In 1996, the CHEST Foundation was founded as CHEST’s charitable foundation to serve CHEST members, their patients, and the public by supporting clinical research, patient education, and community service. From our early focus on tobacco prevention in young girls and women, to our new disease awareness campaigns that reach millions, we’ve had a significant impact in the last 20 years. More than 95 cents of every dollar raised goes toward advancing our mission-based programming. Since our inception, we’ve provided more than $10 million in funding for clinical research and community service, with a reach that spans more than 60 countries. It is with sincere appreciation that we acknowledge our tireless volunteers and the committed donors who have worked with us over the years to champion lung health.

Additional Resources

Content does not replace professional medical care and physician advice, which should always be sought. Medical treatments vary based on individual facts and circumstances.

The information provided herein is not intended to be medical advice.

The CHEST Foundation specifically disclaims all responsibility for any liability, loss, or risk, personal or otherwise, which is incurred as a consequence, directly or indirectly, of the use and application of any of the material herein.

Authors

Vijay P. Balasubramanian, MD, FCCP
UCSF - Fresno Medical Education and Research

Wassim H. Fares, MD, FCCP
Yale University

Deborah Jo Levine, MD, MS, FCCP
UT Health Science Center at San Antonio

Sandeep Sahay, MD, FCCP
Houston Methodist Hospital

Victor J. Test, MD, FCCP
Duke University School of Medicine
# Table of Contents

What Is PAH? .......................................................... 3
How Does PAH Affect Your Body? ................................. 6
Symptoms ................................................................. 9
Causes ................................................................. 10
Risk Factors ......................................................... 10
Diagnosing PAH ...................................................... 11
How Is PAH Treated? ................................................ 15
  Medications
  Using Oxygen
  Lung Transplantation and PAH
Living With PAH ..................................................... 20
  Exercise and PAH
  Depression and Anxiety
  Good Nutrition
  Travel and PAH
Managing PAH ....................................................... 25
Finding Support ..................................................... 26
Questions to Ask Your Doctor About PAH ....................... 27
Key Statistics

There are about 50,000 to 100,000 people affected by PAH in the U.S.

The average age of patients with PAH is 36 years old but it can affect both younger and older patients.

PAH strikes women twice as often as it does men.

Roughly 200,000 hospitalizations happen each year in the U.S. because of PAH.

Each year about 10 to 15 people per million are diagnosed with PAH.
What Is PAH?

_Pulmonary arterial hypertension_ (PAH) is a rare and serious disease that happens when there is high blood pressure in the lungs. PAH is caused when the small blood vessels in your lungs become narrowed and scarred. PAH is one of five types of _pulmonary hypertension_ (PH). PAH can be hard to diagnose and treat unless you work closely with your doctor and medical team.

**Key Facts**

- While PAH is caused by a variety of diseases, different causes have their own, specific best treatment options.
- Because the symptoms of PAH are similar to other diseases, such as shortness of breath, it can be difficult to diagnose.
- PAH will get worse without the right treatment.
- In most cases, PAH is treatable, but not curable.
To understand how PAH affects the body, you first need to understand the way lungs contribute to the normal flow of blood. The heart is divided into two sides: the left and the right. Each side has an *atrium* (a receiving chamber) and a *ventricle* (a pumping chamber). Each side is responsible for pumping blood, but for different reasons. The left side of the heart pumps blood through *arteries* to supply oxygen and vital nutrients to cells and tissues throughout the body. Because the blood has to go a long distance from the heart, it needs a lot of pressure. When you get your *blood pressure* checked, it is measuring the pressure that pushes the blood throughout the body.
After the blood delivers the oxygen, it returns through veins on the right side of the heart and is pumped back to the lungs. The pump in the right side of the heart doesn’t need a lot of pressure since the heart and lung are so close together. This is called pulmonary pressure and it is measured differently from the blood pressure in the left side of the heart.

