

# Pulmonary hypertension

## What is pulmonary hypertension?

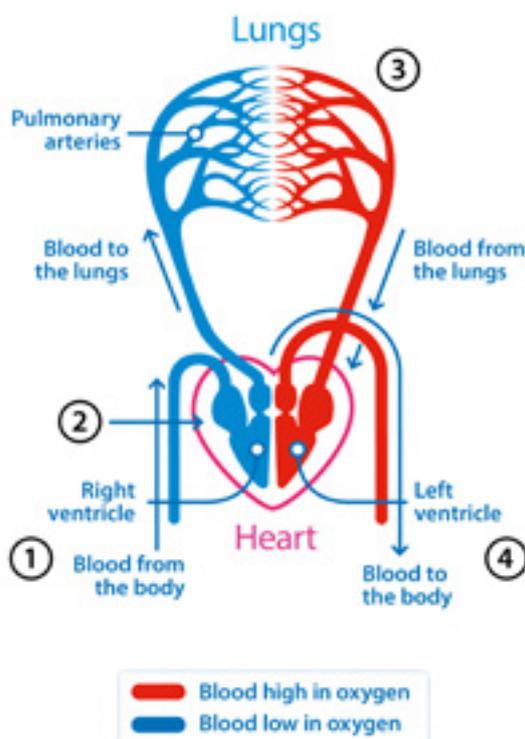
Pulmonary hypertension is high blood pressure in the blood vessels that carry blood to your lungs. They are called the pulmonary arteries.

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

This is not the same blood pressure measured by a cuff on your arm. To measure and diagnose pulmonary hypertension, health care professionals use a key test called right heart catheterisation.

## How do the heart and lungs work together?

### Pulmonary circulation



1. Blood from the body comes back to your heart in large veins. This blood has a low oxygen level, because some of the oxygen has been used by your muscles and other organs.
2. The veins take the blood into the right side of your heart. The right side of the heart is a low pressure system and it pumps blood to the lungs. The main pump on the right side is called the right ventricle.
3. In the lungs, the blood picks up oxygen (and the waste gas carbon dioxide is removed). Blood coming back from the lungs goes to the left side of the heart.
4. The left side of the heart is a higher pressure system and pumps blood high in oxygen to the rest of your body.

## What happens if you have pulmonary hypertension?

Pulmonary hypertension means the right side of your heart has to work harder to push blood through the lungs. This is because the blood vessels in the lung (pulmonary arteries) are blocked, narrowed or destroyed. The result is higher pressure in the right ventricle that pumps blood round your lungs.

Over time, the right ventricle begins to struggle with this extra work. It can become weaker and pump less effectively. This causes symptoms of pulmonary hypertension.

## Who gets pulmonary hypertension?

It can affect people of any age.

But one type of pulmonary hypertension, pulmonary arterial hypertension, is more common in women. Some research suggests that this is due to differences between male and female hormones.

Pulmonary hypertension can happen by itself, be caused by another heart or lung condition or be associated with other medical conditions such as connective tissue conditions.

## What are the symptoms of pulmonary hypertension?

The main symptom of pulmonary hypertension is **breathlessness**. Other symptoms are:

- feeling tired
- feeling dizzy or faint, especially when you exercise
- a racing heartbeat
- leg, ankle and tummy swelling
- chest pain, especially after exercise

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

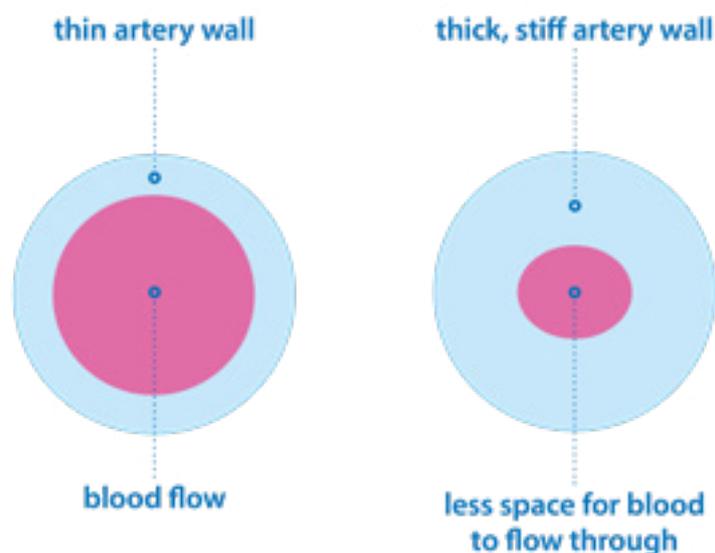
## What causes pulmonary hypertension?

