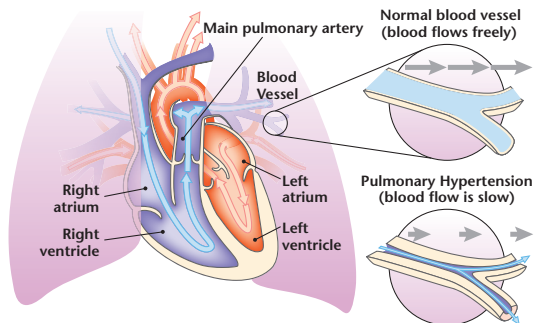


What Is Pulmonary Hypertension?

Pulmonary hypertension (PH) is a general term that is used to describe high blood pressure in the lungs from any cause. This is a different problem than high blood pressure in the whole body (systemic hypertension). There are five different groups of pulmonary hypertension that are based on the causes. Different types of PH can require different treatment but all forms of pulmonary hypertension are serious and can be life-threatening. Pulmonary hypertension can develop in children or adults at any age. This fact sheet will focus on adults and describe what happens in PH and groups. For more information about pulmonary hypertension in children and the diagnosis and treatment of pulmonary hypertension see the Patient Information Series fact sheets at www.thoracic.org/patients.



How does blood flow in the lungs?

To understand pulmonary hypertension (PH) it helps to understand how blood flows throughout your body. While the heart is one organ, it works like two pumps that are connected to one another. There is a left side and a right side of the heart, each with two different jobs.

The left side of the heart (left atrium) takes oxygen-rich blood coming from your lungs and the left ventricle pumps this blood throughout your body. Since the left side of your heart has to pump blood such a great distance, the left side of your heart is designed to pump against a fairly high pressure. This pressure is easily measured with a blood pressure cuff and is called your blood pressure. When your blood pressure is too high, it is called systemic hypertension or simply, hypertension.

After your blood has delivered oxygen to the tissues of your body, the blood needs to come back to the lungs to get more oxygen. It does this by returning the blood to the right side of the heart (right atrium) and then the right ventricle pumps the blood into your lungs, so the process can start over again. The blood does not need to travel very far to get from the right side of your heart to your lungs. Therefore, the right side of your heart pumps against less pressure than the left side of your heart. The right side of your heart is therefore normally a low-pressure system. The pressure that the right side of your heart is pumping against is called your pulmonary pressure.

What happens in the lungs and body with PH?

When the pulmonary pressure is too high, it is called pulmonary hypertension (PH). This puts stress on the right side of the heart because the muscles on the right side are not used to pushing blood out to the lungs against such high pressures. Over time, the right side of the heart is strained and begins to fail.

The heart can lose its ability to pump enough blood through the lungs to meet the needs of the rest of the body. Blood gets backed up into the veins of the lungs. Because the blood has

difficulty getting through the lungs to pick up oxygen, your blood oxygen level may be lower than normal. This can put a strain not only on your heart, but also decrease the amount of oxygen getting to your brain. These problems can lead to death.

What are the different groups of pulmonary hypertension?

There are 5 different groups, based on their causes:

Group 1—Pulmonary arterial hypertension (PAH)

PAH differs from other forms of PH in that the artery walls in the lungs are directly diseased. The arteries remodel and become narrow, thick and/or stiff. There are several types of PAH:

- ▶ Idiopathic PAH—occurs without any clear cause
- ▶ Hereditary PAH—due to genes that are passed down (inherited) in families
- ▶ PAH occurring with other medical conditions including:
 - congenital heart disease
 - liver disease/cirrhosis
 - HIV infection
 - schistosomiasis infection
 - connective tissue diseases—such as scleroderma and lupus
- ▶ PAH occurring with past or present drug use:
 - use of prescription amphetamines or certain diet pills
 - use of illicit drugs such as cocaine and methamphetamines

Group 2—Pulmonary hypertension due to left heart disease

When the left heart has problems that limit how well it can pump blood out to the body, it leads to a “backup” of blood in the lungs that raises pressure in the lungs. Group 2 is the most common form of PH. The left heart can have problems with weakened heart muscles that can’t squeeze blood as well, stiff heart muscles that can’t relax normally, or with the valves on the left side of the heart.

