Pulmonary hypertension

Pulmonary hypertension is high blood pressure in the lungs. It can affect people of any age. It's unusual to get pulmonary hypertension on its own – it's usually caused by another lung or heart condition.

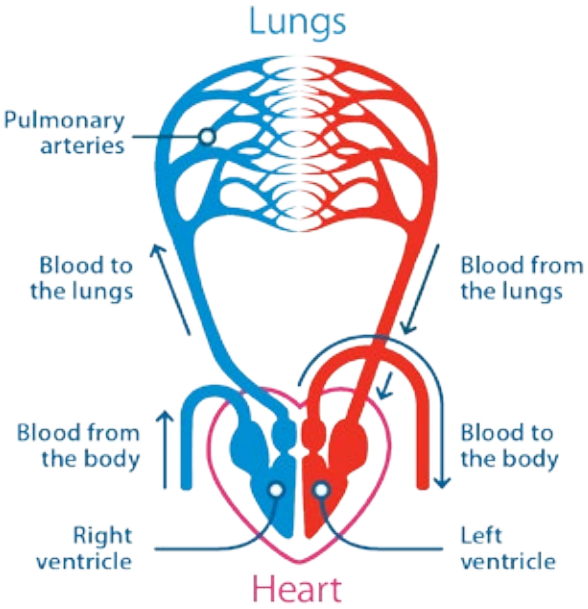
In the UK, around 6,000-7,000 people have pulmonary hypertension. It's thought that more people have the condition and haven't been diagnosed.

What does pulmonary hypertension mean?

Pulmonary: to do with your lungs
Hypertension: high blood pressure

If you have pulmonary hypertension, you have high blood pressure in the pulmonary arteries. These are tubes that carry blood to your lungs.

The right side of your heart pumps blood that's low in oxygen through the pulmonary arteries to your lungs. Here the blood picks up oxygen. The oxygen-rich blood is then pumped to the left side of your heart, which pumps it to the rest of your body (see diagram below).



If you have pulmonary hypertension, the high blood pressure in your lungs means that the right side of your heart has to work harder to pump blood. Over time, the right side of your heart begins to struggle with this extra work. It can become weaker and pump less effectively. This causes the symptoms of pulmonary hypertension.

What are the symptoms of pulmonary hypertension?

The most common symptoms of pulmonary hypertension are:

- feeling short of breath
- feeling tired
- feeling dizzy or faint
- chest pain, especially after exercise
- a racing or irregular heartbeat
- leg, ankle and tummy swelling

These symptoms may get worse when you exercise, or when you do activities like walking up hill or up the stairs.

What causes pulmonary hypertension?

There are five main types of pulmonary hypertension:

Type 1: Pulmonary arterial hypertension (PAH)
This type is rare. It's caused by changes to very small arteries which take blood from the right side of your heart to your lungs. The walls of the arteries get thicker and stiffer. This makes the space for blood to pass through narrower which increases the blood pressure.

It can be genetically inherited. It's also associated with connective tissue diseases, liver disease, congenital heart defects, HIV and certain drugs.

Type 2: Pulmonary hypertension due to left heart disease
If there are problems with the left side of your heart, it can stop blood flowing easily through the lungs. The right side of your heart has to work harder to pump blood through your lungs. This increases the blood pressure in your pulmonary arteries.

This is one of the most common causes of pulmonary hypertension.

Type 3: Pulmonary hypertension due to lung conditions or lack of oxygen
The common causes in this group are:

- chronic obstructive pulmonary disease (COPD)
- interstitial lung disease (such as pulmonary fibrosis)
- obstructive sleep apnoea (OSA)

These conditions reduce the amount of oxygen getting into your lungs. When there's a low level of oxygen, your pulmonary arteries get narrower. This increases the blood pressure.

This is another very common cause of pulmonary hypertension.

Type 4: Pulmonary hypertension due to blood clots (chronic thromboembolic pulmonary hypertension)
When blood clots are carried into the pulmonary arteries, they can sometimes form scars. The scars block the flow of blood and this increases the blood pressure.

Type 5: Pulmonary hypertension due to a range of causes
This is a mixed group of rare causes. They're grouped together because their causes are less clear.

How is pulmonary hypertension diagnosed?

Pulmonary hypertension can be difficult to diagnose, because the symptoms are similar to many other heart and lung conditions.

If your doctor thinks you have pulmonary hypertension, they may suggest you have some tests. The most common are:

Echocardiogram: An echocardiogram, often called an echo, uses high-frequency sound waves to create an image of your heart. This can be used to estimate the pressure in your pulmonary arteries. It can also test how well the right and left sides of your heart are pumping.

