

WHAT IS PULMONARY HYPERTENSION?

Pulmonary Hypertension or PH, is a complex health problem of abnormally high blood pressure in the arteries of the lungs. If the pressure in the pulmonary artery (the blood vessel that leads from the heart to the lungs) rises above normal levels, it may become life threatening.



It is a disease that affects people of all ages and ethnic backgrounds.

PH starts when the small vessels that supply blood to the lungs constrict, or tighten up, making it more difficult for blood to get through to the lungs resulting in the heart having to pump harder to overcome the resistance. As time passes, scarring (or fibrosis) make the vessels stiffer and thicker, and some may even become completely blocked. The extra stress causes the heart to enlarge and become less flexible. Less and less blood is able to flow out of the heart through the lungs and into the body, and more and more symptoms start to show.

DIFFERENT TYPES OF PH

In 1998, the World Health Organization (WHO) sponsored a meeting of PH specialists with the goal of creating a standardised system for defining different types of PH. The committee proposed that the disease be divided into the following five categories of PH:

1. Pulmonary arterial hypertension (PAH)

The pulmonary artery is the large vessel that carries blood from the heart into the lungs to be oxygenated. Most cases of PH affect this artery and the hundreds of tiny blood vessels that branch off from it.

This category includes two types of PH:

Primary pulmonary hypertension (PPH) can occur at random, without any apparent cause. Other times, the disease can be inherited; it is estimated that at least 10 percent of PPH cases occur in families where at least one other person

PAH, associated with other causes appears to be similar to PPH, but is actually associated with exposure to toxins or the effects of other diseases. These can include:

- Collagen vascular disease (scleroderma, lupus, rheumatoid arthritis). This is a type of disease that affects collagen within the body. Collagen helps give the structures within the body their shape allowing them to function properly.
- Congenital (In-borne) heart disease.
- Chronic liver disease.
- Human immunodeficiency virus (HIV).
- Use of diet drugs such as fenfluramine or dexfenfluramine.
- Use of cocaine, methamphetamine, or other street drugs.

2. Pulmonary hypertension associated with disorders of the respiratory system

- Emphysema.
- Interstitial lung disease.
- Sleep apnea.
- Chronic exposure to high altitude.

3. Pulmonary hypertension due to chronic thrombotic or embolic disease

- Blood clots in pulmonary arteries.
- Pulmonary embolism (caused by clot, tumour or foreign matter in the lungs).
- Sickle cell disease.

4. Pulmonary hypertension due to disorders directly affecting the pulmonary blood vessels

- Inflammatory diseases such as schistosomiasis or sarcoidosis, an unusual disorder characterised by clusters of inflamed (swollen) cells forming within the lungs.
- Pulmonary capillary disease.

5. Pulmonary venous hypertension (PVH)

PVH is caused by diseases of the left side of the heart, such as mitral valve disease or a poorly performing left ventricle. This can cause increased pulmonary artery pressures, but rarely leads to severe PAH.

HOW TO CONFIRM A DIAGNOSIS?

PH is often not diagnosed in a timely manner because its early symptoms can be confused with those of many other conditions.

Some of the symptoms of PH include:

- Breathlessness or shortness of breath on exertion.
- Feeling tired all the time.
- Dizziness, especially when climbing stairs or upon standing up.
- Fainting - often the symptom that brings people to doctors.
- Swollen ankles and legs.
- Chest pain, especially during physical activity.

To establish a diagnosis of PH, a series of tests are performed that show how well a person's heart and lungs are working. These tests may include:

- Echocardiogram, sound waves to map the structure of the heart.
- Electrocardiogram (ECG), a record of the electrical activity produced by the heart.
- Right heart catheterisation, a precise measure of blood pressure in the right side of the heart and in the pulmonary artery.
- Six-minute walk test.
- Cardiopulmonary Exercise Test.
- Assessment of function in tasks of daily living.

