

a closer look

at living with pulmonary arterial
hypertension (PAH)

Insights into PAH

If you or someone you are caring for has been diagnosed with PAH, you may be seeking information to help you better understand this complex disease. This brochure has been created to help you learn more about PAH. Here you'll find:

- An overview of PAH—including symptoms, diagnosis, and treatment
- A list of resources you may find helpful as you search for additional information and support

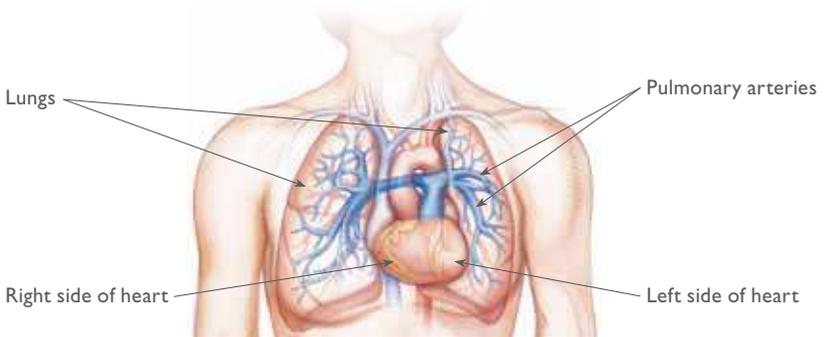
Be sure to talk to your doctor if you have further questions.



You are not alone.

PAH is a disease that affects people of all ages and backgrounds. In the United States, approximately 1,000 new cases of PAH are diagnosed each year.

What do I need to know about PAH?



Pulmonary arterial hypertension, or PAH, is high blood pressure in the arteries of the lungs (the pulmonary arteries).

Your pulmonary arteries transport blood from the right side of your heart to your lungs, where the blood picks up oxygen needed for physical activity. After passing through your lungs, blood returns to the left side of your heart, where it is pumped to the rest of your body.



magnified view of pulmonary arteries

If you have PAH, your pulmonary arteries become much narrower than usual. As a result, blood cannot easily pass through your lungs. The right ventricle of your heart must pump harder, and pressure rises inside your pulmonary arteries. Over time, this extra work weakens your heart, and it loses its ability to pump as much blood as your body needs.



How is pulmonary hypertension (PH) different from PAH?

PH is a general term for any condition in which pressure is too high in the blood vessels of the lungs. There are many types of PH with many different causes. **PAH** is a rare form of PH that is defined by abnormal changes in the walls of the arteries of the lungs. The treatment for one type of PH may differ from the treatment for another, so it is important for your doctor to know which type you have.

What causes PAH?

There are many causes of PAH. Some of these are known and some have yet to be identified.

If no specific cause for a person's PAH can be identified, then it is referred to as *idiopathic PAH (IPAH)*.

PAH that runs in families and can be inherited is called *heritable PAH (HPAH)*.

PAH associated with another disease or condition is called *associated PAH (APAH)*.

Diseases and conditions associated with APAH include:

- Collagen vascular disease (also known as connective tissue disease), which occurs when the body's immune system mistakenly attacks its own tissues and cells (scleroderma is an example)
- Congenital heart disease (a birth defect in the heart)
- Chronic liver disease, such as cirrhosis (scarring of the liver)
- Human immunodeficiency virus (HIV)
- Drugs and toxins, such as amphetamines (for example, "speed" or methamphetamine) and cocaine

The list shows only some common associations. PAH can also be associated with other, more rare conditions.

What are the symptoms of PAH?

The symptoms of PAH may not be recognized at first because they can be similar to those of other, more common conditions.

PAH can develop for some time before symptoms appear. Symptoms of PAH may include:

- Shortness of breath, especially with physical activity
- Feeling tired
- Chest discomfort or pain
- Dizziness
- Light-headedness or fainting
- Swelling of the arms and legs
- Discoloration of the fingers and toes with exposure to cold (Raynaud's phenomenon)



How is PAH diagnosed?

PAH is a complex disease and can be difficult to diagnose.

