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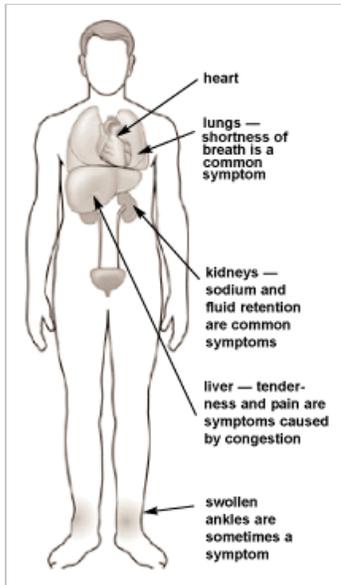
## Diseases & Conditions

### Pulmonary Hypertension: Causes, Symptoms, Diagnosis, Treatment

*(Also Called 'Secondary Pulmonary Hypertension', 'Secondary Pulmonary Hypertension')*

#### What is pulmonary hypertension?

Pulmonary hypertension is a rare lung disorder in which the arteries that carry blood from the heart to the lungs become narrowed, making it difficult for blood to flow through the vessels. As a result, the blood pressure in these arteries -- called pulmonary arteries -- rises far above normal levels. This abnormally high pressure strains the right ventricle of the heart, causing it to expand in size. Overworked and enlarged, the right ventricle gradually becomes weaker and loses its ability to pump enough blood to the lungs. This could lead to the development of right heart failure.



Pulmonary hypertension occurs in individuals of all ages, races, and ethnic backgrounds although it is much more common in young adults and is approximately twice as common in women as in men.

#### Why do the pulmonary arteries narrow?

Scientists believe that the process starts with injury to the layer of cells that line the small blood vessels of the lungs. This injury, which occurs for unknown reasons, may cause changes in the way these cells interact with the smooth muscle cells in the vessel wall. As a result, the smooth muscle contracts more than normal and narrows the vessel.

#### What are the symptoms of pulmonary hypertension?

Symptoms of pulmonary hypertension do not usually occur until the condition has progressed. The first symptom of pulmonary hypertension is usually shortness of breath with everyday activities, such as climbing stairs. Fatigue, dizziness, and fainting spells also can be symptoms. Swelling in the ankles, abdomen or legs; bluish lips and skin, and chest pain may occur as strain on the heart increases. Symptoms range in severity and a given patient may not have all of the symptoms.

In more advanced stages of the disease, even minimal activity will produce some of the symptoms. Additional symptoms include irregular heart beat (palpitations or strong, throbbing sensation), racing pulse, passing out or dizziness, progressive shortness of breath during exercise or activity, and difficulty breathing at rest. Eventually, it may become difficult to carry out any activities as the disease worsens.

#### What causes pulmonary hypertension?

The following are some known causes of pulmonary hypertension:

- **The diet drug "fen-phen."** Although the appetite suppressant "fen-phen" (dexfenfluramine and phentermine) has been taken off the market, former fen-phen users have a 23-fold increase risk of developing pulmonary hypertension, possibly years later.
- **Liver diseases, rheumatic disorders, lung conditions.** Pulmonary hypertension also can occur as a result of other medical conditions, such as chronic liver disease and liver cirrhosis; rheumatic disorders such as scleroderma or systemic lupus erythematosus (lupus); and lung conditions including tumors, emphysema, chronic obstructive pulmonary disease (COPD), and pulmonary fibrosis.
- **Certain heart diseases.** Heart diseases including aortic valve disease, left heart failure, mitral valve disease, and congenital heart disease can also cause pulmonary hypertension.
- **Thromboembolic disease.** A blood clot in a large pulmonary artery can result in the development of pulmonary hypertension.
- **Low-oxygen conditions.** High altitude living, obesity, and sleep apnea can also lead to the development of pulmonary hypertension.
- **Genetic predisposition.** Pulmonary hypertension is inherited in a small number of cases. Knowing that someone in the family had or has pulmonary hypertension should prompt you to seek early evaluation should symptoms occur.

Pulmonary hypertension may also be caused by other conditions, and in some cases, the cause is unknown.

### How is pulmonary hypertension diagnosed?

Because pulmonary hypertension may be caused by many medical conditions, a complete medical history, physical exam, and description of your symptoms are necessary to rule out other diseases and make the correct diagnosis. During the physical exam, your health care provider will:

- listen for abnormal heart sounds such as a loud pulmonic valve sound, a systolic murmur of tricuspid regurgitation, or a gallop due to ventricular failure.
- examine the jugular vein in the neck for engorgement.
- examine the abdomen, legs, and ankles for fluid retention.
- examine nail beds for bluish tint.
- look for signs of other underlying diseases that might be causing pulmonary hypertension.

