

Living With Pulmonary Arterial Hypertension



Lung
Foundation
Australia



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Title: Living With Pulmonary Arterial Hypertension

First published: August, 2019

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ISBN: 978-0-6480834-2-9

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Printer: AllClear

This resource was developed with the input of Lung Foundation Australia Pulmonary Arterial Hypertension working group, including respiratory physicians, rheumatologists, cardiologists, PAH nurse consultants and people in the PAH community.

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About Pulmonary Arterial Hypertension

What is Pulmonary Arterial Hypertension?

Pulmonary Arterial Hypertension (PAH) is a chronic condition that affects the blood vessels (pulmonary arteries) that carry blood from the heart to the lungs. PAH is caused by narrowing or blockage of the blood vessels that connect the right side of the heart to the lungs. This narrowing or blockage of blood vessels restricts the flow of blood and oxygen through the lungs and as a result, the pressure in these blood vessels increases. The heart needs to pump against this higher pressure to maintain blood flow in the lungs and to the rest of the body, making it harder for the heart to work. Over time, this can affect the ability of the heart to work effectively and may eventually lead to heart failure.

PAH forms part of the overarching disease Pulmonary Hypertension (PH). PAH is only one type of PH (increased blood pressure within the blood vessels of the lungs).

PAH is uncommon, affecting less than 50 in one million persons¹, however it can affect males and females of all ages and ethnic backgrounds. Very rarely, PAH occurs in childhood.

¹ Hoeper, M. M. & Gibbs, J. S. R., 2014. The changing landscape of pulmonary arterial hypertension and implications for the patient. *European Respiratory Review*, 23(134), 450-457.

The role of the heart and lungs

To understand PAH, it helps to understand how the heart and lungs work.

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 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, the 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 PH.

What happens to the heart in PAH?

The heart has to work harder to push blood through the blood vessels that have become thicker or stiffer. Over time with the heart working harder, like any overworked muscle it gets 'tired'. As the strain on the heart increases, it affects the ability of the heart to work effectively and may eventually lead to 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.

What is the difference between PAH and PH?

There is an important 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 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 PH when the arteries of the lungs are directly diseased and become narrowed. 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 Pulmonary Arterial Hypertension (PAH), which may include:

- Idiopathic: meaning the cause is unknown
- Genetic: PAH can be genetically inherited in some cases
- Complication of other diseases including:
 - Connective tissue disease such as scleroderma, lupus and rheumatoid arthritis
 - HIV infection
 - Congenital (present from birth) heart disease
 - Liver disease
 - Chronic Thromboembolic Pulmonary Hypertension (CTEPH) (blood clots in the lungs)
- Medicine related: illicit drug related, and rarely chemical toxin exposure.

Symptoms

The first symptoms of Pulmonary Arterial Hypertension (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 daily routine.

It's important to know each person experiences PAH differently.

Common symptoms include:

- Shortness of breath
- Feeling tired or fatigued
- Dizziness
- Chest discomfort or pain
- A fast and/or irregular heartbeat
- Light-headedness or fainting
- Swelling in your legs and /or abdomen
- Lips and fingers turning purple or blue.

Diagnosis

It is often difficult to diagnose Pulmonary Arterial Hypertension (PAH) because breathlessness and fatigue are common symptoms that are often 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.

It is important to get an accurate diagnosis by a doctor who is an expert in this area, so you can get the correct treatment.

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.

Diagnostic tests can include:



Chest X-ray

Takes pictures of the tissues in the lungs and surrounding organs.



Electrocardiography (ECG)

Records the electrical activity of the heart using patches on your skin.



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.



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 test is used as part of the assessment of functional capacity and for monitoring progression of disease.



Echocardiogram (echo)

A specialised ultrasound examination of the heart.



Ventilation / perfusion (V/Q) scan

Compares blood and air circulation in the lungs.



