

# NAVIGATING

## PULMONARY HYPERTENSION



A Guide for  
Newly Diagnosed  
Patients





**Pulmonary Hypertension Association**  
Empowered by hope

# Welcome to the Pulmonary Hypertension Community

You probably have a lot of questions and emotions after you or a loved one received a pulmonary hypertension (PH) diagnosis. This resource aims to answer those questions and help you learn more about PH.

The Pulmonary Hypertension Association (PHA) provides this publication to help you find education, support and hope during the first few weeks, months or years after diagnosis. Although this guide won't address every question you encounter in your journey, it provides an overview of diagnosis, treatment, symptoms and quality of life. Inside, you'll find checklists, resources and questions to ask your doctor, and other information about PH. You'll also learn about ways to tap into a network of support, become involved in research and address some of the many issues that come up in day-to-day living with a chronic disease.

We don't expect you to read the entire publication from beginning to end at once. Instead, take your time to browse the contents, and select the sections or chapters that are meaningful to you on any given day. Take your time, be gentle with yourself and others, and contact PHA or your PH team anytime you have a question or feel worried, unsure or overwhelmed.

Your PH journey is unique to you. If this guide seems overwhelming, set it aside and return to it another time. When you're ready to pick it up again, use the notes section to write down additional questions you have for your care team and support network.

PHA is here to help you. The association was founded in 1991 by patients and caregivers to help individuals affected by PH find support, become better informed, learn how to advocate for themselves, find quality care, help promote research and to further our mission: to improve and extend the lives of those affected by PH. Whether you have PH or you help care for a PH patient, *Navigating Pulmonary Hypertension: A Guide for Newly Diagnosed Patients* will help you begin to navigate this new journey.

If you or a family member need to ask questions or get support, you can reach us at [Support@PHAssociation.org](mailto:Support@PHAssociation.org) or 800-748-7274.

Your PH journey is unique to you.



# Your PH Journey

Your journey is your own. While this publication lists chapters in a certain order, a different path might work better for you. Some patients and caregivers prefer to address self-care and managing emotions early on. Others want to

start by learning about the disease because it empowers them when choosing their care teams. Think of these sections as stepping stones that lead to new discoveries, no matter where you begin.



## Learn About Pulmonary Hypertension

Get more information about disease, symptoms, causes and types of PH.



## Acknowledge Your Feelings

Medical treatment is only one component of caring for yourself. Learn about the benefits of working through your emotions about your diagnosis.



## Assemble Your Care Team

You might need specialists you've never heard of, so make sure you're comfortable with your care team, including caregivers. Ask questions and ensure you're involved in decision-making.



## Understand How PH Is Diagnosed and Treated

Learn about the diagnostic tests to confirm PH and identify types, as well treatment options and how they work.



## Understand Your Treatment Plan

Learn how your medications, exercise and diet can improve your quality of life.



## Navigate Daily Life

Discover strategies to maximize your energy, manage stress and prepare for emergencies. Find resources to manage insurance and medication challenges, get support and connect with others affected by PH.

# Inside This Guide

## Learn About Pulmonary Hypertension

Now that you know the name for your condition, what does it mean? This section is a step in your journey to becoming an informed patient. Explore the tests and procedures you might encounter, and learn about the symptoms and physiological characteristics of various types of PH.

|   |    |
|---|----|
| What Is Pulmonary Hypertension?                   | 9  |
| Heart and Lungs: Basics of the Circulatory System | 10 |
| Common Symptoms                                   | 11 |
| Types of Pulmonary Hypertension                   | 12 |
| Diagnosing PH                                     | 16 |

## How Is PH Treated?

Your treatment will vary based on your type and severity of PH and other factors. As with many illnesses – from the common cold to other rare conditions – there’s no universal treatment that works the same for every patient. Your care team will help you determine the medications to best manage your symptoms and improve your quality of life.

|                               |    |
|-------------------------------|----|
| PAH Treatment Options         | 25 |
| How Does Your Treatment Work? | 26 |
| Functional Classes            | 29 |
| Assemble Your Care Team       | 30 |
| Find a PH Specialist          | 32 |

## Managing Your PH

Part of managing your illness is setting goals for your health and wellness. You’ll work with your care team and support network to develop a plan to keep your treatment on track, boost your energy and help you live your best life.

|                                     |    |
|-------------------------------------|----|
| Understand Your Treatment Plan      | 35 |
| Diet, Exercise and Energy           | 36 |
| How Much Exercise Is Right for You? | 38 |
| Maximize Your Energy                | 38 |

## Coping With PH

It's OK to be overwhelmed, angry, sad or confused about your diagnosis or the diagnosis of someone you love. Maintaining your health means making sure you get support for your emotional needs related to PH.

|                           |    |
|---------------------------|----|
| Acknowledge Your Feelings | 41 |
| Ways to Reduce Stress     | 43 |

## Navigating Potential Problems

Be prepared by creating emergency plans and knowing what to do when the unexpected occurs. Discover tips and resources to help you maneuver potential challenges associated with medication, insurance and emergency situations.

|   |    |
|---|----|
| Prepare for Medication Challenges             | 45 |
| Insurance and Financial Assistance            | 46 |
| Prepare for Emergencies                       | 48 |
| Information for EMTs and Emergency Responders | 49 |
| What If I Need More Help?                     | 51 |

## Expanding Your Knowledge

Familiarize yourself with PH-related terminology, and discover how the Pulmonary Hypertension Association can help you connect with the broader PH community, find support, participate in events and research, and deepen your knowledge of PH.

|                         |    |
|-------------------------|----|
| Participate in Research | 53 |
| PHA Resources           | 55 |
| Glossary                | 57 |

# Learn About Pulmonary Hypertension



What Is Pulmonary Hypertension? **9**

Heart and Lungs: Basics of the Circulatory System **10**

Common Symptoms **11**

Types of Pulmonary Hypertension **12**

Diagnosing PH **16**



# What Is Pulmonary Hypertension?

Pulmonary hypertension (PH) is a complex, often misunderstood disease. PH simply means high blood pressure in the lungs. With PH, blood vessels in the lungs become stiff and narrow, impeding blood flow. As a result, the right side of the heart must work harder to pump blood.

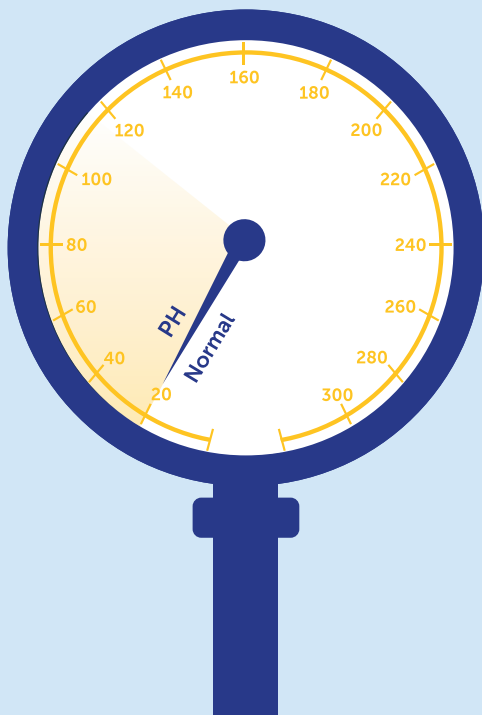
Normal pulmonary artery pressure ranges from 8 to 20 mmHg at rest. People with PH generally have an average resting pulmonary artery pressure above 20 mmHg.\*

PH differs from "regular," or systemic, hypertension, which most people refer to as high blood pressure. With systemic hypertension, pressure is higher than it should be in arteries throughout the body. With PH, pressure is higher in the lungs.

PH affects people of all ages, from newborns to seniors, and people all over the world. Some people are diagnosed with PH as infants or children and others as teens, adults or senior citizens.

There are five types of PH, based on underlying causes. Each type is different, as is each patient. It's important for newly diagnosed patients to find PH specialists as soon as possible after diagnosis to pinpoint the cause and develop a targeted treatment plan.

People with PH can live long, meaningful lives under the care of PH-treating health care teams and appropriate therapy. In fact, research has improved the outlook for people living with this disease and one day might lead to a cure.



Pulmonary hypertension is defined as a resting mean pulmonary artery pressure above **20 mmHg**.

My pulmonary artery pressure

Date

|       |       |
|-------|-------|
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |
| _____ | _____ |

# Heart and Lungs: Basics of the Circulatory System

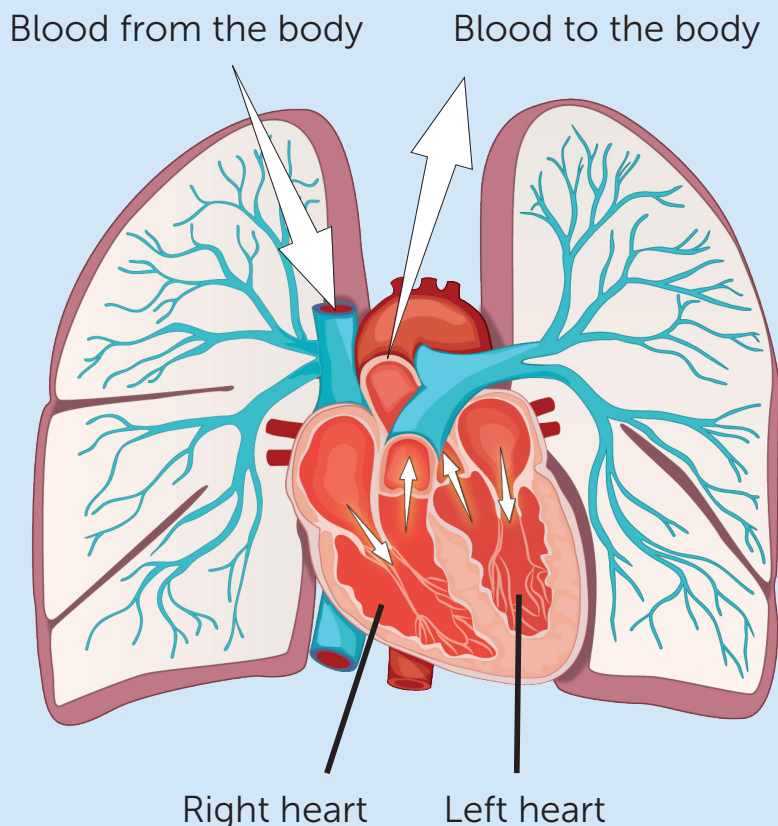
To better understand why PH is so serious, it's helpful to understand how blood circulates between the heart and lungs.

The heart and lungs work together to carry oxygen throughout the body. The heart is a muscle made up of two halves that pump blood to different areas of the body. As deoxygenated blood returns from the rest of the body, it first goes into right side of the heart, which pumps it into the lungs.

The lungs take carbon dioxide from the blood — which the body releases as you exhale — and replace it with oxygen you inhaled. After the blood picks up oxygen, it is considered “oxygenated” and is ready to return to other

areas of the body. The blood then travels from the lungs to the left side of the heart. The left side of the heart pumps blood to the rest of the body, and the process starts over again with each heartbeat.

