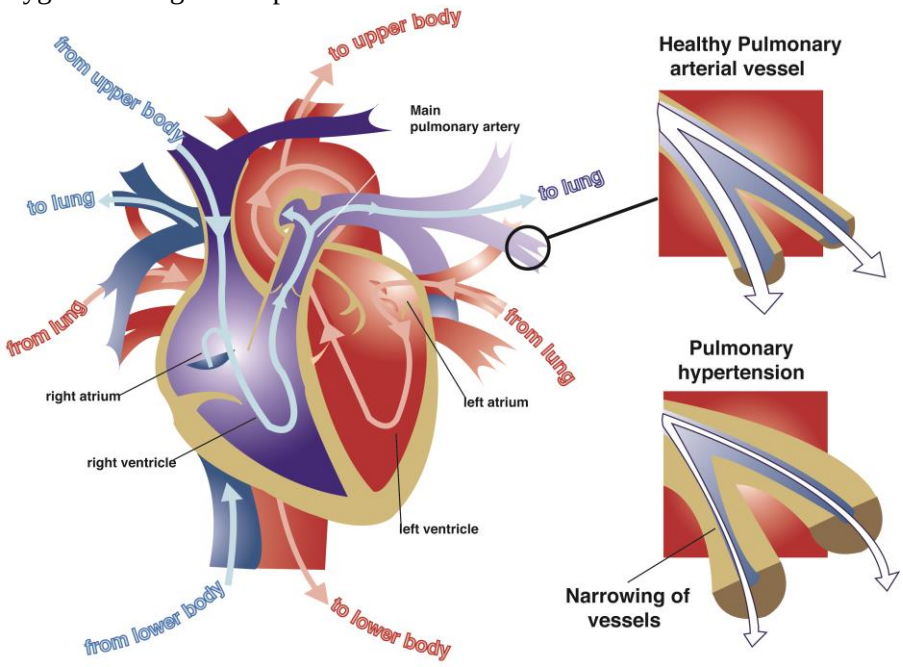
Autoimmune Resource & Research Centre
Information Sheet

Pulmonary Artery Hypertension (PAH): A Patient Overview

What is PAH?

PAH is a rare disease that is diagnosed when medical tests reveal increased resistance to flow in the pulmonary artery, pulmonary vein, or pulmonary capillaries due to narrowing of the blood vessels running from the right-side of the heart to the lungs. This resistance is shown by elevated pulmonary artery pressure. The pulmonary arteries are major blood vessels that carry blood from the heart to the small blood vessels in the lungs where oxygen exchange takes place.



PAH develops over several years and causes a restricted blood flow from the heart to the lungs. The pulmonary arteries are lined with endothelial cells which become damaged or do not function as they normally should, causing constriction or dilation. As the pressure in the pulmonary arteries increases, the right side of the heart must pump harder to force the blood into the lungs which puts a strain on the heart. PAH directly effects the pulmonary arteries and the heart and results in less oxygen supply to the body which causes the signs and symptoms of PAH.

Signs and Symptoms of PAH

People with developing PAH may feel tired and breathless and not know why and are often mistaken for lack of fitness. As the PAH develops these symptoms worsen and are not only present when the patient is exerting themselves but also at rest. Simple activities of daily living may become difficult, limiting a persons ability to go to work, play sport, garden, do housework or walk short distances.
PAH should be considered whenever breathlessness or dizziness on exertion occur. It should also be considered when palpitations, chest discomfort, swelling or unexplained fatigue occur.

Dsypnoea- dsypnoea is the medical term for shortness of breath and is usually the first noticeable symptom of PAH

Exercise tolerance – An inability to maintain endurance during light to moderate exercise or to perform short intense workouts.

Chest pain- Chest pain is a symptom of PAH that results from the heart over working.

Dizziness- PAH can lower the amount of oxygen in the blood that is sent to the brain which causes dizzy spells.

Fatigue- An overall feeling of tiredness caused by poor oxygen supply

Cyanosis- cyanosis is the medical term for blueness of the lips and skin caused from lack of oxygen.

Oedema- oedema is the medical term for swelling and can occur around the ankles, in the legs, hands and abdomen.

Palpitations- This is when the heart beats faster or irregular and harder.

Breathlessness (Dyspnoea) is often classified using the "WHO Functional Classification", which grades the impact of breathing limitation on everyday activities, Table 1.

