

Pulmonary Hypertension

Helpful information for
patients and families



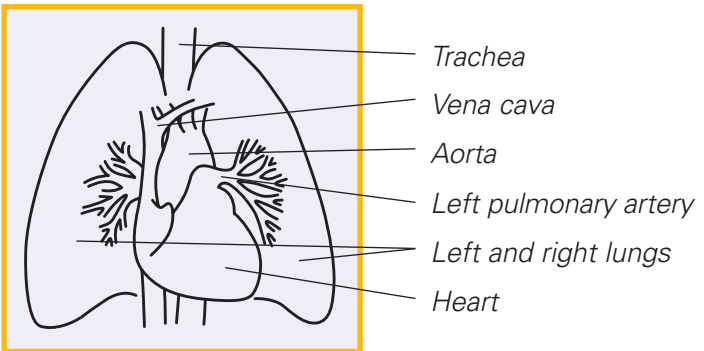
Pulmonary Hypertension Association

Understanding pulmonary hypertension

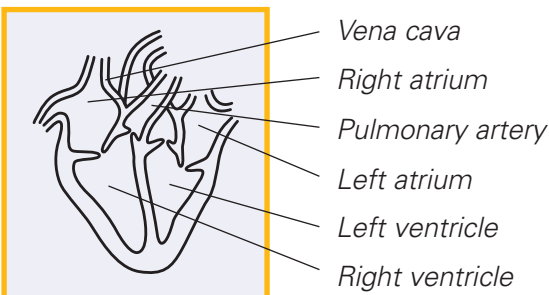
Pulmonary hypertension, or PH, is a simplified name for a complex health problem: high blood pressure in the lungs. The name comes from *pulmonary*, or having to do with the lungs, and *hypertension*, which means high blood pressure. It is a disease that affects people of all ages and ethnic backgrounds.

One thing that most types of PH have in common is that they are often not recognized until they are quite advanced. As a result, many people may have PH without knowing it—so higher awareness and earlier diagnosis can help people get earlier treatment. There are several different kinds of PH, and this brochure will describe them, and explain how they are different.

Basic parts of the lungs and heart



The inside of the heart





Understanding the different types of PH

PH starts when the small vessels that supply blood to the lungs constrict, or tighten up. It is more difficult for blood to get through to the lungs, and the heart must pump harder to overcome the resistance. As time passes, scarring (or *fibrosis*) of the vessels makes them stiffer and thicker, and some may be completely blocked. The extra stress causes the heart to enlarge and become less flexible. Less and less blood is able to flow out of the heart, through the lungs, and into the body, and more and more symptoms start to show.

In 1998, the World Health Organization (WHO)* sponsored a meeting of PH specialists with the goal of creating a standardized system for defining different kinds of PH. The committee proposed that the disease be divided into the following five categories of pulmonary hypertension:

1. Pulmonary arterial hypertension (PAH)

The pulmonary artery is the large vessel that carries blood from the heart into the lungs so it can pick up oxygen. Most cases of PH affect this artery and the hundreds of tiny blood vessels that branch off from it. This category includes two types of PH:

Primary pulmonary hypertension (PPH) can occur at random, without any apparent cause. Other times, the disease can be inherited; it is estimated that at least 10 percent of PPH cases occur in families where at least one other person has had it.

PAH related to other causes appears to be similar to PPH, but is actually associated with exposure to toxins or the effects of other diseases. These can include:

- Collagen vascular disease (scleroderma, lupus, rheumatoid arthritis)
- Congenital heart disease
- Chronic liver disease

*The World Health Organization is considered the international authority on health definitions and disease information. Visit the WHO website at www.who.int.

- Human immunodeficiency virus (HIV)
- Use of diet drugs such as fenfluramine or dexfenfluramine
- Use of cocaine, methamphetamine, or other street drugs
- Persistent pulmonary hypertension of the newborn

2. Pulmonary hypertension associated with disorders of the respiratory system

- Emphysema
- Interstitial lung disease
- Sleep apnea
- Chronic exposure to high altitude

3. Pulmonary hypertension due to chronic thrombotic or embolic disease

- Blood clots in pulmonary arteries
- Pulmonary embolism (caused by clot, tumor, or foreign matter in the lungs)
- Sickle cell disease

4. Pulmonary hypertension due to disorders directly affecting the pulmonary blood vessels

- Inflammatory diseases such as schistosomiasis or sarcoidosis
- Pulmonary capillary disease

5. Pulmonary venous hypertension (PVH)

is caused by diseases of the left side of the heart, such as mitral valve disease or a poorly performing left ventricle. This can cause increased pulmonary artery pressures, but rarely leads to severe PAH.