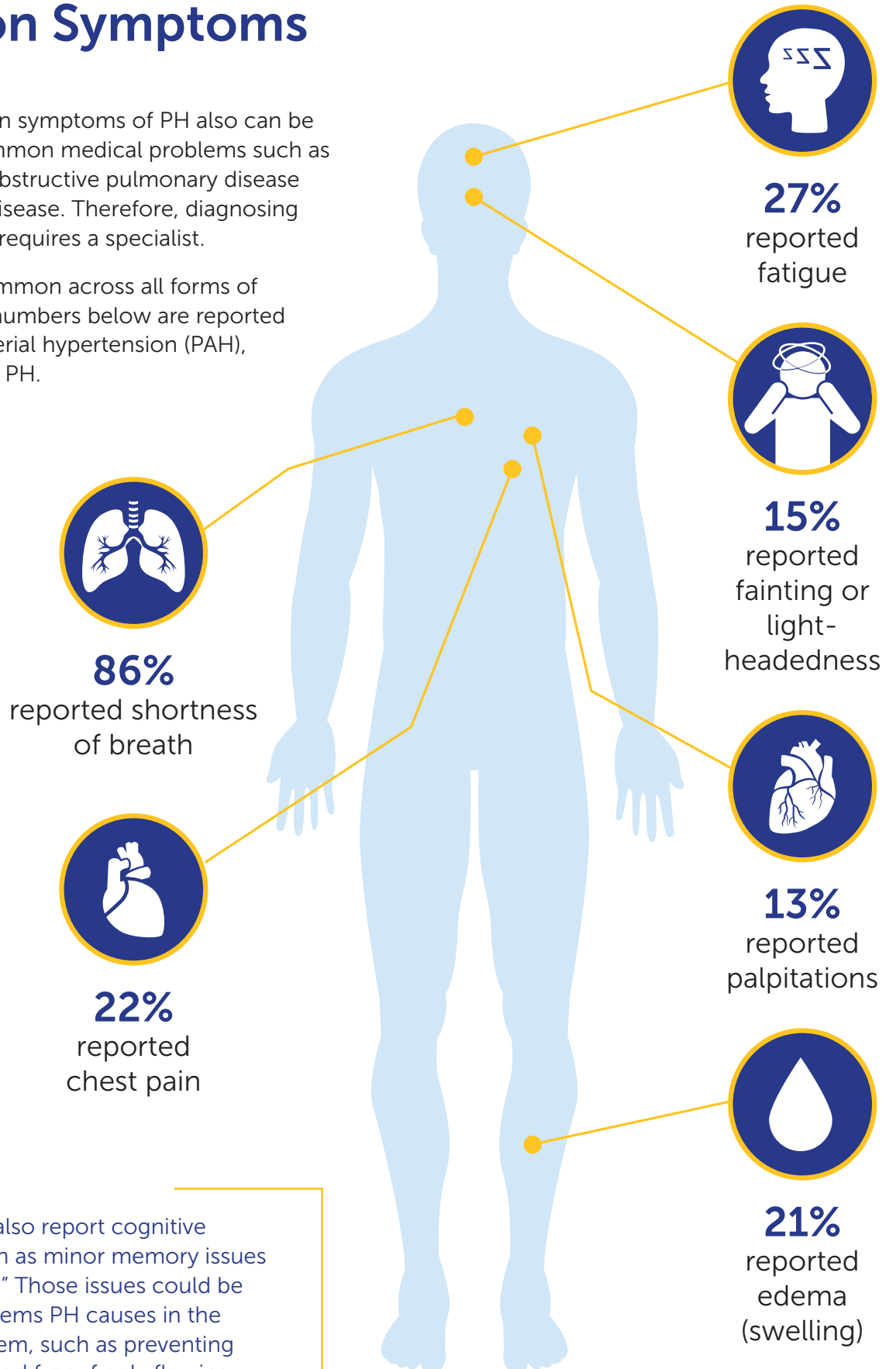
The left side of the heart is more muscular because it has to pump blood through the rest of the body, against gravity to the head, and at the same time, to the toes and back up again. The right side of the heart is smaller and less muscular than the left side because it pumps blood only through the lungs, normally a low-pressure system.



# Common Symptoms

The most common symptoms of PH also can be signs of more common medical problems such as asthma, chronic obstructive pulmonary disease (COPD) or heart disease. Therefore, diagnosing PH is difficult and requires a specialist.

Symptoms are common across all forms of PH, however the numbers below are reported for pulmonary arterial hypertension (PAH), known as Group 1 PH.



Some patients also report cognitive symptoms, such as minor memory issues and "brain fog.\*" Those issues could be related to problems PH causes in the circulatory system, such as preventing oxygenated blood from freely flowing throughout the body, including to the brain.

\* Brown LM, et al. CHEST. 2011;140(1):19-26.

# Types of Pulmonary Hypertension

Not all PH is the same. PH is a general term for high blood pressure in the lungs from any cause. There are five groups of PH based on different causes. Physicians review the classifications every five years at the World Symposium on Pulmonary Hypertension. This classification originally was developed by the World Health Organization, so sometimes you might see references to WHO groups.

## Group 1

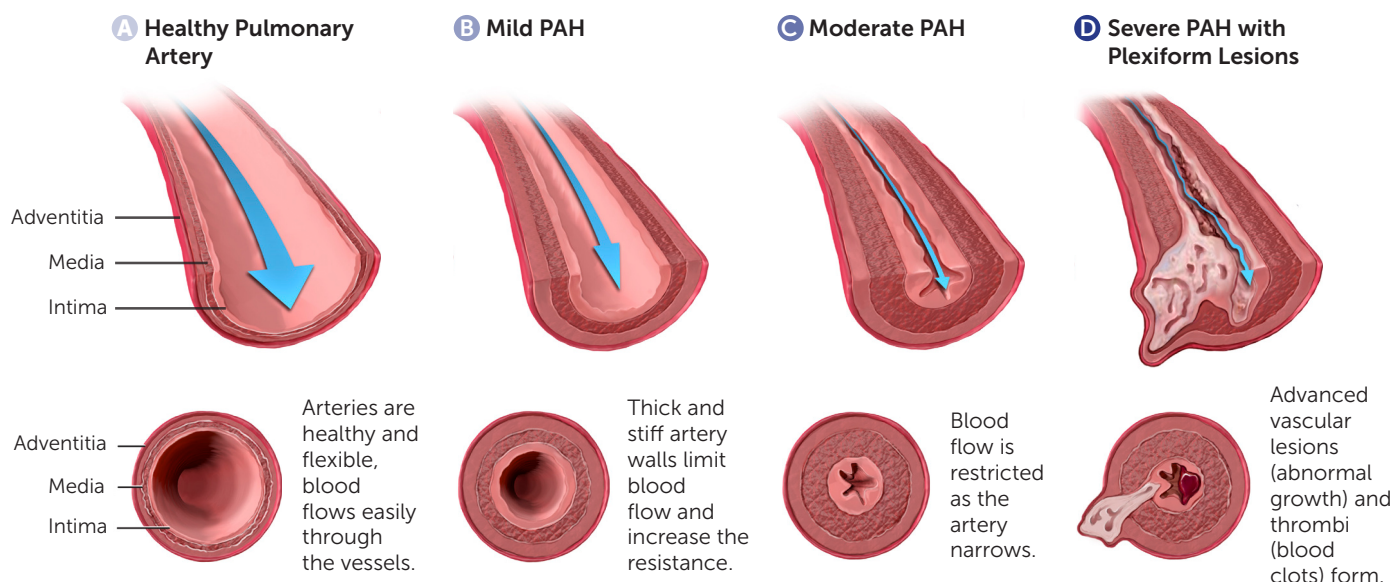
Group 1 refers to pulmonary arterial hypertension (PAH). It is caused when arteries in the lungs narrow, thicken and become rigid. The right side of the heart must work harder to push blood to the lungs through those arteries. The extra stress can cause the heart to lose the ability to pump enough blood throughout the lungs to meet the needs of the rest of the body.

There are several types of PAH. Idiopathic PAH (IPAH) occurs without a clear cause. PAH also can develop in association with other medical conditions, including congenital heart disease, liver disease, HIV and connective tissue diseases such as scleroderma and lupus. PAH can be associated with previous or ongoing use of some drugs, including methamphetamine or certain diet pills.

Heritable PAH (HPAH) is linked to genes inherited from family members. HPAH is rare. Of the small percentage of people who carry a gene associated with increased risk of PH, few will develop the disease. Genetic testing can help you find out whether you carry one of those genes. Before pursuing genetic testing, discuss the pros and cons with your PH specialist.

While several treatment options are available for PAH, there is no known cure.

Some people use the terms PH and PAH interchangeably, but they aren't the same. All PAH is PH, but not all PH is PAH.

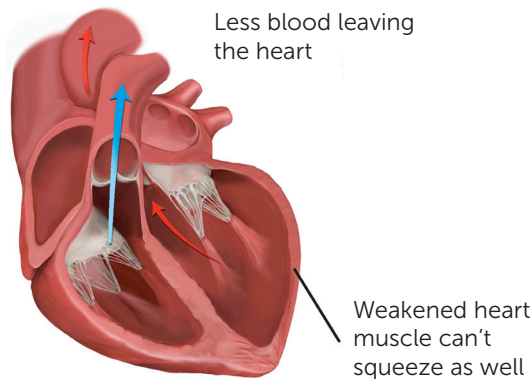


## Group 2

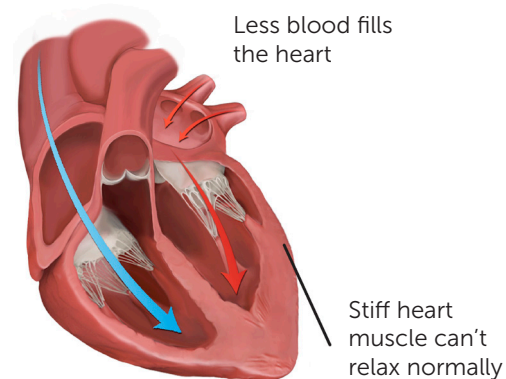
Group 2 PH is the most common form of PH in developed countries. It includes PH due to left heart disease. Although the arteries in the lungs aren't as thick and rigid as they would be with

Group 1 PH. That causes a "backup" of blood, which raises pressure in the lungs. Improving left heart function can also improve Group 2 PH.

### Systolic Dysfunction



### Diastolic Dysfunction



## Group 3

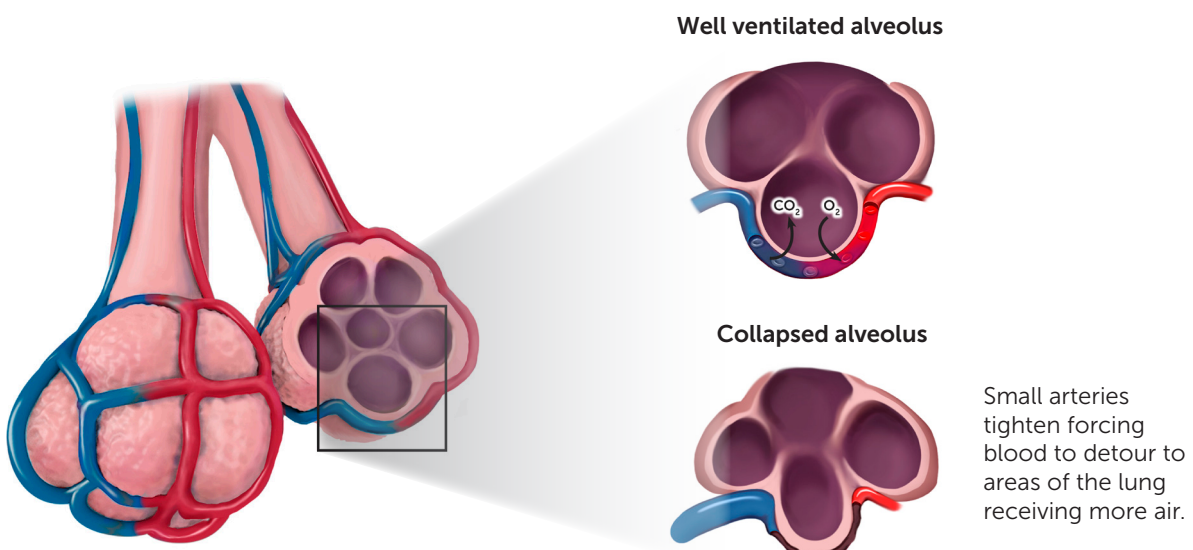
Group 3 includes PH due to chronic lung disease and/or hypoxia (low oxygen levels). These lung disorders include:

- Obstructive lung disease, where the lung airways narrow and make it harder to exhale (e.g., COPD/emphysema).
- Restrictive lung disease, in which the lungs have a tough time expanding with inhalation (e.g., interstitial lung disease, pulmonary fibrosis).

