

# Your **PAH** Guide

Navigating life with pulmonary arterial  
hypertension (PAH)



**PAH Initiative**

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# You

probably have a lot of questions . . .

We're here to help. How? With clear and accurate information about PAH so that you can better understand what's happening to you, why it's happening, and what some of your options are. Hopefully, this information will help you have good discussions with your healthcare provider, manage your PAH, and reach your treatment goals.



**Tip:** It helps to think long-term about PAH. The steps you take today may set you up for better outcomes in the future.

## Something doesn't seem right

### At first you may have barely noticed your symptoms.

You may have had trouble breathing, or maybe you felt tired and worn out quicker than you used to. Perhaps you felt chest pain or noticed swelling in your legs or ankles. Maybe you became dizzy or you fainted.

If you did notice your symptoms, you might have told yourself that you're just "out of shape" or you're "getting older."



**Tip:** Sometimes, people close to you will notice changes in your symptoms before you do.

Over time, your symptoms may have made it **harder for you to do everyday activities, like:**



Yard work



Shopping



Doing laundry



Walking up stairs



Walking the dog



Preparing a meal

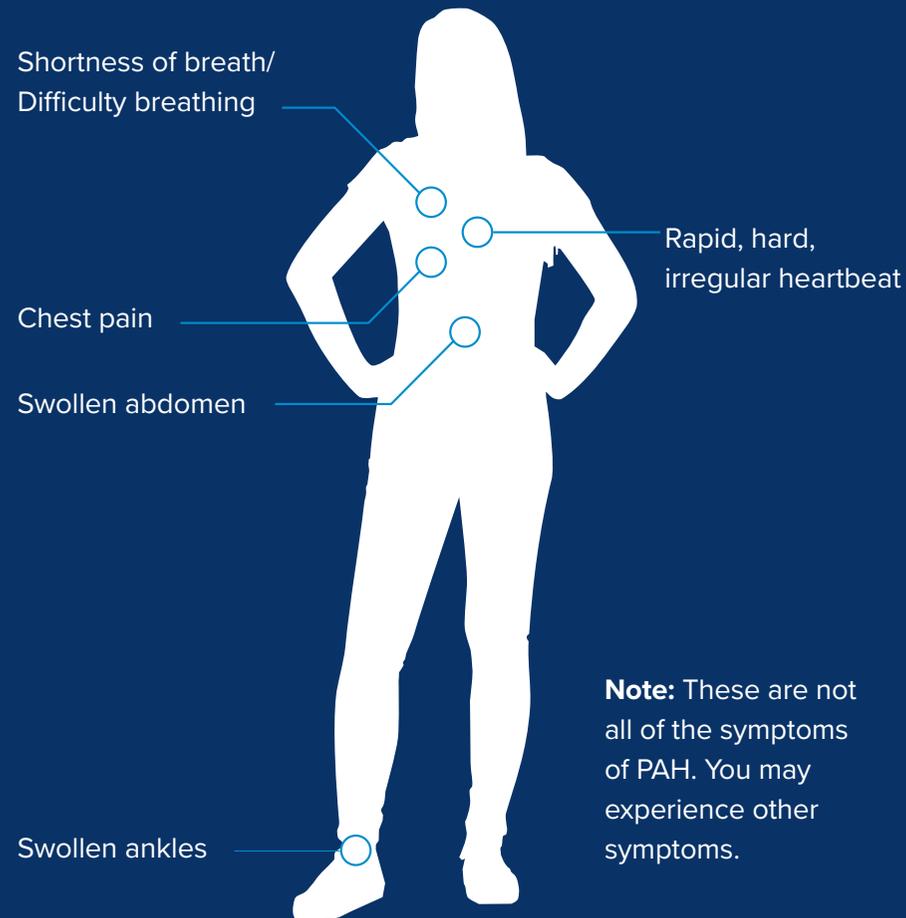
You might have questioned yourself and your own judgment, wondering, "What's happening to me?" Eventually, you learned the answer. **You were experiencing symptoms of PAH.**

## What are the symptoms of PAH?

You can expect your healthcare team to closely watch your symptoms. This helps them know whether your disease is improving, staying the same, or getting worse—and what to do about it.



