Pulmonary Arterial Hypertension (PAH)

What is pulmonary arterial hypertension?

Pulmonary hypertension (PH) refers to high blood pressure in the arteries of the lungs. Pulmonary arterial hypertension (PAH) is one form of PH in which the pulmonary arteries that carry blood from the heart to the lungs, where it picks up oxygen, constrict abnormally, forcing the heart to work faster and causing blood pressure within the lungs to rise. There are several types of PAH. It can occur in response to a variety of associated disorders and taking certain medicines. There also is a form with no known cause, called idiopathic pulmonary arterial hypertension or IPAH.[1]

PAH is progressive and life-threatening because the pressure in a patient's pulmonary arteries rises to dangerously high levels, putting a strain on the heart. At rest, blood pressure in a normal pulmonary artery is 15 mmHg (15 millimeters of mercury), and rises during exercise. In a person who has PAH, the average blood pressure in a pulmonary artery is more than 25 mmHg at rest and more than 30 mmHg during exercise.[2]

The most common symptoms of PAH include shortness of breath, excessive fatigue, dizziness, fainting, weakness, ankle swelling, chest pain and bluish lips, hands or feet.[3]

Certain factors appear to increase the chances of developing PAH. They include use of appetite suppressant drugs (especially fenfluramine and dexfenfluramine), chronic use of cocaine or amphetamines, HIV infection, liver disease and connective tissue diseases such as scleroderma or lupus erythematosus.[4]

Studies estimate that the use of certain appetite suppressant drugs increases the risk of getting PAH more than six times.[5] Fenfluramine and dexfenfluramine were taken off the market in September 1997 after being linked to heart valve damage.[6] Some other possible causes of PAH may include a genetic predisposition, immune system disease or chemical exposures.[7]

Want to know more about PAH? Please view the fact sheet at
www.lungusa.org/pahfactsheet

Who has pulmonary arterial hypertension?

PAH affects men and women of all ages and all ethnic and racial backgrounds. While the true incidence of PAH is unknown, it is a relatively rare disease, affecting 1 in 100,000 to 1,000,000 people.[8] PAH is likely to get worse during labor and delivery, resulting in a high maternal death rate. PAH also is found more often in people with a family history of pulmonary hypertension or sudden death.

IPAH most commonly occurs in women in their mid-30s.[9] The average age at diagnosis is 36 years old. About twice as many cases are reported in women as in men.[10]

Persistent pulmonary hypertension of the newborn (PPHN), another type of PAH, is a condition involving acute or sudden respiratory failure. It is seen more commonly in full-term infants who have underlying diseases such as respiratory distress, sepsis or lung hypoplasia (below normal size or immature). PPHN affects approximately 1 in 1,250 live-born full-term infants.[11]

What is the health impact of pulmonary arterial hypertension?

In 2004, 314 deaths were caused in the United States by IPAH; 241 were female and 73 were male.[12]

The prognosis for PAH patients is quite poor. Currently, approximately 50 percent of people diagnosed with PAH die within five years.[13] The average period of survival is only about three years for those who do not receive treatment. Prognosis is worse for patients who have heart failure, severe PAH or are over the age of 45 when diagnosed.[14] However, many patients report that some lifestyle changes allow them to go about many of their daily tasks.

Are you suffering from PAH? To find a PAH support group in your area, visit
www.phassociation.org/connect

Even with treatment, the pressure in the lungs caused by PAH will continue to worsen and cause the right ventricle or right side of the heart to fail. As the right ventricle gets larger, the patient can develop irregular heart rhythms, which can lead to sudden death. As their PAH progresses, patients get weaker and more easily fatigued, so that their quality of life is affected.

How is pulmonary arterial hypertension diagnosed and managed?

Diagnosis may be delayed for several years since initial symptoms of PAH may be subtle and it is difficult to detect PAH in a routine medical examination. Even when the disease has progressed, the symptoms may be confused with other conditions that affect the heart and the lungs. Additionally, there is no specific test for PAH. Health care providers looking for possible PAH may do a variety of tests, including chest x-ray, electrocardiogram, echocardiogram, stress test, spirometry and cardiac catheterization.[15]

PAH is treated with a number of drugs. None of the drugs can cure or halt PAH progression, but they may relieve symptoms and slow the disease. Some patients take diuretics, although caution should be used with these drugs. Anticoagulants or blood thinners also may be used to keep blood from clotting internally; warfarin is the recommended type. Another drug, digoxin, helps the heart beat more regularly and strongly and can be helpful for some PAH sufferers.[16]

Many of the drugs used to treat PAH are vasodilators, which help to reduce blood pressure in the lungs by enlarging the blood vessels and decreasing cell growth. These drugs include prostanoids, endothelin receptor antagonists and phosphodiesterase-5 inhibitors. Calcium channel blockers also are used, but only help a small number of PAH patients.[17]

Although some patients do well with medication, others may need and be able to receive a heart-lung transplant. It is the only true cure for the heart problems associated with PAH. As technology advances, these transplants are becoming more successful.

The diagnosis of persistent pulmonary hypertension of the newborn is usually made within 24 hours after birth. Therapy for PPHN can include 100 percent supplemental oxygen, assisted ventilation, surfactant, sedation, inhaled nitric oxide or an artificial heart-lung machine (extracorporeal membrane oxygenation or ECMO). Unfortunately, inhaled nitric oxide and ECMO are expensive and many newborns are born in facilities that do not have these options available.[18] Infants with PPHN treated with a drug called sildenafil were more likely to survive than those given a placebo, although more research is needed before this treatment can be recommended.[19]

Want to know more about treatment options for PAH? Please visit PAH/PPH Treatment Options at **http://www.uptodate.com/patients/index.html**. Search under "PAH".

What is new in pulmonary arterial hypertension research?

The National Heart, Lung and Blood Institute conducts new clinical and other research on PAH. These efforts hopefully will lead to a better understanding of the disease.

Researchers are seeking quicker ways to diagnose PAH. In the United States, PAH patients are being referred to health centers too late. Although survival rates have increased, they remain low.[20] A recent pilot study looked into different options for diagnosing PAH. A new non-invasive tool to measure the pulmonary artery distensibility (stiffness) using magnetic resonance imaging (MRI) may be useful.[21]

What is the American Lung Association doing about pulmonary arterial hypertension?

Currently, the American Lung Association is working with leading researchers to identify new interventions for PAH. Studies being conducted include *Role of Uric Acid in Primary Pulmonary Hypertension, Exploring Hereditary Basis for Developing Pulmonary Hypertension, Uncovering Enzyme's Role in Pulmonary Hypertension* and *Seeking Strategies to Stop Process That Leads to Pulmonary Hypertension*. These studies are aimed at better understanding PAH and identifying successful treatment strategies.

Thousands of advocates have joined with the American Lung Association to tell Congress that more needs to be done to fight PAH. Join us to win the battle against lung disease by visiting http://lungaction.org.