Other Group 3 causes include:

- Sleep apnea.
- Living in a high altitude for a long time. In that situation, arteries in the lungs tighten so blood goes only to areas of the lungs that receive the most air and oxygen. Over time, that tightening leads to high blood pressure throughout the lungs.

Group 3 PH can be improved through treating underlying causes such as sleep apnea or COPD.



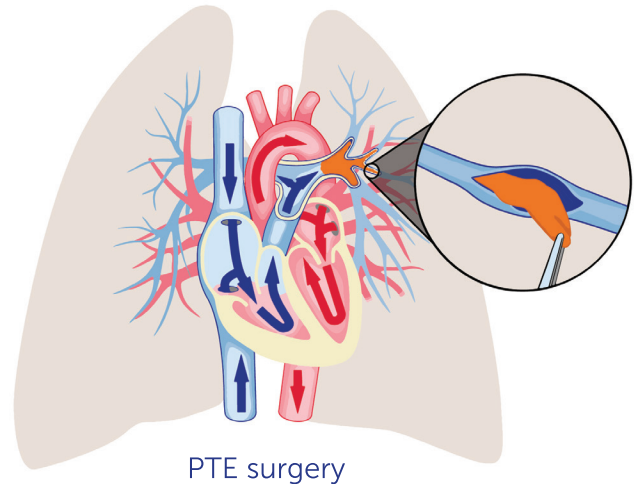
## Group 4

Group 4, known as chronic thromboembolic pulmonary hypertension (CTEPH), occurs when the body can't dissolve a blood clot in the lungs. An undissolved clot can cause scarring in the blood vessels of the lungs. The scarring impedes blood flow and makes the right side of the heart work harder than usual.

CTEPH differs from other forms of PH because it potentially can be cured. For that reason, people with PH should be screened for CTEPH with a test called a ventilation-perfusion scan.

Some patients are eligible for pulmonary thromboendarterectomy (PTE) surgery, which removes blood clots that cause CTEPH.

However, the surgery isn't appropriate for all people with CTEPH. If a doctor determines someone isn't a candidate for PTE surgery, the patient might be able to have a balloon

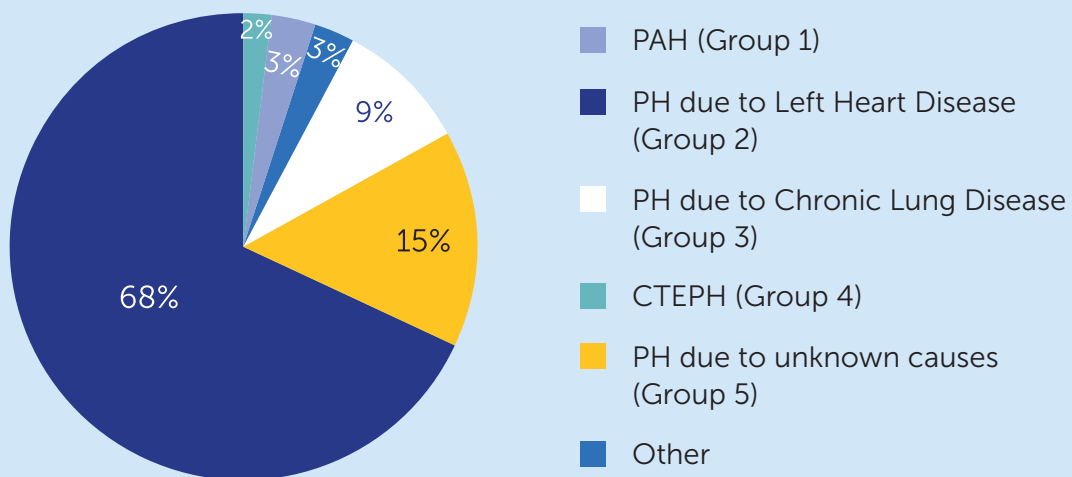


pulmonary angioplasty to open the lung blood vessels. Others might need more than one treatment. Your doctor might recommend medication instead of or in addition to these other treatments.

In some cases, PH remains after PTE surgery. Those patients also might be treated with medication. All CTEPH patients should be evaluated at specialized CTEPH centers.

Between 0.5% and 5% of people who experience a PE can develop CTEPH.

People With PH by Type



Strange G, et al. *Heart*. 2012;98(24):1805-11.



## Group 5

Group 5 refers to PH that occurs with other diseases. In those cases, PH is secondary to the associated conditions in ways that aren't well understood. The associated conditions include, but aren't limited to sarcoidosis, sickle cell anemia, chronic hemolytic anemia, end-stage kidney disease and some metabolic disorders.

No matter the type, PH is a serious disease. If untreated, PH can lead to right heart failure and death.

I have:

- Group 1
- Group 2
- Group 3
- Group 4
- Group 5

My PH is due to one of these associated conditions:

- Left heart disease
- Congenital heart disease (or "defects")
- Scleroderma
- Lupus
- Sickle cell disease
- Chronic obstructive pulmonary disease (COPD)
- Hereditary hemorrhagic telangiectasia (HHT)
- Human immunodeficiency virus (HIV)
- Sarcoidosis
- Sjogren's Syndrome
- Methamphetamine use
- Sleep apnea
- Pulmonary embolism
- Other \_\_\_\_\_

# Diagnosing PH

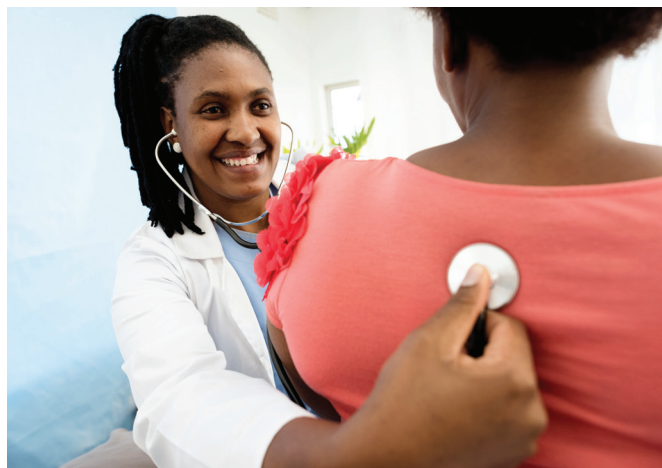
PH can be difficult to diagnose in routine medical exams because the most common symptoms of PH — breathlessness, fatigue and dizziness — also are associated with many more common conditions.

When doctors suspect someone has PH, they will review the patient's medical and family history and perform physical exams and diagnostic tests.

Before conducting diagnostic tests, your doctor will conduct a physical examination. The doctor will look for visible and/or enlarged veins along the sides of the neck, irregular heart sounds and swelling in your legs, ankles and abdomen (edema).

The doctor then will perform a series of tests to identify the type of PH you have, what might have caused it and guide your treatment plan.

Diagnostic tests can vary from clinic to clinic. They include pulmonary function, exercise tolerance tests (six-minute walk or cardiopulmonary exercise), blood tests and echocardiogram. But the only way to definitively diagnose PH is through right heart catheterization.



Right heart catheterization (RHC or right heart cath) is considered the gold standard test to confirm or rule out PH. It is the only test that directly measures the pressure and flow inside the heart and blood vessels of the lungs. All patients should undergo at least one RHC before starting PH-targeted treatments.

All other tests that give pressure readings (such as echocardiograms) provide estimated pressure. The estimates can't be used to diagnose PAH and must be confirmed with a right heart cath.

The only way to definitively diagnose PH is through right heart catheterization.

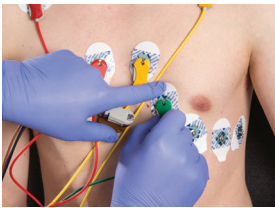


## PH detection tests



### Chest X-ray

A chest X-ray is a non-invasive test that uses radiation to take a picture of the structures inside the chest. Doctors can look at the heart, lungs, blood vessels and bones. When examining patients for PH, doctors will look at the shape of the heart to see if the chambers are larger than expected. They will check whether the pulmonary arteries are more visible than usual. And they will look for signs of lung disease, infection or fluid in the lungs (e.g., pleural effusion). With chest X-rays, doctors may see evidence of heart disease (Group 2 PH) or lung disease (Group 3 PH).



### Electrocardiogram (EKG or ECG)

An electrocardiogram is a simple, painless test that shows the electrical activity of the heart. In most cases, 12 sticky pads are placed on the chest, arms and legs and connected to a machine that prints the electrical activity of the heart. Doctors use EKGs to check how fast the heart beats, whether the beats are regular, the strength of the electrical pulse in the heart that causes the “beat” and prior heart damage.



### Echocardiogram (echo)

An echocardiogram is an ultrasound of the heart. During an echo, a sticky gel is placed on the chest near the heart, and a technician or doctor will glide a wand connected to a computer over the gel. The wand transmits sound waves to the computer. The computer translates the sound waves into pictures and videos of the heart. Doctors use the images to assess the size of the heart, how well it pumps, how fast the blood moves and how the right side of the heart looks (the side that pumps blood into the lungs).

Sometimes doctors inject saline or dye into veins during echocardiograms to better see the heart and detect abnormal blood flow. Doctors also use echocardiograms to look for evidence of left heart disease (Group 2 PH), but the tests can't definitively diagnose PH.

Echocardiograms provide rough estimates of pressure in the pulmonary artery. They estimate only systolic pulmonary artery pressure (when the heart is squeezing), which will be higher than the mean pulmonary artery pressure measured with a right heart cath. The mean pulmonary artery pressure is an average of systolic and diastolic pressures (when the heart relaxes between beats). It will be lower on a right heart cath than an estimated pressure from an echo.



## Cardiac magnetic resonance imaging (MRI)

If images from an echocardiogram are unclear, or doctors want more precise measurements of right heart size and function, they might use cardiac MRIs to look at heart function. The procedure provides images of the heart made by a powerful magnetic field and radio waves.

## PH group identification tests



## Pulmonary function tests

Pulmonary function tests (PFTs) are non-invasive tests that measure how well the lungs work. PFTs measure how much air the lungs can hold and how much and how quickly someone can blow air. They also provide information about how oxygen and carbon dioxide are exchanged between the air you breathe in and the blood circulating through the lungs.

PFTs help health care providers check for chronic lung diseases that cause Group 3 PH. They include diseases that make it harder for air to flow out of the lungs and those that prevent the lungs from easily expanding and taking in air.



## Computerized tomography (CT)

A CT chest scan is a non-invasive test that uses radiation to create many precise pictures of the structures in the chest, including the heart and lungs. Sometimes, a dye is injected into the arm (i.e., a contrast) to make the images clearer.

The test allows health care providers to see the lungs in far greater detail than with chest x-rays. Doctors can see the size of the lungs, larger blood vessels in the lungs and lung tissue. The test looks for evidence of chronic lung diseases (Group 3 PH).



## Pulmonary angiogram

A pulmonary angiogram provides images of blood vessels in the lungs after a dye is injected to make them appear more clearly. Health care providers use the test to look at the structure and branching of the blood vessels of the lungs to find evidence of chronic blood clots (Group 4 PH).



## Ventilation/perfusion scan (V/Q scan)

A V/Q scan helps doctors rule out whether you have undissolved clots in the blood vessels of your lungs. It is the gold standard screening test for Group 4 PH (CTEPH). The test compares two lung scans that show ventilation (air flow) and perfusion (blood flow). Both scans use nuclear imaging to take pictures of your lungs.

During the ventilation scan, patients use masks to breathe in a detectable gas mixed with oxygen. The scanners take pictures as the patient breathes so the health care provider can see the sections of the lungs that receive air.

During the perfusion scan, a traceable substance is injected into a vein in the arm. The scanner looks at which sections of the lungs receive blood flow. If there is a blockage in a pulmonary artery because of a blood clot, that section won't appear dark on the image, but the ventilation scan could still look normal. Doctors then will decide whether to perform more tests to identify chronic blood clots in that section of the lungs and determine whether they could be removed with PTE surgery.



