Lung Foundation Australia

Living with **Pulmonary Arterial Hypertension**

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About Pulmonary Arterial Hypertension

What is Pulmonary Arterial Hypertension?

Pulmonary Arterial Hypertension (PAH) is the name of a long-term health condition due to increased pressure in the lung arteries. PAH is only one type of Pulmonary Hypertension (PH), which means there is increased blood pressure within the blood vessels of the lungs. PAH is uncommon, affecting less than 50 in one million persons[1], however it can affect males and females of all ages and ethnic backgrounds. Very rarely, PAH occurs in childhood.

The role of the heart and lungs

The heart is responsible for pumping blood to the lungs and around the body. It is divided into four chambers: two on the right-hand side and two on the left. The right side of the heart pumps blood into the lungs where oxygen moves into the blood; it then returns to the left side of the heart to pump the oxygenated blood out to the body.

The pressure the right side of the heart pumps against to push blood into the lungs, is known as your **pulmonary pressure**. Normally, your pulmonary pressure is very low. When our heart and lungs are healthy, blood moves easily through the lungs, allowing oxygen to enter the blood.

Heart and lungs when well

- Low pressure
- Heart doesn't have strain
- Oxygen can move from lung into blood stream

Heart and lungs in PAH

- High pressure
- Heart is strained over time
- Insufficient oxygen and blood supply to body over time

[1] Emmons-Bell S et al. Prevalence, incidence, and survival of pulmonary arterial hypertension: A systematic review for the global burden of disease 2020 study. Pulm Circ. 2022 Jan 18;12(1):e12020.

In PAH, the blood vessels become thicker and less flexible, causing very high pulmonary pressure, affecting the function of the heart. The heart needs to pump against this higher pressure to maintain blood flow in the lungs and to the rest of the body, making the heart work harder.

Over time, with the heart working harder, like any overworked muscle it gets 'tired'. 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.

What is the difference between PAH and PH?

PH is a general term and simply means high blood pressure in the lung. High blood pressure in the lungs (pulmonary 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 type of PH. The important difference is that PAH is caused directly by disease in the blood vessels leading to high pulmonary pressure. It is very important that the exact cause of PH is determined since treatment must be directed at the underlying condition.

Causes

There are many causes of PAH, which may include:

Idiopathic, meaning the cause is unknown

Genetic: A family history of PAH

Complication from other diseases including:

- Connective tissue disease such as scleroderma, lupus and rheumatoid arthritis
- HIV infection
- Congenital heart disease (present from birth)
- Liver disease
- Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (blood clots in the lungs)

Drug or toxin (chemical) induced.

Symptoms

The first symptoms of PAH can be subtle and you may only notice them when you exercise - climbing stairs, running, walking uphill or carrying bags. As PAH progresses, the symptoms may become more noticeable with normal daily activity and interfere with your day-to-day routine.

Each person experiences PAH differently.

Every PAH patient's journey is different, everyone responds differently to the treatments, so it's important not to compare yourself to others. We do however learn from each other and it helps to feel like we are not alone.

- Kylie, lives with PAH, NSW

Common symptoms include:

Shortness of breath

A fast and/or irregular heartbeat

Feeling tired or fatigued

Light-headedness or fainting

Dizziness

Swelling in your legs and /or belly

Chest discomfort or pain

Lips and fingers turning purple or blue.