Unlike regular (systemic) hypertension, PAH cannot be measured with a blood pressure cuff on your arm. Instead, special tests are required. If your doctor thinks you may have PAH, he or she will schedule a series of these tests, which may include the following:

- **Echocardiography**—shows blood flow through your heart. An “echo” may be done in your doctor’s office
- **Right heart catheterization**—measures blood pressure in your pulmonary arteries and how much blood your heart can pump every minute. This test is usually done in a hospital under light sedation, and takes about an hour. For more information, please see the *Insights Right Heart Catheterization* brochure

Your doctor may also ask you to have one or more of the following tests:

- Electrocardiogram
- Chest x-ray
- Chest CT or MRI
- Pulmonary function test
- Ventilation/perfusion scan
- Oximetry/sleep study

These tests help your doctor determine if your symptoms are the result of PAH or another illness.

The tests listed here do not necessarily represent all of the tests your doctor may wish to perform or the order in which they may occur.

What factors determine treatment?

Treatment for PAH depends on each individual's needs and the severity of his or her PAH.

WHO functional class.

The World Health Organization (WHO)* divides people with PAH into four groups (called *functional classes*), depending on how severe their symptoms are. Your doctor will determine which functional class you are in as part of planning how best to treat your PAH.

WHO Functional Class	Definition
I	Physical activity is not limited. Ordinary physical activity does not cause shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
II	Comfortable at rest, but ordinary physical activity causes shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
III	Comfortable at rest, but less than ordinary physical activity causes shortness of breath, feeling tired, chest pain or discomfort, or feeling faint.
IV	Unable to perform any physical activity. Shortness of breath and/or feeling tired may be present at rest, and symptoms increase with almost any physical activity.

*The World Health Organization is considered the international authority on health definitions and disease information.

6-Minute walk test.

The *6-minute walk test* is another tool your doctor may use to evaluate your PAH and decide on the best treatment for you. The 6-minute walk test measures how far you can walk in 6 minutes.

How is PAH treated?

The goals of treatment for PAH are to relieve symptoms and slow some of the damage that PAH causes.

Over the past 20 years, many advances have been made in the treatment of PAH.

Available treatment options:

- **Oral therapy**—medications that are taken by mouth
- **Inhaled therapy**—medications that are inhaled
- **Continuous intravenous (into a vein) or subcutaneous (under the skin) infusion**—medications that are delivered continuously to your bloodstream through a vein or a needle placed under the skin
- **Surgery**—including lung or heart-lung transplantation; considered for the most severe disease in people for whom medication is not successful

Your treatment may also include:

- **Blood thinners** to prevent blood clots
- **Diuretics** to reduce swelling
- **Supplemental oxygen** to make sure your body has enough oxygen

Talk to your doctor to learn about the treatment options that are best for you.

Some helpful tips

Share your feelings.

You may find it helpful to participate in a local or national PAH support group and talk to other people who have PAH. You may also find comfort in sharing your thoughts and feelings with a friend, relative, or healthcare professional. Your doctor can help you find a support group or professional counselor in your area. The Pulmonary Hypertension Association also maintains a list of support groups. For more information, visit www.phassociation.org.

Find an activity you enjoy.

Find an activity or hobby that brings you pleasure without overtaxing you physically. Here are some ideas to consider:

- Listening to music
- Painting
- Participating in crafts
- Gardening
- Cooking

Get involved.

Taking an active role in your PAH treatment can give you the confidence to help manage this disease. If there are things about PAH or about your treatment you do not understand, ask your doctor to explain them to you. You'll find a number of helpful resources listed at the end of this brochure. *A Patient's Survival Guide*, available from the Pulmonary Hypertension Association (www.phassociation.org), is filled with practical information on PAH, and is an excellent place to begin.





Additional resources

The following organizations may help you in your search for more information about PAH:

National Institutes of Health—an online library produced by the US Department of Health and Human Services, where you can research health topics, including PAH (www.nih.gov)

PHCentral—PAH information and support for people with PAH and healthcare providers (www.phcentral.org)

Pulmonary Hypertension Association—an organization dedicated to providing support, education, advocacy, and awareness to the PAH community (www.phassociation.org)

Scleroderma Foundation—a national organization for people with scleroderma; provides support and education to patients and funds research into the disease (www.scleroderma.org)

Insights

Your PAH Resource

Insights is an educational program designed exclusively for individuals with pulmonary arterial hypertension and those who care for them—brought to you by Gilead Sciences, Inc.

www.InsightsOnPAH.com