## Leg ultrasound

Doctors might use an ultrasound of the legs to check for blood clots that contribute to Group 4 PH (CTEPH).



## Sleep studies

Overnight oximetry is a test that measures oxygen levels in the blood. A plastic clip with a sensor is placed over the fingertip and worn overnight to measure how much oxygen is in the blood. This non-invasive test can take place at home. If oxygen levels drop below a certain point (generally 89%, set by insurance companies), for a specific length of time, oxygen therapy is recommended.

Polysomnography is a recording of brain waves during sleep. Doctors use it to determine what causes low oxygen levels. They look at how often and how long breathing stops during sleep. They also screen for restless leg syndrome. The test generally takes place overnight at a sleep center.

## Testing for PAH Type

While there are many types of PH, there also are different forms of PAH (Group 1 PH). The following tests can help determine which type you have and influence how it's treated.



### Blood tests and immunology

A health care provider will draw a small amount of blood to assess a patient's general health and look for markers of specific diseases associated with PAH (Group 1 PH). Blood tests measure kidney, liver and thyroid function and can identify HIV, connective tissue diseases or other disorders. They also can identify anemia and evidence of stress on the heart.

Doctors sometimes examine arterial blood gas (ABG) to measure arterial oxygen and carbon dioxide levels.



### Abdominal ultrasound scan

If health care providers suspect a patient has liver disease, they will order an ultrasound of organs in the abdomen, including the liver, kidneys, gallbladder, spleen and pancreas. Health care providers likely will focus on the liver to check for any evidence of liver disease, which can lead to a type of PH called portopulmonary hypertension (PoPH).

POPH occurs as a result of advanced liver disease. It has the same characteristic symptoms as those found in cases of Group 1 PAH not associated with liver disease.

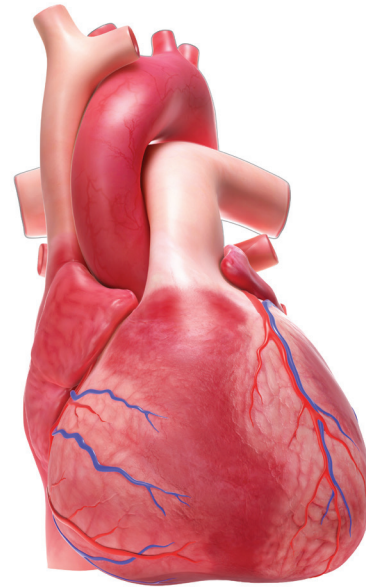
After doctors confirm a PH diagnosis and identify the type, they will use other tests to determine how best to treat a patient's PH, monitor how he or she responds to treatments and obtain other information to forecast disease progression.

## PH Hemodynamics: Testing the Heart Muscle

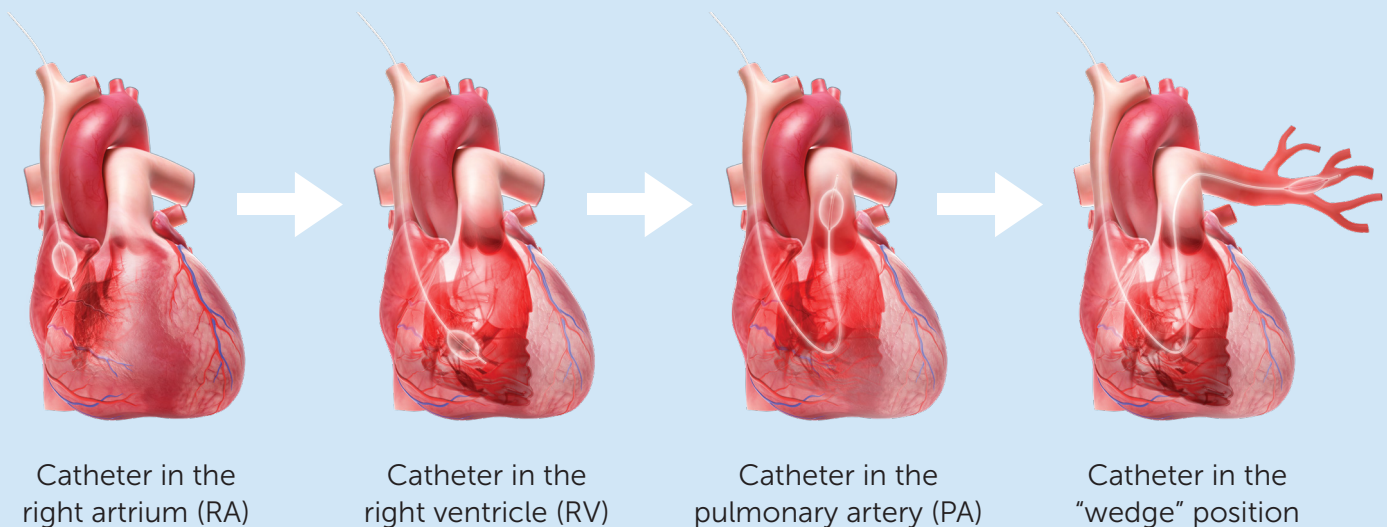
### Right heart catheterization (RHC or right heart cath)

In this test, a doctor inserts a small tube into a vein in neck or groin and guide it into the right atrium of the heart. As the vein is guided through the right ventricle into the pulmonary artery, the practitioner will take several pressure readings and sometimes withdraw blood from the catheter tip.

The doctor will calculate how much blood the heart can pump in a minute (cardiac output) and take a reading called a "wedge" pressure. In this test, a balloon is inflated at the end of the catheter and wedged into a smaller section of one of the pulmonary arteries. The reading gives insight into the pressure on the left side of the heart and can indicate left heart disease that contributes to Group 2 PH. Patients with PAH (Group 1 PH) will have normal wedge pressure.



The only way to definitively diagnose PH is through right heart catheterization.



Catheter in the right atrium (RA)

Catheter in the right ventricle (RV)

Catheter in the pulmonary artery (PA)

Catheter in the "wedge" position

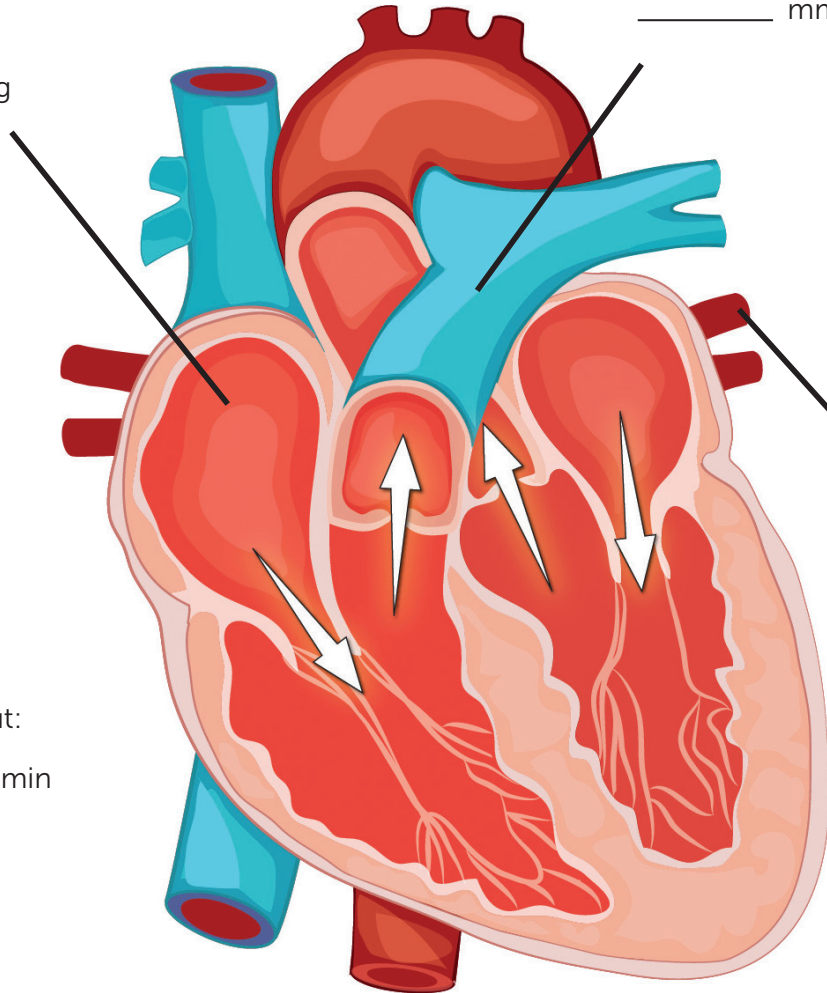
# My Right Heart Catheterization

Date: \_\_\_\_\_

Right Atrium:  
\_\_\_\_\_ mmHg

Mean (Average) Pulmonary  
Artery Pressure:

\_\_\_\_\_ mmHg



"Wedge" Pressure or  
Left Ventricular End  
Diastolic Pressure:

\_\_\_\_\_ mmHg

Cardiac Output:  
\_\_\_\_\_ L/min

Pulmonary Vascular  
Resistance (PVR):

\_\_\_\_\_ Wood Units

Additional notes on my right heart cath:

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

## Acute vasoreactivity test

During the right heart cath, doctors sometimes want to see how the heart and pulmonary pressures change after certain short-acting drugs. The drugs, administered by IV or inhaled, cause the pulmonary arteries to relax.

With the catheter in the pulmonary artery, a doctor will inject the drug or give the patient a gas called nitric oxide. The doctor will take several pressure measurements within a few

minutes. Doctors look for significant drops in pulmonary artery pressures, known as a positive vasodilator response. They check whether patients would benefit from drugs called calcium channel blockers (CCB).

Most patients don't have positive vasodilator responses so they receive PAH-specific therapies.

## PAH Exercise Capacity



### Six-minute walk test

This test measures how far a patient can walk in six minutes. It also measures perceived exertion, heart rate and oxygen saturation (how much oxygen is in the blood). Doctors compare test results from each clinic visit as one way to measure whether PH is improving or worsening.



### Cardiopulmonary exercise test

Health care providers use a CPET to measure blood pressure and oxygen consumption when patients exercise (often on a stationary bike). The test identifies how well both heart and lungs respond to exercise.

### What can I do now?

Track your test results in a binder or electronic file to help you understand what happens with your health. Get started with this worksheet. [PHAssociation.org/patienttools/](https://www.phasociation.org/patienttools/).

Many of the tests that diagnose PH also identify the PH group and functional class. Your providers might require a right heart catheterization every few months or a six-minute walk test at every visit. Those repeated tests help them evaluate your health and – with your input – make the best decisions about your treatment and care.

Some reasons doctors order those tests include changes to your health status, events like hospitalizations or medication changes. If you ever wonder why a test is being run or what its results mean, ask your PH team.

# How Is PH Treated?



PAH Treatment Options **25**

How Does Your Treatment Work? **26**

Functional Classes **29**

Assemble Your Care Team **30**

Find a PH Specialist **32**



# PAH Treatment Options

Although no cure yet exists for PH, with the possible exception of operable CTEPH, treatments are available for Group 1 PH, and more are in the drug-development pipeline. Treatments include conventional medical therapies, such as diuretics and oxygen therapy. They also include oral, inhaled, subcutaneous (under the skin) and intravenous (into the vein) medications for some types of PH.

Other forms of PH (Groups 2 and 3) are managed by treating the underlying heart and/

or lung conditions. Depending on the severity of PH, a heart or lung transplantation also might be an option.

People with Group 4 can benefit from PTE surgery to remove clots. Those who aren't candidates for surgery or whose PH remains after surgery might benefit from balloon pulmonary angioplasty or PH-targeted therapy.

Each patient is different, so talk to your PH doctor about the best treatment options for your specific diagnosis.

