

# What is pulmonary hypertension?

This leaflet offers more information about pulmonary hypertension, including what it is, how it is diagnosed and how it can be treated. If you have any further questions or concerns, please speak to the staff member in charge of your care.

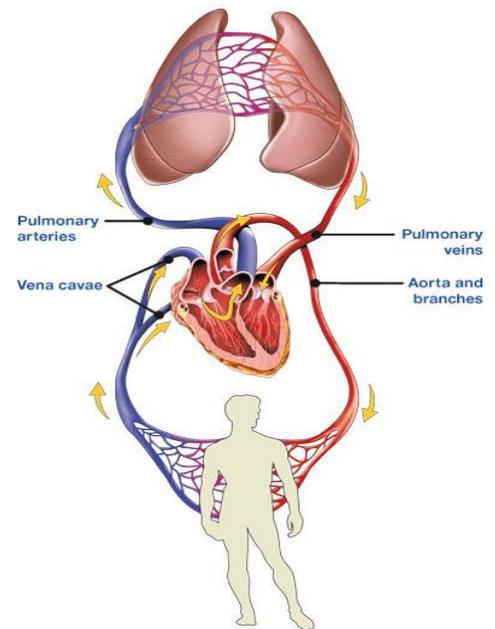
## What is pulmonary hypertension?

Pulmonary hypertension is raised blood pressure within the **pulmonary arteries**, which are the blood vessels that supply the lungs. It is important to know that pulmonary hypertension is very different to the more common type of "high blood pressure" that is measured using your arm. Rather, pulmonary hypertension is a serious medical condition that can damage the right side of the heart, making the heart less efficient at pumping blood around the body and getting oxygen to the muscles.

To understand pulmonary hypertension, it is useful to have an understanding of the normal flow of blood through the heart and lungs:

The **right ventricle** is one of the big pumping chambers of the heart. It pumps blood that has been circulated around the body through the pulmonary arteries and into the lungs. In the lungs, the blood picks up oxygen and flows back to the left side of the heart, where it is pumped around the body, beginning the cycle again.

**Pulmonary hypertension** is where the pressure in the pulmonary arteries becomes higher than normal, causing the walls of the blood vessels to become thicker and less flexible. Over a period of time the right ventricle also becomes thick and stiff and has to work harder to pump blood through the lungs and the heart. This means the heart eventually works less efficiently.



## What are the symptoms of pulmonary hypertension?

The most common symptoms are:

- shortness of breath
- excessive tiredness/fatigue
- chest pains
- dizziness
- fainting or near fainting episodes
- palpitations or a racing, thumping or fluttering feeling in the chest.

These symptoms can occur at rest or during periods of physical activity, and can have an impact on a person's day to day life.

## What causes pulmonary hypertension?

There are many different types of pulmonary hypertension. Often it is linked to diseases such as:

- congenital heart disease (heart disease that you are born with)
- connective tissue disease (for example, systemic lupus erythematosus)
- thromboembolic disease (blood clots in the lungs)
- human immune-deficiency virus (HIV)
- liver disease (for example, cirrhosis).

Sometimes the cause of PH is unknown – this is known as **idiopathic pulmonary arterial hypertension** (IPAH). In very rare cases, PH can be inherited.

## How is pulmonary hypertension diagnosed?

Patients are referred to one of the seven designated specialist centres in the United Kingdom. St George's works in collaboration with The Royal Brompton Hospital. Information is shared between these centres during the investigation process.

In order to help diagnose your problem, you may be asked to undergo one or a number of the following tests:

- Blood tests
- Chest x-ray
- Echocardiogram (using ultrasound to create images of the heart)
- Computerised tomography (CT) scan
- Magnetic resonance imaging (MRI) scan
- Lung function tests
- Electrocardiogram (ECG, which tests for problems with the electrical activity of your heart)
- V/Q scan (measuring air and blood flow in your lungs)
- Sleep study
- Six-minute walk test (to assess your capacity for exercise)

A confirmed diagnosis of pulmonary hypertension is usually made by carrying out a test called a **right heart catheterisation** (RHC), which will be performed at St George's Hospital. The RHC is used to assess how well the heart is pumping and to measure the pressures in the heart and lungs. The procedure involves having a small injection of local anaesthetic to numb the skin on your neck. Occasionally the test can be performed via the groin. Once the area is numb, a small cut (incision) is made and the doctor places a catheter (a thin, flexible tube) under your skin and either into the internal jugular vein (if the incision is made to the neck) or the femoral vein (if the incision is made in the groin). This tube is then guided into your heart. When the test is over the tube is gently removed and pressure is applied to the site of insertion to stop any bleeding. The test takes about 30 minutes to complete.

## What are the treatments available?

There is no cure for most types of pulmonary hypertension, but treatment can be given to help you manage your condition and reduce symptoms. The treatment available depends on the type of pulmonary hypertension – if it has been caused by an underlying condition, then the condition must also be treated.

Depending on the cause of your pulmonary hypertension, you may have conventional therapy such as oxygen, warfarin (blood thinning medication) and/or diuretics (water tablets).

Alternatively, you may start on more specific therapies, such as:

- sildenafil
- tadalafil
- riociguat
- bosentan
- ambrisentan
- macitentan
- inhaled prostacyclin therapy (iloprost)
- intravenous prostacyclin therapy (epoprostenol).

These disease-specific treatments help to relax the arteries in the lungs, allowing blood to flow through these vessels more easily, and reducing the blood pressure in your lungs. These treatments will be discussed with you in more detail if treatment is required.

If your pulmonary hypertension is caused by thromboembolic disease (blood clots in the lungs) you may be offered surgery to remove the clots. This is called a pulmonary endarterectomy (PEA) and will be discussed further if necessary.

In some cases of pulmonary hypertension, disease-specific therapies may not be suitable. If this is the case the reasons for this will be discussed with you.

## What can I expect during my consultation and hospital visits?

Your visits to the pulmonary hypertension clinic are very important. Not only do they ensure that your symptoms are controlled adequately and that you are getting the best possible care, they are also a chance for you to ask questions and/or voice concerns.

The team is made up of a range of specialist nurses and doctors, who are happy to answer your questions and address any worries you may have.

## Contact us

For advice or information regarding pulmonary hypertension, please contact the **pulmonary hypertension nurse specialist** on **020 8725 1094**.

Further information is available from **The Pulmonary Hypertension Association (PHA) UK**, which publishes a number of free information booklets and DVDs for people with pulmonary hypertension, their carers, family and friends. To learn more visit their website at **[www.phassociation.co.uk](http://www.phassociation.co.uk)**.

## Additional services

### Patient Advice and Liaison Service (PALS)

PALS can offer you on-the-spot advice and information when you have comments or concerns about our services or the care you have received. You can visit the PALS office between 9.30am and pm, Monday to Friday in the main corridor between Grosvenor and Lanesborough Wing (near the lift foyer).

**Tel:** 020 8725 2453 **Email:** [pals@stgeorges.nhs.uk](mailto:pals@stgeorges.nhs.uk)

### NHS Choices

NHS Choices provides online information and guidance on all aspects of health and healthcare, to help you make decisions about your health.

**Web:** [www.nhs.uk](http://www.nhs.uk)

### NHS 111

You can call 111 when you need medical help fast but it's not a 999 emergency. NHS 111 is available 24 hours a day, 365 days a year. Calls are free from landlines and mobile phones.

**Tel:** 111