There are five main groups of pulmonary hypertension, as it can be caused by different things.

### Group 1: Pulmonary arterial hypertension (PAH)

This group 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, in turn, increases the blood pressure.

It can be inherited. It's also associated with connective tissue diseases, liver disease, congenital heart defects, HIV and certain drugs. When no cause is found, it is called 'idiopathic'.



**pulmonary artery in a healthy person (left) and in someone with pulmonary arterial hypertension**

## **Group 2: Pulmonary hypertension caused by left heart disease**

The left side of your heart is the high-pressure system that pumps blood to the rest of your body. If there are problems with how the left heart works, blood can't flow easily through your lungs into the left hand side of your heart.

As a result, 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.

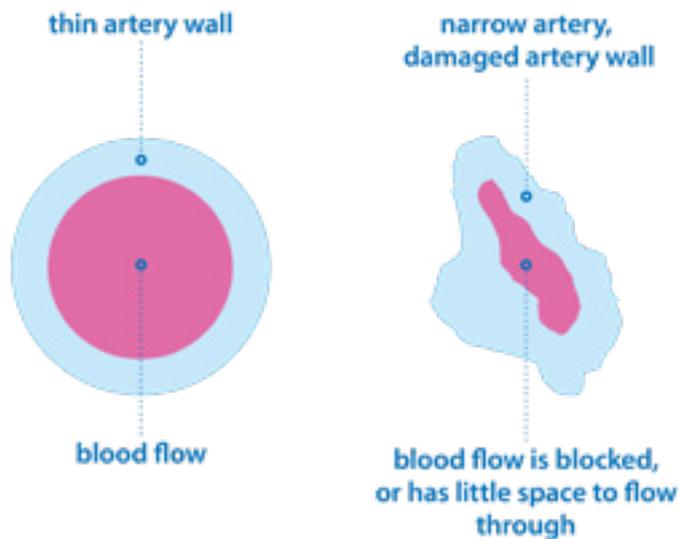
## **Group 3: Pulmonary hypertension caused by lung conditions or lack of oxygen**

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 lung blood pressure. Lung diseases can also damage blood vessels directly.

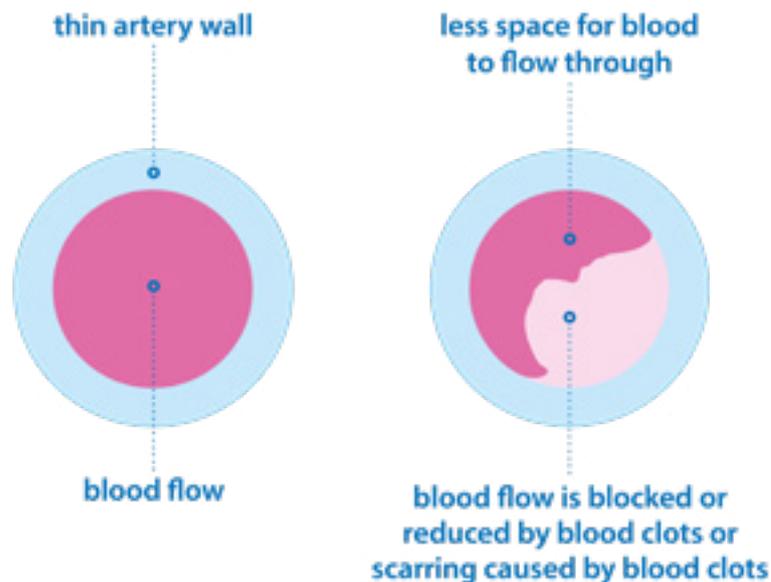
This is another common cause of pulmonary hypertension.



pulmonary artery in a healthy person (left) and in someone with a lung condition

### Group 4: Pulmonary hypertension caused by blood clots (chronic thromboembolic pulmonary hypertension, CTEPH)

This is a rare condition. It occurs when blood clots (pulmonary embolism) block the flow of blood in arteries and blood pressure in the lungs increases. Read more about pulmonary embolism at [blf.org.uk/support-for-you/pulmonary-embolism](http://blf.org.uk/support-for-you/pulmonary-embolism). Normally, blood clots dissolve once you get anticoagulant drugs. But rarely blood clots fail to dissolve and form scars instead, blocking the flow of blood.



pulmonary artery in a healthy person (left) and in someone with blood clots

## Group 5: Pulmonary hypertension caused by 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. Most people will have a range of tests to get an accurate diagnosis.

