Pulmonary hypertension

Summary

- Pulmonary hypertension is high blood pressure on the lungs.
- High blood pressure on the lungs can be due to a number of different causes.
- Some forms of pulmonary hypertension can be treated, reducing symptoms and improving quality of life.
- If you have pulmonary hypertension, it is important to be reviewed in a specialist centre regularly as the treatment is very different depending on the cause.
- Pulmonary hypertension is best managed by a team of healthcare workers including cardiologists, respiratory physicians, rheumatologists, specialist nurses, and physiotherapists.

Pulmonary hypertension is high blood pressure on the lungs. This can have flow-on effects for your heart. The right side of the heart pumps blood through the extensive network of arteries (blood vessels) in the lungs, and the left side pumps blood everywhere else. If the pressure in the lungs is too high, the right side of the heart has to work harder. This can lead to the heart muscle changing size and shape, becoming less efficient and resulting in abnormal function.

In the past, pulmonary hypertension was associated with high mortality. The development and increased availability of new medications has led to significant improvements in life expectancy for people with pulmonary hypertension. Although the condition is still incurable, there are now many treatment options available.

How pulmonary hypertension develops

Pulmonary hypertension begins when there is damage to the blood vessels of the lungs at a molecular level. Over time these problems build up and, if not corrected, can lead to permanent changes in the makeup of those blood vessels. Pulmonary hypertension is usually a progressive disease (becomes worse over time), which can be slowed with treatment. Part of the difficulty with treating people with pulmonary hypertension is the subtle and non-specific nature of their symptoms. This often leads to a delay in diagnosis.

Classification of pulmonary hypertension

There are many different reasons a person may develop pulmonary hypertension. The World Health Organisation (WHO) classifies pulmonary hypertension by these underlying causes. These classifications include:

- **Group 1**: Pulmonary hypertension from genetic or unknown (‘idiopathic’) causes, drug-induced, or related to connective tissue diseases
- **Group 2**: Pulmonary hypertension from failure of the left side of the heart
- **Group 3**: Pulmonary hypertension from lung diseases and chronic low levels of oxygen
- **Group 4**: Pulmonary hypertension from chronic blood clots on the lungs
- **Group 5**: Pulmonary hypertension from ‘mixed or miscellaneous’ causes.

Pulmonary hypertension is also classified by its severity and the impact it has on a person’s activities. These classifications include:

- **Functional Class I**: symptoms do not limit the person’s physical ability
- **Functional Class II**: symptoms result in slight limitation of physical ability, but the person is comfortable at rest. Ordinary physical activity may cause shortness of breath
- **Functional Class III**: symptoms result in marked disability, with less than ordinary physical activity bringing on breathlessness. Comfortable at rest
- **Functional Class IV**: symptoms at rest. Severely limited by shortness of breath.
Risk factors for pulmonary hypertension

Risk factors for pulmonary hypertension include:

- family history of pulmonary hypertension – especially a first-degree relative such as a parent, sibling or children
- connective tissue disorders or autoimmune diseases, including scleroderma, lupus, and rheumatoid arthritis
- living at high altitudes for extended periods of time
- obesity
- female sex
- obstructive sleep apnoea, especially untreated
- congenital heart disease
- underlying lung disease
- chronic liver disease
- infectious diseases including human immunodeficiency virus (HIV), hepatitis B and hepatitis C
- use of certain drugs and medications including methamphetamines, chemotherapy, and some diet drugs or ‘appetite suppressants’.

Screening for pulmonary hypertension

There is no currently recommended screening for pulmonary hypertension. If a member of your immediate family (mother, father, sibling, or child) is diagnosed, it is recommended that you have an echocardiogram (ultrasound of the heart) to screen for the condition.

The gene most closely associated with inheritable pulmonary hypertension is BMPR2. However, many people without this gene develop pulmonary hypertension, and some people who do have the gene lead healthy lives without ever suffering from high blood pressure on the lungs. For these reasons, genetic testing is only done in very specific circumstances, often for academic purposes such as research.

If you have scleroderma, you may be monitored yearly for development of pulmonary hypertension.

Symptoms of pulmonary hypertension

Symptoms of pulmonary hypertension include:

- shortness of breath (dyspnoea), especially on exertion
- chest pain (angina)
- fatigue, tiredness, or weakness
- blue tinge to lips or skin (cyanosis)
- dry cough
- light-headedness or loss of consciousness (syncope)
- swollen legs (peripheral oedema)
- weight gain over a short period of time
- abdominal bloating.

