



A lifetime of specialist care

Royal Brompton & Harefield 
NHS Foundation Trust

Royal Brompton Hospital

What is pulmonary hypertension?





Contents

• What is pulmonary hypertension?	3
• What are the symptoms of PH?	4
• What causes PH?	5
• How is PH diagnosed?	5
• What are the treatments available?	6
• What to expect during your consultation and hospital visits	8
• Our team	9
• Shared care	11
• Useful contacts	12
• More information	13

This leaflet provides you with information about pulmonary hypertension. It does not replace the need for personal advice from a qualified health professional. Please ask us if you have any questions.

You have been referred to Royal Brompton Hospital's pulmonary hypertension service as your doctor thinks there is a possibility that you may have pulmonary hypertension. You need to have some tests in order to confirm if this is the case.

This leaflet aims to provide you with some information about this condition and about what to expect during your time in hospital.

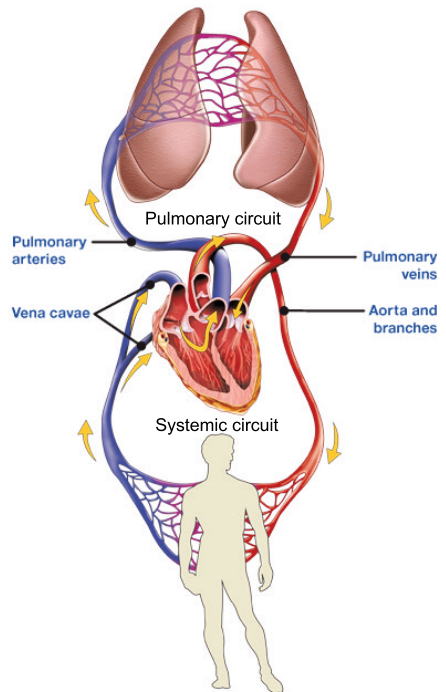
What is pulmonary hypertension?


Pulmonary hypertension (PH) is a rare, serious heart and lung condition where the pressure in the pulmonary arteries is higher than normal. The pulmonary arteries are the blood vessels that carry blood from the heart to the lungs.

It may be referred to as "high blood pressure in the lungs". It is important to know that PH is very different to the more common type of "high blood pressure" that is measured using your arm.

To understand PH 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.





If the pressure in the pulmonary arteries becomes higher than normal, the walls of the blood vessels may 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 effectively. This can take several years.

What are the symptoms of PH?

The most common symptoms are:

- Shortness of breath
- Excessive tiredness / fatigue
- Chest pains
- Palpitations (a racing, thumping, or fluttering feeling in your chest)
- Dizziness
- Fainting or near fainting episodes (known as syncope)

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

What causes PH?

There are many different types of PH. Sometimes the cause of PH is unknown, and this is called idiopathic pulmonary arterial hypertension (IPAH).

More commonly, it is linked to other diseases. Some examples are:

- Heart diseases that you are born with (congenital heart disease)
- Connective tissue disease such as systemic sclerosis
- Thromboembolic disease (blood clots in the lungs)
- Human immuno-deficiency virus (HIV)


In very rare cases it can be inherited.

How is PH diagnosed?

Patients should be referred to one of seven designated specialist centres in the United Kingdom. We are one of them. Information is shared between these centres and well thought out protocols (guidelines) have been developed to investigate patients for PH.

Tests include:

- Blood tests
- Cardiopulmonary exercise – a test to see how your lungs, heart and muscles react together when you exercise on treadmill or an exercise bicycle
- Chest X-ray
- Echocardiogram – an ultrasound scan of your heart. This scan shows the structure of the valves and chambers in the heart and shows how well your heart is working

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- CT scan (computerised tomography) – a type of X-ray that gives detailed pictures of your lungs
 - MRI (magnetic resonance imaging) – a scan using magnets and radio waves to create images of the structure of your heart and find out how well it is working
 - Lung function – tests that measure how well your lungs are working
 - ECG (electrocardiogram) – a tracing of the electrical activity of your heart
 - V/Q scan – a scan that measures ventilation (breathing) and perfusion (blood flow) in your lungs
 - Sleep study – a test that measures your oxygen levels and breathing overnight.