Your GP will refer you to a hospital heart or lung clinic for tests, and if health care professionals there think you may have pulmonary hypertension, you will be referred to a specialist centre for pulmonary hypertension. Find out about these specialist centres at [pulmonaryhypertensioncentres.co.uk/centres/index.html](https://pulmonaryhypertensioncentres.co.uk/centres/index.html)

Tests for pulmonary hypertension include:

**Echocardiogram:** An echocardiogram, often called an **echo**, uses ultrasound to show how your heart is working. An echo can give a rough estimate of the pressure in your pulmonary arteries and how well the chambers of the heart are working. It can also tell you if you have been born with problems (called 'congenital') with your heart that may cause pulmonary hypertension.

### Right heart catheterisation

This is the definitive test. It gives accurate information about blood pressure in the right side of the heart and the pulmonary arteries. It's used to confirm a diagnosis of pulmonary hypertension. A long, thin, flexible tube is inserted into a vein in your groin or neck, and fed through to your pulmonary artery. You might need to stay in hospital for a few days for this procedure and other tests. Find out more about the procedure at [phauk.org/what-is-ph/tests-you-might-have/cardiac-catheterisation/](https://phauk.org/what-is-ph/tests-you-might-have/cardiac-catheterisation/).

Other tests you might have are:

- an electrocardiogram or ECG. This records the electrical activity of your heart. It provides evidence of a possible problem with the heart's rhythm
- blood tests
- lung function and breathing tests. Find out more at [blf.org.uk/support-for-you/breathing-tests/tests-to-measure-breathing](https://blf.org.uk/support-for-you/breathing-tests/tests-to-measure-breathing)
- a sleep study to measure oxygen levels, heart rate and breathing patterns while you're asleep. Read more at [blf.org.uk/support-for-you/breathing-tests/tests-to-measure-your-breathing-during-sleep](https://blf.org.uk/support-for-you/breathing-tests/tests-to-measure-your-breathing-during-sleep)
- other types of scan. Read about them at [blf.org.uk/support-for-you/breathing-tests/other-tests#imaging](https://blf.org.uk/support-for-you/breathing-tests/other-tests#imaging). They may include:
  - a CT scan of your lungs to look for lung disease and blood clots
  - a V/Q scan, which is a type of scan that looks specifically for blood clots
  - an ultrasound scan to look at your liver
  - an MRI scan to look at the function of your heart
- exercise or walking tests. Find out more at [blf.org.uk/support-for-you/breathing-tests/tests-to-measure-your-exercise-capacity](https://blf.org.uk/support-for-you/breathing-tests/tests-to-measure-your-exercise-capacity)

## What's the treatment for pulmonary hypertension?

It's important to treat pulmonary hypertension to stop it getting worse. You will be treated by 1 of 8 specialist centres in the UK and your treatment will depend on what group it is, what's causing it and how severe it is. The Pulmonary Hypertension Association (UK) has more detail about individual treatments at [phauk.org/treatment-for-pulmonary-hypertension/](http://phauk.org/treatment-for-pulmonary-hypertension/).

### Group 1: Pulmonary arterial hypertension (PAH)

Your specialist will offer you treatments called **pulmonary vasodilators**. They 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 include medications called:

- phosphodiesterase 5 (PDE5) inhibitors ([phauk.org/treatment-for-pulmonary-hypertension/phosphodiesterase-5-inhibitors/](http://phauk.org/treatment-for-pulmonary-hypertension/phosphodiesterase-5-inhibitors/))
- endothelin receptor antagonists ([phauk.org/treatment-for-pulmonary-hypertension/endothelin-receptor-antagonists/](http://phauk.org/treatment-for-pulmonary-hypertension/endothelin-receptor-antagonists/))
- soluble guanylate cyclase stimulators (SGCSs) ([phauk.org/treatment-for-pulmonary-hypertension/soluble-guanylate-cyclase-stimulators-sgcss/](http://phauk.org/treatment-for-pulmonary-hypertension/soluble-guanylate-cyclase-stimulators-sgcss/))
- prostanoids ([phauk.org/treatment-for-pulmonary-hypertension/intravenous-prostanoids/](http://phauk.org/treatment-for-pulmonary-hypertension/intravenous-prostanoids/))

The choice of medication is complex and your specialist will discuss this with you.

Other treatments will help with your symptoms, called **background (or supportive) therapy**.