**Tip:** It's important to find the right healthcare provider for you. PAH is a complex disease, and it requires the expertise of a PAH specialist.



**Note:** These are not all of the symptoms of PAH. You may experience other symptoms.

## The importance of tracking your symptoms

Lab work and tests are important. But your healthcare provider does not know what you do or how you feel each day. So, it's important to be open about all your symptoms.

Knowing your symptoms and how they change with daily activities can help your healthcare provider determine which treatments might be effective for you.

**One important step you can take is to track your symptoms and share them with your healthcare team so they can:**

- Figure out whether your condition is improving
- Manage your disease and find the right treatment plan



**Tip:** Keep a symptom journal on paper or on your computer, tablet, or phone. Be sure to bring it with you to every appointment. That way, you can share specific details about your symptoms with your healthcare provider.

## What can you do now?

You already may be on one or more medications for your PAH. Hopefully, you have seen some improvement.

But if you are still having symptoms, you still might find yourself

Participating in fewer activities

Spending less time with family and friends

“Sitting on the sidelines” and feeling isolated from life

Like some patients, you may wonder, “Why am I not feeling better?”

“Is there anything else I can do, or is this as good as it gets?”

These are important questions to discuss with your healthcare provider.

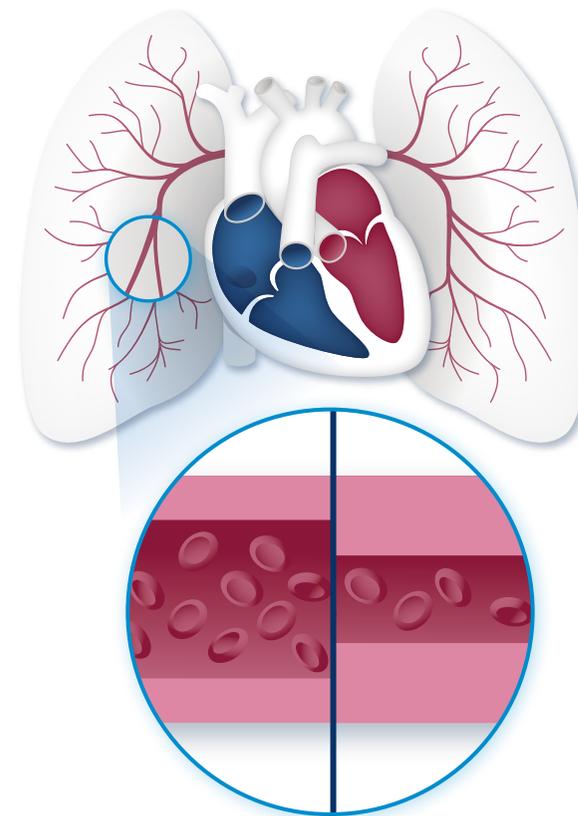
**In the next section, let’s look at what’s happening in your body and why you may still be having symptoms.**

## What’s happening in your heart and lungs?

Understanding how the heart and lungs work together will help you understand how PAH affects your body.

### Blood vessels in the lungs narrow

The heart and the lungs work together to deliver oxygen-rich blood throughout your body. PAH begins when the walls of the blood vessels in the lungs thicken and become narrower. This makes it harder for blood to flow through your lungs.