A confirmed diagnosis of PH is usually made by carrying out a test called a right heart catheterisation, also known as cardiac catheterisation.

This test involves having a small injection to numb the skin in your groin, arm or neck. The doctor makes a small cut and places the catheter (a thin flexible tube) under your skin and into a blood vessel that goes to your heart. While the catheter is in place the doctor can see how well blood flows through your heart and measures pressures in the heart and lungs. When the test is over the tube is gently removed and pressure applied to where the cut was made to stop any bleeding.

What are the treatments available?

There is no cure for most types of PH but treatment can be given to manage your condition and help reduce your symptoms. The treatments you have depend on your type of PH. If it is caused by another disease, then this must also be treated.

Depending on the cause of PH, you may have conventional therapy such as:

- Oxygen
- Warfarin (blood thinning medication)
- Diuretics (water tablets)


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

- Sildenafil
- Tadalafil
- Riociguat
- Bosentan
- Ambrisentan
- Macitentan
- Selexipag
- Inhaled prostacyclin (Iloprost)
- Intravenous prostacyclin (Epoprostenol)

These disease-specific treatments called “pulmonary vasodilators” help to relax the arteries in the lungs. This allows blood to flow through these blood vessels more easily and reduces the blood pressure in your lungs. These treatments will be discussed with you in more detail during your consultation.

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

If your PH is caused by thromboembolic disease (blood clots in the lungs) you may be offered surgery to remove the blood



clots. This is called a pulmonary endarterectomy (PEA) and you would be referred to Papworth Hospital in Cambridge. The staff at Papworth will meet with you and decide whether this treatment is right for you.

What to expect during your consultation and hospital visits

Royal Brompton Hospital is part of the National Pulmonary Hypertension Service and specialises in the diagnosis, care and treatment of people with pulmonary hypertension.

The team consists of experienced consultants and clinical nurse specialists who will discuss your condition with you and support you through the diagnosis and treatment options.

It may be necessary for you to be admitted to the hospital for between two and four days. During this time you will have further tests carried out to confirm if you have pulmonary hypertension. The results of your tests will be discussed with you during this admission. Depending on the results of the tests, and if a diagnosis of PH is made, you may then start treatment. After starting your treatment you will need regular reviews of your condition. This will be done in outpatient clinics every three to six months.

Our team

Pulmonary hypertension consultants

- **Dr John Wort** clinical lead for respiratory and pulmonary hypertension
- **Professor Kostas Dimopoulos** cardiology, adult congenital heart disease (ACHD) and pulmonary hypertension
- **Dr Laura Price** respiratory and pulmonary hypertension
- **Dr Aleksander Kempny** cardiology, ACHD and pulmonary hypertension
- **Dr Colm McCabe** respiratory and pulmonary hypertension
- **Dr Aimee Brame** respiratory, intensive therapy unit and pulmonary hypertension
- **Dr Bhaskar Mukherjee** respiratory and pulmonary hypertension

Associated consultants

- **Professor Michael Gatzoulis** cardiology, ACHD and pulmonary hypertension
- **Dr Anna Pires** ACHD and pulmonary hypertension
- **Dr Isma Rafiq** ACHD and high risk pregnancy
- **Dr Wei Li** echocardiography
- **Dr Simon Padley** radiology



- **Dr Thomas Semple** radiology
- **Dr Jayne Wood** palliative care
- **Dr Anna Reed** transplant
- **Dr Tessa Homfray** genetics

Clinical nurse specialists and pharmacy

- **Carl Harries** pulmonary hypertension
clinical nurse specialist (CNS)
- **Stuart Craig** pulmonary hypertension CNS
- **Deepa Lakhani** pulmonary hypertension
pharmacist
- **Deepti Ghelani** pulmonary hypertension
pharmacy technician

Administration and research teams

- **Sweeta Dhakan-Oakey** service coordinator
- **Selina Iuliano** service coordinator
- **Della Stokes** data manager
- **Margaret Hollis** medical secretary
- **Geraldine Faulkner** inpatient and day case
scheduler
- **Rosa Da Costa** research coordinator

Shared care

We have developed a shared care service with a number of NHS hospital trusts (see list of trusts and health professionals below). Consultants from Royal Brompton regularly attend clinics at these hospitals.

