British Lung Foundation

Pulmonary hypertension

What is pulmonary hypertension?

Pulmonary hypertension (PH) is a rare blood vessel disorder of the lung. Pressure in the pulmonary artery (the blood vessel that leads from the heart to the lungs) rises above normal levels and may become life threatening.

What are the symptoms of PH?

Symptoms of pulmonary hypertension include:

- shortness of breath
- fatigue
- chest pain
- dizzy spells
- fainting.

What causes PH?

If we don't know what causes pulmonary hypertension, it is called **idiopathic pulmonary arterial hypertension** (PAH). There is a family history in about ten per cent of cases. PAH is extremely rare and occurs in about two in every million people per year.

Associated forms of pulmonary hypertension (PH) means the cause is known. Common causes are:

- emphysema
- bronchitis.

Other less common causes are:

- inflammatory or collagen vascular diseases such as scleroderma, CREST syndrome or systemic lupus erythematosus (SLE)
- congenital heart diseases that increase the movement of blood through the lungs like ventricular and atrial septal defects
- chronic pulmonary thromboembolism (old blood clots in the pulmonary artery)
- HIV infection
- liver disease
- diet drugs like fenfluramine and dexfenfluramine

How is PH diagnosed?

Pulmonary hypertension is frequently misdiagnosed and is often severe by the time it is accurately diagnosed.

The best way to screen for the presence of PH is by echocardiography. An echocardiograph uses ultrasound to build a picture of your heart. This picture can be used to estimate the blood pressure in your pulmonary artery and how well your heart is pumping blood.

The diagnosis must then be confirmed by measuring the blood pressure in the right side of your heart and your pulmonary artery**.** This is done by **right-heart cardiac catheterisation.** A doctor will insert a thin, flexible tube, or catheter, into your arm, leg, or neck vein. The catheter is then threaded into the right ventricle and pulmonary artery.

During catheterisation, your doctor can also check the right side of your heart's pumping ability. This is done by measuring the amount of blood pumped out of the right side of your heart with each heartbeat.

The diagnosis of idiopathic PAH is made by excluding all known causes of associated PH.

Pulmonary hypertension has been historically chronic and incurable with a poor survival rate. However, treatments are now more effective. Recent data indicate that the length of survival is continuing to improve, with some patients able to manage the disorder for 15 to 20 years or longer.

What treatments are available?

Current treatments include:

Calcium channel blocking drugs (CCBs)

Calcium causes muscle cells to contract. CCBs block the movement of calcium into the cells of your heart and blood vessels. This helps your blood vessels to relax and increases the supply of blood and oxygen to your heart while reducing its workload. Less than 10 per cent of people with PH respond to CCBs.

CCBs may be dangerous in some people so special testing needs to be carried out at the time of right heart catheterisation to assess suitability.

CCBs are also prescribed for systemic hypertension, but in much lower dosages than when they are prescribed for PH. CCBs can cause fluid retention.

Prostacyclin receptor antagonists

Prostacyclin is a steroid that is produced naturally in the body of a healthy person. It causes blood vessels in the lungs to relax and allows blood to flow through them more easily. People with pulmonary hypertension do not produce enough prostacyclin, so the blood vessels in the lungs are constricted. Prostacyclin, also known as epoprostenol, is used to treat this condition.

Prostacyclin therapy was initially used as a bridge to lung transplantation although it has also emerged as an alternative to transplantation in some patients. Prostacyclin, or its longer lasting derivatives, can be given by continuous infusion and sometimes by other routes. It has been shown to improve breathlessness and probably survival in patients with idiopathic PAH.

Endothelin receptor antagonists

These are a class of oral drugs that are now licensed for the treatment of pulmonary arterial hypertension. Endothelin is a peptide made by the body in the

endothelium (a layer of cells which line the heart and blood vessels). It constricts blood vessels and increases blood pressure. It plays an important role in blood flow.

In PH, the body produces too much endothelin. This leads to the constriction of blood vessels and also affects blood pressure in the lungs.

Endothelin must connect with an endothelin receptor in order to be activated. Endothelin receptor antagonists block endothelin receptors. This limits the harmful excess endothelin in your blood vessels.

Controlled trials have demonstrated that endothelin receptor antagonists improve symptoms and quality of life in PH. They may also have a beneficial effect on survival.

Phosphodiesterase inhibitors
A further class of drugs that now available are those that act by inhibiting the activity of the type 5 phosphodiesterase. An example of this class of drugs is sildenafil, also known as Viagra. These drugs enhance the production of an important molecule in the walls of blood vessels that dilates blood vessels, known as cyclic GMP.

Trials have now shown that these drugs improve symptoms of breathlessness in patients with PAH.

Lung transplantation
Quality of life can be moderately to substantially improved by lung transplantation, and life may be extended beyond your life expectancy prior to transplantation. It is impossible to predict how long you may survive after transplantation. The most critical period is the year after transplantation; this is the period when surgical complications, rejection, and infection are the greatest threat to survival.

Patients who survive the first year are more likely to survive three years or longer after transplantation. There are people alive today who had lung transplantation five or even more years ago.

Life expectancy after lung transplantation is shorter than for heart, liver, or kidney transplantation, particularly for PAH patients. Rejection and infection are the two major complications of lung transplantation. Immunosuppressive (anti-rejection) medications prescribed by your doctors will help keep the rejection process "turned off." Other medications may be necessary to control and treat rejection if your immune system breaks through the immunosuppressive blockade. Following your doctor’s orders and taking all medications as prescribed help to prevent or control rejection.

Because you will be taking immunosuppressive medications, your immune system will be less able to fight off invading bacteria and viruses. You will be much more susceptible to infections, which are more likely to become severe.

Other therapies
Therapies that may be used alongside treatments include:

Diuretics - used to control an excessive amount of watery fluid in cells, tissues or serous cavities (such as the abdomen). Pulmonary hypertension can lead to right heart failure and an excess of fluid in the lower and upper extremities and abdomen. An excess of fluid can also be caused by high-dose calcium channel blockers. Diuretics will cause frequent urination.

Digitalis medicines (Digoxin, Lanoxin) - used to improve the strength and efficiency of the heart or to control the rate and rhythm of the heartbeat. This leads to better blood circulation and reduced swelling of hands and ankles in patients with right heart problems. Its value for patients with PH has not yet been fully examined.

Supplementary oxygen - sometimes prescribed for PH patients if they have a lack of blood oxygen (hypoxaemia) at rest or with physical activity. It can also treat people with PH under special conditions, such as when hospitalized with a respiratory infection, or at high altitudes or sometimes when travelling by air.

Oral anticoagulant therapy - blood clots are potential complications of PH. Oral anticoagulant therapy (Coumadin, Warfarin) is widely recommended for people with PH because it probably prolongs survival.

Further Information and Contacts

The Pulmonary Hypertension Association (UK) aims to provide support, understanding, and information for all those people whose lives are touched by Pulmonary Hypertension.

Tel: 0800 389 8156; Fax: 0701 976 0265
Email: office@phassociation.uk.com
Website: www.phassociation.uk.com

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© British Lung Foundation, 73-75 Goswell Road, London EC1V 7ER
Helpline 08458 50 50 20 www.lunguk.org enquiries@blf-uk.org
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