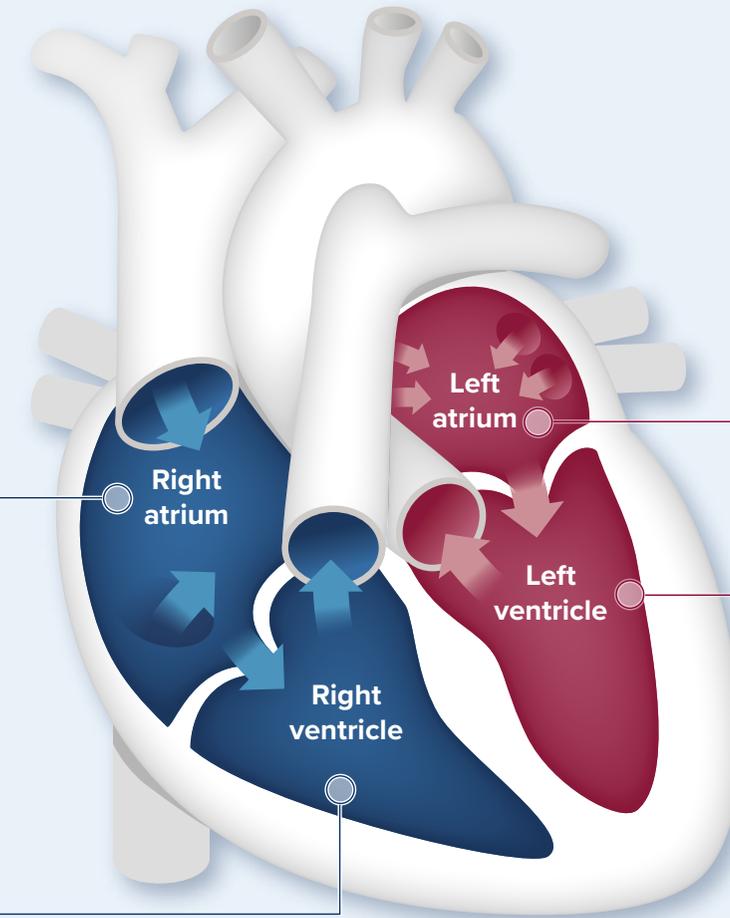
**Tip:** Learn more about how PAH affects the heart and lungs in the **PAH Basics** video, which you can find on the flash drive included with this booklet.

## How does the heart work?

The heart is divided into 2 sides, the left and the right. Each side has an atrium (that receives blood) and a ventricle (that pumps blood). So, both right and left ventricles pump blood, but for different reasons.

**The right atrium** receives oxygen-poor blood from the body.

**The right ventricle** pumps oxygen-poor blood to the lungs so it can pick up oxygen. It pumps blood only to the lungs, which are close to the heart. So, it's lower pressure on the ventricle.