## Oral medications

Endothelin receptor antagonists (ERA)

- Ambrisentan (Letairis)
- Bosentan (Tracleer)
- Macitentan (Opsumit)

Phosphodiesterase Type-5 Inhibitors (PDE-5)

- Sildenafil (Revatio)
- Tadalafil (Adcirca)

Prostacyclin analog

- Oral Treprostinil (Orenitram)

Selective IP Receptor Stimulator

- Selexipag (Uptravi)

Soluble Guanylate Cyclase Stimulator

- Riociguat (Adempas)

## Inhaled medications

Prostacyclin analog

- Iloprost (Ventavis)
- Inhaled Treprostinil (Tyvaso)

## Subcutaneous medications

Prostacyclin analog

- Treprostinil (Remodulin)

## Intravenous medications

Intravenous treatments will be started in a hospital. They include:

Prostacyclin analog

- Epoprostenol (Flolan)
- Room temperature stable epoprostenol (Veletri)
- Treprostinil (Remodulin)

# How Does Your Treatment Work?

Fortunately, major advancements in the past two decades have led to therapies for Group 1 PH that target the pulmonary arteries (PH-targeted therapies). Medication can relieve symptoms, improve quality of life and slow progression. Your treatment – by pill, inhaler, injection or IV – depends on your type of PH, your functional class and anticipated chemical reactions in your body, among other considerations.

## Pathways

Changes in body chemistry can have a profound impact on overall health. A change in the chemical balance affects how cells communicate with each other. That imbalance can cause cells and organs to react abnormally.

For PH patients, three chemical families or pathways are known to be abnormal in the blood vessels of the lungs. The abnormalities contribute to PH by causing the blood vessels in the lungs to tighten.

PH-targeted medications help blood vessels relax so blood flows more easily through them to the lungs. They work by modifying the chemical imbalance in three pathways.



## Supplemental oxygen

Low oxygen levels in the lungs can cause further narrowing of the blood vessels in your lungs, making it harder for blood to flow through your lungs. Your PH team might call this “an increase in resistance.” Supplemental oxygen can help some patients by relaxing the blood vessels in the lungs, which can improve how you feel.

If prescribed by your PH team, supplemental oxygen use is just as important as the pills,

inhaled or infused medications your PH team prescribes. There are many different oxygen device options. Your health care team will work to balance oxygen flow and portability depending on your medical needs and insurance coverage.

The goal is to keep my oxygen saturation level above: \_\_\_\_\_

## 1 Endothelin pathway

Too much endothelin-1



Endothelin is a family of chemicals made by the cells in the innermost layer of the blood vessels. Endothelin can tell the blood vessels to relax or tighten. One form of endothelin, called ET-1, tells the blood vessels to tighten by binding to a specific receptor on a nearby cell's surface.

It can be thought of as a key (ET-1) fitting into a lock (receptor). Many PH patients make too much ET-1. Endothelin receptor antagonists (ERA) block the receptors from the increased ET-1, decreasing its effects and allowing the blood vessel to relax and widen.

## 2 Nitric oxide pathway

Too little nitric oxide



Nitric oxide is a gas naturally produced by the body that circulates in the blood. Nitric oxide acts as a "key" for a "lock" called soluble guanylyl cyclase (sGC). sGC can be thought of as a factory (called an enzyme) that produces an important chemical called cyclic GMP (cGMP).

Cyclic GMP is an important chemical because it helps blood vessels relax. It is broken down by another enzyme called phosphodiesterase type-5 (PDE-5). Many PH patients don't have enough nitric oxide to fuel production of cGMP.

PH-targeted treatments help increase cGMP in two ways to keep blood vessels more relaxed. The first: PDE-5 inhibitors, which stop PDE-5 from breaking down the important cGMP. The second is with a sGC stimulator – or agonist – which makes the sGC "factory" produce cGMP with less fuel (nitric oxide).

## 3 Prostacyclin pathway

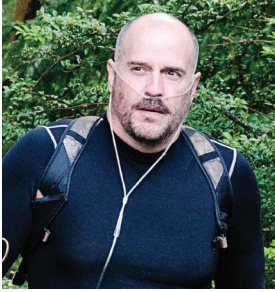
Too little prostacyclin



Prostacyclin also is produced in the cells of the innermost layer of blood vessels. It also helps those blood vessels relax and widen. Prostacyclin also can be considered a "key" that fits into a "lock" called the prostacyclin IP receptor.

Many PH patients don't make enough prostacyclin. PH-targeted treatments help increase prostacyclin in two ways. The first is prostacyclin analogs, synthetic forms of

prostacyclin that "unlock" prostacyclin IP receptors. The second is prostacyclin IP receptor stimulators. The receptor stimulators make prostacyclin IP receptors more sensitive to prostacyclin so they are easier to "unlock" with prostacyclin chemicals.



Doctors also might prescribe non-PH-specific medication, such as:

- Calcium channel blocker: Decreases blood pressure (appropriate only for people who had positive acute vasodilatory tests during the right heart cath).
- Digoxin: Helps the heart pump blood.
- Diuretics: Remove excess fluid that puts pressure on the heart.
- Oxygen: Inhaled by patients via nasal cannulas or face masks. Can relax and widen pulmonary blood vessels.
- Blood thinners, such as Warfarin (Coumadin) or direct oral anticoagulants.

Your treatment depends on your type of PH, your functional class and anticipated chemical reactions in your body, among other considerations.



A PH specialist will perform a risk assessment to determine the best treatment. They will consider:

- Signs of right heart failure.
- How quickly symptoms worsen.
- History of fainting/passing out.
- PH functional classes.
- Six-minute walk test results.
- Significant changes in six-minute walk test results from previous visits.
- Cardiopulmonary exercise test results.
- Levels of the NT-proBNP hormone produced by the heart, which can be released in response to pressure changes.
- Appearance of heart from echocardiogram or cardiac MRI images.
- Pressure measurements and cardiac output during right heart catheterization.

### What can I do now?

Participate in research. Being involved in research can be empowering for patients and caregivers. Learn more about how to get involved on Page 53.

# Functional Classes

Doctors rate the severity of PH symptoms through four functional classes. The classes describe the severity of a patient's symptoms and can help doctors map out treatment plans. Many patients fall under higher classes at diagnosis because many people aren't diagnosed in early stages of PH. However, one goal of treatment is to move into a lower class.

While many people initially receive a dismal prognosis, many find their health improves with treatment and self-management. The descriptions are modified from World Health Organization functional assessments for pulmonary hypertension.



## Class I

You have no symptoms during ordinary physical activity.



## Class II

You are comfortable at rest, but your ordinary physical activity is somewhat limited because of breathlessness, chest pain, fatigue or dizziness.



## Class III

You usually have no symptoms at rest, but breathlessness, chest pain, fatigue or dizziness greatly limits routine activity.



## Class IV

You are often breathless and tired even while resting. You can't do any physical activity without symptoms. You show signs of right heart failure. Anyone prone to fainting would be in this class.

As your functional class or risk status changes, your doctor might change your medication type, amount and/or delivery method.

# Assemble Your Care Team

Your care team consists of health care professionals who care for people with PH, caregivers, and most importantly, you. Each manages specific areas of your care. In addition to doctors, nurses, physician's assistants and medical technicians found in a general medical practice, your PH care team can include

respiratory therapists, rheumatologists, specialty pharmacists and physicians who provide specialized care, such as cardiologists and pulmonologists.

**Remember:** You are the most important person on your care team. As the patient, you are at the center of your care. Your participation in your care planning helps other members of your care team identify and address your specific needs.

## Caregivers

Your care team may include a caregiver, often an unpaid nonprofessional who provides support, comfort or care. Typically, caregivers are spouses, parents, children or other relatives. But anyone can be a caregiver including friends and neighbors. Caregivers simply are people patients rely on to make life a little easier. Many patients rely on more than one person, although they might have a primary caregiver who is more

intimately involved than anyone else with the patient's care.

A friend or a spouse can help by joining a patient's diet and exercise regimen. Caregivers can keep patients company at the hospital during extended stays. Caregivers also can lift spirits by participating in and encouraging activities that bring the patient joy.



### What do caregivers do?

Some caregivers provide many services, while others focus on one role. Those roles include:

- Running errands.
- Attending doctor visits, as a chauffeur and a second set of eyes and ears.
- Preparing meals.
- Performing household tasks.
- Helping with medications and dressings.

## Specialized Care

Many doctors are unfamiliar with PH, a relatively rare disease. That's why it's important to find a physician with specialized training and experience to accurately diagnose and treat PH. PH patients should seek care providers with PH expertise to receive the best care possible.

Specialists typically include cardiologists, pulmonologists, rheumatologists and some internists, pharmacists, physician assistants, nurses and nurse practitioners with special PH training. Not all pulmonologists and cardiologists are PH specialists.

The diagnostic process can be long, complicated and emotional. After undergoing many tests and sometimes receiving misdiagnoses, patients often obtain second opinions.

PHA encourages all patients to ask for second opinions. You have the right to ask for a second



opinion. This is commonly done; Don't worry about offending your provider. PHA offers several resources to help you find PH specialists.

## Accredited PH Care Centers

PHA's PH Care Centers (PHCCs) are accredited medical centers and hospitals that have gone through an extensive review process. PHA evaluates and recognizes the centers for their ability to properly diagnose PH and provide expert care. The health care teams at these centers provide excellent comprehensive care for adult and pediatric PH patients.

Comprehensive care is the treatment and management of patients "as a whole" and not just their diagnosis. Care teams, which sometimes

include social workers, work with patients to manage symptoms and side effects, prevent hospital stays, answer questions and talk to loved ones about the patients' health. In addition to comprehensive PH care, patients also should have primary care physicians who work closely with the PH specialists to manage care not related to PH.

To find out whether there's a PHCC near you, see a list at [PHAssociation.org/PHCareCenters](https://www.phasociation.org/PHCareCenters).

# Find a PH Specialist

PHA provides a list of doctors with an interest in PH who belong to PHA's medical membership network, PH Clinicians and Researchers (PHCR). PHCR is a membership-based network of physicians and Ph.D.-level researchers. See the "Find a Doctor" directory at PHAssociation.org.

The directory helps patients search for specialists. PH doesn't endorse specific physicians. Unlike the PH Care Centers, PHA hasn't evaluated the doctors in the directory for their ability to treat PH patients. However, these are doctors who have chosen to focus on PH and join PHA.

## Questions to Ask Potential Doctors

Finding the right physician depends on personal preference, rapport and feeling comfortable — the right "fit." Consider interviewing more than one doctor to determine who will provide the best care.

Here are some questions to ask potential doctors, but there are no right or wrong answers — just what feels right to you.

How often should I see you?

---

What tests will you do at my follow-up appointments?

- Functional class assessment
- ECG/EKG
- Six-minute walk test
- Cardiopulmonary exercise test
- Echocardiogram
- Blood work

How often do I need a right heart catheterization?

---

What medications will I be taking for my PH and why?

---

What are possible side effects of the medications?

---



(If you wear a pump for intravenous medication) What do I do if the line leaks, breaks or comes out?

---

Whom should I call for an after-hour emergency?

---

When should I contact my medical team?

---

When should I contact the specialty pharmacy?

---

How should I change my diet?

---

Can I exercise with PH?

---

Do you work with a pulmonary or cardiac rehab program? Is that right for me?

---

What other lifestyle changes do you recommend?

---

Can you recommend a support group that your other PH patients attend?

---

Additional questions:

---

---

---

---

---

Keep a list of questions between doctor visits. Don't be afraid to ask why your doctor is running a test, scheduling a procedure or changing a medication. Find more information on all of these topics at [PHAssociation.org](http://PHAssociation.org).

# Managing Your PH

Understand Your Treatment Plan **35**

Diet, Exercise and Energy **36**

How Much Exercise Is Right for You? **38**

Maximize Your Energy **38**



# Understand Your Treatment Plan

After you assemble a health care team, the next step is to create a treatment plan in collaboration with your team. Throughout the process, it's important to ask questions and share goals for treatment with your PH care team. Their goal is to help you have the best quality of life, but be clear what quality of life means to you.

