



Pulmonary hypertension

Overview

Pulmonary hypertension is a type of high blood pressure that affects the arteries in your lungs and the right side of your heart.

In one form of pulmonary hypertension, tiny arteries in your lungs, called pulmonary arterioles, and capillaries become narrowed, blocked or destroyed. This makes it harder for blood to flow through your lungs, and raises pressure within your lungs' arteries. As the pressure builds, your heart's lower right chamber (right ventricle) must work harder to pump blood through your lungs, eventually causing your heart muscle to weaken and fail.

Some forms of pulmonary hypertension are serious conditions that become progressively worse and are sometimes fatal. Although some forms of pulmonary hypertension aren't curable, treatment can help lessen symptoms and improve your quality of life.

Symptoms

The signs and symptoms of pulmonary hypertension in its early stages might not be noticeable for months or even years. As the disease progresses, symptoms become worse.

Pulmonary hypertension symptoms include:

- Shortness of breath (dyspnea), initially while exercising and eventually while at rest
- Fatigue
- Dizziness or fainting spells (syncope)
- Chest pressure or pain
- Swelling (edema) in your ankles, legs and eventually in your abdomen (ascites)
- Bluish color to your lips and skin (cyanosis)
- Racing pulse or heart palpitations

Causes

Your heart has two upper chambers (atria) and two lower chambers (ventricles). Each time blood passes through your heart, the lower right chamber (right ventricle) pumps blood to your lungs through a large blood vessel (pulmonary artery).

In your lungs, the blood releases carbon dioxide and picks up oxygen. The oxygen-rich blood then flows through blood vessels in your lungs (pulmonary arteries, capillaries and veins) to the left side of your heart. Ordinarily, the blood flows easily through the vessels in your lungs, so blood pressure is usually much lower in your lungs.

With pulmonary hypertension, the rise in blood pressure is caused by changes in the cells that line your pulmonary arteries. These changes can cause the walls of the arteries to become stiff and thick, and extra tissue may form. The blood vessels may also become inflamed and tight.

These changes in the pulmonary arteries can reduce or block blood flow through the blood vessels. This makes it harder for blood to flow, raising the blood pressure in the pulmonary arteries.

Pulmonary hypertension is classified into five groups, depending on the cause.

Group 1: Pulmonary arterial hypertension

- Cause unknown, known as idiopathic pulmonary arterial hypertension
- A specific gene mutation that can cause pulmonary hypertension to develop in families, also called heritable pulmonary arterial hypertension
- Certain drugs — such as certain prescription diet drugs or illegal drugs such as methamphetamines — or certain toxins
- Heart abnormalities present at birth (congenital heart disease)
- Other conditions, such as connective tissue disorders (scleroderma, lupus, others), HIV infection or chronic liver disease (cirrhosis)

Group 2: Pulmonary hypertension caused by left-sided heart disease

- Left-sided valvular heart disease, such as mitral valve or aortic valve disease
- Failure of the lower left heart chamber (left ventricle)

Group 3: Pulmonary hypertension caused by lung disease

- Chronic obstructive pulmonary disease, such as emphysema
- Lung disease such as pulmonary fibrosis, a condition that causes scarring in the tissue between the lungs' air sacs (interstitium)
- Sleep apnea and other sleep disorders
- Long-term exposure to high altitudes in people who may be at higher risk of pulmonary hypertension

Group 4: Pulmonary hypertension caused by chronic blood clots

- Chronic blood clots in the lungs (pulmonary emboli)

Group 5: Pulmonary hypertension associated with other conditions that have unclear reasons why the pulmonary hypertension occurs

- Blood disorders
- Disorders that affect several organs in the body, such as sarcoidosis
- Metabolic disorders, such as glycogen storage disease
- Tumors pressing against pulmonary arteries

Eisenmenger syndrome and pulmonary hypertension

Eisenmenger syndrome, a type of congenital heart disease, causes pulmonary hypertension. It's most commonly caused by a large hole in your heart between the two lower heart chambers (ventricles), called a ventricular septal defect.

This hole in your heart causes blood to circulate abnormally in your heart. Oxygen-carrying blood (red blood) mixes with oxygen-poor blood (blue blood). The blood then returns to your lungs instead of going to the rest of your body, increasing the pressure in the pulmonary arteries and causing pulmonary hypertension.

Risk factors

Your risk of developing pulmonary hypertension may be greater if:

- You're a young adult, as idiopathic pulmonary arterial hypertension is more common in younger adults
- You're overweight
- You have a family history of the disease
- You have one of various conditions that can increase your risk of developing pulmonary hypertension
- You use illegal drugs, such as cocaine
- You take certain appetite-suppressant medications
- You have an existing risk of developing pulmonary hypertension, such as a family history of the condition, and you live at a high altitude

Complications

Pulmonary hypertension can lead to a number of complications, including:

- **Right-sided heart enlargement and heart failure (cor pulmonale).** In cor pulmonale, your heart's right ventricle becomes enlarged and has to pump harder than usual to move blood through narrowed or blocked pulmonary arteries.

At first, the heart tries to compensate by thickening its walls and expanding the chamber of the right ventricle to increase the amount of blood it can hold. But this thickening and enlarging

works only temporarily, and eventually the right ventricle fails from the extra strain.

- **Blood clots.** Clots help stop bleeding after you've been injured. But sometimes clots form where they're not needed. A number of small clots or just a few large ones dislodge from these veins and travel to the lungs, leading to a form of pulmonary hypertension that can generally be reversible with time and treatment.

Having pulmonary hypertension makes it more likely you'll develop clots in the small arteries in your lungs, which is dangerous if you already have narrowed or blocked blood vessels.

- **Arrhythmia.** Irregular heartbeats (arrhythmias) from the upper or lower chambers of the heart are complications of pulmonary hypertension. These can lead to palpitations, dizziness or fainting and can be fatal.
- **Bleeding.** Pulmonary hypertension can lead to bleeding into the lungs and coughing up blood (hemoptysis). This is another potentially fatal complication.

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