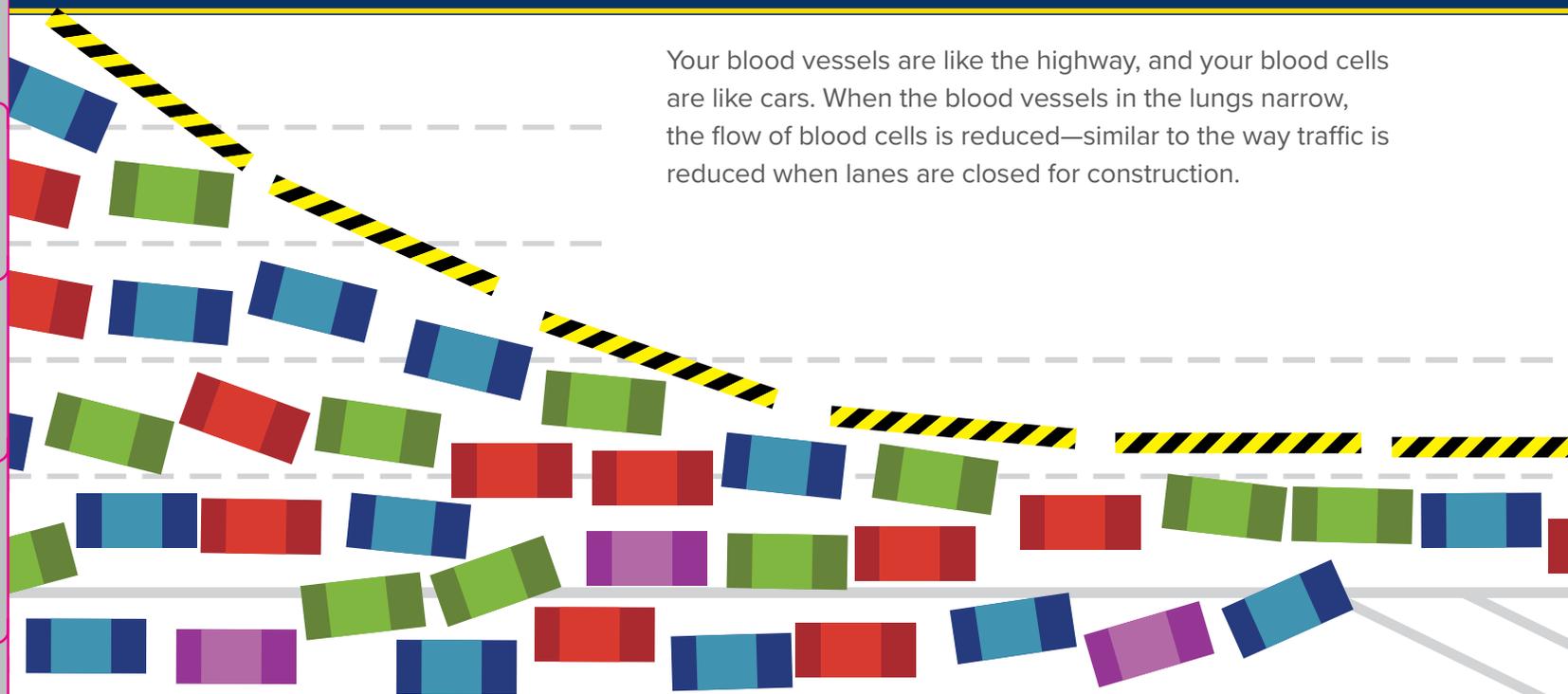


**The left atrium** receives red, oxygen-rich blood from the lungs.

**The left ventricle** pumps oxygen-rich blood to the cells and tissues throughout your body. It has a hard job because it pumps blood a long distance from the heart. So, it's higher pressure on the ventricle.

## Think of PAH like traffic on a highway under construction

Your blood vessels are like the highway, and your blood cells are like cars. When the blood vessels in the lungs narrow, the flow of blood cells is reduced—similar to the way traffic is reduced when lanes are closed for construction.



## Bringing it all together

So, the blood vessels in your lungs are narrower, which causes the “traffic backup.”

**As a result, the right side of your heart wears down because it works harder and harder to get blood to your lungs.**

The “traffic backup” of blood cells is the reason you feel symptoms like being lightheaded or short of breath—less blood and oxygen are reaching parts of your body where they are needed.

**To keep track of how you’re doing, your healthcare provider has you perform tests such as the 6-minute walk test. The information from this and other assessments helps establish your “risk status,” which is the focus of the next section.**

## What is risk status?

To treat you most effectively, your healthcare provider must understand your risk status.

### Risk status



Tells your healthcare provider how you're doing today—whether your disease is getting better, staying the same, or getting worse

# 5 YEARS

Predicts how your PAH may change over the next 5 years

## How is your risk status determined?

You are probably undergoing tests at every appointment. Listed below are just some of the tests that PAH specialists now use to understand your risk status and how it might be changing.



### The 6-minute walk test

Measures the distance you walk in 6 minutes



### Functional Class

Indicates the level of activity you can do without feeling symptoms



### Cardiopulmonary exercise test (CPET)

Assesses the performance of your heart and lungs while at rest and during exercise



### BNP (or NT-proBNP)

A blood test that indicates how much strain is on your heart



**Tip:** It's important for you to talk to your healthcare provider about your symptoms. The information you provide will help to accurately determine your Functional Class.



## 6-minute walk test

The 6-minute walk test gives clues about how your heart, lungs, and blood vessels are doing. It might not be much fun, but completing the 6-minute walk test at each visit provides an important view of your PAH over time.