They may include:

- **diuretics or water tablets:** These remove excess fluid from your body. They treat symptoms like swollen ankles.
- **oxygen therapy:** This will increase the level of oxygen in your blood and may make you feel less breathless. Read more at [blf.org.uk/oxygen](http://blf.org.uk/oxygen)
- **anticoagulation:** With some forms of PAH you have a greater risk of getting blood clots. Anticoagulant medicines like warfarin help prevent blood clots forming.
- **pulmonary rehabilitation or PR:** PR is a programme of exercise and advice to help cope with breathlessness. Find out more at [blf.org.uk/pr](http://blf.org.uk/pr)

Your specialist will advise you:

- not to become pregnant
- not to travel to high altitude (above 1500-2000m). You may also need oxygen when flying – ask your health care professional about a fitness-to-fly test. Find out more at [blf.org.uk/support-for-you/breathing-tests/other-tests#hypoxic](http://blf.org.uk/support-for-you/breathing-tests/other-tests#hypoxic)
- to keep up to date with flu and pneumonia vaccinations

### Groups 2 and 3: Pulmonary hypertension caused by left heart disease or lung conditions

In both groups, pulmonary hypertension is a **secondary condition**. This means it's caused by another lung or heart condition - the **primary condition**. Examples of primary lung conditions are COPD, interstitial lung disease such as pulmonary fibrosis, and OSA.

If your pulmonary hypertension is a secondary condition, your health care professional will focus on treating the primary condition. If the primary condition improves with treatment, it should improve your pulmonary hypertension too. It's very unlikely that you'll be treated with pulmonary vasodilators if pulmonary hypertension is not your primary condition.

#### Group 4: Pulmonary hypertension caused by blood clots (chronic thromboembolic pulmonary hypertension, CTEPH)

If your pulmonary hypertension is caused by blood clots, your specialist will treat you with anticoagulant medicine. This medicine stops more blood clots forming. You might be offered:

- warfarin, which is taken as a tablet. Read more about warfarin at [phauk.org/treatment-for-pulmonary-hypertension/warfarin/](http://phauk.org/treatment-for-pulmonary-hypertension/warfarin/)
- a new group of medicines called NOACs (novel oral anticoagulants) or DOACs (direct oral anticoagulants). Read more about these medicines at [phauk.org/treatment-for-pulmonary-hypertension/noacs-or-doacs/](http://phauk.org/treatment-for-pulmonary-hypertension/noacs-or-doacs/)

If your blood clot has caused scar tissue in your pulmonary arteries, you may be offered a **pulmonary endarterectomy (PEA)**. Find out more at [phauk.org/treatment-for-pulmonary-hypertension/pulmonary-endarterectomy/](http://phauk.org/treatment-for-pulmonary-hypertension/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.

If a pulmonary endarterectomy isn't suitable, you may be offered a new procedure called **balloon pulmonary angioplasty**. Find out more about this procedure at [phauk.org/new-cteph-procedure-now-available-nhs/](http://phauk.org/new-cteph-procedure-now-available-nhs/). In this procedure, a tiny balloon is guided into a narrowed lung artery and inflated for a few seconds to widen the artery. This can be done several times, and in different parts of the lung during one procedure. The aim is to improve blood flow through the lungs and reduce pressure on the heart. Early use of this technique has showed it can lower blood pressure in the lung arteries, improve breathing and allow you to exercise more.

Both are specialist procedures currently performed in the UK only at Papworth Hospital.

If you're not suitable for surgery or you still have some pulmonary hypertension after the operation, you may be offered a drug that is a **soluble guanylate cyclase stimulator (SGCS)**, which has recently been found to be effective in slowing the progression of CTEPH. You may also be given another **pulmonary vasodilator**.

#### Group 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 specialist will discuss the best treatment for you.

##### Transplant surgery

If your pulmonary hypertension doesn't respond to treatment, a double lung or a heart-lung transplant might be an option.

## 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. Pulmonary hypertension is a relatively rare disease, so it can be difficult to find enough people for a trial. Clinical trials are usually available at specialist centres, so you may be asked if you'd be willing to take part.

## Further information

The Pulmonary Hypertension Association (UK) provides support and detailed information for people affected by pulmonary hypertension. Find out more about them at [phauk.org/](http://phauk.org/).

Get in touch with us to find support near you.

Helpline: 03000 030 555

Monday to Friday, 9am-5pm

Ringling our helpline never costs more than a local call and is usually free, even from a mobile.

[helpline@blf.org.uk](mailto:helpline@blf.org.uk)

[blf.org.uk](http://blf.org.uk)

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