Computed Tomography (CT) scan

Takes more detailed pictures than an X-ray to look at the lungs and surrounding organs.



Right heart catheterisation (RHC)

Measures pressure in the heart and lungs. This safe and painless test is usually needed to confirm the diagnosis of PAH.

Measuring the severity of PAH

You may have heard your specialist doctor 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 four levels and your specialist doctor will use this system to assess how severe your PAH symptoms are.

How Symptoms Affect You

1

You have no symptoms with normal daily activities.

2

You have some symptoms with normal daily activities AND some limits when doing physical activity.

3

You may feel symptoms when doing less than your normal activity. You may also feel more limits when doing physical activity.

4

You may feel symptoms with any kind of activity, including resting.

Management

While there is currently no cure for Pulmonary Arterial Hypertension (PAH), early treatment and self-management will help improve your condition and your quality of life.

In general, management of PAH is divided into treatment and self-management options.



Treatment options

Everyone's disease is different, and treatment options will depend on the cause of PAH and the stage at which it is diagnosed.

The treatment options for PAH have changed dramatically over the last ten 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. Ongoing monitoring of the condition is required, such as regular blood tests to monitor haemoglobin, liver function and iron levels.

As a progressive disease, PAH will generally worsen over time and early treatment is important as it can advance quickly, especially if left untreated. Taking your treatment as prescribed can help slow worsening of symptoms. Different treatments have different side effects.

As PAH is uncommon and its treatment is often complex, it should be managed by an experienced treatment centre.

There is ongoing research into the causes and management of PAH, and new medications that are being studied can sometimes be accessed by participating in a clinical trial. Clinical trials are research studies that test new and potentially better ways of improving people's health.



Medications

There are several different medications that PAH patients can take to 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 tablets by mouth but can also be given via a needle into the vein, under the skin or breathed in as a mist. Many people may need more than one medication.

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 and iloprost. Epoprostenol is given as a continuous drip into the vein. Iloprost is taken through a nebuliser and breathed in five or six times a day.

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 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.

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



Clinical trials

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 pulmonary hypertension expert centre to see if you are eligible to participate in a trial.



Oxygen therapy

Home oxygen may be prescribed by your doctor or specialist to assist with shortness of breath and to help you stay active. Some patients only use oxygen when they feel breathless (e.g. walking or exercising) but other patients need to use oxygen continuously during the day and night. While oxygen therapy may relieve breathlessness in some people, in many cases it does not.



Transplant surgery

Although most people with PAH do well with medications, in some cases complicated treatments like transplantation may need to be considered. Transplant surgery involves replacing a heart and lungs, or just lungs, from a donor.



Self Management Options

Although there is currently no cure for PAH, effective self-management 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.

Medications

It is important to take your medications as prescribed. Medication should not be stopped unless advised by your doctor; your PAH may worsen if you stop suddenly. Bring your prescriptions to your appointments, and make sure you take your medications with you if you are admitted to hospital, as not all hospitals have these medications available.

Pulmonary rehabilitation

Pulmonary rehabilitation is an exercise and education program provided by specially trained health professionals that teaches you the skills needed to manage your breathlessness and to stay well and out of hospital. The program may run for up to ten weeks and then most patients are given exercises to do at home.

Quit smoking

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

Stay healthy and active

Even minor illnesses or sickness 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 well-being 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.

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



Ensure your vaccinations are up-to-date

Support your immune system and reduce the risk of symptoms by having your vaccinations. This may include discussing seasonal influenza (flu) vaccination and a pneumonia vaccination with your doctor.

Contraception

It is important to talk to your specialist doctor about contraception. It can be very dangerous to become pregnant if you have PAH.

Eat well

Because everyone is different, there is no set diet to help to reduce PAH symptoms. 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, such as chicken
- Reduce alcohol intake.

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 to help reduce fluid swelling. Your healthcare team may ask you to measure how much fluid you drink each day.

Some foods and drinks can affect medications, 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 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. Get to know your body and understand the signs it gives. 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.