Group 3—Pulmonary hypertension due to lung disease

This group includes PH due to chronic lung disease and/or hypoxia (low oxygen levels).

This can occur in lung diseases like COPD, cystic fibrosis, and interstitial lung disease. It can also occur with sleep apnea, particularly if not treated. It could also occur in a person who has been living in an area of high altitude for a long period of time because of the lower oxygen level in the air. Arteries in the lungs tighten so that blood can only go to areas of the lungs that are receiving the most air and oxygen.

GROUP 4—Pulmonary hypertension due to chronic blood clots in the lungs.

In this group, there are blood clots in the pulmonary arteries that the body has trouble dissolving. These clots block blood flow in the lungs causing high blood pressures. This form of pulmonary hypertension is called chronic thromboembolic pulmonary hypertension (CTEPH). For more information on CTEPH, see the ATS Patient information Series fact sheet at www.thoracic.org/patients.

Group 5—Pulmonary hypertension due to unknown causes

In this group, PH is secondary to other diseases in ways that are not well understood. These associated conditions include, but are not limited to, sarcoidosis, sickle cell anemia, chronic hemolytic anemia, splenectomy (spleen removal) and certain metabolic disorders.

What are the symptoms of pulmonary hypertension?

There may be no signs or symptoms of PH in its early stages. Over time, the high pressure in the lungs can put a stress on the heart and low oxygen levels cause changes throughout the body. Some symptoms start out mild and get worse slowly while others can come on all of a sudden.

You can help your healthcare provider in diagnosing your condition by telling them what kind of symptoms you are having and if there is any worsening of these symptoms. For example, let your healthcare provider know if you notice any of the following:

- New or increased shortness of breath
- Dizziness
- Feeling like you might faint
- Fainting/passing out (syncope)
- Chest pain
- Heart palpitations (feeling like your heart is racing or pounding)
- New or worsening swelling of your feet, legs or belly
- Lips and/or fingers turning blue

Pulmonary hypertension (PH) can be difficult to diagnose in a routine medical exam because the most common symptoms of PH, such as breathlessness, fatigue and dizziness, are also associated with many other conditions. If your healthcare provider suspects that you have PH, he or she will want to review your medical and family history, perform a physical exam and perform one or more diagnostic tests.

Because the different kinds of pulmonary hypertension are treated differently, it is important that your healthcare provider takes the time and orders the necessary tests to find

out what kind of pulmonary hypertension you have. There are treatments available which can stop it from getting worse and help symptoms. At this time, there is no cure for most people with PH. Centers specializing in PH (www.phassociation.org/Patients/FindADoctor) may be the best option for care as they have dedicated doctors, nurses, and other staff to assist in your care.

For more information about the diagnosis and treatment of pulmonary hypertension, go to the ATS Patient Information Series fact sheet at www.thoracic.org/patients.

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Rx Action Steps

- ✓ Talk to your healthcare provider if you have symptoms that might suggest pulmonary hypertension
- ✓ If you have chronic heart or lung disease work with your healthcare provider to keep it in good control to reduce your risk of developing pulmonary hypertension
- ✓ If you have other health issues that may lead to PH, ask your healthcare provider what to do and how to watch for any problems.
- ✓ If you have PH, work closely with a specialist in managing pulmonary hypertension and take all medications regularly.

Healthcare Provider's Contact Number:

For More Information**American Thoracic Society**

- www.thoracic.org/patients/
 - COPD
 - CTEPH
 - Diagnosis & Treatment of PH
 - OSA in Adults

Pulmonary Hypertension Association

- www.phassociation.org

PHAware Global

- www.phaware.global

PHA Europe—European Pulmonary Hypertension Association

- <http://www.phaeurope.org/>

US National Library of Medicine

- <https://medlineplus.gov/pulmonaryhypertension.html>

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