Diagnosis

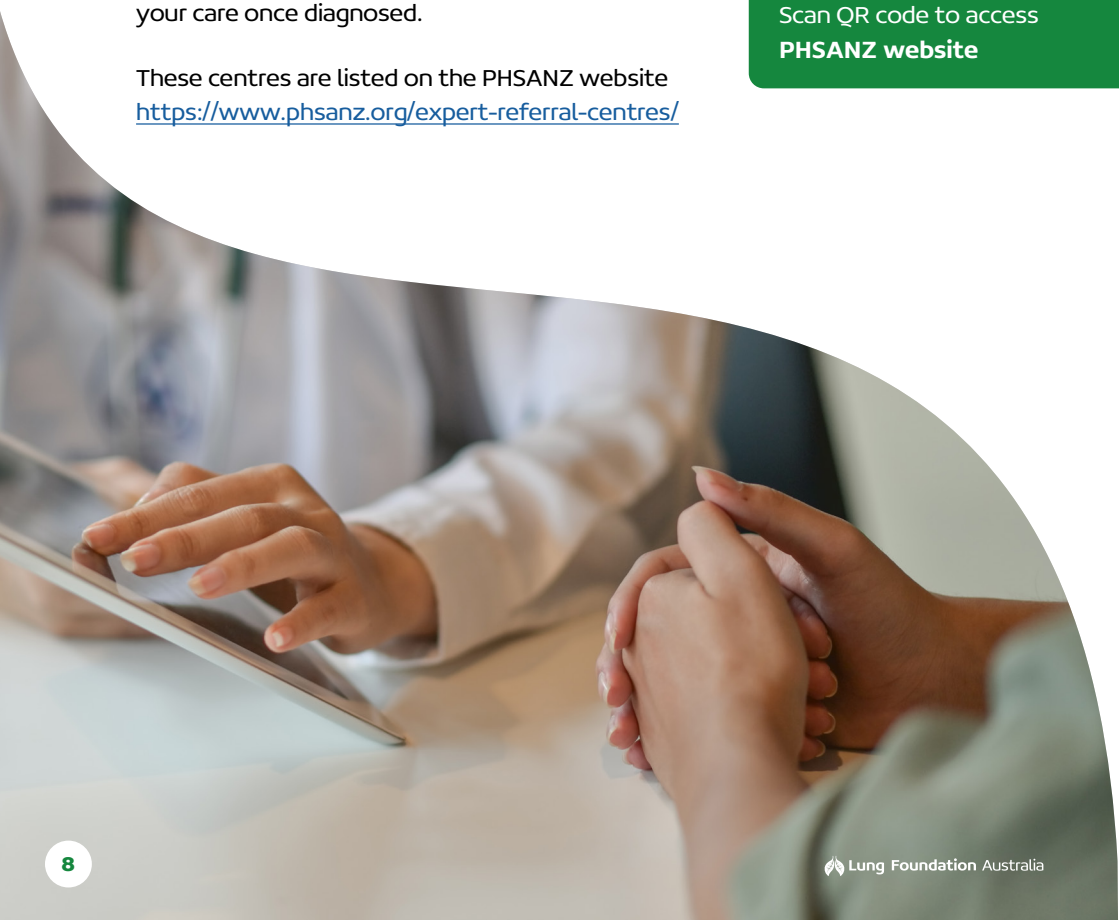
It is often difficult to diagnose PAH because breathlessness and fatigue are common symptoms that may be overlooked or thought to be caused by other heart and lung diseases. The symptoms can also be mistaken for signs of getting older, being overweight or unfit.

Diagnostic tests

Once PAH is suspected by your general practitioner (GP), it is preferable to be referred to a PAH Expert Referral Centre to get an accurate diagnosis by a specialist doctor who is an expert in this area. If testing is not conducted in a specialist centre, you should be referred to a PAH specialist to oversee your care once diagnosed.

These centres are listed on the PHSANZ website https://www.phsanz.org/expert-referral-centres/

Scan QR code to access **PHSANZ website**



Diagnosing PAH can be complex, and usually involves a series of tests to rule out other possible conditions before confirming the diagnosis. These diagnostic tests often include:

Electrocardiography (ECG)
Records the heart's electrical activity using patches on your chest.

Echocardiogram (echo)
A specialised ultrasound examination of the heart.

Ventilation/perfusion (V/Q) scan
Compares blood and air circulation in the lungs.

Lung function tests (breathing tests)
Full lung function testing is done to assess for any lung causes contributing to the elevated pulmonary pressures, to look at blood oxygen levels, and to check the lung diffusing capacity. A 6-Minute Walk Test is used as part of the assessment of functional capacity and for monitoring progression of disease.

Right heart catheterisation (RHC)
Measures pressure in the heart and lungs. This safe and vital test is usually needed to confirm the diagnosis of PAH.

Other tests/investigations may include:

Chest X-ray
Takes pictures of the tissues in the lungs and surrounding organs.

Various blood tests
These are not used to diagnose PAH, but may be used to rule out other conditions or to work out the cause of your symptoms.

