



## Pulmonary arterial hypertension

**Pulmonary arterial hypertension (abbreviated to PAH) is a rare condition affecting about 15 people out of every million. It is caused by very high blood pressure in the arteries that lead from the heart to the lungs, which are called "the pulmonary arteries". This increase in blood pressure puts strain on the heart, and will eventually cause the right side to stop working. Pulmonary arterial hypertension is a very serious disease, for which there is unfortunately no cure. However, there are many treatments now available that can benefit people with the condition and improve their quality of life.**

### What are the pulmonary arteries?

The pulmonary arteries are the blood vessels that carry blood which has very little oxygen in it from the bottom right of the heart (right ventricle) to the small arteries and capillaries in the lungs. The blood in these capillaries is then oxygenated by the lungs and carried back to the left side of the heart, before it is pumped around our whole body.

### Who gets pulmonary arterial hypertension?

Pulmonary arterial hypertension can affect anyone of any age, of any sex and of any race. However, it is most common in women aged between 30 and 50 years.

### What are the symptoms of pulmonary arterial hypertension?

As pulmonary arterial hypertension needs to be treated as soon as possible, it is essential that it is detected very early. However, it is very difficult to identify in its early stages because its symptoms are similar to those of other heart and lung diseases.

Breathlessness, especially when exercising, is the first and main symptom. It may feel as if you are not able to get enough air into your body. Other symptoms may develop as the condition gets worse.

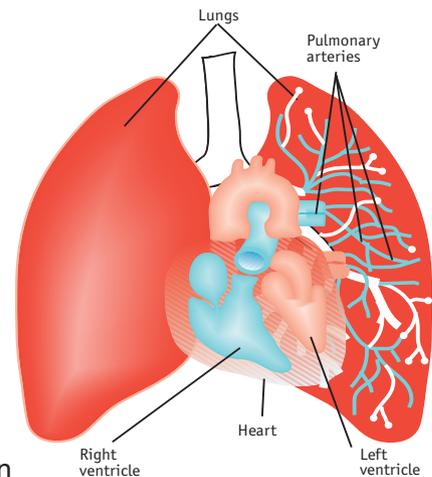
These symptoms, which may occur when exercising or at rest, include:

- Tiredness – "fatigue"
- Dizziness
- Fainting – "syncope"
- Chest tightness or pain, especially when exercising
- Swelling in the ankles or legs – "oedema"
- A fast or irregular pulse
- Cough

### Are there different types of pulmonary arterial hypertension?

Pulmonary arterial hypertension is classified into different groups depending on what causes it.

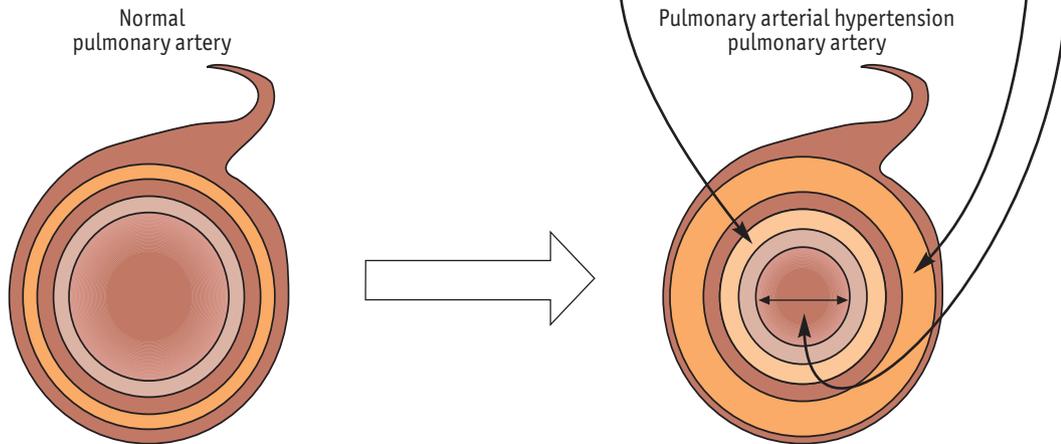
1. **"Idiopathic" pulmonary arterial hypertension** is pulmonary arterial hypertension with no obvious cause.
2. **"Familial" pulmonary arterial hypertension** is when the disease is shown to be caused by a problem with a gene that runs in your family. Some genes have been found to play a role in pulmonary arterial hypertension and treatment will improve and advance as more is understood about them. The most important genetic mutations associated with familial pulmonary arterial hypertension affect the gene *BMPR2*, which contains the code for a protein that is found at the surface of the cells lining the pulmonary arteries.
3. **Associated pulmonary arterial hypertension** is when the disease occurs in patients with other conditions. Examples of these include being infected with HIV, having a liver disease and having a disease associated with connective tissue (such as scleroderma or lupus erythematosus). Using appetite suppressants, cocaine or amphetamines can also increase the probability of getting pulmonary arterial hypertension.