If these tests show that a person definitely has PH, other types of diagnostic tests may be ordered to aid in identifying the cause. These include:

- A computed tomography (CT or CAT) scan to rule out pulmonary embolism or lung disease.
- A pulmonary function test to rule out obstructive lung disease.
- A formal sleep study to rule out sleep apnea.
- Laboratory tests to rule out hepatitis, collagen disease, HIV, or other conditions.

When no cause can be found, the diagnosis is primary pulmonary hypertension (PPH).

TREATMENT FOR PULMONARY HYPERTENSION

PH of almost any type is a chronic condition that can become life-threatening. Although there is no cure, there are several treatment options available:

- Anticoagulants to prevent blood clots in the lungs.

- Calcium channel blockers to relieve constriction in pulmonary arteries.
- Digoxin to help the heart pump blood more effectively.
- Diuretics to reduce fluid in the system.
- Inhaled oxygen to make more oxygen available to the blood.
- Bosentan is an oral endothelin receptor antagonist, or ERA. It affects endothelin, a natural chemical in the body that plays an important role in the development of PAH. Bosentan works by reducing the high blood pressure in the lungs, thereby enabling the heart to pump blood more effectively.
- Prostacyclin analogue such as Epoprostenol or Treprostinil or Iloprost to help open up constricted lung blood vessels, thereby reducing the high blood pressure in the lungs.
- Phosphodiesterase-V inhibitors (such as Sildenafil) to increase nitric oxide levels in the blood and increase pulmonary blood flow.

For patients who do not respond to any of these treatments, a lung transplant may be recommended.

LIVING OPTIMALLY WITH PH

A diagnosis of PH does not necessarily mean you cannot have an active, fulfilling life provided appropriate measures and precautions are taken. PH is a lifelong illness that can be made worse by a variety of factors, such as smoking or going to high altitude.

If you have PH, you should be as active as physically possible. However, as physical activity can be associated with marked increases in pulmonary artery pressure, isometric exercises and activities that produce dangerous symptoms, such as chest pain or dizziness should be avoided.

Care should be taken both in taking medications and in using over-the-counter drugs as this may affect levels of certain drugs like warfarin. Any anaesthetics or sedatives can be very hazardous; ask your physician which medications are safe.

Pregnancy and delivery produce dramatic changes that can seriously endanger your life. Thus, avoid pregnancy by practicing a safe and effective method of contraception. Avoid oral contraceptives as these can aggravate PH. The most effective form of contraception for people with significant PH is surgical sterilisation.

Additional precautions are often taken with PH patients. These include supplemental oxygen during air travel, antibiotic therapy for significant respiratory tract infections, once-off pneumococcal pneumonia vaccine and yearly flu vaccines (since pneumonia can be very serious with PH patients). Also avoid conditions in which the ambient oxygen concentration may be decreased, such as high altitude and travel in unpressurized airplane cabins. Before starting an exercise programme, ask your physician what activities are appropriate for you.

Finally, if you've begun medical treatment for your PH, stopping any of your medicines without your physician's approval can be extremely dangerous. Medical therapy has significantly improved the outlook for most PH patients, but it doesn't "cure" PH. So do not stop medical therapies unless recommended by your physician.

People with PH and underlying congenital heart defect or acquired valvular abnormalities will need antibiotic before certain dental or surgical procedures to prevent endocarditis.

THE PROMISE OF THE FUTURE

Today there is a lot of exciting research that is rapidly advancing our understanding of the causes and mechanisms of PH. These discoveries mean that other important new treatments are on the horizon - and they offer bright hope for the future.

DISCLAIMER

This information is given as a guide only and does not replace medical advice from your doctor. You should seek the advice of your doctor before starting any treatment or if you have any questions related to your health, physical fitness or medical condition.

UNDERSTANDING PULMONARY HYPERTENSION ADULT CONGENITAL HEART DISEASE PROGRAMME



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