A comprehensive treatment plan will address many aspects of daily life, from medication to lifestyle to diet. Make sure you communicate clearly and frequently with your PH care team, caregivers and other people whose help or support you need.

## Care Plan Adherence: Following Doctor's Orders

After your care team proposes a treatment plan, the next step is adherence. That means taking your medicines, following the diet and fluid management plan your doctor prescribes, and exercising or seeking pulmonary rehabilitation.

Adherence can be challenging and burdensome. You might have to take multiple pills at different times each day, including medications to offset the side effects of your PH medications. You might have to push yourself on days when you have low energy.

Your care is your choice. Sometimes, treatment plans can seem too difficult to follow, and you might be tempted to take shortcuts. If following your care plan seems challenging, let your PH team know right away. Sometimes they can adjust your plan, but they need time to explore other options before making changes.

### Here are a few tips to remember:

- Take your medication as prescribed, and make sure you have it with you when you're not at home.
- Use supplemental oxygen, which should be viewed as medication, as prescribed by your doctor.
- Ask your PH doctor or nurse for help if you're struggling to follow your medication regimen. Be honest, so they can work with you to improve your medication adherence.
- Consider setting reminders on your phone.
- Don't stop taking your medication without talking to your care team. List your questions and describe any side effects.

## Oxygen saturation

Make sure you know your normal range of oxygen saturation. If your oxygen saturation drops below your normal range, you may notice:

- Unusual shortness of breath.
- Headache.
- Dizziness.
- Blue lips, fingernails or earlobes.
- Rapid heart rate.

# Diet, Exercise and Energy

How you manage your diet, exercise and energy is an important aspect of your care plan. Your care team will ask you to limit your sodium intake, help you create an exercise plan, and provide tips to conserve your energy.

This section explains how those factors affect PH and provides tips to help you adhere to your doctor's recommendations.

## Diet and Fluid Retention

Your PH care team will recommend dietary changes to reduce PH symptoms and improve heart health. One of the most important recommendations is a low-sodium diet. Sodium causes fluid retention, which can worsen symptoms and put more pressure on the heart – a serious risk for people with PH.

Buildup of excess fluid or swelling is called edema. You will notice it when your legs, ankles or abdomen swell, and it will show up as weight gain. That's why it's important to weigh yourself daily, and track any changes in your symptoms. Weigh yourself at the same time every day with the same scale. The best time to weigh yourself is in the morning after using the bathroom. Ask your care team when to notify them that your weight is increasing.



Sodium causes fluid retention, which can worsen symptoms and put more pressure on the heart.

## Limiting Sodium

Sodium is a naturally occurring element in many foods, especially table salt. That's why many people with heart issues ask to "hold the salt" when eating away from home. At home, you can reduce sodium intake by not adding salt when cooking, avoiding salty snacks and checking nutrition labels on prepared foods.

Instead of using salt, try:

- Lemons
- Limes
- Garlic
- Other herbs and spices

## Knowing your limit

Many PH patients must restrict their daily sodium intake to 2 grams, or 2,000 milligrams. They typically must limit fluid to 2 liters per day. Ask your PH team about your “max” for sodium and fluid intake.

### Finding sodium on nutrition labels

Make sure to check serving size, servings per container and sodium amounts on nutrition labels. Most packages usually contain more than one “serving” as described on the nutrition label.

### Watch for:

Low salt foods contain 5% or less of your daily requirement.  
High salt foods contain 20% or more of your daily limit.

| Nutrition Facts                 |      |                                   |
|---------------------------------|------|-----------------------------------|
| Serving Size 3/4 cup (29g)      |      |                                   |
| Servings Per Container about 17 |      |                                   |
| Amount Per Serving              |      | Cereal with 1/2 cup Dry skim milk |
| <b>Calories</b>                 | 110  | 150                               |
| Calories from Fat               | 5    | 10                                |
| % Daily Value**                 |      |                                   |
| <b>Total Fat</b> 0.5g*          | 1%   | 1%                                |
| Saturated Fat 0g                | 0%   | 0%                                |
| Trans Fat 0g                    |      |                                   |
| <b>Cholesterol</b> 0mg          | 0%   | 1%                                |
| <b>Sodium</b> 210mg             | 9%   | 11%                               |
| <b>Potassium</b> 160mg          | 5%   | 10%                               |
| <b>Total Carbohydrate</b> 24g   | 8%   | 10%                               |
| Dietary Fiber 5g                | 20%  | 20%                               |
| Sugars 5g                       |      |                                   |
| Other Carbohydrate 14g          |      |                                   |
| <b>Protein</b> 3g               |      |                                   |
| Vitamin A                       | 25%  | 30%                               |
| Vitamin C                       | 25%  | 25%                               |
| Calcium                         | 0%   | 15%                               |
| Iron                            | 50%  | 50%                               |
| Vitamin D                       | 10%  | 25%                               |
| Vitamin E                       | 25%  | 25%                               |
| Thiamine                        | 25%  | 30%                               |
| Riboflavin                      | 25%  | 35%                               |
| Niacin                          | 25%  | 25%                               |
| Vitamin B <sub>6</sub>          | 100% | 100%                              |
| Folate                          | 100% | 100%                              |
| Vitamin B <sub>12</sub>         | 100% | 110%                              |
| Phosphorus                      | 15%  | 25%                               |
| Magnesium                       | 10%  | 15%                               |
| Zinc                            | 25%  | 25%                               |

### Common high-sodium foods:

- Bacon
- Bread
- Canned beans, vegetables and soup
- Cereal
- Cheese
- Cottage cheese
- Frozen meals, such as pizza, burritos, hash browns
- Instant noodles
- Ketchup
- Prepackaged mixes for cake, pancakes, pasta, rice
- Smoked, cured (cold cuts/sausage), salted or canned meat, poultry and fish
- Salsa
- Soy sauce
- Tomato sauce/paste/juice



### Low-sodium alternatives:

- Cream cheese
- Dried fruit
- Eggs
- Fresh or frozen fruit and vegetables with no additives
- Fresh or frozen meat, poultry and fish
- Mayonnaise
- Milk
- Mozzarella
- Ricotta
- Unsalted butter
- Yogurt

# How Much Exercise Is Right for You?

Regular exercise can improve lung capacity, muscle function and quality of life for people with PH. Even a small amount of exercise, such as walking slowly to the opposite side of the room, can have positive health benefits.

Exercise recommendations differ for each patient based on the severity of PH and other health factors. Those recommendations might change over time depending on symptoms and response to treatment.

Always talk with your PH care team before beginning an exercise regimen. Your PH specialist might recommend cardiac or pulmonary

rehabilitation or help you create an exercise plan that works for you. Pulmonary and/or cardiac rehabilitation is a structured program that teaches you to exercise safely within the parameters of your PH. Ideally, you will learn to exercise with less shortness of breath.

## What can I do now?

Discover PHA's "Introduction to Exercise" videos on strength training, cardio, yoga and pulmonary rehab. Visit [PHAssociation.org/exercise](http://PHAssociation.org/exercise).

## Maximize Your Energy

Finding the right balance of activity and rest can be challenging for people with PH. You know better than anyone how much PH drains your energy and affects your mobility. Sometimes, even a long walk is out of the question.

Consider these strategies to conserve your energy:



### Lifestyle

Not being able to do the things you did before PH isn't a sign of failure. Because of your PH, you need to be more selective about what you can do and what's important to you. Take time to re-examine priorities.

- Say no, and don't feel guilty for saying it. If you tend to wear yourself out by helping other people, take a hard look at how you're doing before accepting requests, invitations or activities.
- Don't be afraid to cancel plans. Some people with PH say it took a few years to understand it was OK to cancel. That way, they didn't need multiple days of rest to make up for one day of overdoing things.
- Listen to your body.

- Take breaks or naps as often as you need.
- Don't be afraid to ask for and accept help. Ask people to help you carry things, perform chores or run errands. Many people feel good when they help someone else.



## Home and Office

- Keep anything you use often at hip level. Avoid stooping, crouching or bending. If you have to bend, get up slowly.
- Elevate trash cans, washers, dryers, garden boxes and furniture, especially your bed.
- Keep a high stool in the kitchen to use while cooking.
- Use a shower chair and movable shower head.
- Use a robotic vacuum and a grabber.
- Set a timer during activities to remind yourself to take a break. For example, if you have to do physical work, set a timer for 10 to 15 minutes. When the timer goes off, stop working, hydrate and rest at least a minute for every minute worked.
- Use a wagon, cart or wheeled item to transport items.
- Shop online and use delivery services. Why wander store aisles and wait at the register when you can do most of your shopping online?



## On the Go

- Research new destinations as much as possible.
- Avoid parking on inclines so you use less energy when walking to or from your car.
- Obtain a disabled parking permit. Ask your doctor for certification to send to your state or local department of transportation.
- Shop strategically to avoid extra walking.
- Make your shopping list according to the store layout.
- Ask the checkout clerk to bag items according to the way you put them away at home.
- Ask the clerk to make each bag is as light as possible.
- Keep a collapsible chair with you to make waiting in lines easier.
- Use mobility devices in places that require a lot of walking. Save your energy to enjoy the experience.

# Coping With PH



Acknowledge Your Feelings 41

Are You a Caregiver 42

Ways to Reduce Stress 43



# Acknowledge Your Feelings

Every new patient responds differently when learning that they have PH. You might feel angry, frightened, lonely, frustrated, overwhelmed, worried, numb or a combination of those feelings.

Remember: There are no right or wrong reactions. Those feelings are normal, and you aren't alone. Treat yourself with kindness and

compassion by giving yourself permission to feel and respond to your diagnosis without judgment.

Medical treatment is only one component of caring for yourself. You also need to work through your emotions to rediscover meaning and purpose in your life. The emotional and mental components of your health need time and attention to allow you to feel your best.

Healing means "to make whole" and refers to the process of exploring and accepting aspects of your physical, emotional, mental, social and spiritual self.

## Dealing with Emotions



### Stress

Stress is the feeling of being overwhelmed, worried or run-down. Stress is a natural part of life that affects people of all ages, genders and circumstances. It can lead to physical and psychological health issues. When combined with a chronic illness such as PH, stress can become difficult to manage. It's important to seek appropriate help to manage your stress.



### Anxiety

Anxiety is characterized by feelings of tension, worried thoughts and physical changes, such as increased blood pressure. People with anxiety often have recurring thoughts or concerns that interfere with daily life. You might avoid certain situations because of worry. You also might have physical symptoms such as sweating, trembling, dizziness or rapid heartbeat. Ask your doctor about appropriate resources.



### Burnout

Burnout is a state of mental exhaustion that results from ongoing stress and inadequate or lack of self-care. Burnout can accompany increased anxiety and sometimes is an early sign of depression.



## Depression

Depression is a common, serious medical illness that negatively affects how you feel, the way you think and how you act. Fortunately, it also is treatable. Depression causes feelings of sadness and/or loss of interest in previously enjoyable activities. Many people affected by PH suffer from periods of depression. Therefore, it's important for you and your family to learn its warning signs.



## Denial

Some people experience denial, where they protect themselves by refusing to accept the reality of distressing situation. Although denial is a coping mechanism that gives you time to adjust to that situation, prolonged denial can interfere with treatment or your ability to manage aspects of your disease.

If you experience any of these emotions more often or more intensely than usual, ask your PH care team or your primary care physician to connect you with a mental health professional. A therapist can properly assess what you're experiencing and feeling and help you live your best life with PH.

Keep in mind that caregivers and family members can be affected by the diagnosis of a loved one and also experience these emotions. It's important for patients and caregivers to understand those feelings and find strategies and resources to deal with them.

Call the National Suicide Prevention Lifeline at 800-273-8255 if you have thoughts of self-harm or suicide. If you need to speak with someone who will listen and understand, call PHA's Support Line at 800-748-7274.

## Are you a caregiver?

If you are a caregiver, you'll spend a lot of time taking care of the person with PH. You can begin to feel emotionally or physically exhausted, putting your own health at risk. It's important to prioritize your well-being, not just for yourself but also for your loved one with PH.