Diagnosis of pulmonary hypertension

Pulmonary hypertension is diagnosed via a procedure called a right heart catheterisation. Other tests including CT scans, echocardiograms, and chest x-rays may also be used but a right heart catheterisation is necessary to confirm the diagnosis.

Investigation into pulmonary hypertension

The series of tests required to confirm a diagnosis of pulmonary hypertension can be quite extensive. Your first appointment with your specialist might just be spent organising these tests, which may include:

- **blood tests** – your doctor will order a targeted set of blood tests for different causes of pulmonary
hypertension, including low iron, altered thyroid, kidney, or liver function, and certain viruses (including hepatitis and HIV)

- **electrocardiogram (ECG)** – a tracing of the electricity in the heart which may help your team identify issues with its conduction
- **chest x-ray** – a plain x-ray of the chest to screen for lung disease
- **high resolution CT of the chest** – a higher definition lung scan to screen for structural diseases not detected at the resolution seen on the chest x-ray. Not always required
- **CT pulmonary angiogram (CTPA)** – a chest scan that maps the arteries of the lungs, looking for clots. Although it picks up some pictures of the surrounding tissue, it is not as good as the high resolution scan at assessing lung tissue, so in some circumstances your doctor may request both kinds of CT test
- **ventilation perfusion nuclear medical scan (V/Q scan)** – this involves breathing a gas which is taken up by the lung tissue before having special images of the chest taken. A ‘mismatch’ between the areas supplied by blood vessels and taking up gas may represent a lung clot. This is better than the CTPA at picking up lung clots which have been there a long time
- **echocardiogram** – this is an ultrasound of the heart. It provides information including the pressure, size, and function of the right side of the heart. These tests may be performed regularly to monitor pulmonary hypertension non-invasively
- **six minute walk test** – this is a test where you walk at a gentle pace for six minutes while measuring your oxygenation with a clip on your finger. This test is performed at most clinics
- **respiratory function test** – specialist breathing tests which determine the capacity and function of the lungs. Additional measurements such as lung volumes may be performed
- **sleep study** – if indicated by a sleepiness score or other measurement, a sleep study may be requested. This involves sleeping overnight in a sleep lab and having oxygenation and a few other parameters measured overnight with interpretation from a respiratory and sleep physician
- **right heart catheterisation** – a very specialised test performed in a cardiology catheterisation lab where a cardiologist makes a small cut in the femoral or jugular vein and guides a special tool called a catheter to the heart to take measurements of the pressures at different, specific places. This is a routine procedure performed as a ‘day case’, with the majority of patients going home the same day.

If your doctor asks you to have any tests before the appointment, please do so because it will speed up the process of determining what the nature of your pulmonary hypertension may be, and how to treat it.

**Treatment for pulmonary hypertension**

If you are diagnosed with pulmonary hypertension you will have three- to twelve-monthly reviews, depending on:

- the severity of your illness
- the cause of your pulmonary hypertension
- the treatment you are receiving.

You will be encouraged to make healthy lifestyle changes. You can reduce your risk of worsening pulmonary hypertension by maintaining a healthy weight, exercising regularly and avoiding smoking. A healthy diet and moderate amounts of exercise are proven to be of benefit in improving effort tolerance and reducing symptoms.

Your treating doctor will issue the appropriate referrals to ensure that any underlying medical conditions contributing to your pulmonary hypertension are managed by the appropriate specialists.

Your GP will help you to lower your cardiovascular risk by treating high cholesterol, diabetes, or high blood pressure.

It is important to tell your GP or specialist if you are feeling overwhelmed, as they may be able to help you or refer you to someone who can. They may refer you to a counsellor or psychologist who can help you cope with the changes your symptoms make to your everyday life.

**Medication for pulmonary hypertension**

If you are diagnosed with pulmonary hypertension it is important that you see a specialist pulmonary hypertension
service so that you can start appropriate medications as soon as possible

Many of the medications used for pulmonary hypertension are very expensive, and are supplied to the public through government and private company 'compassionate access' schemes. Because of this, you may be asked to repeat some procedures such as six-minute walk tests, echocardiograms, and even right heart catheterisation, to prove the medications are having a continued effect.