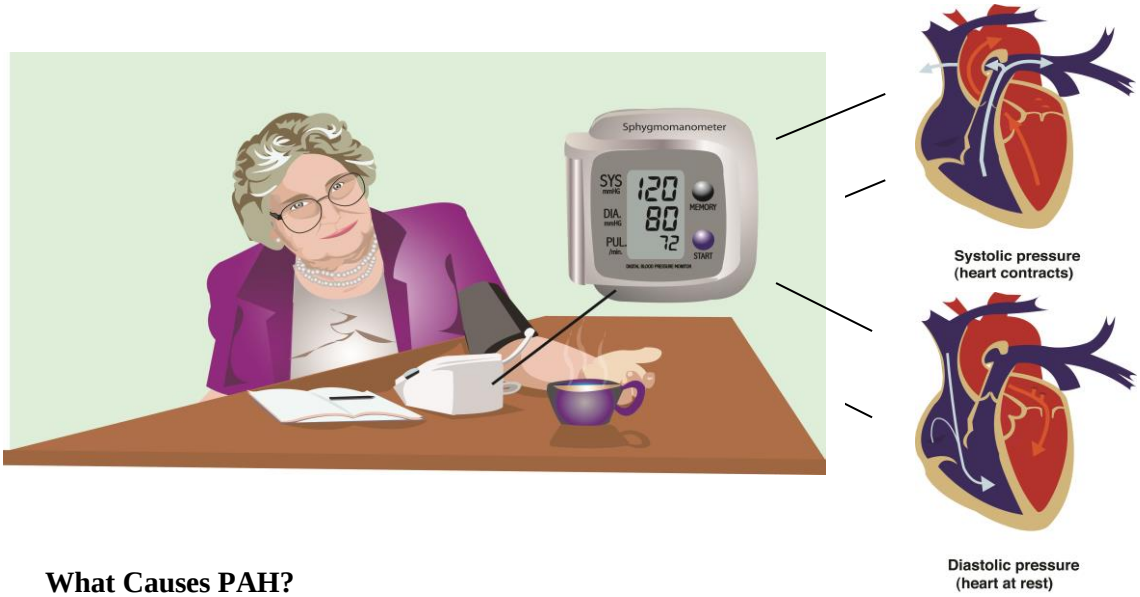
Table 1. WHO Functional Classification of Pulmonary Arterial Hypertension

Class	Description
I	No limitation of usual physical activity; ordinary physical activity does not cause increased dyspnoea (breathlessness), fatigue, chest pain, or presyncope (dizziness).
II	Mild limitation of physical activity; no discomfort at rest, but normal physical activity causes increased dyspnoea (breathlessness), fatigue, chest pain, or presyncope (dizziness).
III	Marked limitation of physical activity; no discomfort at rest, but minimal ordinary activity causes increased dyspnoea (breathlessness), fatigue, chest pain, or presyncope (dizziness).
IV	Inability to perform any physical activity at rest and possible signs of right ventricular failure; dyspnoea (breathlessness), fatigue, or both may be present at rest, and symptoms are increased by almost any physical activity.

What PAH Is Not

PAH is different to what is commonly referred to as "blood pressure"(elevated systemic blood pressure (SBP), or systemic hypertension (SHT)), which is a raised pressure in the vessels supplying blood to the body from the left-side of the heart through the aorta and its branches, as reflected by readings detected using the "blood pressure cuff".

Pulmonary Arterial Pressure measurements (PAP) can also go up in people with lung disease, hypoxia, thromboembolism (clots), and left heart muscle or valve disease, which must be ruled out before PAH can be diagnosed.



What Causes PAH?

The reasons why PAH develops in some people are often unclear. PAH is not just due to muscle contraction in the vessel walls, but also reflects disturbed function of cells lining the inner vessel walls (endothelial cells), vessel narrowing due to cell proliferation and scar tissue formation, along with more general changes in immune function (inflammation) and clotting. When no clear cause is found for PAH, it is called "idiopathic"; other causes are listed in Table 2. Note that there may be a genetic component in some people (as reflected by PAH running in the family).

Table 2: PAH Classification
Idiopathic (IPAH) Familial (the term "heritable" is now preferred) (FPAH) "Associated" (APAH): seen with autoimmune conditions (lupus, scleroderma), congenital heart shunts, portal hypertension, HIV, some diet-suppressant & other drugs, and a range of other conditions

How is PAH Detected?