When the pumping pressure is too high in the left side of the heart, it is called hypertension or high blood pressure. Similarly, when the pumping pressure is too high in the right side of the heart, it is called pulmonary hypertension (PH).

PAH (pulmonary arterial hypertension) is one form of PH. In this case, high blood pressure in the right side of the heart is caused from blood vessels in the lungs that have thickened and narrowed. Because of this narrowing of blood vessels, the right side of the heart has to pump harder – meaning use more pressure – to get blood to pass through. It also leads to a back-up of blood returning from the veins to the heart. As a result, the right side of the heart works too hard to keep up with the blood flow, which can weaken the heart and, over time, make it fail. Additionally, it takes longer for the blood to be cleansed through the lungs and pick up oxygen, which can lead to a lower-than-normal blood oxygen level. Oxygen is vital not only to cells and tissue, it is a critical component for healthy brain function. That’s why diagnosing and treating PAH is so critical.
Measuring Blood Pressure

You’re probably familiar with getting your blood pressure checked with a blood pressure cuff. This measures the pressure in the left side of the heart and is recorded in millimeters of mercury (mm Hg). There are two parts to this blood pressure measure. The first number reflects the systolic measure, which measures the pressure when your heart beats. The second number is the diastolic measure, which measures the pressure when your heart rests between beats. A normal blood pressure level is 120 (systolic) over 80 (diastolic).

In the right side of the heart, the pressure measures the blood vessels in the lungs, not the heart, and is called pulmonary artery pressure (PAP). Normal pulmonary artery pressure is 8 to 20 mm Hg at rest. If this measure is greater than 25 mm Hg at rest or 30 mm Hg during physical activity, it is considered high and qualifies as pulmonary hypertension.
Symptoms

It is hard to diagnose PAH because it’s symptoms can be similar to those of other diseases including:

- Asthma
- Emphysema
- Chronic obstructive pulmonary disease (COPD)
- Obesity
- Heart failure

Common symptoms for PAH include:

- Shortness of breath, first noticeable with exercise, but as the disease progresses, even during rest
- Feeling tired (fatigue)
- Swelling in the feet, legs, belly or neck
- Chest pain
- Heart pounding (palpitations)
- Fainting or dizziness

In rare cases, patients may cough up blood or have a change in their voices.
Causes

Some of the most common diseases or conditions associated with PAH are:

- Connective tissue and/or autoimmune diseases, such as scleroderma, lupus, or rheumatoid arthritis
- Cirrhosis of the liver
- HIV infection
- Methamphetamine and cocaine use
- Congenital heart defects
- Rarely, an inherited disease

If no other disease are associated with PAH, it is referred to as idiopathic pulmonary arterial hypertension (IPAH).

Risk Factors

Risk factors for PAH include:

- Obesity
- Illegal drug use
- Family history of pulmonary hypertension
- Taking some appetite-suppressant medications
- Living in high altitudes
- Sleep apnea
If your doctor suspects you have PAH, you will require several tests to prove first, that you have PH and specifically PAH and second, what type it is so that you can be treated appropriately. These tests help your doctor examine and directly measure the blood pressure in your lungs.
Tests commonly used to diagnose PAH are:

**Blood tests**: Your doctor will likely test for potential diseases that are associated with PAH as well as other signals of PAH. These include an HIV test; thyroid tests; autoimmune disease panels to test for systemic lupus erythematosus and scleroderma; liver tests; as well as standard tests like a complete blood count (CBC) and chemistry tests. Your doctor may also include a test to measure a hormone called *brain natriuretic peptide* (BNP) that helps evaluate the amount of stress on the heart.

**Chest X-ray**: A chest X-ray gives your doctor a picture of your heart, lung, and chest to look for signs of pulmonary hypertension.