Make sure everyone knows the signs of burnout and takes time to care for themselves. Just like those who are newly diagnosed, caregivers must find time to care for themselves and focus on activities or techniques to reduce stress.

# Ways to Reduce Stress



## Find Support

Connecting with others who also have PH is a great way to get answers to specific questions or just come to terms with a new diagnosis. PHA has more than 200 support groups all around the U.S., a telephone support line, Facebook groups and individuals you can email for one-to-one support. Groups exist for certain types of PH, caregivers and families with children. Contact PHA at [Support@PHAssociation.org](mailto:Support@PHAssociation.org). Learn more in PHA Resources, Page 55.



## Take Care of Yourself

Whether you are a newly diagnosed individual or a caregiver, journaling, meditating, eating well and exercising can reduce stress and improve mental and physical health. For more ideas on coping, exercise, and day-to-day living, visit [PHAssociation.org/Living-With-PH](http://PHAssociation.org/Living-With-PH).



## Get Involved in PHA

For those who feel better when they're actively doing something, there are so many ways to get involved as a member of the PHA community. If you'd like to tell your story, you can submit a story for The Right Heart blog ([PHAssociation.org/TheRightHeart](http://PHAssociation.org/TheRightHeart)) or learn how to contact elected officials who will be voting on insurance and health care issues that affect the patient community. You can also organize or join special events that raise money to further the PHA mission.



## Seek Professional Help

Treating your mental health is as important as treating your physical health. Untreated depression, anxiety or both can affect your physical and emotional health and interfere with your ability to care for yourself or others. Consult your PH health care team or primary care provider about medication, counseling and other treatment options. Your health care team might refer you to a psychiatrist, social worker or other mental health professional.

PH can change the way you view yourself and your body, affecting your body image, dating and long-term relationships. To address these issues, order a free copy of PHA's "Living With PH: A Guide to Intimacy" at [PHAssociation.org/brochures/](http://PHAssociation.org/brochures/).

# Navigating Potential Problems

- Prepare for Medication Challenges 45
- Insurance and Financial Assistance 46
- Prepare for Emergencies 48
- Information for EMTs and Emergency Responders 49
- What If I Need More Help? 51

# Prepare for Medication Challenges

Your PH specialist will prescribe therapy targeted to your PH. If you encounter roadblocks like these, don't give up.

- **Insurance roadblocks.** Your health insurance plan might require prior approval before covering the cost of your medication. Or you might have to try a cheaper medicine before getting the one your physician prescribed. Or the plan could deny coverage for your PH therapy. Your health care team can help you navigate those obstacles to get treatment that works for you.

- **REMS requirements.** Some PH therapies cause birth defects or have toxicity risks. Some treatments might require enrollment in safety monitoring programs approved by the Food & Drug Administration, known as Risk Evaluation and Mitigation Strategies.

- **Delivery delays.** Some PH therapies are available from retail pharmacies, while others are delivered by mail via specialty pharmacies. Ask your health care team how your medication will be delivered. Specialty pharmacies offer nursing support and resources to help you adapt to your new normal, but mail-order medication might pose challenges. You can give feedback to your specialty pharmacy about billing or delivery challenges through a form on PHA's website.

- **Brand vs. generic therapy.** Whether you receive brand or generic therapy depends on many factors, including physician preference or where

you live. It's important to know that generic PH therapy isn't always substantially cheaper than brand therapy. PHA's website answers questions about brand vs. generic. For additional information, talk with your medical team.

- **Starting medication.** Some PH treatments require a nurse to visit your home and teach you to use your medication. Others require a brief hospital stay. Your health care team or specialty pharmacy will help you understand what to expect.

- **Delayed benefit and side effects.** Some PH medications must be taken for weeks or months before you feel the benefits. Others have significant side effects when you start or when your doctor increases your dose. But the side effects decrease over time. It's important to take your medications as instructed and communicate your concerns to your PH care team. Don't stop your PH medications without notifying your care team. PHA can connect you with other PH patients who can share their experiences with you and support you during this process.

## What can I do now?

Find more tools to address issues about managing PH. Download PHA's Empowered Patient Toolkit at [PHAssociation.org/patienttools/](https://PHAssociation.org/patienttools/).

## Organization tips

Managing medication dosages, specialty pharmacy information and insurance plans can make PH time-consuming. These tools can help you plan and stay organized:

- Track symptoms, medications, weight, mood, food and fluid intake, vitals and oxygen saturation.

- Keep important medical information in one place, such as a binder or a flash drive, so you can access it when you need it.

- Keep contact information available for your PH team, specialty pharmacy, insurance and other important members of your care team.

# Insurance and Financial Assistance

Now that you've been diagnosed with PH, your existing insurance coverage might no longer cover all of your medical needs. Some people change plans after diagnosis, such as a shift from employer-provided coverage to Medicare. Regardless of your situation, you deserve prompt, affordable access to the treatment prescribed by your medical team. PHA is here to help.

## Insurance Words to Know

### Coinsurance

An amount you pay out of pocket for covered medical care. The amount is a percentage of the total cost.

### Copay

A fixed amount you pay out of pocket for covered medical care.

### Deductible

A threshold you must reach before your insurance plan pays for medical care or prescriptions. For example, you might pay \$1,500, \$5,000 or another amount annually for services or medication before your coverage kicks in, depending on your plan.

### Durable medical equipment

Medical equipment that can be used repeatedly and is not disposable: such as wheelchairs, pumps and oxygen equipment.

### Formulary

An approved list of prescription drugs covered under a specific insurance plan.

### Medicaid

Government health insurance program for eligible, low-income people. Eligibility varies by state.

### Medicare

A federal health insurance program primarily for people 65 and older. If you qualify for social security disability assistance, you also might qualify for Medicare after a 24-month waiting period regardless of your age. Medicare coverage is broken into several parts:

- Part A covers hospitalization and inpatient care.
- Part B covers outpatient care including doctor's visits and health screenings. It also covers durable medical equipment, such as oxygen, scooters, wheelchairs, nebulizers and pumps.
- Part C, i.e., Medicare Advantage, combines parts A and B coverage into one plan. Some Advantage plans include prescription drug coverage. Medicare recipients can choose Medicare Advantage coverage instead of Medicare Parts A and B
- Part D covers prescription drugs.

### Premium

A monthly payment made by an employer or individual to buy insurance.

### Step therapy

Insurance might require you to "try and fail" a specific, less expensive medication before your plan will approve the originally prescribed medication.

## Financial Assistance



PHA lists financial assistance resources from a variety of sources at [PHAssociation.org/HELP](https://www.phassociation.org/HELP).

I have spoken with my medical team about the following resources to help pay for my medication:

- Nonprofit assistance funds
- Drug manufacturer assistance programs
- Specialty pharmacy
- Social Security Disability assistance and other government programs

## Finding Help

People who might be able to help if you're having problems with your insurance:

- Your PH care team.
- Your PH treatment center's billing department.
- Friends or family members.
- PHA: Call 301-565-3004, ext. 749.
- Constituent services staff in the offices of your elected officials.

Visit [PHAssociation.org/insurance](https://www.phassociation.org/insurance) for more information.

# Prepare for Emergencies

Emergency preparedness is critical for people with PH and their caregivers. Work with your support network to create an emergency plan that reflects the severity of your PH, other medical conditions, your location, support systems and other personal considerations. Then discuss and share your emergency plan with your PH care team.

Here are a few tips to get started:

- Stock up on necessities, especially drinking water, food, first aid kits, flashlights and batteries.
- Keep a supply of extra distilled water for inhaled therapy devices or CPAP machines.
- Ask your PH care team if you can get extra medication and/or supplies (cassettes, syringes, etc.). Some insurance companies will allow early refills for emergencies or disasters.
- If you use oxygen, ask your oxygen company to have your tanks filled so you don't run short. Also ask about extra batteries.
- Keep a list of all your medications with you at all times. Infusion patients should state that your infused medication should never be interrupted or discontinued.
- Keep with you a list of numbers for your specialty pharmacy, PH doctor/clinic and emergency contacts.
- Consider investing in a generator, and learn how to use it. Power outages are dangerous for those on infusion medication and those who use oxygen.
- Contact your power company about your health condition so it lists you as high priority in a power outage. Work with your health provider to complete/submit paperwork to ensure your power stays on in an outage.
- If you have a central IV line (catheter), keep your cleaning and dressing supplies with you including gloves, mask, alcohol pads, dressings and tape.
- Be sure you have a safe place to go and a way to travel if you need to evacuate. If you are driving, obtain a full tank of gas as soon as you learn of an impending storm.
- Sign up for emergency alerts.
- Contact your specialty pharmacy if you're running low on medication, need to change your medication shipment address or have other therapy-access concerns.
- Keep your insurance cards with you when you leave home.
- If you are a member of a PHA support group, exchange numbers with the leader and other members to keep in touch during unexpected events.

## Emergency situations

If you experience any of these symptoms or situations, seek emergency help or dial 911.

- Loss of consciousness.
- Coughing up blood.
- Pump has stopped.
- You run out of medication for your pump.
- Dislodged line/damaged or non-functioning catheter.



# Information for Emergency Service Providers

Pulmonary arterial hypertension (PAH) is a rare disorder of the blood vessels in the lungs. As pulmonary arteries narrow, blood pressure in the arteries rises above normal limits. This causes strain on the right side of the heart and can be life threatening.

As with any population, PAH patients vary greatly. Some might not look sick, while others need wheelchairs, oxygen or medicine continuously delivered by a pump (see below).

- Listen to the patient.
- Don't administer nitroglycerin for chest pain.
- Never remove or stop an infusion line.

Patients might be on one or more medications to treat PH. Bring the medications listed below to the hospital with the patient:

- PH medication list
- PAH medication
- Dosage/Infusion rates
- Ambrisentan (Letairis) oral
- Bosentan (Tracleer) oral
- Tadalafil/Alyq (Adcirca) oral
- (Orenitram) oral
- Selexipag (Uptravi) oral
- Macitentan (Opsumit) oral
- Sildenafil (Revatio) oral
- Riociguat (Adempas) oral
- Treprostinil (Tyvaso) inhaled
- Iloprost (Ventavis) inhaled
- Treprostinil (Remodulin) subcutaneous or intravenous\*
- Epoprostenol (Flolan and Veletri) intravenous\*



## \*About PH Medication

IV epoprostenol (also known as Flolan and Veletri) are given by continuous infusion through a central venous catheter and pump. The half-life of these medications is three to five minutes.

IV Remodulin is delivered by continuous infusion through a central venous catheter and pump. The half-life of Remodulin is about four hours, although symptoms such as shortness of breath can occur in less time.

Subcutaneous Remodulin is delivered through continuous infusion with a CADD MS-3 pump. The infusion site may be reddened. This is normal due to vasodilation.

Carefully tear out along the perforated edge and post on your refrigerator or download a copy from [PHAssociation.org/emergency](http://PHAssociation.org/emergency).

If a patient is on intravenous Flolan, Veletri or Remodulin, and the catheter is dislodged or damaged, or if the pump stops working, it is an emergency.

## When Treating Patients on Flolan, Veletri, or Remodulin:

- Don't turn off the pump. It could be fatal.
- Involve a PAH specialist as soon as possible.
- If there is a problem with the line or pump, start an IV in the patient's arm. When the IV line is in place, attach the pump tubing directly to the IV, and make sure the pump is running. Pump tubing should be connected to as short as possible IV tubing.
- Don't prime or flush the IV line. Don't give a large volume (bolus) of IV fluids. It can worsen heart failure.
- Don't infuse other medication into the PH medication line. Use a second peripheral IV for additional medications or IV fluids.
- Don't change the infusion pump without first talking to the patient's PAH specialist (unless patient is capable).
- Don't draw blood from the IV.
- Note: Fever and/or drainage from the catheter can indicate a sudden, serious line infection onset.

