PAH

GET TO KNOW PULMONARY ARTERIAL HYPERTENSION (PAH)

Your guide to understanding what PAH is, how it is diagnosed, and how approved treatments may help.

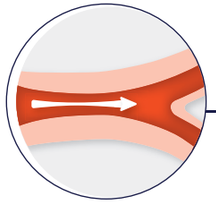
UNDERSTANDING PAH

Learning about your diagnosis and symptoms is important so you can receive effective treatment that may help improve your quality of life.

What is PAH?

PAH is a rare and progressive disease where there is high blood pressure in the arteries of your lungs and the right side of your heart.

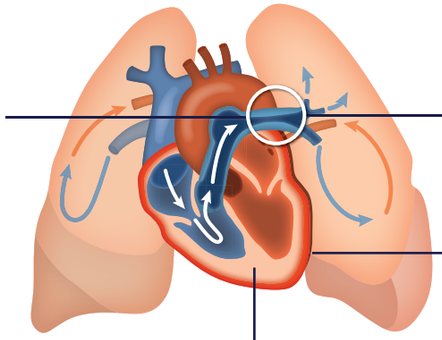
Normal pulmonary artery



Narrow pulmonary artery

1. In PAH, the pulmonary arteries become narrow and stiff, which means there's less space for blood to flow.

Pulmonary arteries
(blood vessels that carry blood from the right side of your heart to your lungs)



2. As a result, blood pressure in your pulmonary arteries goes up.

3. This causes your heart to work harder to pump blood through the lungs.

4. High blood pressure causes the right side of the heart to become enlarged and weakened.

What are the signs and symptoms of PAH?

PAH can lead to a wide range of symptoms that may develop slowly over time. Most people with PAH begin to notice symptoms as the condition progresses.

Symptoms of PAH include:

TIREDNESS

SHORTNESS OF BREATH

CHEST PAIN OR PRESSURE

FAINTING

DIZZINESS

RAPID HEARTBEAT

SWOLLEN ABDOMEN

SWOLLEN LEGS AND ANKLES

DIAGNOSING PAH

Your doctor may perform several tests to see what's happening inside your body:

- **Electrocardiogram (EKG or ECG)** to measure the electrical activity of your heart
- **Pulmonary function test (PFT)** to see how well your lungs are working
- **6-minute walk test (6MWT)** to see how far you can walk in 6 minutes
- **Echocardiogram (echo)** to check the size and function of your heart
- **Chest x-ray** and **ventilation perfusion scan (VQ scan)** to get images of your heart, lungs, and blood vessels
- **Right heart catheterization (RHC)** to measure the pressure inside your heart and the blood vessels of your lungs

How are the effects of PAH on everyday activities measured?

A functional class assessment can help your doctor understand how much PAH affects your everyday activities.

The functional classes are defined as follows:

NYHA Functional Class I	No limitations on physical activity. Ordinary physical activity doesn't cause shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class II	Some limitations on physical activity. Although comfortable at rest, ordinary activity causes shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class III	Clear limitations on physical activity. Although comfortable at rest, even less than ordinary activity causes shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class IV	Any level of physical activity is uncomfortable. There may be signs of heart failure, and shortness of breath and/or fatigue may be present even when resting.

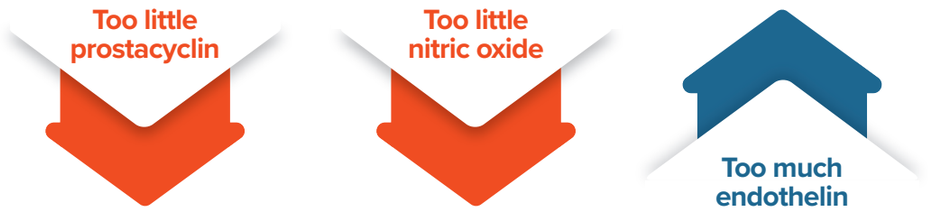
NYHA = New York Heart Association.

TREATING PAH

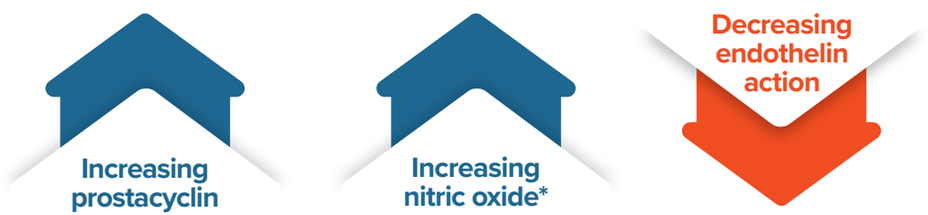
There are several types of therapy available to treat PAH.

Doctors treat PAH using medicines that aim to restore balance among 1 or more of 3 substances that are produced by your lungs: **prostacyclin**, **nitric oxide**, and **endothelin**. The right amount of each substance helps keep pulmonary arteries open and blood flowing through the lungs.

IN PAH, 1 OR MORE OF THE 3 SUBSTANCES ARE OUT OF BALANCE, CAUSING PULMONARY ARTERIES TO NARROW:



PAH THERAPIES HELP OPEN PULMONARY ARTERIES BY:



*Two types of therapies, called phosphodiesterase-5 inhibitors and soluble guanylate cyclase agonists, work in a similar way to nitric oxide to open blood vessels.

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WHAT IS PAH?

PAH is a rare and progressive disease where there is high blood pressure in the arteries of your lungs and the right side of your heart.

WHAT ARE THE RISKS?

High blood pressure from PAH causes the right side of the heart to become enlarged and weakened.

WHAT CAN I DO?

If you are diagnosed with PAH, there are treatment options that directly address the disease that could be right for you.

LEARN MORE ABOUT A TREATMENT THAT COULD BE RIGHT FOR YOU.

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