Other tests that might be ordered include:

- **Blood tests:**
  - Complete metabolic panel (CMP): Examines liver and kidney function
  - Autoantibody blood tests, such as ANA, ESR, and others: Screens for collagen vascular diseases
  - Thyroid stimulating hormone (TSH): A screen for thyroid problems
  - HIV: A screen for human immunodeficiency virus
  - Arterial blood gases (ABG): Determines the level of oxygen in arterial blood.
  - Complete blood count (CBC): Tests for infection, elevated hemoglobin, and anemia
  - B-type natriuretic peptide (BNP): A marker for heart failure
- **Doppler echocardiogram:** Uses sound waves to show the function of the right ventricle, to measure blood flow through the heart valves, and then calculate the systolic pulmonary artery pressure.
- **Chest X-ray:** Shows an enlarged right ventricle and enlarged pulmonary arteries.
- **6 minute walk test:** Determines exercise tolerance level and blood oxygen saturation level during exercise.
- **Pulmonary function tests:** Evaluates for other lung conditions such as chronic obstructive pulmonary disease and idiopathic pulmonary fibrosis among others.
- **Polysomnogram or overnight oximetry:** Screens for sleep apnea (results in low oxygen levels at night).
- **Right heart catheterization:** Measures various heart pressures (ie, inside the pulmonary arteries, coming from the left side of the heart), the rate at which the heart is able to pump blood, and finds any leaks between the right and left sides of the heart.
- **Ventilation perfusion scan (V/Q scan):** Looks for evidence of blood clots along the pathway to the lungs.
- **Pulmonary angiogram:** Looks for blood clot blockages in the pulmonary arteries.
- **Chest CT scan:** Looks for blood clots and other lung conditions that may be contributing to or worsening pulmonary hypertension.

### How is pulmonary hypertension treated?

Appropriate diagnosis and analysis of the problem is necessary before starting any treatment. Treatment varies per individual based on the different underlying causes but generally includes taking medications; making lifestyle and dietary changes; having surgery, if necessary; and seeing your doctor regularly. Listed below are medication and surgical treatment approaches.

## Medications

Many different types of medications are available to treat pulmonary hypertension. Treatment choices, such as those listed below, depend on the severity of pulmonary hypertension, the likelihood of progression, and individual drug tolerance.

- **Oxygen** — replaces the low oxygen in your blood.
- **Anticoagulants or "blood thinners"** such as warfarin sodium (Coumadin) — decreases blood clot formation so blood flows more freely through blood vessels. **Note:** when taking anticoagulant medications, it is important for you to monitor bleeding complications and have regular lab work to monitor the level of medication in your bloodstream.
- **Diuretics or "water pills"** [such as furosemide (Lasix®), spironalactone (Aldactone®)] — removes extra fluid from the tissues and bloodstream, which reduces swelling and makes breathing easier.
- **Potassium** (such as K-dur®) — replaces potassium (an essential nutrient) that may be lost with increased urination when taking diuretics.
- **Inotropic agents** (such as digoxin) — improves the heart's pumping ability.
- Vasodilators [such as nifedipine (Procardia®) or diltiazem (Cardizem®)] — lowers pulmonary blood pressure and may improve the pumping ability of the right side of the heart.
- **Bosentan (Tracleer®), ambrisentan (Letairis®)** — helps block the action of endothelin, a substance that causes narrowing of lung blood vessels. These medications require monthly labwork to monitor liver function.
- **Epoprostenol (Flolan®), treprostinil sodium (Remodulin®, Tyvaso®), iloprost (Ventavis®)** — dilates pulmonary arteries and helps prevent blood clots from forming.
- **Sildenafil (Revatio®), tadalafil (Adcirca®)** — relaxes pulmonary smooth muscle cells, which leads to dilation of the pulmonary arteries.

## Surgical therapies

**Pulmonary thromboendarterectomy:** If present, a large clot in the pulmonary artery may be surgically removed to improve blood flow and lung function.

**Lung transplantation:** Currently, this is the only cure for primary pulmonary hypertension. Transplantation is reserved for advanced pulmonary hypertension that is not responsive to medical therapy. The right side of the heart will generally return to normal after the lung/lungs have been transplanted. About 1,000 lung transplants are performed annually in the United States. Many people are on the waiting list, yet a shortage of donors is the major limiting factor. Your health care provider will discuss transplantation if it is an appropriate treatment option for your condition.

**Heart/lung transplantation:** This type of double organ transplant is very rare but is necessary for all patients who have combined lung and left heart failure.

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