## What happens in pulmonary arterial hypertension?

Blood pressure rises in pulmonary arterial hypertension because the blood is unable to flow properly through the blood vessels. The reasons for this include:

- narrowing of blood vessels due to muscles around the blood vessels tightening up, which is termed "vasoconstriction"
- changes to the shape and thickness of the blood vessel walls, because of an increase in the amount of muscle
- swelling of the blood vessel walls caused by inflammation
- the formation of small blood clots within the blood vessels



## How can pulmonary arterial hypertension be diagnosed?

Doctors will perform a number of different tests to diagnose pulmonary arterial hypertension, many of which will be used to rule out other diseases with similar symptoms.

Further tests may then be carried out, including lung function tests, exercise tests and blood tests.

<b>ECG (electrocardiogram)</b>	Measures the rate of your heartbeat and can tell whether you have any conditions of the heart. If you have pulmonary arterial hypertension, the ECG will show problems with the right side of your heart.
<b>Chest X-ray</b>	A picture is taken of your chest to show if the right side of your heart or your pulmonary arteries are bigger than usual. It can also help to rule out some other lung and heart diseases.
<b>Doppler echocardiogram</b>	With this test the doctor can form a moving picture of your heart from sound waves that are directed at your heart. It will be able to tell the size and shape of your heart, and assess how well it is working, as well as estimating the pulmonary artery pressure. It is important for screening for pulmonary arterial hypertension, as well as seeing how severe the condition is.
<b>Right-heart catheterisation</b>	This is essential to confirm the diagnosis of pulmonary arterial hypertension. It provides a really precise measure of the blood pressure on the right side of the heart and in the pulmonary arteries. It also shows the doctor how well the right side of your heart is pumping blood. The average blood pressure in the pulmonary arteries in a resting person is 14 mmHg. However, in PAH it is more than 25 mmHg.

## ► How can pulmonary arterial hypertension be treated?

Although pulmonary arterial hypertension can not be cured, in the past decade great improvements have been made in the understanding of the disease, which has resulted in significant improvements in treatment. Treatment can now allow you to exercise longer and improve your quality of life and outcome.

Most drug treatment aim to stop or reduce the processes which block the pulmonary arteries and cause pressure to rise. Different treatments will help different people.