### How far should you walk in this test?

A goal for many patients is about 440 meters (a little under 500 yards). What's important, though, is to improve *your* walk distance. Your results in this test may indicate whether your treatment is working effectively or if an adjustment may be needed.

Ask your healthcare provider what your goal should be for the 6-minute walk test.



**440 meters is equal to a little less than 5 football fields.**



**Tip:** The goal of the 6-minute walk test is to walk as *far* as you can, not as fast as you can. You should not run or jog.



## Functional Class

Functional Class has to do with physical activity and your symptoms. Your Functional Class is determined by information you give to your healthcare provider.

### Functional Class is divided into 4 groups



#### Class 1

Can you ride a bike or go for a long walk without getting breathless?



#### Class 2

Do you need to pause to catch your breath when you climb stairs or go shopping?



#### Class 3

Can you walk to the end of your driveway without stopping to rest?



#### Class 4

Do you experience symptoms of PAH even when resting?

**Your assessment results inform how you are doing today.**

Keep reading to learn how risk status predicts how your PAH may change over the next 5 years.

## Risk status is divided into 3 categories

Risk status is either low risk, intermediate risk, or high risk.

Low risk means that there's a low chance of your PAH getting worse over the next 5 years. If your risk level is intermediate or high, there is more of a chance that your disease will get worse. When PAH gets worse, it's called *disease progression*.



If your risk status is high or intermediate, your healthcare provider may consider changes in your treatment plan to help improve your status.

Remember, if your risk status is low today, chances are you'll be doing better down the road, too. For more information about risk status, watch the *PAH Treatment Goals* video on the attached flash drive.

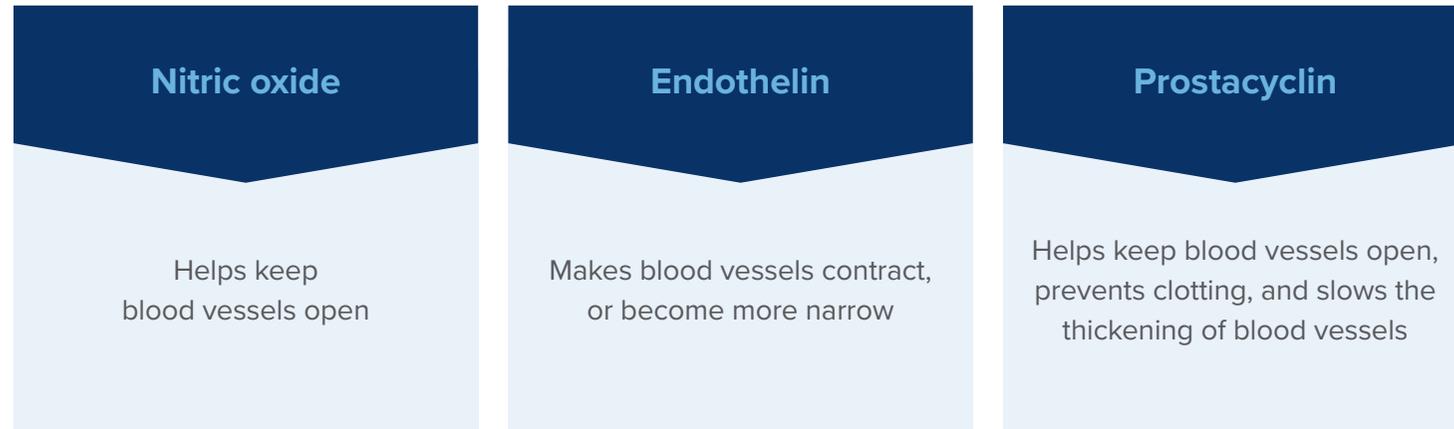
**Over time, your healthcare provider may adjust your treatment plan by adding therapies or changing medications, which is the focus of the next section.**



**Tip:** Changes in your 6-minute walk test or Functional Class help show how well your treatment plan is working—and whether adjustments are needed.

## Understanding the balance

The small blood vessels in your lungs produce  
3 natural substances that are important for healthy blood vessels.



