

Definition

Causes

Diagnosis

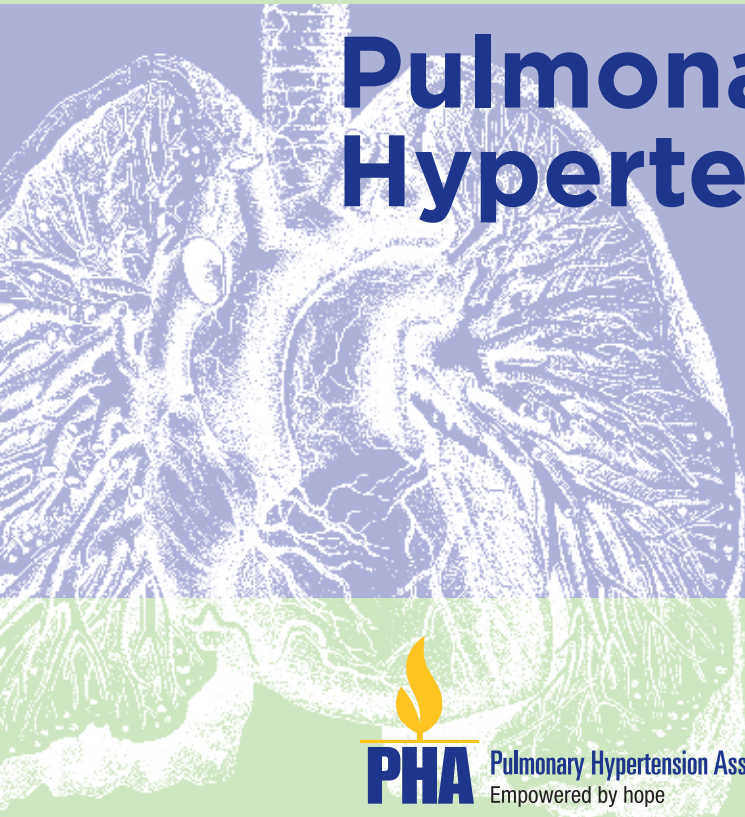
Finding a doctor

Treatment

Living, coping and more

# UNDERSTANDING

# Pulmonary Hypertension



Pulmonary Hypertension Association  
Empowered by hope

[www.PHAssociation.org](http://www.PHAssociation.org)



# THE BASICS

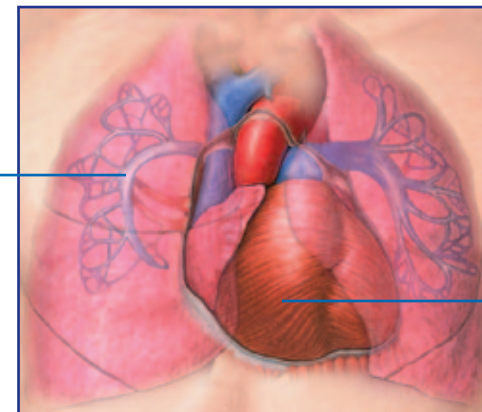
## What is pulmonary hypertension?

Pulmonary hypertension is a complex and often misunderstood disease. The term pulmonary hypertension (PH) refers to high blood pressure in the lungs. While in regular hypertension (also known as high blood pressure), the arteries throughout the body are constricted, PH affects only the blood vessels in the lungs and the right side of your heart.

This brochure focuses primarily on *pulmonary arterial hypertension* (PAH), a chronic and currently incurable disease that causes the walls of the arteries of your lungs to tighten and stiffen. In someone with PAH, the right side of the heart has to work harder to push blood through narrowed arteries in the lungs. Eventually, the extra stress causes the heart to enlarge and become less flexible, compromising the heart's ability to push blood out of the heart, through the lungs, and into the rest of the body.

### Symptoms of pulmonary hypertension may include:

- breathlessness
- fatigue
- dizziness
- chest pain
- fainting
- swelling of the arms, legs or abdomen



Narrowing of pulmonary artery

Enlarged right ventricle of the heart



## What causes PH?

In many cases, no cause can be identified, and in these cases the disease is referred to as *idiopathic PAH*. Sometimes PH can develop due to another medical condition, including connective tissue disease, congenital heart disease, liver disease, HIV, and others. This type of PH is known as *associated PAH*. Some families have a form of PH that can be inherited. This is known as *heritable PAH*.

**Could PH run in my family?**  
Yes, but heritable PAH is relatively uncommon. Of the small percentage of people who do carry the PH gene, only a small number of carriers will develop the disease. Genetic testing is available to find out if you carry the PH gene. To learn more, visit [www.PHAssociation.org/AboutPH/Genetics](http://www.PHAssociation.org/AboutPH/Genetics).