**CT scan**: A CT scan provides your doctor with a more detailed, visual picture of your lungs, blood vessels and heart.

**Electrocardiogram**: This noninvasive test shows the electrical activity of the heart and can detect abnormal heart rhythms. It is a useful test to identify different causes for your symptoms associated with PAH.
Pulmonary function tests: These simple breathing tests measure how much air you can hold in your lungs and how much air moves in and out of your lungs. Pulmonary function tests may signal other lung diseases, like asthma or pulmonary fibrosis, which often causes scarring that can narrow lung blood vessels.

Exercise tolerance test (also known as the 6-minute walk test): This test is used to compare your exercise capacity, oxygen levels and symptoms over time and to evaluate how these characteristics change over time and with therapy.

Cardiopulmonary exercise testing (CPET or CPX): This test measures how well your heart and lungs are performing both at rest and during exercise. CPET helps your doctor understand the amount of oxygen your body is using, the amount of carbon dioxide your body is producing, and your breathing pattern. Results can help determine whether or not you may have PAH.

Ventilation-perfusion scan (VQ scan): This test examines air and blood flow to the lungs and creates images doctors use to look for blood clots in the lungs.
**Echocardiogram**: This noninvasive test – a type of ultrasound of the heart – looks at the chambers and valves of the heart, determining their size and function. An echocardiogram will detect some signs of PH in most patients with the disease, making it very practical. While it does not directly measure pulmonary arterial pressure (PAP), it does give doctors an idea if PH is present and how severe it is. Unfortunately, it cannot guarantee the diagnosis of PH by itself, which means further testing is likely.

**Cardiac MRI** (Magnetic Resonance Imaging): This is an MRI test that evaluates the heart size and function more accurately than the echocardiogram and gives a better picture of the heart muscles, valves, and blood work. It may also show any congenital heart disease (an abnormality in the heart that developed before birth).

A cardiac MRI also helps doctors identify and quantify any shunting of blood that may be happening between the right and left sides of the heart. Normally, the right and left sides of the heart function completely separately and no blood is exchanged between them. Sometimes PH or PAH can cause the blood to flow in a different pattern, which can impact how well the heart and lungs work.

**Right-sided heart catheterization**: Your doctor will likely recommend a right-sided heart catheterization because it is the clearest test for showing which of the five forms of PH you have. This test involves placing a small tube, known as a catheter, into a large vein in the neck, arm, or groin. This catheter is then threaded through the different chambers of the heart and lung to measure the pressure in each. How much blood the heart pumps each minute, known as cardiac output, is also measured. The amount of resistance to blood flow in the lungs, known as pulmonary vascular resistance, can be calculated from these measurements and are important indicators of how severe your PAH is.

During heart catheterization, it is likely that you will undergo a trial to assess if a medication is effective at dilating your blood vessels. There are two types of medications that can be tested:

- Medications delivered by breathing (Nitric oxide), or
- Medications given through an IV (prostacyclin or adenosine).

The results help guide your doctor’s choice of medications that can be used to manage your PH. Mostly the trial tells your doctor whether or not regular high blood pressure medications, known as calcium channel blockers, will be helpful for you.
How Is PAH Treated?

In most cases, PAH is treatable, but not curable. Treatment for PAH is very individualized and is designed to relieve symptoms and slow the progress of the disease. The exact course of your PAH treatment depends on you, the severity of your symptoms, the test findings, and your support system at home. Fortunately, the treatment of PAH has changed during the last 20 years. Several medications have been approved for PAH that don’t work for other forms of PH. In all likelihood, you will need to take more than one medication to get the best results. In addition to medications, some patients also need oxygen to make breathing easier.

Medical treatment is different for every patient and is usually adjusted to enable the patient to meet goals that are important to the patient and the physician. These goals, both from the lab and clinical standpoints, are often adjusted to the individual and often require discussion with the physician.