When PAH is suspected, a diagnosis is confirmed by performing some or all of the following tests:

Echocardiogram- The echocardiogram uses sound waves to examine the structure of the heart by placing a microphone –like device on the pts chest. The sound waves that are sent into the heart are reflected back to form a moving image of the beating heart. This test allows the doctor to measure the thickness of the heart muscle and to some extent, the pressure in the pulmonary arteries and any leaking of the tricuspid valve. Generally, in pts with PAH, the right heart is enlarged while the left side of the heart is normal or even squashed by the right ventricle and reduced in size. The echocardiogram is also useful in monitoring the response to treatment.

Chest X-ray – Is a projection radiograph of the chest used to diagnose conditions affecting the chest, its contents and nearby structures. Chest radiography employs ionizing radiation in the form of x-rays to generate images of the chest.

Pulmonary Function tests – These tests examine the function of the lungs. The pts nose is clipped while they are asked to breath in and out of a mouth piece. Measurements of lung volume (amount of air in the lungs) and airflow are recorded.

In Pts with PAH, there is a restriction to gas movement through the small sacs within the lungs.

Right Heart Catheterisation – Catheterisation is performed by a cardiologist. A thin plastic tube is inserted through a vein in the neck, arm or leg, under local anaesthetic and is gently guided into the right ventricle and pulmonary artery. The pt is awake during the procedure and discomfort is minimal.

The catheter provides the pressures within the right ventricle and pulmonary artery. The ability of the heart to pump blood into the pulmonary artery and the presence of unsuspected holes in the heart can be established.

Exercise test (6 minute walk test) this is an easy way to determine exercise performance. The pt is asked to walk up and down on a flat surface for 6 minutes and the distance walked is measured. The amount of oxygen in the blood (oxygen saturations) may also be recorded.

Pulmonary angiogram- An angiogram provides a picture of your pulmonary arteries which can help to determine to what degree they are blocked. This procedure will exclude the presence of pulmonary hypertension due to emboli. An iodine dye is injected into the bloodstream and outlines the arteries including the pulmonary artery.

Sleep studies- The sleep study is performed in a sleep lab where various sensor leads are attached to the pt that collect data on heart and breathing rate and rhythm, duration of sleep, number of waking episodes and blood oxygen levels.

What Other Settings Should Prompt a Search for PAH?
PAH should be considered not only when symptoms such as breathlessness occur, but also it should be screened for in people recognised as being "at risk" for PAH, such as individuals with scleroderma, lupus, congenital heart disease and HIV.

Do PAH Treatments Work?
The development of PAH involves a complex process and is not fully understood. PAH has no cure at this stage however there are many medications now available to treat the disease. There is ongoing extensive research to investigate the reasons why PAH develops with the aim of developing improved treatment options and ultimately a cure.

Evidence suggests that management within a specialised multidisciplinary PAH clinic (incorporating expertise of nursing, allied health, cardiology, respiratory, rheumatology and immunology specialists) also improves patient outcomes and satisfaction

PAH Medications
The three major classes of PAH-specific therapies include prostacyclin-analogues (such as IV epoprostenol and inhaled Iloprost), endothelin-receptor blockers (also called endothelin-receptor antagonists, or "ERAs": including Bosentan and Ambrisentan), and phosphodiesterase-5 (PDE5)-inhibitors (such as sildenafil).

Recent pooled studies of the use of these therapies in PAH revealed that they reduced death rates by around 40% with good side-effect profiles. The choice of treatment involves considerations including dosage-convenience (once or twice daily dosing preferable), potential drug interactions (e.g. sildenafil and nitrates), and risks of side-effects (e.g. liver function changes with ERAs). Traditional supportive therapies, including warfarin and diuretics are often appropriate, and selected circumstances may warrant treatment with calcium blockers, digoxin and oxygen. On average, PAH-specific treatments increase the 6MWD by 40m.

Techniques to help manage your illness?
In addition to treatments/medications prescribed by your specialist there are other helpful techniques to help manage your illness.

Get enough rest
Fatigue is a warning signal from your body; listen, slow down. Resting, even for five minutes, will be beneficial. It is a sign of strength to recognise and do as your body requests.
If you have a strenuous task to do, break it up with relaxing activity. Don't feel you have to complete a task after you have started it, eg: Cleaning the house. Dust, sit down and plan what you will have for dinner. Vacuum, and then have a cup of tea. Clean the bathroom tomorrow.

Learn how to share the load, ask for and encourage family co-operation. Set tasks for each member of the family. Let them know why you are doing this, and then stick to the plan.

