



## Glossary of Terms



**PAH Initiative**

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**Atrium (right and left):** The upper chambers of the heart. The atria receive blood from the veins, while the ventricles pump blood to the veins.

**Blood vessels:** Blood vessels are part of the circulatory system and function to transport blood throughout the body.

**Chronic:** Chronic means ongoing or occurring over an extended time. Chronic diseases generally cannot be prevented by vaccines or cured by medication, nor do they just disappear.

**Disease progression:** When a condition gets worse over time, it's called disease progression.

**Echocardiogram:** This test measures the size and shape of the heart. It also measures how well the heart squeezes and relaxes.

**Endothelin:** One of 3 natural substances produced by small blood vessels in the lungs that helps to keep blood vessels healthy. Endothelin helps contract blood vessels.

**High blood pressure:** Also known as “hypertension,” this is when blood pressure—the force of blood flowing through the blood vessels—is consistently too high. The primary way that high blood pressure causes harm is by increasing the workload of the heart and blood vessels, making them work harder and less efficiently.

**Nitric oxide:** One of 3 natural substances produced by small blood vessels in the lungs that helps to keep blood vessels healthy. Nitric oxide helps keep blood vessels open.

**NT-proBNP (or BNP blood test):** A test that measures the amount of a hormone found naturally in the body. The hormone is called B-type natriuretic peptide, or BNP. When the heart works harder than usual, it releases high levels of BNP. If this test shows high levels of BNP, it means the heart is straining and working harder than normal. The strain may be caused by narrow blood vessels in the lungs due to PAH.

**PDE-5 inhibitor:** A type of targeted therapy used to treat pulmonary hypertension and pulmonary arterial hypertension. PDE-5 is an enzyme found in the blood vessel walls. It helps control blood flow to the arteries in the lungs. PDE-5 inhibitors stop PDE-5 from working properly. By stopping PDE-5 from working, PDE-5 inhibitors cause the blood vessels to relax, which increases blood flow to the lungs and lowers blood pressure.

**Prognosis:** This is a medical term for predicting the likely course of a disease, including whether signs and symptoms will improve, remain stable, or worsen (and how quickly that may happen). Prognosis also can include the likelihood of survival.

**Prostacyclin:** One of 3 natural substances produced by small blood vessels in the lungs that helps to keep blood vessels healthy. Prostacyclin helps keep blood vessels open, prevents clotting, and slows the thickening of blood vessels

### **Pulmonary arterial hypertension (PAH):**

A condition where the blood vessels in the lungs become narrower. This causes the heart to work harder to pump blood to the lungs.

**Pulmonary hypertension:** A type of high blood pressure that affects arteries in the lungs and the heart. (Pulmonary arterial hypertension is a specific and more serious form of pulmonary hypertension.)

**Right heart catheterization:** A procedure that reveals how much blood the heart pumps through the lungs. In addition, it helps determine the severity of PAH. This is currently the only test that can diagnose PAH.

**Risk assessment:** Part of the standard guidelines for treating PAH. In a risk assessment, the healthcare provider looks at all test results and other factors to help predict how a patient will be doing over the next 5 years. Healthcare providers use the information from the risk assessment to determine risk status.



## Glossary of Terms (cont)



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**Risk status:** A designation that healthcare providers assign to patients with PAH based on the results of a risk assessment. Risk status can be low, intermediate, or high. Risk status enables healthcare providers to predict the probability of life expectancy over the next 3 to 5 years.

**Ventilation perfusion scan:** This 2-part test measures breathing and blood flow in the lungs. The test is used to determine if there are blood clots in the lungs, which can cause high blood pressure.

**Ventricles:** The lower 2 chambers of the heart. The ventricles receive blood from the atria and pump it to the lungs (right ventricle) or to the entire body (left ventricle).

**WHO Functional Class:** A system developed by the World Health Organization (WHO) to help classify PAH. The 4 Functional Classes are based on the severity of PAH.

**6-minute walk test:** A common test that evaluates how far a patient with PAH can walk in 6 minutes. Normal goals vary for each person, and risk status will affect each patient's goal. The low-risk goal is walking more than 440 meters in 6 minutes. The 6-minute walk test is the best predictor of how patients will be doing over the next 3 to 5 years.