---

Stick to Your Treatment Plan

It is important that you follow all the instructions your doctor gives you for taking medications and making lifestyle improvements to keep you in the best health possible. Finding the right medications can take some trial and error, but you need to stay the course. Stopping the medications can be very dangerous and should only be done under your doctor’s supervision.
**Medications Used to Treat PAH**

PAH medications come in pill, inhaled, intravenous (through the veins), and subcutaneous (just below the skin) forms. The medications are often used in a variety of combinations. Following is a breakdown of medications for PAH and what each one does to reduce the symptoms of the disease.

### Oral Medications

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>NAME</th>
<th>WHAT IT DOES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inotrope</td>
<td>Digoxin</td>
<td>Helps strengthen pumping in the heart with a more regular rhythm to heartbeats</td>
</tr>
<tr>
<td>Blood thinners</td>
<td>Warfarin (Coumadin®)</td>
<td>Help prevent blood from clotting</td>
</tr>
<tr>
<td></td>
<td>Rivaroxaban (Xarelto®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Dabigatran (Pradaxa®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Apixaban (Eliquis®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Heparin</td>
<td></td>
</tr>
<tr>
<td>Thiazides diuretics</td>
<td>Chlorothiazide (Diuril®)</td>
<td>Helps the body get rid of excess fluid that put extra pressure on the heart</td>
</tr>
<tr>
<td></td>
<td>Chlorthalidone (Hygroton®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Indapamide (Lozol®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hydrochlorothiazide (Hydrodiuril®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Methyclothiazide (Enduron®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Metolazone (Zaroxolyn®, Diuret®, Mykrox®)</td>
<td></td>
</tr>
<tr>
<td>Loop diuretics</td>
<td>Bumetanide (Bumex®)</td>
<td>Helps the body get rid of excess fluid that put extra pressure on the heart</td>
</tr>
<tr>
<td></td>
<td>Furosemide (Lasix®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ethacrynate (Edecrin®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Torsemide (Demadex®)</td>
<td></td>
</tr>
<tr>
<td>Potassium sparing diuretics</td>
<td>Amiloride hydrochloride</td>
<td>Helps the body get rid of excess fluid that put extra pressure on the heart</td>
</tr>
<tr>
<td></td>
<td>Spironolactone (Aldactone®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Triamterene (Dyrenium®)</td>
<td></td>
</tr>
<tr>
<td>Endothelin receptor antagonists (ERAs)</td>
<td>Ambrisentan (Letairis®)</td>
<td>Block a hormone that causes scarring in the blood vessels of the lung and improve blood flow through the lung</td>
</tr>
<tr>
<td></td>
<td>Bosentan (Tracleer®)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Macitentan (Opsumit®)</td>
<td></td>
</tr>
<tr>
<td>Phosphodiesterase inhibitors (PDE5 inhibitors)</td>
<td>Sildenafil (Revatio™, Viagra®)</td>
<td>Cause the blood vessels in the lung to relax which may improve blood flow through the lungs</td>
</tr>
<tr>
<td></td>
<td>Tadalafil (Adcirca®, Cialis®)</td>
<td></td>
</tr>
</tbody>
</table>
### Oral Medications

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>NAME</th>
<th>WHAT IT DOES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calcium channel blockers</td>
<td>Amlodipine</td>
<td>A traditional blood pressure medicine that relaxes blood vessels but only works in a small number of patients with PAH</td>
</tr>
<tr>
<td></td>
<td>Nifedipine</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Diltiazem</td>
<td></td>
</tr>
<tr>
<td>Prostacyclin analogue</td>
<td>Oral treprostinil (Orenitram®)</td>
<td>Causes the blood vessels in the lung to relax and limits platelet clumping to improve blood flow through the lungs</td>
</tr>
<tr>
<td>Selective IP receptor agonists</td>
<td>Selexipag (Uptravi®)</td>
<td>Activates specific receptors in the blood vessels to cause the vessels to relax and limits platelet clumping to improve blood flow through the lungs</td>
</tr>
<tr>
<td>Soluble guanylate cyclase (sGC) stimulators</td>
<td>Riociguat (Adempas®)</td>
<td>Acts in two ways to cause the blood vessels of the lung to relax which may improve blood flow through the lungs</td>
</tr>
</tbody>
</table>