**The right amount of each of these 3 substances keeps the blood vessels healthy.**

This balance allows red blood cells to flow freely through the lungs and carry oxygen to the rest of the body. When you have PAH, the amount of the 3 substances changes and they become imbalanced.

## PAH medications

PAH medications work to help correct the imbalance. Each medication works on 1 of the 3 substance “pathways.”

**So, if you take only one medication, you’re treating only one pathway.**

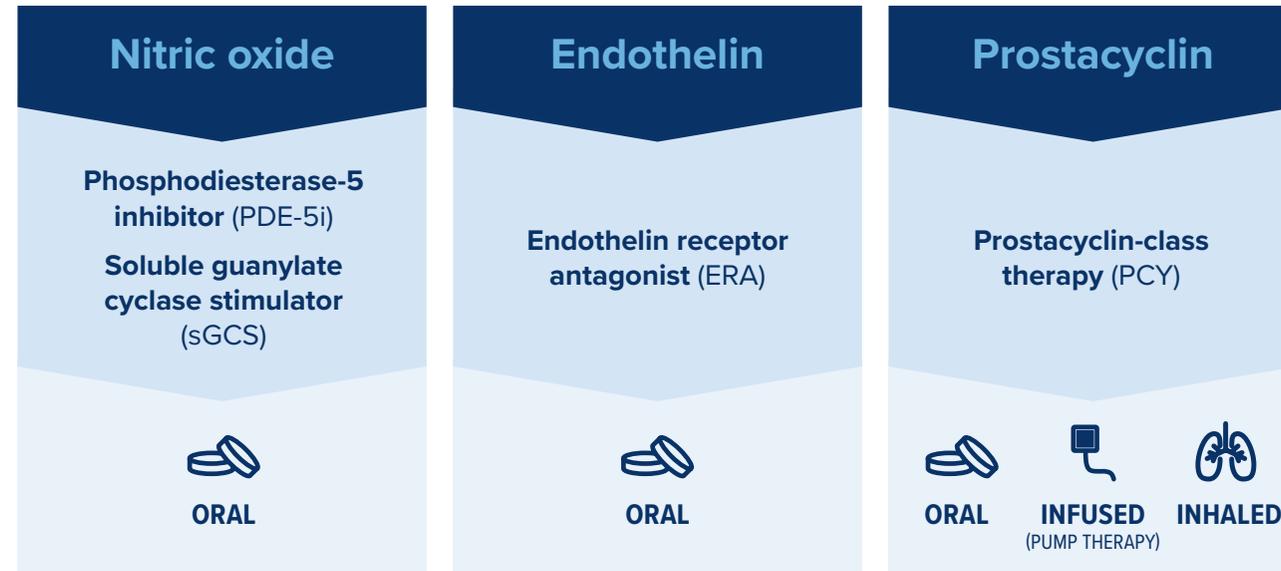
Currently there is no test available to determine which of the 3 substances needs to be adjusted. You could have more than one pathway out of balance. If you’re still experiencing symptoms, or you think you still have room for improvement, talk with your doctor about your options.

**Today, PAH patients typically are treated with more than one medication.**

Research shows that patients who receive more than one PAH medicine generally have better outcomes than patients who receive only one treatment. This holds true whether you are just starting treatment, or you’ve been taking PAH medicine for a while.

## Finding the right treatment plan

You and your healthcare provider will decide which PAH medicines will work best for you.



Treating PAH through only one pathway may not be enough for many patients.

