Pulmonary Hypertension & HIV

Pulmonary Hypertension Association
Empowered by hope
Pulmonary hypertension (PH) is a complex and often misunderstood disease. The term PH simply means high blood pressure in the arteries of the lungs. In regular hypertension — also known as high blood pressure or systemic hypertension, which you can check with a blood pressure cuff — the pressure in the arteries of the entire body is higher than it should be. In PH, the blood vessels specifically in the lungs are affected. They can become stiff and narrow, and the right side of the heart must work harder to pump blood through them. There are five groups of PH based on different causes.

Each form of PH is different, so it is important for newly diagnosed patients to find a PH specialist who can accurately pinpoint what is causing their PH. They then can develop a treatment plan for that specific type as soon as possible after a confirmed diagnosis. Every individual with PH is different, and new research with the potential to improve the outlook for people living with this disease is conducted on a continual basis. Once in the care of a PH-treating health care team and on appropriate therapy, individuals with PH can live for many years.

*Normal mean pulmonary artery pressure is between 8-20 mmHg at rest.*
*Pulmonary hypertension is defined as a resting mean pulmonary artery pressure at or above 25 mmHg.*

PH, or high blood pressure in the lungs, frequently results from a narrowing of the small blood vessels in the lungs, which leads to a larger right side of the heart.
HIV-ASSOCIATED PULMONARY HYPERTENSION

Human immunodeficiency virus (HIV) is a chronic infection that can lead to acquired immunodeficiency syndrome (AIDS) if untreated. Unlike other viruses, the body can never completely get rid of HIV. It attacks cells in the body’s immune system — specifically T cells (a type of lymphocyte or white blood cell, also called CD4 cells) — that help the body fight infections. If untreated, the number of T cells in the body is reduced, making it more likely for HIV-positive individuals to develop infections and infection-related cancers.

Effective therapies, specifically antiretroviral therapy (ART), have extended and improved the lives of those with HIV, as well as reduced the risk of HIV-positive individuals passing the virus to others. Today, with early detection and treatment, individuals with HIV can live nearly as long as those without the virus.
HIV is a disease known to be associated with World Health Organization Group 1 PH (pulmonary arterial hypertension, or PAH). In the U.S., between 1 million and 1.5 million people age 13 and older are thought to be HIV positive, with approximately 35,000-40,000 new diagnoses annually.¹ As HIV treatments improve and patients live longer, more people with HIV are developing cardiovascular diseases, including PAH. Scientists continue to study the connection between these two diseases.

The first case of PAH in an HIV-infected individual was reported in 1987.² Scientists believe that approximately 0.5 percent (or 1 out of every 200) of people with HIV develop PAH.³,⁴ The cause of increased risk of people with HIV developing PAH is unknown, but some researchers think this could be due to the consequences of aging, HIV-related inflammation and the effects of antiretroviral drugs.⁵ HIV-positive individuals have roughly 2,500 times the risk of developing PAH than those without HIV. HIV-associated PAH (HIV-PAH) occurs more frequently in males compared to individuals with idiopathic pulmonary arterial hypertension (IPAH), in which the cause of the PAH is unknown.⁶

HIV-PAH patients have higher mortality than patients with IPAH; however, patients with HIV-PAH do improve with PAH medications. The symptoms of HIV-PAH are similar to those in other individuals with PAH, with shortness of breath being the most common.

DIAGNOSING HIV-PAH

Careful compliance with medical care is critical for the best possible long-term health in HIV-positive individuals, and any changes in their condition should be brought to the attention of their health care providers. Changes that may have to do with PH also should be discussed with a person’s PH and HIV health care teams and can include shortness of breath with activity, increased fatigue, swelling of the ankles, legs or abdomen, chest discomfort or pain, and light-headedness and fainting.

An echocardiogram is an ultrasound of the heart that provides a rough estimate of the pressure in the lungs and assesses the function of the right ventricle, in addition to providing information on whether the heart is squeezing and relaxing as expected. An echocardiogram is not accurate enough to make the diagnosis of HIV-PAH. Right heart catheterization remains the most accurate PH diagnostic test because it directly measures the pressures and flow in the heart and provides other information about the heart’s condition. HIV-positive individuals with shortness of breath should be screened for PAH by a PH expert with an echocardiogram. A PH expert may consider following up with a right heart catheterization if patients have otherwise unexplained shortness of breath or signs of right heart dysfunction, even if the pressure estimates on the echocardiogram appear to be normal.
TREATMENT OPTIONS FOR HIV-PAH

To date, there have been few studies done specifically in HIV patients who have PAH because the number of patients with HIV-PAH is small. Another problem has been that the long-term survival of HIV-PAH patients is lower than HIV-negative individuals who have PH. Therefore, PAH treatment recommendations for HIV-PAH must be taken from other PAH studies. If HIV-PAH is diagnosed, PAH-specific therapies can be carefully prescribed. However, HIV-PAH patients should identify a PH specialist for their diagnosis and treatment who will work closely with their HIV doctor, since there are drug interactions between certain HIV drugs and PH-targeted therapies that need to be carefully managed.
ABOUT THE PULMONARY HYPERTENSION ASSOCIATION

The Pulmonary Hypertension Association (PHA) is a community of individuals with PH, caregivers, families, medical professionals and researchers who work together to empower people battling the disease, while supporting research toward improved treatments. PHA was the first organization in the world dedicated to providing comprehensive PH patient and caregiver support, medical education, research and services that improve patients’ quality of life.

PHA facilitates more than 200 active support groups around the country and serves as a model for more than 85 international PH organizations. In addition to in-person support, PHA offers a telephone support line, an email mentorship program, educational patient programs and a variety of print and online publications. Every two years, PHA hosts its International PH Conference and Scientific Sessions, which brings together nearly 1,500 individuals with PH, caregivers and medical professionals to share information on the latest research, treatments and lifestyle coping tips.

PHA also has provided more than $8 million for PH research and offers continuing education programs for medical professionals through the PHA Online University and an in-person CME program.

PHA’s mission is to extend and improve the lives of those affected by PH. PHA’s vision is a world without PH, empowered by hope.

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