### Inhaled Medications

<table>
<thead>
<tr>
<th>NAME</th>
<th>WHAT IT DOES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iloprost (Ventavis®)</td>
<td>Helps relax and widen blood vessels in the lungs, reducing the pressure on the heart and improving blood flow</td>
</tr>
<tr>
<td>Inhaled treprostinil (Tyvaso®)</td>
<td>Helps the body dilate narrowed blood vessels in the lungs, relaxing and widening them for better blood flow and lower pulmonary blood pressure. It also helps limit blood platelets from clumping together</td>
</tr>
</tbody>
</table>

### Intravenous Treatments

<table>
<thead>
<tr>
<th>NAME</th>
<th>WHAT IT DOES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intravenous terprostinil (Remodulin®)</td>
<td>Causes the blood vessels in the lung to relax and limits platelet clumping to improve blood flow through the lungs</td>
</tr>
<tr>
<td>Epoprostenol (Flolan®, Veletri®)</td>
<td>Causes the blood vessels in the lung to relax and limits platelet clumping to improve blood flow through the lungs</td>
</tr>
</tbody>
</table>

### Subcutaneous Treatments

<table>
<thead>
<tr>
<th>NAME</th>
<th>WHAT IT DOES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Subcutaneous treprostinil (Remodulin®)</td>
<td>Causes the blood vessels in the lung to relax and limits platelet clumping to improve blood flow through the lungs</td>
</tr>
</tbody>
</table>
Using Oxygen

Some people with PAH may need to use extra oxygen to help them breathe better and improve their daily life. Oxygen is not addictive, so you don’t have to worry about how much you use. Oxygen can help:

- Make breathing easier
- Improve sleep, mood, and mental ability
- Your body work better during the day

There are three types of oxygen:

- **Compressed gas oxygen**, which comes in steel or aluminum containers in different sizes for home use and traveling
- **Liquid oxygen**, which changes liquid into oxygen that you can breathe
- **Oxygen concentrators**, electrical devices that remove nitrogen from the air, which makes oxygen more concentrated

Questions to ask If your doctor prescribes oxygen include:

- How do I make sure I receive the right amount of oxygen?
- When and how often should I use my oxygen container?
- Which type of oxygen equipment will be best for my lifestyle?
Lung Transplantation and PAH

Lung transplantation is a final treatment option for some patients with PAH when other medical therapy is no longer effective.

Lung transplantation may prolong survival and improve quality of life. However, it should be considered a last resort because there are other complications associated with lung transplantation, such as infections, kidney failure, rejection. Although long-term outcomes with lung transplantation are improving, they are still far from ideal. Therefore, it is often a very complex decision and needs to be made along with the family, caregivers, your PAH medical team, as well as a transplant team.

Traveling with Oxygen

Airlines have rules about using oxygen. You cannot travel on an airplane with an oxygen canister. You must use either an airline-supplied oxygen canister or an FAA-certified portable oxygen container. There may be other rules, so check with your airline and/or oxygen supplier at least 2 weeks before you plan to travel.
Living with PAH can be difficult to manage. You will need to be active in your care. It is important that you know as much about your disease and medications as possible and work with your doctor to find the best treatment plan for you.

Because PAH is a rare and complex disease, good care for PAH patients involves a close relationship with a PAH specialist as well as support from family and friends. Be sure you work with a clinical team with lots of experience treating patients with PH and PAH. They will understand the variety of diagnostic tests and the most current treatment advances that might improve your situation. They will also have access to medical facilities experienced in doing the more complex diagnostic procedures, like right heart catheterization.
If you have PAH, your life may change a lot. The treatments for PAH can range from pills taken one to three times a day to medicine that has to be inhaled several times a day to a pump that delivers medicine all the time. It is also hard to know how you will respond to a particular medicine. It depends how serious your PAH is and what caused it. Patients who are not treated will get worse and possibly die. Treatment helps improve your survival and your symptoms.

