAH please visit the suggested links below:

**National Organisations**
- **Lung Foundation Australia** www.lungfoundation.com.au | Ph: 1800 654 301
- **Pulmonary Hypertension Association Australia** www.phaustralia.com.au
- **Pulmonary Hypertension Network Australia** www.phwa.info | Ph: 08 9302 3078
- **Scleroderma Australia** www.sclerodermaaustralia.com.au | Ph: 02 9990 5159
  Call Scleroderma Australia to obtain contact details of Statewide organisations.

**International Organisations**
- **Pulmonary Hypertension Association (PHA) USA** www.phassociation.org
- **Pulmonary Hypertension Association United Kingdom (PHAUK)**
  www.phassociation.uk.com
About Lung Foundation Australia

Information and Support Centre
The Centre provides the following services and can be contacted on the toll free number 1800 654 301 or via email enquiries@lungfoundation.com.au

Patient Education Resources
Brochures are available covering a range of lung conditions, treatment options and diagnostic procedures. The Lung Foundation has produced additional resources such as handbooks, booklets and DVDs to assist patients in understanding and managing their lung condition.

Newsletter
Published on a quarterly basis, LungNet News is a free publication which provides useful articles on lung health and keeps readers informed of the work of Lung Foundation Australia.

Patient Support Groups
Located in all States and Territories, Patient Support Groups meet regularly to provide a welcoming, informal environment for those with a lung condition, their families and/or carers. Group activities may include guest speakers, social activities, exercise programs and peer support.

Education Days
Held annually in each state and territory, Patient Education Days provide an opportunity for patients to hear first-hand from health professionals about the latest developments in lung health.
This resource is one of a series produced by Lung Foundation Australia to provide information on lung disease, its treatment and related issues.

The information published by the Lung Foundation is designed to be used as a guide only, is not intended or implied to be a substitute for professional medical treatment and is presented for the sole purpose of disseminating information to reduce lung disease. Please consult your healthcare professional if you have further questions relating to the information contained in this booklet.

If you would like to support Lung Foundation Australia in its important work, please call 1800 654 301 for details on how you can make a donation. All donations over $2 are tax deductible.