Medications used for pulmonary hypertension include:

- **vasodilators** – these make the blood vessels in the lungs relax and expand. They are often the first-line of treatment for pulmonary hypertension. If appropriate, a combination of vasodilators may be used to treat pulmonary hypertension. Examples include sildenafil (Revatio™) and tadalafil (Adcirca™)

- **endothelin receptor antagonists** – these block a receptor, forcing the blood vessels to relax and expand. Examples include ambrisentan (Volibris™), macitentan (Opsumit™), and bosentan (Tracleer™)

- **riociguat** (Adempas™) – this is a newer medication. It works by directly acting on the blood vessel walls

- **blood thinners (anticoagulants)** such as warfarin (Coumadin™ or Marevan™), apixaban (Eliquis™), rivaroxaban (Xarelto™), or dabigatran (Pradaxa™) may be used to treat pulmonary hypertension caused by blood clots on the lungs. Depending on your risk factors, this medication is usually taken lifelong

- **fluid tablets (diuretics)** – these may be used to help remove excess fluid and reduce ankle swelling. You might be asked to take a regular low dose, and ‘top up’ with an extra tablet if your weight increases two days in a row. Regular weight recording (preferably daily) is an important part of management. Some people using diuretics may be asked to practise ‘fluid restriction’ – drinking less than a certain amount (normally 1.5 l) per day. Examples of diuretics include furosemide (Lasix), hydrochlorothiazide (HCT), or spironolactone (Spiractin™ or Aldactone™).

**Surgery**

In special cases of Group 4 pulmonary hypertension, a procedure called a pulmonary endarterectomy (PEA) may be performed to remove blood clots on the lungs. This procedure can potentially cure some people’s pulmonary hypertension by improving the function of the right side of the heart and lowering the lung pressures. Patient selection is very important, as this procedure carries some risk and does not work for all people.

In rare situations, patients may be referred for a lung transplant. Choosing the right candidates for this high-risk surgery is very important, and all patients must be trialled on appropriate medical therapy before consideration of referral to a transplant centre. See ‘Lung transplantation’ for more information.

**Oxygen therapy**

Home oxygen (also known as supplemental or domiciliary oxygen) is when a patient is given canisters of air with extra oxygen in it to inhale during time spent at home. This may be helpful for some people with pulmonary hypertension who have particularly low oxygen levels in their blood. It can help improve symptoms of breathlessness and tiredness by causing the arteries in the lungs to ‘relax’.

Home oxygen may be used continuously over 24-hours, or only during times of higher activity. Unfortunately it has not been shown to prolong life expectancy, but does improve people’s quality of life.

**Research into pulmonary hypertension**

Pulmonary hypertension is an area of intense research, particularly with respect to:

- the correct type of medication to use in the different groups of pulmonary hypertension, and in which combinations

- management of right heart failure, and possible new treatment targets or therapies

- treatments at the molecular level, such as monoclonal antibodies, which may be useful tools to slow disease progression

- further genetic markers that may identify people at high-risk of developing pulmonary hypertension.

**Caring for someone with pulmonary hypertension**

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Pulmonary hypertension usually progresses slowly, and eventually results in shortness of breath and a decreased ability to perform the activities of daily living at home.

For most of the course of the illness you will not need any extra assistance at home. However, at the end stage of pulmonary hypertension, you may need services such as home oxygen, council cleaning, and meals on wheels. These can be arranged through your GP or local volunteer services.

**Pregnancy and pulmonary hypertension**

If you have pulmonary hypertension, before considering conception, please talk to your specialist. Pregnancy in women with pulmonary hypertension is very high-risk, both to the mother and the baby.

Many of the medications used to treat pulmonary hypertension are toxic to developing foetuses or untested in pregnant women. Unfortunately, for many women with pulmonary hypertension, carrying a baby to term may not be possible.

**Where to get help**

- Your GP
- Your pulmonary hypertension specialist – for example your cardiologist, respiratory physician or rheumatologist
- Pulmonary rehabilitation centre
- Psychologist
- **Pulmonary Hypertension Society of Australia and New Zealand (PHSANZ)**
- **International CTEPH Association**
- **The Lung Foundation**
- **Scleroderma Australia**
- **Pulmonary Hypertension Association (USA)**

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