

Pulmonary Hypertension

What is pulmonary hypertension?

Pulmonary hypertension refers to a condition in which high blood pressure exists within the vessels of the lungs. Normally, venous (low oxygen) blood returns from the body to the right side of the heart. The blood is pumped to lungs via the pulmonary arteries. Breathing brings oxygen to venous blood in the lungs, turning it into arterial (high oxygen-containing) blood. Arterial blood returns through the pulmonary veins to the left side of the heart, where it is pumped to the rest of the body to deliver oxygen to organs and tissues.



How is pulmonary hypertension classified?

Pulmonary hypertension has been classified as primary (without obvious cause, or idiopathic) or secondary (occurring as a result of another disease). Although this is still referenced in medical texts, the revised World Health Organization (WHO) classification system does away with these definitions.

The WHO divides pulmonary hypertension into five categories. These categories include:

- WHO Group I: Pulmonary hypertension due to a disease of the arteries themselves, such as idiopathic, or hereditary, or related to autoimmune disease.
- WHO II: Pulmonary hypertension due to increased pressure on the left side of the heart.
- WHO III: Pulmonary hypertension due to chronic lung disease, low oxygen levels or sleep disorder.
- WHO IV: Pulmonary hypertension due to blood clots in the lung.

- WHO V: Pulmonary hypertension associated with diseases that don't fall into the other four categories, such as sickle cell disease.

What are associated conditions?

Pulmonary hypertension can occur in isolation or, more commonly, with diseases of the lungs and heart. Pulmonary hypertension in the absence of other diseases is very rare and generally idiopathic or familial in nature. This kind of pulmonary hypertension is referred to as pulmonary arterial hypertension (PAH).

Pulmonary arterial hypertension can also be associated with autoimmune diseases; drug use such as methamphetamines or diet drugs; human immunodeficiency virus (HIV); liver disease and congenital heart disease.

Pulmonary hypertension is commonly associated with a variety of lung conditions with low oxygen levels. These include COPD, emphysema, interstitial lung disease, chronic pulmonary blood clots or sleep apnea. When pulmonary hypertension arises from cardiac conditions such as heart failure or heart valve disease, it is referred to as a pulmonary venous hypertension. However, it is important to remember that pulmonary hypertension can be associated with multiple causes.

What are symptoms of pulmonary hypertension?

Healthy pulmonary arteries of the lungs are elastic. They expand and contract with each beat of the heart. In pulmonary hypertension, arteries stiffen and thicken. This leads to increased resistance to blood passing through the vessel, thereby increasing pressure. Higher pulmonary pressure can lead to symptoms of pulmonary hypertension. These symptoms of pulmonary hypertension usually involve the heart. Common symptoms may include:

- Shortness of breath, especially with exertion
- Low oxygen levels
- Chest pain or pressure
- Near-fainting/fainting
- Fatigue
- Palpitations
- Swelling of the ankles or abdomen
- Heart failure (in advanced cases)

How is pulmonary hypertension diagnosed?

The diagnosis of pulmonary hypertension can be difficult and is often delayed until the disease has progressed. Pulmonary hypertension cannot be diagnosed non-invasively.

When diagnosing pulmonary hypertension, an ultrasound of the heart, or echocardiogram, can provide an estimate of the pressure in the heart.

Only a procedure called a right heart catheterization (RHC) can directly measure blood pressure in the lungs and determine if pulmonary hypertension is present. A right heart catheterization can also be used to determine if pulmonary hypertension is responsive to intravenous vasodilator medication. This will help determine whether or not a person is a candidate for chronic medication treatment.

Other tests are performed to screen for associated diseases, including blood tests, EKG, chest X-ray, pulmonary function testing, and a test for lung blood clots called a ventilation/perfusion scan (VQ scan). A 6-minute walk time is typically performed to assess a person's exercise capacity and need to oxygen therapy.

How is pulmonary hypertension treated?

Treatment of pulmonary hypertension is directed toward improving symptoms, improving exercise capacity and delaying progression of the disease. The treatment used for pulmonary hypertension depends upon its underlying cause. Pulmonary hypertension can improve with treatment of associated heart or lung disease.

Non-medication Treatments

Important non-medication treatment includes:

- Oxygen therapy
- Quitting smoking
- Removal of deleterious drugs
- Routine exercise (with the guidance of your pulmonary hypertension doctor)

Medication and other Treatments

People whose pulmonary blood pressure “responds” to intravenous vasodilator treatment during right heart catheterization may be candidates for calcium channel blocker therapy. People with idiopathic pulmonary arterial hypertension that does not respond to vasodilator challenge during right heart catheterization or who do not improve with calcium channel blocker treatment, or people who have inherited, or connective tissue-associated pulmonary hypertension are candidates for oral medications.

These oral medications include:

- Phosphodiesterase-5 (PDE-5) inhibitors (sildenafil or tadalafil),

- Soluble guanylate cyclase stimulator (riociguat),
- Endothelin receptor antagonists (ERAs) (bosentan, ambrisentan, macitentan).

PDE-5 inhibitors and ERAs can improve exercise capacity, and sometimes, slow the progression of the disease. It is unclear if these medication provide benefit to people who have pulmonary hypertension associated with lung or heart disease.

Prostanoid Medications

Medications called prostanoids may improve symptoms and, sometimes, survival. Prostanoids can be given by inhalation (treprostinil), subcutaneous injection (treprostinil) or via IV infusion (epoprostenol, treprostinil). There are also oral medications as well (selexipag, treprostinil). While the prostanoids can provide significant improvement, they have many side effects. They should be managed at pulmonary hypertension centers experienced with their use.

In people who have chronic thromboembolic pulmonary hypertension (CTEPH), surgical removal of blood clots or balloon pulmonary artery angioplasty is always considered, as this can be of greatest benefit.

When pulmonary hypertension develops into heart failure, diuretics, digitalis and sodium restriction can be helpful. In people who have pulmonary hypertension due to chronic blood clots, treating with a blood thinner is very important to prevent more blood clots.

Finally, lung or combined heart/lung transplantation may be appropriate in people who have progressive disease and have failed traditional therapy.

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