Confirming a diagnosis

PH is often not diagnosed in a timely manner because its early symptoms can be confused with those of many other conditions. Some of the symptoms of PH include:

- Breathlessness or shortness of breath
- Feeling tired all the time
- Dizziness, especially when climbing stairs or upon standing up
- Fainting—often the symptom that brings people to their doctors
- Swollen ankles and legs
- Chest pain, especially during physical activity

To establish a diagnosis of PH, a series of tests are performed that show how well a person's heart and lungs are working. These tests may include:

- Echocardiogram
- Electrocardiogram (ECG)
- Right heart catheterization
- Six-minute walk test
- Assessment of function in tasks of daily living

If these tests show that a person definitely has PH, other kinds of diagnostic tests may be ordered to aid in identifying the cause, including:

- A computed tomography (CT or CAT) scan to rule out a pulmonary embolism or lung disease
- A pulmonary function test to rule out obstructive lung disease
- A formal sleep study to rule out sleep apnea
- Laboratory tests to rule out hepatitis, collagen disease, HIV, or other conditions

When no cause can be found, the diagnosis is primary pulmonary hypertension (PPH).

A comprehensive treatment plan

PH of almost any type is a chronic condition that can become life-threatening. Although there is no cure, there are several treatments available, and patients have more options than ever before

Most of the current treatments for PH are prescribed for specific problems caused by the disease. If you have a combination of symptoms, you may receive different types of medication alone or combined with others, such as:

- Anticoagulants to prevent blood clots in the lungs
- Calcium channel blockers to relieve constriction in pulmonary arteries
- Digoxin to help the heart pump blood more effectively
- Diuretics to reduce fluid in the system
- Inhaled oxygen to make more oxygen available to the blood
- Bosentan (brand name Tracleer®) is an oral *endothelin receptor antagonist*, or ERA. It affects endothelin, a natural chemical in the body that plays an important role in the development of PAH. Bosentan works by reducing the high blood pressure in the lungs, thereby enabling the heart to pump blood more effectively. Bosentan is administered in simple pill form.
- Epoprostenol (brand name Flolan®) is a prostacyclin analogue that helps open up constricted lung blood vessels, thereby reducing the high blood pressure in the lungs. This medication is delivered by a constant intravenous infusion.
- Treprostinil (brand name Remodulin™) is also a prostacyclin analogue used to open constricted blood vessels. It is administered through a continuous subcutaneous (under the skin) infusion.

For patients who do not respond to any of these treatments, a lung transplant may be recommended.



The promise of the future

Today there is a lot of exciting research that is rapidly advancing our understanding of the causes and mechanisms of PH. These discoveries mean that other important new treatments are on the horizon—and they offer bright hope for the future.

Right now, innovative new therapies that are under investigation in clinical trials may soon be available to patients. These include a new generation of oral and inhaled prostacyclin drugs that are easier for patients to take.

Be informed, be aware, be active

Whether you are reading this brochure because you think you or a loved one might have PH, or because you are living with PH now, you should know that today you have more choices—and more opportunities—than ever. Be proactive about making the right choices; work closely with your family, friends, and healthcare providers to make sure you have the emotional support you need, as well as the most appropriate treatment.

Your healthcare team can also guide you to resources in your area where you can find out more about PH and the many new developments in research and treatment.

One important resource you should know about is the Pulmonary Hypertension Association (PHA). This organization offers information and support to PH patients and their families; it also works to build awareness of PH an

professionals. Please read the rest of this brochure for details, and for several easy ways you can contact the PHA.



About the Pulmonary Hypertension Association

This unique organization was founded by and for PH patients. Its mission is to seek a cure, to provide hope, support, and education, to promote awareness, and to advocate for the pulmonary hypertension community. The PHA fulfills its mission through:

- Funding for research
- Conferences for physicians and patients
- Support for PH support groups
- 800# patient response help-line
- PHA website with PH discussion and message boards

*To reach the PHA patient
HELP-LINE call **800-748-7274**.*

*For more information about
PH or PHA, contact the
PHA office at **301-565-3004**,
or email pha@phassociation.org.*



Pulmonary Hypertension Association

www.phassociation.org

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