Access emotional support

PAH is an emotionally challenging disease, and it can take its toll on your emotional wellbeing. 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. Living with PAH is a different experience for everyone. 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. Anxiety and depression are not uncommon in PAH, and being a carer can also be very stressful.

It is very important to find support to help you manage your condition. Support can be found in many places: your healthcare team, health and community services, family and friends and broader social networks. Talk to your doctor or contact Lung Foundation Australia for a referral to an appropriate support service.

Have regular dental checks

Dental hygiene is particularly important for people living with PAH. 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 you are taking.

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.

Helpful Tips

There are some small things you can do to help manage life with Pulmonary Arterial Hypertension (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.
- Have a **list of your doctors**, including their phone numbers and addresses, kept by the phone.
- **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.
- **Take your PAH medications with you** when travelling or are admitted to hospital. Stopping your PAH medications may suddenly cause your symptoms to increase.
- **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.

The healthcare 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.
- Situationally required specialists include **rheumatologists, immunologists, liver specialists, and infectious diseases specialists.**
- **General practitioners** (GPs) are qualified in general practice medicine. GPs are often a person's first point of contact with the healthcare system when feeling unwell. They provide long-term, continuous care, and can refer patients to specialists.
- **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 dispensing and providing advice about medications, including how to take them, what reactions may occur and answering patient questions.

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 medications, including any side effects
- Other conditions you may have and their impact on your health and wellbeing
- Strategies to self-manage your PAH
- Environmental risk factors you may have recently been exposed to such as smoking, dust or fumes.

Support

Useful websites

Lung Foundation Australia - lungfoundation.com.au

Other organisations*

Advanced Care Planning	advancecareplanning.org.au 1300 208 582
Beyond Blue	beyondblue.org.au 1300 224 636
Carer Gateway	carergateway.gov.au 1800 422 737
Carers Australia	carersaustralia.com.au 1800 242 636
Department of Human Services	humanservices.gov.au
Dietitians Association of Australia	daa.asn.au 1800 812 942
Eating Well: Australian Dietary Guidelines	eatforhealth.gov.au
Lifeline Australia	lifeline.org.au 13 11 14
My Aged Care	myagedcare.gov.au 1800 200 422
PHuman	ph-human.com
Pulmonary Hypertension Association Australia	www.phaaustralia.com.au
Pulmonary Hypertension Network Australia	phna.info
Pulmonary Hypertension Society of Australia and New Zealand	phsanz.org
Quitline	quitnow.gov.au 137 848
Scleroderma Australia	www.sclerodermaaustralia.com.au
The Department of Health	health.gov.au 1800 020 103

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

About Lung Foundation Australia

We are inspired by hope. The hope of our community of patients, health professionals, colleagues and the millions of people affected by lung disease who are fighting to change the lung health space for the better.

As Australia's leading lung health charity, we fund life-changing research and programs to reduce the prevalence of lung disease and lung cancer and improve support and care for all Australians.

Our mission is to ensure lung health is a priority for all – from promoting lung health and early diagnosis, to supporting people with lung disease. We champion equitable access to treatment and care, and support health professionals through education and resources.

We stand for compassion, respect and collaboration, and we are fuelled by courage and driven by hope for change.

You can contact Lung Foundation Australia for more information and to access our support services including:

- Information and support line
- Lung disease information and resources
- Education seminars and webinars
- Lung cancer support nurse
- Respiratory nurse
- Peer support
- Referral to pulmonary rehabilitation
- Lungs in Action exercise programs
- Newsletter.

For more information about the services listed, please call 1800 654 301 or email enquiries@lungfoundation.com.au

The development of this resource has been funded by
Lung Foundation Australia.

Lung Foundation Australia's Rare Lung Disease program receives, in addition to monies from the sale of educational resources, competitive grants, donations and bequests, sponsorship from the following pharmaceutical company:



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