Computerized Tomography (CT) scan
Takes more detailed pictures than an X-ray to look at the lungs and surrounding organs.

While there is currently no cure for PAH, research shows that early diagnosis combined with treatment at the early stage of the condition can improve quality of life, slow progression, reduce mortality and keep people out of hospital.

Measuring the severity of PAH

You may have heard your specialist doctor refer to PAH Functional Class or stage of the disease. This is a way to measure the impact that PAH is having on your day-to-day living. The Functional Class system has four levels and your specialist doctor will use this system to assess the severity of your PAH symptoms.

How symptoms affect you:

Functional class

1. No symptoms with normal daily activities
2. Some symptoms with normal activities and some limitations with physical activity
3. Symptoms doing normal activities such as dressing, showering and cooking
4. Symptoms with any kind of activity and at rest

Treatment for PAH

Although there is currently no cure for this disease, early treatment with medications and a focus on healthy living can help improve your condition and your quality of life. Everyone's disease is different, and treatment and self-management options will depend on the cause of PAH and the stage at which it is diagnosed.

Medications

There are several different PAH medications that open up the blood vessels of the lung, improve blood flow to the lungs, and reduce the strain on the heart. These are usually taken as oral tablets but can also be given intravenously (via a small plastic tube inserted into a vein), or breathed in as a mist. Often a combination of medications to treat PAH is required.

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, iloprost and selexipag. Epoprostenol is given as a continuous drip into the vein. Iloprost is taken through a nebuliser to deliver the medication as a mist five or six times a day. Selexipag is an oral medication that helps manage PAH by targeting the prostacyclin pathway. It is used alongside other PAH treatments to help slow disease progression.

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, bosentan and ambrisentan. 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 and tadalafil.

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.

Other medications can be used as supportive medications and may include fluid tablets, blood thinners and medications to help relieve symptoms.

Diuretics help reduce excess fluid in the body to reduce swelling and improve breathing. Some of the examples include frusemide, bumetanide, or spironolactone. Your doctor may prescribe these tablets as well as a low salt and low fluid diet to control excess fluid. Learning about managing fluid balance can be an important way of relieving symptoms in PAH.

Key tips for PAH medications

Do

- ✓ Take your medications as prescribed
- ✓ Bring your prescriptions or a list to every medical appointment
- ✓ Bring your medications to hospital if being admitted

Don't

- ✗ Stop medications unless advised by your GP or PAH team
- ✗ Run out of scripts

Oxygen therapy

Many people with PAH will have shortness of breath, but not everyone will have low oxygen levels and need oxygen therapy. To determine if you need home oxygen therapy, 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 oxygen therapy, 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, overnight or with exercise).

Clinical Trials

Clinical trials are research studies that test new and potentially better ways of improving people's health. There have been many advances in prevention, treatment and management as a direct result of clinical trials and research initiatives. There can be both advantages and challenges when participating in research studies and clinical trials. It's important you understand what's involved before you agree to take part in a research study. Talk to your healthcare professional at a PAH Expert Referral Centre to see if you are eligible to participate in a trial.

Transplant surgery

Although most people with PAH do well with medications, in very severe cases, your treating doctor may consider transplantation. Transplant surgery involves replacing the heart and lungs, or just lungs, from a donor.

Self-management options

Effective self-management of your PAH is important to help improve how you feel. By taking the following steps you can improve your overall quality of life, slow the progression of symptoms, and keep your PAH well managed.

Stay healthy and active

Even minor illnesses such as common colds can worsen PAH symptoms. There are lots of things you can do to make sure you stay healthy and have energy to do the things you enjoy, including:

- Eat a healthy, nutritious diet
- Be physically active
- Get enough rest and good quality sleep
- Look after your wellbeing by enjoying friends, family and hobbies. Practicing relaxation techniques can also help to reduce feelings of stress or anxiety.

Exercise

There are many benefits to being physically active. Exercise can boost energy levels, decrease fatigue, increase strength, and relieve stress as well as lower anxiety and depression. Regular exercise also helps the heart and body work smarter, not harder. It keeps your blood vessels in good shape and eases inflammation. The right exercise program can have a positive impact on your PAH.