With PAH, you will need to see the doctor at least every 3 to 4 months at first. You may also receive regular tests, such as echocardiograms and 6-minute walk testing. Some doctors will want yearly right heart catheterizations to see if treatments are helping your PAH and heart function.

**Special Considerations**

**PAH and pregnancy**: Patients with PAH must not get pregnant. PAH is a risk for the mother and the baby. Also, many PAH medicines can be harmful to the baby as it grows.

**PAH associated with scleroderma**: Scleroderma is a chronic connective tissue disease. Patients with scleroderma-associated PAH are at higher risk for more rapid deterioration.

**PAH and surgery**: Patients with PAH are at a higher risk when having any type of surgery. Your doctor may recommend avoiding any elective surgery and possibly some other, more serious surgeries depending on the risks and benefits of the surgery and the potential risk of heart failure. If you do require a surgery, it is best to undergo surgery at a center where they specialize in patients with PAH.
Exercise and PAH

Patients with well-controlled PAH find that carefully structured exercise is helpful. However, for patients who don’t have their PAH under control and continue to experience a lot of symptoms, any exertion or physical activity may not be possible until treatment starts working.

*Pulmonary rehabilitation* is a very important part of treatment for PAH because it helps you reduce breathlessness and makes your body stronger.

This comprehensive, Medicare-funded outpatient program offers:

- Structured and monitored exercise training
- Nutrition advice
- Detailed explanation of your how to take your medications
- Techniques to reduce and control breathing problems
- Education about maintaining and improving body function
- Help to quit smoking
- Emotional and psychological support

Pulmonary rehabilitation improves the quality of life for many people with PAH. Be sure ask your doctor about pulmonary rehabilitation to keep you active in a healthy way.
Depression and Anxiety

Many people with PAH may feel depressed or anxious. This doesn’t mean that you are crazy or defective. Depression is the rule — not the exception — in patients with complex medical problems like PAH. It also can be hard to keep up with personal relationships and maintain your regular levels of activity when you are sick.

For some people, medication helps with depression, especially if the person wants to hurt themselves. Some treatments for depression and anxiety that don’t involve medicine are talk therapy, exercise, and meetings with a support group. You should never be afraid to talk to your doctors and other members of your care team about your feelings.

Your diagnosis of PAH can be difficult for the entire family. Try to remember that your family members may have feelings of depression and anxiety too. Support groups can help both you and your family.
Good Nutrition

Good nutrition – eating the right foods – can also help you better manage your PAH. Some people with CTEPH have trouble keeping weight on. When you lose weight, you lose muscle mass, which can weaken your strength and cause other physical limitations. Other people with CTEPH may be overweight, which adds strain to your heart and lungs. Talk to your doctor about good nutrition and a good weight for you to maintain.

Here are some helpful tips for maintaining a healthy weight:

- Eat several smaller meals throughout the day instead of three large ones.
- Drink plenty of fluids.
- Eat slowly and chew food thoroughly.
- Make sure you are on a low salt diet.

Travel and PAH

It can be hard to travel with PAH, which is why it’s important that you make plans in advance. Some things to think about include:

- Review your travel plans with your doctors to see if you need special supplies, medication, and/or extra oxygen. Even if you don’t usually need oxygen, you may need it if you are traveling to a high altitude.
- You need to know where to get medicine and oxygen on your trip.
- You should know in advance where you can receive care on your trip in case of emergency.
Managing PAH

Managing your PAH takes commitment and consistency. There are several challenges related to living with PAH:

- **Medication management:** Your medication schedule may be complicated and will likely require paying close attention each day and for every dose. Generally, once a medication is started, it won't be stopped. It is also very important that you make sure not to run out of any of your medications.