## How serious is PH?

PH is a serious disease that can take a significant toll on your energy and activity levels and, if left untreated, may lead to right heart failure. Fortunately, due to major advancements in the past two decades, PH-specific therapies are available to help relieve symptoms, improve your quality of life, and slow disease progression. To benefit from these advances, it is essential for new patients to find a PH-treating doctor who can develop a treatment plan as soon as possible after diagnosis.

Every PH patient is different, and new research with the potential to improve the outlook for this disease is being conducted all the time. Your journey with PH depends on many factors, including the severity of your disease and how you respond to treatment. Once in the care of a PH-treating healthcare team and on PH-specific treatment, patients can live for many years.

## How does my doctor know I have PH?

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. To determine if you have PH, your medical team will schedule specialized tests, possibly to include a pulmonary function test and an echocardiogram. If the results of these tests point to PH, your doctor will schedule a right heart cardiac catheterization, commonly referred to as a "right heart cath." Right heart catheterization is considered the gold standard for diagnosing PH.

**Right heart catheterization** is one of the most accurate and useful tests to get a definitive diagnosis for PH. This is the only test that directly measures the pressure inside the pulmonary arteries, and it should be done in all patients at least once.



# FINDING A DOCTOR

## What kinds of doctors treat PH?

PH is a rare but serious disease, and many doctors, including many cardiologists and pulmonologists, have little training or experience in diagnosing and treating PH. Patients are encouraged to see a cardiologist, pulmonologist or, in some cases, a rheumatologist who treats many PH patients. These doctors are familiar with PH medications and side effects, and can determine which treatments or combination of therapies are right for you.

## How can I find a doctor to treat my PH?

One way to begin your search for a PH-treating doctor is through the Find a Doctor directory on the PHA website at [www.PHAssociation.org/FindaDoctor](http://www.PHAssociation.org/FindaDoctor). People without Internet access can call PHA's Patient-to-Patient Support Line (800-748-7274) and ask the volunteer at the end of the line to give you the names and contact information for doctors in your area. Once you've identified the physicians closest to you who treat PH, interview them carefully to determine which center will offer you optimal care.

## What questions should I be asking my medical team?

Keep a list of your questions between appointments, and don't be afraid to ask why your doctor is running a test, scheduling a procedure, or changing a medication.

At your first appointment, you might consider asking:

- How often should I come to see you?
- How often do I need to have cardiac catheterization?
- What are the potential side effects of my medication?
- (If you wear a pump for intravenous medication) What do I do if the line leaks or comes out?
- Who should I contact in the event of an after-hours emergency?
- What lifestyle changes (nutrition, exercise, etc.) do I need to make?

## Here are some questions to pose to doctors you're considering for PH treatment:

- When did you begin caring for PH patients, and how many PH patients do you currently treat?
- Do you have at least one nurse who works with patients on PH-specific medications?
- Do you require all patients to undergo right heart catheterization with **vasodilator testing** prior to prescribing a therapy for PH?
- What PH therapies do you prescribe, and how many patients are on IV prostacyclin? (While you may ultimately be placed on a different therapy, a center's familiarity with treating patients on the most advanced PH therapy is a good reflection of its level of PH expertise.)
- Do you conduct clinical trials on PH medications?
- Have you referred patients for lung or heart/lung transplant?
- Have you referred patients for **pulmonary thromboendarterectomy**?
- Which educational meetings regarding PH have you attended in the last two years?

*Vasodilator testing* is used to determine how much the pulmonary blood vessels can relax over a brief period of time.

*Pulmonary thromboendarterectomy (PTE)* is a complex surgical procedure performed on eligible patients with chronic pulmonary thromboembolic disease (CTEPH), whose PH is caused by blood clots in the lungs. PTE surgery is used to remove blood clots and restore blood flow to the lungs. This surgery is performed only at select, specialized centers.



# TREATMENT

## How is PAH treated?

A number of FDA-approved medications are available for the treatment of PAH. These medications are administered in several ways: directly into the vein (intravenously), beneath the skin (subcutaneously), orally, and by inhalation. Medications to treat PAH work in three distinct ways, and may be prescribed alone or in combination:

**Prostacyclins** tell the blood vessels to relax. They mimic prostaglandin, a substance that is often deficient in PH patients. Treatments in this category include *epoprostenol* (Flolan™ and Veletri™), *treprostinil* (Remodulin™ and Tyvaso™), and *iloprost* (Ventavis™).

**Endothelin receptor antagonists** block endothelin, a substance that tells the blood vessels to constrict. PH patients tend to have this substance in excess. Medications in this category include *bosentan* (Tracleer™) and *ambrisentan* (Letairis™).