It may be hard to know how much or what type of exercise is possible with PAH. Ask your PAH specialist doctor about the best exercise options for you. This may involve:

- Booking in to see an Exercise Physiologist or Respiratory Physiotherapist to help design a program with you.
- Attending a pulmonary rehabilitation program

Do not start an exercise program without discussing this with your PAH specialist doctor first.

Pulmonary rehabilitation

Some people may benefit from a structured pulmonary rehabilitation program. Provided by specially trained health professionals, it is a series of exercise and education sessions that teach you the skills needed to manage your breathlessness and to stay well. Ask your healthcare team about the possibility of a referral to your nearest pulmonary rehabilitation program.

Ensure your vaccinations are up-to-date

Vaccinations are an important part of supporting your immune system and reducing the risk of illness. You and your doctor may discuss vaccinations for COVID-19, seasonal influenza (flu) and pneumonia.

Eat well

There is no set diet to help to reduce PAH symptoms. However, it is important to eat a nutritionally balanced and varied diet, and to maintain a healthy body weight. Eating well is especially important for people with lung conditions, as foods and fluids contain essential nutrients to help prevent infections and keep your lungs healthy. Eating well will also keep your energy levels up.

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 dietitian 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.
- Reduce alcohol intake.

Don't feel like you have to tackle your health and your diet all on your own! Partner with your healthcare team to come up with a plan.

If you have problems with fluid retention (swollen legs and abdomen), your healthcare team may talk to you about avoiding food containing lots of salt and making sure you don't drink too much fluid each day. Some patients need to take fluid tablets (diuretics, pg 13) to help reduce fluid swelling. Your healthcare team may recommend 'fluid restriction', where they will ask you to track your fluid intake and weight each morning.

Some foods and drinks can affect medicines, so you need to check with your specialist doctor or pharmacist if there is anything you should avoid.

If you notice any significant weight loss or gain, tell your specialist doctor.

Manage fatigue

A common symptom of PAH can be extreme fatigue. It may feel different to normal tiredness. Most people have good days and bad days. Get to know your body and understand the signs it gives. This will enable you to identify when your body needs to rest.

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 do too much in one day, you may be very, very tired the next. Take this as a sign 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.

Have regular dental checks

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.

Quit smoking

If you do smoke or vape, quitting is the single most important thing you can do to improve your health and lung function. If you continue to smoke or vape, your health and respiratory symptoms will worsen. The sooner you quit, the better your chances of living well and improving your lung function.

Women's reproductive health

Pregnancy and PAH

It can be a significant risk to both the mother and the baby to become pregnant if you have PAH. This is something to discuss with your specialist doctor. Endothelin receptor antagonist (ERA) medications that are often prescribed to treat PAH cannot be taken in pregnancy as they are toxic to foetuses. Talk to your GP about contraception if you are on bosentan, macitentan or ambrisentan.

Contraception

- Recommended long-acting contraceptive options include intrauterine devices (IUDs) like Mirena, which are inserted into the womb, and contraceptive implants like Implanon, placed under the skin of the arm.
- Generally, oestrogen containing contraception, like the combined oral contraceptive pill, should be avoided.

Monitoring your PAH

PAH is a progressive condition, therefore ongoing monitoring is an important part of management to ensure early treatment is prescribed. Ongoing monitoring may include regular blood tests to monitor haemoglobin, liver function and iron levels.

After you have started on therapy, your PAH specialist team will repeat tests every 3-6 months. These tests will include:

- Functional class assessment
- 6-minute walk test (6MWT)
- Electrocardiogram (ECG)
- Blood tests (including NT-proBNP)
- Echocardiogram

Red flag symptoms to specifically mention to your PAH specialist team as soon as possible include:

- Rapid symptom changes (over weeks to months)
- Black outs (called syncope)
- Dizziness / light headedness
- Swelling in ankles
- Difficulty lying flat in bed
- Chest pain
- Heart palpitations

Access emotional support

PAH is a challenging disease and can take its toll on your emotional wellbeing. People living with PAH face many psychological and practical day-to-day demands on top of the physical impacts of the disease and its treatment. Living with PAH is a different experience for everyone. The physical symptoms of PAH can be stressful, and you may experience higher than usual levels of negative emotions like sadness and irritability. People may feel anxious when they experience breathlessness; frustrated or lonely when they can't keep up with others. Anxiety and depression are not uncommon in PAH, and caring for a loved one can also lead to high levels of stress.

