# Pulmonary Arterial Hypertension (PAH)

Lung Foundation Australia

*when you can't breathe... nothing else matters®*

## Overview

Pulmonary Arterial Hypertension (PAH) is a serious condition caused by narrowing or blockage of the blood vessels (arteries) that carry blood from the heart to the lungs. This reduces the flow of blood and oxygen through the lungs which increases the pressure in these blood vessels. Because the heart needs to pump against this higher pressure to keep blood flowing through the lungs, this can make it harder for the heart to work. This may eventually cause heart failure.

**PAH is only one type of Pulmonary Hypertension (increased blood pressure within the blood vessels of the lungs).**

## Who gets PAH?

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

## Symptoms

**The first symptoms of PAH may be very slight, and might only be noticed when you exercise e.g. climbing stairs, running, walking uphill, carrying bags, or doing housework. However as PAH gets worse, you may gradually find it harder to do your normal daily activities.**

**Common symptoms include:**

- Shortness of breath (breathlessness)
- Feeling tired or fatigued
- Dizziness
- Chest discomfort or pain
- Feeling a fast and/or irregular heartbeat (palpitations)
- Light-headedness or fainting
- Swelling in your legs, and/or the belly (fluid retention or oedema)
- Lips and fingers turning purple or blue.

## Causes

There are many causes of PAH, which may include:

- **Idiopathic** (meaning the cause is unknown)
- **Heritable / Genetic** (it is important for other close relatives to be tested if you have heritable PAH)
- **Drug and toxin** (chemical) induced
- **Chronic Thromboembolic Pulmonary Hypertension** (CTEPH) (blood clots in the lungs)
- **Association with other diseases** (this includes connective tissue diseases such as: systemic sclerosis (scleroderma), lupus (SLE) and rheumatoid arthritis)
- **HIV infection**
- **Congenital heart disease** (birth defects of the heart)
- **Liver disease.**

## Diagnosis

PAH has many common symptoms associated with other heart and lung diseases and/or which can be part of ageing, being overweight or unfit. To get the correct treatment, it is important to get an accurate diagnosis by a doctor who is an expert in this area.

**Diagnostic tests can include:**

- **Chest X-ray**
- **Electrocardiography** (ECG) (recording of the heart's electrical activity using skin patches)
- **Lung function tests** (breathing tests)
- **Echocardiogram** (ultrasound of the heart)
- **Ventilation/perfusion lung (V/Q)** scan (compares blood and air circulation in the lungs)
- **CT scan** (looks at the tissue in the lungs)
- **Right heart catheterisation** (a safe and painless procedure to measure pressure in the heart and lungs) - this test is usually needed to confirm PAH.

## Experience

It's important to know each person experiences PAH differently. As it is a progressive disease, it means PAH can advance quickly, especially if left untreated. We do know that PAH will generally worsen over time.

## Treatment

Although there is no current cure for PAH, studies have shown that early treatment is important to help improve how you feel and to keep your PAH well managed for many years.

As PAH is uncommon and its treatment is often complex, it should be managed by an experienced treatment centre. There is at least one centre in each state and your doctor should be able to advise you where they are.

> *Being compliant with medications can be time consuming and takes careful preparation, but without them I would not be able to do all the things I love. This small sacrifice is something I am willing to do to feel as good and as healthy as I can!*
>
> **- Tegan, PAH patient Melbourne**

## Current treatment options include:

- **Medicines**
  There are several different medicines that PAH patients can take to open up the blood vessels to the lungs and reduce the work for the heart. These are usually taken as tablets but can also be given by intravenous infusion (into the vein) or breathed in as a mist (like Ventolin). Some people may need more than one medicine.
- **Oxygen**
  Oxygen therapy may be prescribed 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.
- **Transplant surgery**
  Although most people with PAH do well with medicines, in some cases, complicated treatments like transplantation may need to be considered. This is where your heart and lungs or just your lungs are replaced from a donor.

## Self-management options include:

- **Pulmonary or heart failure 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. Heart failure rehabilitation may run for up to 10 weeks and then most patients are given exercises to do at home.
- **Staying active and healthy**
  Quitting smoking, being physically active, eating well, getting plenty of rest, enjoying friends, family and hobbies, practicing relaxation techniques, joining a support group and keeping a positive attitude, are all things you can do to support managing your PAH.
- **Ensuring your vaccinations are up-to-date**
  This may include discussing seasonal influenza vaccination and a pneumonia vaccine with your doctor, in order to help support your immune system.
- **Watching how much salt you eat**
  If you have problems with fluid retention (swollen legs and belly), your medical team may talk to you about avoiding food containing lots of salt and to make sure you do not drink too much fluid each day. Some patients need to take fluid tablets to help reduce fluid swelling.
- **Contraception**
  It is important to talk to your specialist about contraception. It can be very dangerous to become pregnant if you have PAH.
- **Accessing emotional support**
  Anxiety and depression is not uncommon in PAH and it is important to access support. Talk to your doctor or contact Lung Foundation for referral to an appropriate support service.

## FURTHER INFORMATION AND SUPPORT

Contact Lung Foundation Australia for more information, to access our support services and join our mailing list for regular updates and latest news.

### Lung Foundation Australia Services

- Information and Support Line
- Lung disease information resources
- Education seminars and webinars
- Lung Cancer Support Nurse
- Support groups
- Peer-to-peer connections
- Referral to pulmonary rehabilitation and Lungs in Action exercise programs
- Newsletter

### External Links

**Pulmonary Hypertension Association Australia** www.phaaustralia.com.au

**Pulmonary Hypertension Society of Australia & New Zealand** www.phsanz.org

**Pulmonary Hypertension Network Australia** www.phna.info

**Pulmonary Hypertension Association (USA)** www.phaassociation.org

**Pulmonary Hypertension Association UK** www.phauk.org

**Scleroderma Australia** www.sclerodermaaustralia.com.au

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