Allergies or known drug reactions:

---

---

---

Normal oxygen saturation range for this patient:

---

Oxygen needs and tank settings:

---

---

---

Additional medications:

---

---

---

# What If I Need More Help?

If your symptoms or side effects are becoming a significant burden you might benefit from palliative care.

Palliative care is goal-directed therapy appropriate at any stage of chronic disease that focuses on relieving symptoms, stress and suffering related to illness. Palliative care is designed to improve a patient's quality of life.

A palliative care specialist will work with your PH doctor to improve your symptoms and quality of life. Palliative care can ensure your goals and what's most important to you are part of regular conversations with your PH care team. Palliative

care specialists may ask you about:

- Your symptoms and other medications' side effects.
- Communication about your disease with your family.
- Psychosocial and spiritual issues.
- Your wishes, plans and worries for the future.

Ask your doctor whether a palliative care referral is appropriate for you.



Palliative care isn't the same as hospice, which provides end-of-life care. Palliative care is designed to improve a patient's quality of life. It is appropriate at any stage of a chronic illness.

# Expanding Your Knowledge

Participate in Research **53**

PHA Resources **55**

Glossary **57**

# Participate in Research

Scientists and clinicians need patients and caregivers as partners in research studies and clinical trials. No one knows pulmonary hypertension (PH) like those who live with it every day. You can provide insight about PH that researchers and physicians do not have. Every treatment that exists today is the direct result of individuals who have stepped up to volunteer for research.

Not all research tests a new treatment. Some studies look at the safety or efficacy of a new drug or therapy, while others collect data and samples to better understand disease progression and the biology of PH. To prepare yourself for research:

- Read about PH research and options at [PHAssociation.org/Research/ClinicalTrials](http://PHAssociation.org/Research/ClinicalTrials).
- Participate in the Pulmonary Hypertension Association Registry (see PHA Resources).
- Visit the PHA Clinical Trial Finder at [PHAssociation.org/ClinicalTrials](http://PHAssociation.org/ClinicalTrials) to find a PH study.
- Participate in a research study in the PHA Research Room at PHA International PH Conference and Scientific Sessions.
- Ask your doctor for the trial protocols.
- Talk with your PH health care team about research opportunities. The following list of questions will help you get started:

How could this trial help me?

---

What are the potential short-term and long-term side effects of this treatment?

---

---

How do the possible risks and benefits of this clinical trial compare with my standard of care? How do they compare with other potential experimental treatment options?

---

---

---

Who has reviewed and approved this study for protecting participants rights and safety?

---

What steps will be put in place to monitor my safety?

---

---

How will I know if the treatment is working?

---

If the treatment works for me, can I keep using it after the trial ends?

---

What will I have to do if I join this trial?

---

Will I need to come to the hospital or clinic for specific tests or treatments? If yes, how often?

---

How will this study affect my daily life?

---

Where will I receive my medical care?

---

Who will be managing my care? Will I be able to see my own PH care team?

---

Can anyone find out whether I am participating in this clinical trial?

---

Can I talk to other people who are participating in this trial?

---

Will I receive any follow-up care after the study has ended?

---

Do the researchers/clinicians have any financial or special interests in this clinical trial?

---

Who do I contact if I have questions during the trial?

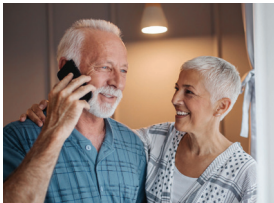
---

For more questions to ask your doctor about clinical trials, visit [PNAAssociation.org/researchquestions/](https://www.pna.org/researchquestions/).

# PHA Resources

Like many people who are recently diagnosed with PH — or are close to someone newly diagnosed — you probably never met anyone else with the disease. It is a huge relief to make connections, share experiences and understand that you're not alone. Other people who have gone through similar journeys and understand what you're going through. And they want to help.

The Pulmonary Hypertension Association (PHA) has a robust support network to ensure no one affected by PH ever feels alone. Patients and loved ones can turn to PHA and others living with PH for support, hope and resources to help them adjust to life with PH. Here are some resources:



## Support

- Support groups are meetings where patients, families, health care providers and friends come together to learn about PH. The meetings, whether in-person or virtual, can relieve the isolation that naturally arises from life with a rare disease. Find a support group meeting on PHA's website at [PHAssociation.org/support](http://PHAssociation.org/support).
- The PHA Support Line offers support calls for patients, caregivers, young adults and parents of children with PH. The calls, facilitated by PHA volunteers, allow you to ask questions and share experiences with people who have similar challenges. Call 800-748-7274.
- Email Phriends: Connect with an email mentor for one-on-one support based on your topic of interest, type of PH and more.
- PHA administers private Facebook groups for patients of various ages and with associated conditions, as well as, family members and caregivers.



## Membership

- PHA was established by people just like you: patients and their families who wanted to connect with others and change the future of PH. A membership in PHA allows you to join our community and support the search for a cure. Your membership also supports our educational programs, services and resources and contributes to a better quality of life for everyone affected by PH. It also includes a subscription to our quarterly print magazine, *Pathlight*, filled with inspiring stories written by and about patients.
- By becoming a member, you increase PHA's clout when reaching out to elected officials, the media and anyone who wants to help advance our cause. Your simple act of joining or renewing your membership makes a big difference.



## PHA on the Road

PHA offers regional education and networking events for people with PH and their families. They feature presentations and interactive sessions with PH-treating health care professionals. PHA on the Road provides opportunities to learn how to manage your health and meet others in the local PH community. The events are free.



## PHA International PH Conference and Scientific Sessions

The Pulmonary Hypertension Association's biennial Conference brings together more than 1,400 people affected by pulmonary hypertension, including patients, caregivers and medical professionals. Conference offers opportunities to learn about advancements in PH research and treatment, participate in clinical trials and connect with others in the PH community. Learn more at [PHAssociation.org/pha2022](http://PHAssociation.org/pha2022).



## Pulmonary Hypertension Association Registry (PHAR)

The PHAR collects patient information and results of diagnostic tests at participating Pulmonary Hypertension Care Centers (PHCCs). PHAR is open to pediatric patients and individuals with pulmonary arterial hypertension and CTEPH. For more information, visit [PHAssociation.org/PHAR](http://PHAssociation.org/PHAR).

### Learn more

Visit [PHAssociation.org](http://PHAssociation.org) for more information about PH, support services and guidance to manage your health.

Sign up for PHA's free e-newsletter, PHA News, to learn about the latest research, educational programs and activities, and support and fundraising events, at [PHAssociation.org/PHANews](http://PHAssociation.org/PHANews).

Contact PHA at 301-565-3004 or [Support@PHAssociation.org](mailto:Support@PHAssociation.org).

Become a member of PHA to receive our quarterly print magazine, Pathlight, and discounted registration to the PHA Conference.



# Glossary

## Arteries

Blood vessels that carry blood away from the heart.

## Cardiac output

The amount of blood the heart pumps per minute.

## Cardiopulmonary

Related to the heart and lungs.

## Chronic

Persisting for a long time or constantly recurring.

## Chronic thromboembolic pulmonary hypertension (CTEPH)

A form of pulmonary hypertension that is secondary to chronic blood clots in the lungs.

## Diastolic

Blood pressure measurement that refers to when the heart is relaxing and refilling with blood.

## Edema

Swelling caused by fluid in the body's tissues. It typically occurs in the feet, ankles and legs but can occur elsewhere in the body, such as the abdomen.

## Functional classifications for PH

A scale that defines the severity of a patient's PH based on how symptoms affect the patient's daily activities.

## Hypertension

Abnormally high blood pressure.

## Hypoxia

Low oxygen levels.

## Idiopathic

A medical term that means a disease has no known underlying cause.

## Immunology

Related to the body's immunity.

## Intravenous

Literally "inside the vein." Used to administer fluids or medication via a catheter or needle placed into the vein. An intravenous administration is called an infusion.

## Left atrium

One of the four chambers of the heart. Oxygenated blood flows from the veins in the lungs into the left atrium and then is pumped into the left ventricle.

## Left ventricle

One of the four chambers of the heart. Oxygenated blood is pumped from the left atrium into the left ventricle and then is pumped throughout the body.

## mmHg

An abbreviation for "millimeters of mercury," a standard measurement for atmospheric pressure used to measure blood pressure.

## Palpitations

Fast or irregular heartbeats that can be felt.

## Pulmonary

Related to the lungs.

## Pulmonary artery

The blood vessels carrying blood from the right ventricle of the heart to the lungs, where blood is oxygenated. It starts with the main pulmonary artery, then branches into the right and left pulmonary arteries feeding the right and left lungs, then continues to branch into smaller and smaller blood vessels, eventually becoming capillaries.

## Pulmonary artery pressure

A measurement of the blood pressure in the pulmonary artery measured in millimeters of mercury (mmHg). Normal pulmonary artery pressure is between 8-20 mmHg at rest. Pulmonary hypertension occurs when the pressure in your pulmonary artery is greater than 20 mmHg at rest or 30 mmHg when active.

## Pulmonary arterial hypertension (PAH)

Pulmonary arterial hypertension (PAH) is more specific than the general term "pulmonary hypertension." PAH is high blood pressure in the pulmonary arteries from disease of medium and small vessels in the lungs, which are narrowed and impede blood flow.

## Pulmonary hypertension (PH)

Pulmonary hypertension is a general term used to describe high pressure in the pulmonary arteries from any cause.

## Pulmonary thromboendarterectomy (PTE)

A surgical procedure to remove blood clots in the pulmonary arteries.

## Right atrium

One of the four chambers of the heart. Deoxygenated blood enters the right atrium from the body and then is pumped into the right ventricle.

## Right ventricle

One of the four chambers of the heart. Deoxygenated blood is pumped from the right atrium into the RV and then into the arteries of the lungs.

## Subcutaneous

Beneath or under all layers of skin. Some PAH medications are delivered subcutaneously.

## Systolic

Refers to blood pressure measurement when the heart is squeezing (contracting).

## Vasoreactivity

Ability of the blood vessels to react to a medication.

## Veins

Blood vessels that carry blood to the heart.

## Wedge pressure

Pressure measurement during right heart catheterization when the balloon-end of the catheter is "wedged" into a small branch of a pulmonary artery.

For a full list of terms, visit [PHAssociation.org/phterms/](https://www.phasociation.org/phterms/).

## Notes:

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

---

## Acknowledgments

Thanks to Murali Chakinala, M.D., Washington University School of Medicine; Anna Hemnes, M.D., Vanderbilt University; Martha S. Kingman, D.N.P., FNP-C, UT Southwestern Medical Center; Susie E. McDevitt, R.N., M.S.N., ACNP, University of Michigan; Ronald Oudiz, M.D., LA Biomedical Research Institute at Harbor-UCLA Medical

Center; and Jeffrey Robinson, M.D., The Oregon Clinic, for their review and expertise.

Additional thanks goes to patients Nicole Creech, Kim Crowell, Janessa Curnow, Quin Folkestad, Bill Kolm, Estella Medina, Linda Morgan, Nikole Nichols and Wendy Griggs.

## About the Pulmonary Hypertension Association

Headquartered in Silver Spring, Md., the Pulmonary Hypertension Association (PHA) is the country's oldest and largest nonprofit patient association dedicated to the pulmonary hypertension (PH) community. PHA's mission is to extend and improve the lives of those affected by PH. To achieve this mission, PHA engages people with PH and their families, caregivers, health care providers, and researchers worldwide

who work together to advocate for the PH community, provide support to patients, caregivers and families, offer up-to-date education and information on PH, improve quality patient care, and fund and promote research. For information, visit [PHAssociation.org](http://PHAssociation.org).



**PHA's mission** is to extend and improve the lives of all those affected by pulmonary hypertension.

8401 Colesville Road, Suite 200  
Silver Spring, MD 20910

Phone: 301-565-3004

[PHA@PHAssociation.org](mailto:PHA@PHAssociation.org)  
[PHAssociation.org](http://PHAssociation.org)

Copyright © 2021 Pulmonary  
Hypertension Association