- **Avoiding strains:** Patients with PAH must avoid things that can put strain on the lungs and heart, such as stimulants (cocaine, methamphetamines), smoking, and pregnancy.

- **Reduce salt in your diet:** Since PAH can lead to a form of heart failure, you will need to stick to a low salt diet and limit your fluid intake.

- **Weight management:** One way to monitor fluid retention is to keep track of your weight. If your weight goes up, you need let your doctor know because you may need a change in your medications.

- **Exercise management:** Since the primary symptoms for PAH are fatigue and shortness of breath, you may need to develop strategies to manage the physical exertion associated with work, your personal life, and taking care of your home. A careful exercise program, especially a pulmonary rehabilitation program, will help you improve your abilities. Remember that while it is important to remain active as much as possible, it is also important to avoid very stressful exercise. Ask your doctor about which activities and levels are right for you.

- **Immunization:** In general, patients with PAH should get the flu and pneumonia vaccines.
There are several organizations and programs to help patients with pulmonary hypertension and pulmonary arterial hypertension as well as their family members and caregivers. The Pulmonary Hypertension Association (www.phassociation.org) helps patients find medical specialists and facilities with expertise in PAH and provides access to support groups.

Here are some additional websites that help patients with PH and PAH find support:

- Living PAH (http://www.livingpah.com/)
- Insights (http://www.insightsonpah.com/)
- Peer Network (http://peernetwork.net/#/Home)
Questions to Ask Your Doctor About PAH

Whether you are going to your first appointment with a primary care physician or a pulmonologist, take the time to prepare for a meaningful discussion about PH and PAH. Consider bringing a trusted friend or loved one to the appointment. This person may remember details of the visit that you may forget or misunderstand, especially related to explanations about your condition and the plans to proceed. Some people also find it useful to record their appointments with a cell phone voice recorder or audio recorder device.

Questions About Your Diagnosis

- Have we confirmed that I have PAH?
- What is the cause of my PAH?
- Are there any unique complicating factors?
- How advanced is my disease?
- How would you grade the severity of my PAH – mild, moderate or severe?
- Will I need a lung transplant?
- Where can I find reliable information about the disease and its treatment?
Questions About Your Treatment

- What are the goals of my treatment plan? What can I realistically accomplish?
- What are your recommendations for which medicines I should take? Why these medications? What are the pros and cons for each?
- Do I need to take blood thinners?
- How each medication is delivered (pill, inhaled, intravenous, and subcutaneous)?
- How frequently will I need to take each medicine?
- What are possible side effects from each medicine?
- Are there medications I should avoid?
- Do any of these medications cause problems with other medications that I currently take?
- What kind of monitoring is required, both for medications and the progress of my disease?
- Where can I turn to get more information about these medications?
- How do I get my medications and refills?
- What therapies do you recommend for me?
- What do you recommend I do for exercise?
- Would I benefit from pulmonary rehabilitation?
- Do I need oxygen therapy?
- How do I use oxygen therapy?
- What should I do if I have an emergency problem?

Questions About Your Lifestyle

- Will I be able to go back to work?
- Can I drive?
- How much daily exertion can I take?
- Can I do housework?
- Can I lift up my children?
- Is it safe for me to travel?
- What changes do you recommend to my diet and nutrition?
- Do I need to reduce salt in my diet? How do I do this?
- Are there patient support groups in this area? Which do you recommend for me?
- Are there support groups for my family and caregivers in this area? Which do you recommend?
- Can I take medication for depression and/or anxiety?