Every person living with PAH or caring for a loved one will face challenges and need different types of support at different times. Acknowledging the mental impact early can help you find the support and gather the tools you will need to look after your emotional wellbeing as well as your physical wellbeing.

It is very important to find support to help you manage your mental health and emotional wellbeing. Help can come in many forms including from family, friends, neighbours or medical and allied health professionals such as psychologists, social workers or occupational therapists. As a starting point, you can talk to your GP about your mental health and wellbeing, including steps you can take to proactively care for and manage your mental health.

Peer 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.

I think peer support is essential. You've got to have someone who knows what you're talking about and going through. If you don't have anyone around who you can contact, you don't get that support and you'll just be left floating with all the worries in the world. The support is the glimmer at the end of the tunnel.

- Morrie, lives with PAH, QLD

To learn more, and express your interest in joining a support group, contact our Information and Support Centre on **1800 654 301**.

Your healthcare team

You will be cared for by a range of healthcare professionals, each of whom specialises in a different aspect of your treatment. Working with you, a multidisciplinary team will develop a treatment plan to ensure you are provided with the best care. 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.

The healthcare team

General practitioners (GPs) are qualified in general practice medicine. GPs are often people's first point of contact with the healthcare system when feeling unwell. They provide long-term, continuous care, and can refer patients to specialists. You will need to see your GP for blood tests and prescriptions regularly, often every three months even when well. Your GP will liaise with your PAH specialists, allied health team and mental health team.

- **PAH Specialist**: A specialist is a doctor who has been specially trained in the diagnosis, treatment and management of PAH. Most PAH specialists are respiratory physicians or cardiologists.
- **Respiratory nurses / PAH nurses** are nurses with special training in lung disease. They provide evidence-based care, information and guidance, emotional support and referral advice to local services.
- **Dietitians** provide advice regarding healthy eating and optimal nutrition.
- **Physiotherapists** use physical techniques to improve movement, reduce pain and stiffness and increase quality of life.
- **Pharmacists** are responsible for the quality of medicines supplied to patients, and advising patients about medicines, including how to take them, what reactions may occur and answering patient questions.
- **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.
- Other specialists that may be required in certain circumstances include **rheumatologists, immunologists, liver specialists, and infectious diseases specialists.**

Having regular contact with your healthcare team will help you to stay in control of your symptoms. You should go to your appointments even if you are feeling well.

You can talk to your healthcare team about:

- Changes in your symptoms, such as new chest pain with exercise
- Your medicines, including any side effects
- Other conditions you may have and their impact on your health and wellbeing
- Lifestyle strategies to self-manage your PAH
- Environmental risk factors you may have recently been exposed to such as smoking, dust or fumes.

Getting the most out of your appointments

There are some small things you can do to help manage the medical aspects of your PAH.

To prepare for appointments, **write down questions** that you have and take them with you – many people have questions but cannot remember them when they see their doctor.

It may help to **bring someone with you** to your appointment, like a partner or family member, to take notes for you.

Keep a diary before your next appointment. 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.

Keep the **contact details of your healthcare team** easily accessible.

Keep a current list of medications, vitamins, supplements and anything else you are taking. Note dosage and how often you take each. Sharing this information with family, specialists and nurses allows them to better understand your PAH symptoms and your treatment.

Attend all of your specialist appointments – you may need tests repeated or an assessment by your specialist doctor before your PAH medications can continue to be prescribed.

Travel with PAH

Many people with PAH 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 changes in altitude.

Support

Lung Foundation Australia	lungfoundation.com.au
Pulmonary Hypertension Association Australia	www.phaaustralia.com.au
PHuman	ph-human.com
Pulmonary Hypertension Society of Australia and New Zealand	phsanz.org
Quitline	quit.org.au 137 848
Scleroderma Australia	www.sclerodermaaustralia.com.au

These links are a guide only. Use your discretion to determine if they are appropriate for your circumstances.

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

We thank the individuals who contributed to the content and expert review of this booklet, in particular, Dr Claire Ellender, Respiratory & Sleep Physician.

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