<b>Anticoagulants</b>	In the basic treatment of pulmonary arterial hypertension, you may be prescribed drugs called anticoagulants, which will reduce the risk of blood clots in your pulmonary arteries.
<b>Calcium channel blockers</b>	A small group of pulmonary arterial hypertension patients will benefit from treatment with calcium channel blockers and the doctor will be able to tell this after right-heart catheterisation. Calcium channel blockers help to relax the muscles in the blood vessels, which will increase the blood and oxygen supply to the heart and reduce its workload.
<b>Prostacyclin therapy</b>	<p>Prostacyclin works by widening the arteries of the lung and preventing blood clots forming. Unfortunately, the tablet versions of prostacyclin, such as beraprost, are not very effective so the treatment has to be given in other ways:</p> <p>Intravenous (i.v.) prostacyclin is called epoprostenol. This can be given into the veins via a pump and is the main treatment for the most severe pulmonary arterial hypertension patients. Epoprostenol has to be given constantly in order to work effectively.</p> <p>As giving epoprostenol intravenously can be quite difficult, a version of the treatment has been developed which can be given under the skin ("subcutaneously"), this is called treprostinil.</p> <p>Recently, prostacyclin has been developed into a form that can inhaled through a nebuliser. This means that it is now easier to use and works on the lungs directly. This is called iloprost and usually needs to be taken 6 times a day. Inhaled treprostinil is also currently being tested.</p>
<b>Endothelin receptor antagonists</b>	Another important form of treatment is the endothelin receptor antagonists. These tablets taken twice a day help to widen the blood vessels and reduce the build up of muscle in the vessels. Bosentan, sitaxsentan and ambrisentan are all drugs in this group.
<b>Phosphodiesterase type 5 inhibitors</b>	Phosphodiesterase type 5 inhibitors, such as sildenafil, work by widening the arteries of the lung. Sildenafil is given by mouth three times per day. New drugs such as tadalafil are currently being tested.
<b>Lung transplantation</b>	If medical therapy has not been effective, surgery to replace one or both lungs and sometimes the heart if necessary with healthy organs from a donor can help some patients.

## Living with pulmonary arterial hypertension

A diagnosis of pulmonary arterial hypertension should not stop you from leading a full and active life, as long as it doesn't cause you pain or problems. However, the more severe your symptoms, the less you may be able to do. There are many things that you can do to help relieve your symptoms without even taking medication:

- Avoid high altitude.
- Avoid getting pregnant as this puts severe strain on your heart and blood vessels.
- Identify the activities that use more of your energy and look for alternative activities.
- Invest time in the preparation of nutritious meals. Your diet should be low in salt and pre-cooked meals should be avoided. Control your liquid intake.
- Have 3 meals a day, eat slowly and while seated. Control your weight.
- Don't smoke or drink alcohol. You can ask for help in quitting both of these.
- Dental hygiene is vital to prevent infections.
- Plan your day so that you can take breaks throughout to rest.
- Take your time for the preparation and intake of the medication so that it can be most effective.
- If you need to start a new treatment, do research, learn about it and ask for educational material.
- When you are taking different medications, be well informed of the effects they may cause.
- Organise your medicine cabinet. Make sure you have extra sets of medication in case of any eventuality.
- If your medication varies according to the day of the week, organise your diary.
- Always keep the telephone number of the medical team that takes care of you at hand.
- If you are going to travel, prepare well in advance anything you may need.

## Useful websites

### PHA Europe

The European Association for Pulmonary Hypertension, PHA Europe, was founded in 2003. It is an umbrella organisation for PH Support Associations throughout Europe. The ultimate goal for PHA Europe is to unite all European countries and to encourage the formation of effective PH Support Associations nationally. You can find your national PH Support Association using this site.

[www.phaeurope.org](http://www.phaeurope.org)

### PHA Association

The mission of the Pulmonary Hypertension Association (PHA) is to find ways to prevent and cure pulmonary hypertension, and to provide hope for the pulmonary hypertension community through support, education, advocacy and awareness. The PHA Association is based in the USA.

[www.phassociation.org](http://www.phassociation.org)

- ▶ For additional information and links go to [www.pah.european-lung-foundation.org](http://www.pah.european-lung-foundation.org)



European Respiratory  
Society

The ELF is the public voice of the European Respiratory Society (ERS), a non-profit-making medical organisation with more than 8,000 members in more than 100 countries. The ELF is dedicated to lung health throughout Europe, and draws together the leading European medical experts to provide patient information and raise public awareness about respiratory disease.

This material was based on information from the following scientific references and with the expert help of ERS PAH expert Prof. Marc Humbert.

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3. Humbert M, Sitbon O, Chaouat A, et al. Pulmonary arterial hypertension in France: results from a national registry. *Am J Respir Crit Care Med* 2006; 173: 1023–1030.