**PDE-5 inhibitors** allow the lungs to produce more of their own natural vasodilators. Vasodilators are substances that tell your blood vessels to relax, improving blood flow. This category of treatment includes *sildenafil* (Revatio™) and *tadalafil* (Adcirca™).

Doctors may also prescribe:

**Blood thinners**, also known as anticoagulants, to prevent blood clots

**Calcium channel blockers** to relieve constriction in the pulmonary arteries

**Diuretics** to reduce fluid retention

**Supplemental oxygen** to help patients breathe

Lung or heart/lung transplant may also be an option for certain patients. Pulmonary thromboendarterectomy (PTE) may be an option for certain patients with chronic pulmonary thromboembolic disease.

## Learn More

PHA offers education and information for patients, family members and medical professionals.

- Materials for people who are newly diagnosed  
[www.PHAssociation.org/Patients/NewlyDiagnosed](http://www.PHAssociation.org/Patients/NewlyDiagnosed)
- Information on PH and treatments  
[www.PHAssociation.org/AboutPH](http://www.PHAssociation.org/AboutPH)
- Education programs and webinars  
[www.PHAssociation.org/EducationPrograms](http://www.PHAssociation.org/EducationPrograms)
- Insurance resources  
[www.PHAssociation.org/Patients/Insurance](http://www.PHAssociation.org/Patients/Insurance)
- *Pulmonary Hypertension: A Patient's Survival Guide*, our 300-page guide for PH patients and caregivers  
[www.PHAssociation.org/Store](http://www.PHAssociation.org/Store) or 301-565-3004 x0
- Continuing medical education for healthcare providers  
[www.PHAOnlineUniv.org](http://www.PHAOnlineUniv.org)

## How will my doctor determine which treatment is best for me?

Your doctor will take into consideration the severity of your illness (referred to as your “functional class”) and the results of your cardiac catheterization to help determine which medication is right for you. As your symptoms and pressures change, your doctor may want to adjust the type and dosage of your medication accordingly.



## How can I adjust to life with PH?

While it can be difficult to imagine in the days and months following diagnosis, many patients develop strategies to cope with the physical and emotional aspects of living with PH and lead happy, fulfilling lives. Making small changes in daily activities can make a big impact on how you feel. Here are a few lifestyle modifications to get you started:



- Listen to your body and rest when you need to
- Be honest with friends and family about how you are feeling
- Avoid activity in outdoor temperature extremes and high humidity
- Work with your healthcare team to develop an exercise plan to keep you upbeat and healthy without overexerting your body
- Monitor your sodium intake to reduce fluid retention (a common problem for PH patients)
- Talk to your doctor about the possibility of supplemental oxygen or pulmonary rehab to help you breathe better
- Be on the lookout for common signs of depression and anxiety in you and your loved ones (these may include feelings of helplessness, difficulty sleeping, or a lack of interest in activities you used to enjoy)
- Seek help from a healthcare professional, counselor or clergy member to discuss coping strategies to help you adjust to life changes and challenges

With a bit of patience, planning, and flexibility, many patients find ways to work, travel, exercise, and do the things they loved before diagnosis. Visit [www.PHAssociation.org/Patients/LivingwithPH](http://www.PHAssociation.org/Patients/LivingwithPH) for more tips on living with PH.

## Find Support

When you're ready, you can reach out to PHA and others living with PH for support, hope and resources to help you cope and adjust to life with PH.

- Patient-to-Patient Support Line **800-748-7274**
- Local and virtual support groups  
[www.PHAssociation.org/Community](http://www.PHAssociation.org/Community)
- PH Email Mentors for patients, parents and caregivers  
[www.PHAssociation.org/Mentors](http://www.PHAssociation.org/Mentors)
- Support and information for family and friends  
[www.PHAssociation.org/Caregivers](http://www.PHAssociation.org/Caregivers)
- Opportunities to get involved and make a difference  
[www.PHAssociation.org/GetInvolved](http://www.PHAssociation.org/GetInvolved)
- Membership in our "Community of Hope" to receive *Pathlight*, our quarterly newsletter, a discount on *Pulmonary Hypertension: A Patient's Survival Guide*, and much more  
[www.PHAssociation.org/Join](http://www.PHAssociation.org/Join)



PHA's mission is to seek ways  
to prevent and cure pulmonary  
hypertension and to provide hope for  
the PH community through support,  
education, advocacy and awareness.

**Newly diagnosed? Order an *Envelope of Hope*.**

Request your free information packet at  
[www.PHAssociation.org/EnvelopeofHope](http://www.PHAssociation.org/EnvelopeofHope)



**Pulmonary Hypertension Association**  
Empowered by hope

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