# 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 the lungs via the pulmonary arteries. Oxygen is transferred into the blood at the alveolus-capillary interface, and then 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?

There are many different types of pulmonary hypertension, and these types are categorized into different groupings, known as the World Health Organization (WHO) classification system. These different groupings reflect the different causes of the disease (and therefore different recommended treatments). These categories include:

- WHO Group I: Pulmonary hypertension due to a disease of the arteries themselves, such as idiopathic (of unknown cause), hereditable, or related to autoimmune disease
- WHO Group II: Pulmonary hypertension due to increased pressure on the left side of the heart.
- WHO Group III: Pulmonary hypertension due to lung disease and/or low oxygen levels(including sleep disorders)
- WHO Group IV: Pulmonary hypertension due to blockages in the pulmonary arteries (e.g., chronic blood clots in the lungs)
- WHO Group 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 is often idiopathic, associated with autoimmune disease or familial in nature. This kind of pulmonary hypertension is

referred as pulmonary arterial hypertension (PAH).

Pulmonary arterial hypertension can also be associated with 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 sometimes to as pulmonary venous hypertension.

And while there are specific types of pulmonary hypertension, it is important to note 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 can involve the heart. Common symptoms may include:

- Shortness of breath, especially with exertion
- Fatigue
- Low oxygen levels
- Chest pain or pressure
- Near-fainting/fainting
- 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 suspecting pulmonary hypertension, the first step is getting an ultrasound of the heart, or echocardiogram, which can provide an estimate of the pressure in the lungs.

Only a procedure called a right heart catheterization (RHC) can directly measure blood pressure in the lungs and determine if pulmonary hypertension is present. Doctors use the pattern of numbers from the heart catheterization, along with the patient's other testing, to determine whether the patient has pulmonary hypertension, and if so, what type. This helps determine the best course of therapy for each patient.

Other tests are performed to look for associated diseases, including blood tests, electrocardiogram (EKG), chest X-ray, pulmonary function tests, and a test for chronic blood clots in the lungs called a ventilation/perfusion scan (V/Q scan). A six-minute walk time is typically performed to assess a person's exercise capacity and need for oxygen therapy. It is also important for patients to have a sleep study or a night-time oxygen test.

While the testing can be intensive, especially at the time of diagnosis, these data are essential to mapping out a personalized plan to help the patient with pulmonary hypertension live and breathe better.

## How is pulmonary hypertension treated?

Treatment of pulmonary hypertension is directed toward improving your symptoms and ability to move; delaying progression of the disease; and, most important, helping you have a better quality of life. The treatment used for pulmonary hypertension depends upon its underlying cause.

### Non-medication Treatments

Important non-medication treatment includes:

- Oxygen therapy
- Quitting smoking
- Removal of deleterious drugs
- Pulmonary rehabilitation
- Healthy nutrition, consisting of real food, low in sodium and sugar (especially if there are issues with fluid retention)
- Routine exercise (with the guidance of your pulmonary hypertension doctor)

### Medication and other Treatments

People whose pulmonary blood pressure “responds” to inhaled vasodilator treatment during right heart catheterization may be candidates for calcium channel blocker therapy. People with pulmonary arterial hypertension (PAH) 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, or PAH due to other causes may be candidates for vasodilator therapy.

Pulmonary hypertension medications come in different forms: oral, inhaled, intravenous (through an IV catheter) and subcutaneous.

These medications include:

- Phosphodiesterase-5 (PDE-5) inhibitors (sildenafil or tadalafil)
- Soluble guanylate cyclase stimulator (riociguat)
- Endothelin receptor antagonists (ERAs) (bosentan, ambrisentan, macitentan)
- Prostacyclin pathway medications (epoprostenol, treprostinil, selexipag

Medications in the prostacyclin pathway may improve symptoms, exercise tolerance and sometimes survival. These medications can be given orally (selexipag, treprostinil), by inhalation (treprostinil), by subcutaneous injection (treprostinil) or via IV infusion (epoprostenol, treprostinil). While these medications can provide significant improvement, they require judicious dosing and have many side effects. They should be managed at pulmonary hypertension programs experienced with their use.

In treating pulmonary arterial hypertension, patients are often started on multiple medications at the same time or in rapid succession to target multiple pathways in this disease and achieve the best response in the shortest amount of time.

In people who have pulmonary hypertension due to chronic blood clots, known as chronic thromboembolic pulmonary hypertension (CTEPH), surgical removal of the scar tissue or balloon pulmonary artery angioplasty is always considered, as this can be of greatest benefit. In patients with CTEPH, lifelong treatment with a blood thinner is crucial to prevent more blood clots.

When pulmonary hypertension develops into heart failure, diuretics, digoxin and sodium restriction can be helpful.

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

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