

Pulmonary Arterial Hypertension

What is PAH?

Pulmonary Arterial Hypertension (PAH) is the medical term for a unique disease process which takes place in the small blood vessels in the lungs (the "pulmonary arteries and arterioles").

Although it is uncommon, it is a disease that can affect males and females of all ages and ethnic backgrounds. PAH can occur in childhood although more commonly first presents in adulthood. The disease process in the pulmonary arteries, which leads to PAH, occurs over several years and results in the progressive obstruction of blood flow 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.

Types of PAH

PAH can occur without obvious cause. Sometimes, there is a familial predisposition (ie it may be seen in several members of the same extended family). These forms of PAH are known as idiopathic PAH and familial PAH. These terms have replaced the previous term of primary pulmonary hypertension (PPH).

PAH can also be associated with other conditions, including:

- Scleroderma (also known as systemic sclerosis)
- Lupus
- HIV/AIDS
- Congenital heart defects
- Some liver disorders
- Certain drugs or toxins.

Signs and Symptoms of PAH

Some of the early symptoms of PAH include:

- Shortness of breath during exertion or normal daily activities
- Fatigue (feeling tired all the time)
- Dizziness, especially when climbing stairs or upon standing up
- Fluid build up in the limbs
- In babies, the first signs may be difficulty with feeding, breathlessness, going blue and failure to gain weight.

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 progresses, the symptoms become more noticeable. Breathlessness and tiredness become more a part of daily life, so that even simple tasks, such as getting dressed and walking short distances, become increasingly difficult. Fluids can build up in the legs and chest pain may also be experienced. These are all signs of the increased stress being placed on the heart as it attempts to adjust to the disease's effects on the body's functioning.

It is usually when these symptoms severely affect normal activities of daily life, that people go to their doctor to identify the cause.

The signs and symptoms of PAH are often hard to distinguish from other diseases or conditions, particularly in the early stages. As a result, formal diagnosis of PAH can be delayed.

Diagnosis

To assist in the diagnosis of PAH, the doctor asks questions to gain a clear and detailed history of all prior and current medical conditions, and those of close relatives. Additionally, the doctor needs to know of any drugs (be they prescription, over-the-counter or illegal) that have been taken in the past, or are currently being taken.

Blood tests and a physical examination are conducted. Additionally, further tests are required to help diagnose PAH. Diagnosis usually results from ruling out the presence of other diseases which may have similar symptoms to PAH.

Your doctor will have more information on each of these tests, such as how they are performed and what they are looking for. Other tests, which are not listed here, may also be used to help diagnose PAH.

An echocardiogram (ultrasound of the heart) is required and if there is a suggestion of PAH, other tests including breathing tests and walking tests, CT scans and a right heart catheter are also required.

Doctors have devised a grading system to help understand the extent to which PAH symptoms are affecting lifestyle and well-being. This is referred to as the PAH "functional class", and helps the doctor know what type of treatment can be given. This functional classification system is based upon the extent of functional limitation caused by symptoms.

Class I describes individuals with PAH who experience no limitation in daily function, progressing to class IV in which individuals are unable to perform any physical activity and have symptoms at rest, indicating more severe PAH.

This brochure is one in a series produced by Lung Foundation Australia to provide information on lung disease, its treatment and related issues. The information published by Lung Foundation Australia 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.

Any information relating to medication brand names is correct at the time of printing. Lung Foundation Australia has no control or responsibility for the availability of medications, which may occasionally be discontinued or withdrawn.

Please consult your family doctor or specialist respiratory physician if you have further questions relating to the information contained in this leaflet. For details of patient support groups in Australia please call 1800 654 301.

Treatment

Without treatment, the outlook for patients with PAH is uncertain. However, there are now several good treatments available. These treatments vary from tablets to nebulisers and for individuals with very severe PAH, continuous intravenous infusion therapy is available.

Most individuals with PAH do very well with this treatment but occasionally and only if these treatments are ineffective, more involved treatments including lung transplantation may need to be considered.

As PAH is uncommon and its treatment complex, all individuals with PAH should be managed by an experienced centre of which there is at least one in each state in Australia. Your doctor should be able to advise you where the centres are in your state and further information is available from the following websites.

Support Information

Looking for support and/or further information on PAH?

- Pulmonary Hypertension Association (PAH) Australia
 - www.phaaustralia.com.au
- Pulmonary Hypertension Association (PHA) USA
 - www.phaassociation.org
- Pulmonary Hypertension Association, United Kingdom (PHAUK)
 - www.phassociation.uk.com
- The Lung Foundation Australia
 - www.lungfoundation.com.au
 - Free call: 1800 654 301
- WA Pulmonary Hypertension Association
 - Call The Lung Foundation Australia on 1800 654 301 for details
- Scleroderma Australia
 - www.sclerodermaaustralia.com.au
 - Ph: 02 9990 5159
 - Call Scleroderma Australia to obtain contact details of Statewide organisations.

Content Updated August 2011

TD0811V2PAH

Last Updated (Thursday, 15 September 2011)

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