

Royal Brompton Hospital

What is pulmonary hypertension?

Information for patients



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This leaflet provides you with information about pulmonary hypertension. It does not replace the need for personal advice from a healthcare 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 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. 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.

Over a period of time the

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)

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)

- 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.

- 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.
- 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.
- 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 in order 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 be treated also.

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
- Bosentan
- Ambrisentan
- Inhaled prostacyclin therapy (lloprost)
- Intravenous prostacyclin therapy (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, disease-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 would find out more about 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 about 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, it may be necessary for you to come back to the hospital for a further few days so you can 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

•	Dr John Wort	Consultant in pulmonary hypertension and respiratory medicine
•	Professor Michael Gatzoulis	Professor of cardiology
•	Dr Konstantinos Dimopoulos	Consultant cardiologist
•	Dr Rafa Alonso	Consultant cardiologist
•	Dr Laura Price	Locum consultant in pulmonary hypertension and respiratory medicine
•	Dr Lorna Swan	Consultant cardiologist
•	Dr Wei Lei	Consultant cardiologist
•	Carl Harries	Clinical nurse specialist
•	Lisa Parfitt	Clinical nurse specialist
•	Sweeta Dhakan	Pulmonary hypertension service co-ordinator
•	Sylvia Dedman	Pulmonary hypertension secretary

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/ph
- Pulmonary Hypertension Association (PHA UK) www.phassociation.uk.com
- 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.

Your notes

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 PALS on 020 7349 7715 or email pals@rbht.nhs.uk. This is a confidential service.

Royal Brompton Hospital Sydney Street London SW3 6NP tel: 020 7352 8121 textphone: (18001) 020 7352 8121

Harefield Hospital Hill End Road Harefield Middlesex UB9 6JH tel: 01895 823 737 textphone: (18001) 01895 823 737

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 Turkçe tercumesi için tedavi goruyor oldugunuz bolume bas vurunuz. Bolum personeli tercumenin gerçeklesmesini en kisa zamanda ayarlacaktir.

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August 2014