Right heart catheterisation: If an echo test suggests you have pulmonary hypertension, you will need a second test to make sure. This test is called a right heart catheterisation. A long, thin, flexible tube is inserted into a vein and fed through to your pulmonary artery. This can be used to accurately measure the blood pressure in the right side of your heart and in your pulmonary arteries.

Other tests you might have are:

- blood tests
- breathing tests
- sleep studies
- other types of scan. These may include:
    - a scan of your lungs to look for lung disease and blood clots
    - a V/Q scan, which is a type of scan that looks for blood clots
    - a scan to look at your liver
- exercise or walking tests

How is pulmonary hypertension treated?

It's important to treat pulmonary hypertension to stop it getting worse. Your treatment will depend on what type it is and what's causing it.

Type 1: Pulmonary arterial hypertension (PAH)
If you have pulmonary arterial hypertension, you'll be referred to a specialist centre for treatment .

You'll usually have background therapy first – treatments to help with your symptoms. Background therapy may include:

- **diuretics**: These remove excess fluid from your body. They treat symptoms like swollen ankles.
- **oxygen therapy**: In oxygen therapy, you breathe air with a higher concentration of oxygen than normal air. This increases your oxygen levels and improves your symptoms.
- **anticoagulation**: With some forms of PAH you have a greater risk of getting blood clots. Anticoagulant medicines like warfarin stop blood clots forming.
- **pulmonary rehabilitation**: This is a programme of gentle physical exercise and advice, which helps you cope with breathlessness. The classes are run by physiotherapists and specialist nurses.

Type 1 continued...

Your health care professional may also offer you treatments called **pulmonary vasodilators**. These aim to lower the blood pressure in your lungs by relaxing and opening up the pulmonary arteries. With more space for the blood to pass through, the right side of your heart is under less strain and should work better. This means that your symptoms should improve.

Pulmonary vasodilators can be taken as tablets, with a nebuliser or as an injection. You can find out more about these treatments on the Pulmonary Hypertension Association website.
Visit **www.phassociation.uk.com**

Your health care professional will advise you:

- not to become pregnant
- not to travel to high altitude. This includes avoiding high altitude destinations like mountains, as well as flying
- to keep up to date with flu and pneumonia vaccinations

Types 2 and 3: Pulmonary hypertension due to left heart disease or lung conditions
In both these cases pulmonary hypertension is a **secondary condition**. This means it's caused by another lung or heart condition, which is the **primary condition**.

If your pulmonary hypertension is a secondary condition, your health care professional will focus on treating the primary condition. If you get the best treatment for your primary condition, it should improve your pulmonary hypertension too.

In these cases, it's very unlikely that you'll be treated directly for pulmonary hypertension.

For information on different types of lung disease and how they're treated go to **www.blf.org.uk/support**

Type 4: Pulmonary hypertension due to blood clots (chronic thromboembolic pulmonary hypertension, CTEPH)
If your pulmonary hypertension is caused by blood clots, your health care professional will treat you with anticoagulant medicine. This medicine stops more blood clots from forming. The most common anticoagulant medicine is warfarin, which is taken as a tablet.

If your blood clot has caused scar tissue in your pulmonary arteries, you may be offered a **pulmonary endarterectomy**. This is an operation to remove scar tissue from the inside layer of the pulmonary arteries. This improves the blood flow and reduces the pressure inside the arteries. This is a specialist operation and is only performed at Papworth Hospital in Cambridgeshire.

If you're not suitable for surgery or you still have some pulmonary hypertension after the operation, you may be given tablets called **pulmonary vasodilators**. These help to open up narrowed pulmonary arteries.

Type 5: Pulmonary hypertension due to a range of causes
Because pulmonary hypertension in this group is caused by a range of different factors, there's no standardised treatment. Your health care professional will decide the best treatment for you, which may include some of the treatments above.

Lung transplant
If your pulmonary hypertension doesn't respond to treatment, a lung transplant might be an option. This is a very rare procedure and not everyone is suitable for a lung transplant. Factors such as age and other medical problems would be considered, as well as the suitability of donors.

Clinical trials
Researchers are constantly working on new drugs to help treat pulmonary hypertension. Clinical trials look at how effective these new drugs are, and are an important part of the research. Clinical trials are nearly always available at specialist centres and you may be asked if you want to take part.

Further Information
The Pulmonary Hypertension Association (UK) provides support and information for all people affected by pulmonary hypertension. Go to **www.phassociation.uk.com** or call **01709 761450**.

You can also call our helpline for support and advice on living with a lung condition. Call **03000 030 555**.

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We value feedback on our information. To let us know your views, and for the most up to date version of this information and references, call the helpline or visit **www.blf.org.uk**