Keep stress under control

- Don't take on the problems of others. You can rarely do anything constructive about people's worries and it only adds to your own.
- Accept what you can't change. It is wasted energy worrying over something that is out of your control.
- You can, however, control your expectations and how you react to situations. It may take time but concentrate on taking one day at a time and not expecting too much of yourself or others.
- No matter how busy you think you will be, build into everyday time to do something you really enjoy. It may be as simple as picking a bunch of flowers to bring inside or sitting in the garden. A little self-indulgence is a wonderful pick-me-up.
- Talk to someone you trust if things are bothering you. Knowing when to seek help, or just to a compassionate listener, can avoid added stress.
- A sense of humour helps keep things in perspective.

Pace Yourself

- Write a list of things each morning that you want to do that day. Now single out those you have to do and give priority to these. This gives you a chance to organise your day allowing time for yourself.
- Build resting periods into your day so that you are not always rushed. This doesn't mean you have to lie down. Just stop for a while; take a few deep breaths, e.g. Break up shopping trips with a lunch break.
- Reassess your need to take 3 course meals for dinner, have an absolutely spotless house, and iron everything. When cooking some meals, make twice as much as you need and freeze half for a day when you feel too tired to cook.
- We tend to stand at tasks when we really don't need to. Consider sitting when you iron, cook, take a shower, and use the telephone. Experiment with chairs and compatible tables to find which suit you. An office chair with castors allows you to move in the kitchen from one workspace to another while seated.
- If you must stand for long periods, consider the shoes you wear. Shoes with heels and pointed toes throw additional weight onto your foot causing pain in your foot more quickly than low-heeled shoes.

Look for devices that will make tasks less tiring

Using assistive devices is not a sign of defeat. If you have difficulty getting out of chairs, raise the height of your chair by using blocks under the legs or cushions on the seat. Avoid stooping. Place things that you constantly use within easy reach. This applies in the workplace as well as the home. Are you organised? Make sure your desk and chair are a compatible height for you. This avoids you having to stretch or being in a constant tiring and unnatural position. Proper posture is very important to the way we feel.

Are Support Groups Helpful?

Support groups provide access to many of the helpful factors mentioned above, and are increasingly becoming available. Current clinical, support, educational and research activities undertaken by Hunter New England Health in the field of PAH are supported by Autoimmune Resource & Research Centre (ARRC) in recognition of this close connection between autoimmunity and PAH. Pulmonary Hypertension Australia and Scleroderma Australia also provide valuable advocacy, educational and support roles.

There are a number of on-line support websites available to people living with PAH.

- Pulmonary Hypertension New South Wales — info@phnsw.org.au, www.phnsw.org
- Pulmonary Hypertension Association (PAH) Australia — www.phaaustralia
- Pulmonary Arterial Hypertension Association (PHA) USA — www.phaassociation.org
- Pulmonary Hypertension Association United Kingdom — www.pha-uk.com
- The Australian Lung Foundation — www.lungnet.com.au

What Can I Expect If I Have PAH?

Searches of the internet and even medical literature can paint a very negative picture. Articles quote "3 year survival rates" which suggest that lifespan is limited. However, the following points should be remembered whenever reading information about health issues, in particular PAH:

- Much published data relies on older studies where less treatment options existed and sicker patients were recruited
- With increased awareness and screening for PAH illness, milder and earlier forms of PAH are being detected, which display better outcomes than those previously reported
- When we look at all these studies and patient outcomes, we see that death rates fall by around 40% when current PAH treatments are used
- Certain individual test results such as a longer 6MWD, milder heart pressure and function changes, and more gradual symptom onset & progression can improve individual health outcomes.
- Maintaining a positive attitude can promote more favourable outcomes.
- Research into PAH is ongoing resulting in improved treatment options.
- "Incurable" does not mean unmanageable: remember that elevated cholesterol is incurable but manageable and "incurable" is not the same as "terminal".

How can ARRC help you?

- ARRC offers information and education to help you live well with your illness.
- ARRC can help you to better understand your illness and what you can do to improve your quality of life.
- ARRC can offer access to innovative and quality research and treatments to help unlock the mysteries of PAH.

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Original Artwork by Morgan Penkae Williams

Website: www.autoimmune.org.au
Email: arrc@autoimmune.org.au
Address: 2nd floor, Pathology North Building, John Hunter Hospital, New Lambton Heights, NSW Australia 2305