Prediagnosis Patient Checklist

- How many PH patients have you worked with?
- Who else is on your medical team and what experience have they got with PH?
- Which medical facilities do you use for diagnostic testing?
- Why do you think I may have PH?
- How do you recommend we make a clear diagnosis?
- Which tests do you recommend I undergo?
- Are any of these tests invasive? Which ones?
- Do you recommend a right heart catheterization?
- Will you get approval from my insurance carrier for any tests you recommend?
- How long will it take to make a diagnosis?
- What happens if I do have PH?
In 1996, the CHEST Foundation was founded as CHEST's charitable foundation to serve CHEST members, their patients, and the public by supporting clinical research, patient education, and community service. From our early focus on tobacco prevention in young girls and women, to our new disease awareness campaigns that reach millions, we've had a significant impact in the last 20 years. More than 95 cents of every dollar raised goes toward advancing our mission-based programming. Since our inception, we've provided more than $10 million in funding for clinical research and community service, with a reach that spans more than 60 countries. It is with sincere appreciation that we acknowledge our tireless volunteers and the committed donors who have worked with us over the years to champion lung health.

Additional Resources

Content does not replace professional medical care and physician advice, which should always be sought. Medical treatments vary based on individual facts and circumstances.

The information provided herein is not intended to be medical advice.

The CHEST Foundation specifically disclaims all responsibility for any liability, loss, or risk, personal or otherwise, which is incurred as a consequence, directly or indirectly, of the use and application of any of the material herein.

Authors

Vijay P. Balasubramanian, MD, FCCP
UCSF - Fresno Medical Education and Research

Wassim H. Fares, MD, FCCP
Yale University

Deborah Jo Levine, MD, MS, FCCP
UT Health Science Center at San Antonio

Sandeep Sahay, MD, FCCP
Houston Methodist Hospital

Victor J. Test, MD, FCCP
Duke University School of Medicine
Navigating PAH
Pulmonary Arterial Hypertension
PATIENT EDUCATION GUIDE

CHEST Foundation
American College of Chest Physicians
2595 Patriot Boulevard
Glenview, IL 60026

chestfoundation.org/pah

Other patient education guides available from the CHEST Foundation in print and on our website:
chestfoundation.org/patienteducation

phaware® global association is a 501(c)3 organization dedicated to raising global awareness and creating innovative technology for research. Our goal is to use technology to create a global footprint and address PH patients’ needs around the world.

Become #phaware. Follow us @phaware.

Learn more at www.phaware.global

Disclaimer:
The American College of Chest Physicians (“CHEST”) and its officers, regents, executive committee members, members, related entities, employees, representatives, and other agents (collectively, “CHEST Parties”) are not responsible in any capacity for, do not warrant and expressly disclaim all liability for, any content whatsoever in any CHEST publication or other product (in any medium) and the use or reliance on any such content, all such responsibility being solely that of the authors or the advertisers, as the case may be. By way of example, without limiting the foregoing, this disclaimer of liability applies to the accuracy, completeness, effectiveness, quality, appearance, ideas, or products, as the case may be, of or resulting from any statements, references, articles, positions, claimed diagnosis, claimed possible treatments, services, or advertising, express or implied, contained in any CHEST publication or other product. Furthermore, the content should not be considered medical advice and is not intended to replace consultation with a qualified medical professional. Under no circumstances, including negligence, shall any CHEST Parties be liable for any DIRECT, INDIRECT, INCIDENTAL, SPECIAL or CONSEQUENTIAL DAMAGES, or LOST PROFITS that result from any of the foregoing, regardless of legal theory and whether or not claimant was advised of the possibility of such damages.

The authors, editors, and publisher have exerted every effort to ensure that drug selection and dosage set forth in this text are in accordance with current recommendations and practice at the time of publication. However, in view of ongoing research, changes in government regulations, and the constant flow of information relating to drug therapy and drug reactions, the reader is urged to check the package insert for each drug for any change in indications and dosage and for added warnings and precautions. This is particularly important when the recommended agent is a new or an infrequently employed drug.

Some drugs and medical devices presented in this publication may have US Food and Drug Administration (FDA) clearance for limited use in restricted research settings. It is the responsibility of the health-care provider to ascertain the FDA status of each drug or device planned for use in his or her clinical practice.