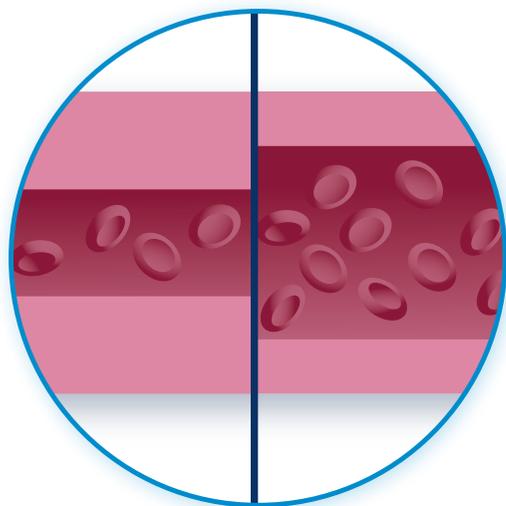
## Finding the right PAH specialist

Because PAH is a rare disease, not all heart and lung specialists in your community have expertise treating it. For this reason, **people living with PAH are encouraged to seek out a healthcare provider and treatment team that specializes in treating PAH.** These specialist providers are committed to treating PAH—they have had specific PAH training, keep up with the latest research, and have offices that support the special needs of people with PAH.

## Is now the time to ask about a prostacyclin-class medication?

### Your body makes prostacyclin naturally.

Prostacyclin is considered a “vasodilator,” which means it helps open your blood vessels. Prostacyclin-class medications mimic some of the effects of natural prostacyclin. If you recall, PAH begins when the walls of the blood vessels in your lungs become narrower. So, vasodilators help open your blood vessels to improve blood flow. If you’re not taking a prostacyclin-class medication already, your healthcare provider may consider one for you.



## How might prostacyclin-class medications help your PAH?

Prostacyclin-class medications have been shown to:

Increase the distance you can walk in the 6-minute walk test

Improve your PAH symptoms, like shortness of breath and fatigue

Reduce pressure in the blood vessels in the lungs

Improve your Functional Class

Keep PAH from getting worse

The benefits of prostacyclin-class medications could help improve your risk status. Find out more in the **PAH Treatment Options** video on the included flash drive.

Prostacyclin-class medications may cause side effects. Your healthcare team should be familiar with these side effects, and they can help you prepare for them. Be sure to ask about these and any other side effects that you might experience.

- Headache
- Diarrhea
- Flu-like symptoms
- Jaw pain
- Muscle spasm
- Flushing
- Nausea

## Things for you to think about

### It's important for your healthcare team to know how you are feeling.

They will also want a clear idea about the activities you struggle with, and the things you want to be able to do.

#### Some things to consider before your next appointment:

- ▶ How have you been feeling lately (since your last visit)?
- ▶ Are you able to finish your planned daily activities?
- ▶ Are there some everyday activities you are unable to do?
- ▶ Are you setting any new goals for yourself, like going shopping with friends?
- ▶ Do you find yourself thinking, "I feel good enough," but then wondering if you could feel better?

You can also think about goals—the goals you have for yourself and the goals your healthcare provider might have for you. To learn how goals can work hand in hand, check out the *PAH Treatment Goals* video on the included flash drive.



#### Examples of personal goals

Experiencing fewer symptoms, such as fatigue and shortness of breath

Spending less time in the hospital, or not being hospitalized at all

Being more active

Spending more time with friends and family



#### Examples of physician PAH treatment goals

Improving right ventricle heart function

Having healthier blood vessels in the lungs

Slowing disease progression

Achieving low-risk status



**Tip:** Talk about your personal goals with your healthcare provider. Consider making a list of your goals and see if they match your healthcare provider's goals.

## Questions to ask your healthcare provider

Make the most of your time with your healthcare providers by staying informed about your PAH and being prepared. To help you get the best care you can, consider discussing some of these questions:

- ▶ What is my risk status?
- ▶ Is low-risk status the right goal for me?
- ▶ Is my 6-minute walk distance improving?
- ▶ How can I do more activities with fewer symptoms?
- ▶ How can I be proactive in managing my PAH?
- ▶ Could prostacyclin-class medications help improve my symptoms?
- ▶ How will I know if my treatment plan is working?



**Remember, getting the most from your treatment today may help you make the most of your tomorrows.**

## Keep up with PAH

For more information and helpful resources,  
visit [www.PAHInitiative.com](http://www.PAHInitiative.com).

Come back often to see what's new.  
You can also sign up for updates and alerts.



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