This shared care system makes it easier for patients to get a diagnosis and treatment. Offering services more locally also means that patients do not have to travel as far for appointments.

Teams from Royal Brompton and the other shared care hospital trusts also meet regularly to discuss the care and management of patients.

Shared care trusts and health professionals

St George's University Hospital NHS Foundation Trust Tooting – London

- **Professor Madden** lead consultant
- **Nordita Bascon** pulmonary hypertension clinical nurse specialist (CNS)

University Hospital Southampton NHS Foundation Trust – Southampton

- **Dr Samantha Fitzsimons** lead consultant pulmonary hypertension and congenital heart disease
- **Melanie Finch** pulmonary hypertension CNS
- **Dr Ahilanadan Dushianthan** lead consultant (pulmonary hypertension and respiratory medicine)
- **Michelle Ritchie** pulmonary hypertension CNS



Oxford University Hospital NHS Foundation Trust (John Radcliffe) – Oxford

- **Dr Elizabeth Orchard** lead consultant ACHD
- **Emma Weingart** pulmonary hypertension CNS

Other centres

Royal Brompton's teams also visit and run clinics at these hospital trust locations:

- Surrey and Sussex Healthcare NHS Trust – Redhill, Surrey
- Royal Berkshire NHS Foundation Trust – Reading, Berkshire
- Frimley Health NHS Foundation Trust – Frimley, Surrey

Occasionally, you will also need to visit Royal Brompton for tests or treatments. We will tell you when that is necessary.

Useful contacts

For general enquiries and appointments, please contact the pulmonary hypertension secretary on **020 7352 8362**.

For enquiries regarding a hospital admission, please contact cardiac bookings on **020 7351 8617**.

Other useful contacts

- Royal Brompton & Harefield NHS Foundation Trust website
www.rbht.nhs.uk/our-services/heart/pulmonary-hypertension-service
- Pulmonary Hypertension Association (PHA UK)
www.phauk.org
- The Somerville Foundation (formerly Grown-up Congenital Heart Patient's Association)
www.thesf.org.uk

For more information

If you would like any further information before your admission, please contact the pulmonary hypertension clinical nurse specialists on **020 7352 8121**, **extension 2156** or **bleeps 1165 or 1170**.



If you have concerns about any aspect of the service you have received in hospital and feel unable to talk to those people responsible for your care, call the Patient Advice and Liaison Service (PALS) on:

- Royal Brompton Hospital – 020 7349 7715
- Harefield Hospital – 01895 826 572

You can also email pals@rbht.nhs.uk. This is a confidential service.

Royal Brompton Hospital
Sydney Street
London
SW3 6NP
Tel: 0330 12 88121

Harefield Hospital
Hill End Road
Harefield
Middlesex
UB9 6JH
Tel: 0330 12 88121

Website: www.rbht.nhs.uk

إذا كنت ترغب في الحصول على ترجمة فورية لمضمون هذه الوثيقة إلى اللغة العربية، يرجى منك الاتصال بأحد مستخدمينا بجناح المصلحة التي يتم فيها استشفائك. أحد موظفينا سيسعى لترتيب إجراءات الترجمة وإتمامها في الوقت المناسب لك.

Brosurteki bilginin Türkçe tercumesi için tedavi görüyor olduğunuz bölüme bas vurunuz. Bölüm personeli tercümenin gerçekleşmesini en kısa zamanda ayarlayacaktır.

