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MEDfacts

An Educational Health Series From National Jewish Health®

Pulmonary Hypertension

What is Pulmonary Hypertension?

Pulmonary hypertension (PH) is a condition in which high blood pressure exists within the vessels of the lungs. Normally, venous (low oxygen) blood returns from the body to the right side of the heart where it is pumped to lungs via the pulmonary arteries. Breathing brings oxygen to venous blood in the lungs, turning it into arterial (high oxygen) blood. Arterial blood returns to the left side of the heart through the pulmonary veins where it is pumped to the rest of the body.

Healthy pulmonary arteries of the lungs are elastic, expanding and contracting with each beat of the heart. In PH, these arteries stiffen and thicken. This leads to increased resistance to blood passing through the vessel thereby increasing pressure. Higher pulmonary pressure can lead to shortness of breath, low oxygen levels, chest pain, near-fainting/fainting, heart rhythm problems, and in its more advanced form, heart failure.

How is pulmonary hypertension classified?

PH used to be classified as primary (without obvious cause, or idiopathic) or secondary (occurring as a result of another disease). Although this terminology is still used, the revised World Health Organization classification system does away with these definitions. Instead PH is divided into 5 categories based upon mechanism of disease.

PH can occur by itself or, more commonly, with diseases of the lungs and heart.

- PH in the absence of other diseases is very rare and generally is idiopathic or familial in nature. This kind of PH is referred to as pulmonary arterial hypertension (PAH).
- PH is commonly associated with a variety of lung conditions characterized by low oxygen levels. These include emphysema, asthma, interstitial lung disease, chronic

pulmonary blood clots, or sleep apnea.

- When pulmonary hypertension arises from cardiac conditions such as heart failure, heart valve disease, or congenital heart disease, it is referred to a pulmonary venous hypertension.
- Other important disease states seen with PH include connective tissue diseases (scleroderma, lupus, rheumatoid arthritis), sarcoidosis, hyperthyroidism, liver disease, sickle cell disease, and bone marrow disorders.
- Amphetamine use such as meth or the diet drug Fen-Phen has been linked to the development of PH. Often times multiple causes of PH are present.

How is Pulmonary Hypertension (PH) Diagnosed?

The diagnosis of PH can be difficult and is often delayed until the disease has progressed. PH cannot be diagnosed non-invasively. An ultrasound of the heart (echocardiogram) can provide an estimate of the pressure in the heart. Only a procedure called a right heart catheterization (RHC) can directly measure blood pressure in the lungs and determine if PH is present. RHC can also be used to determine if PH is responsive to intravenous vasodilator medication. This is useful to determine whether chronic medication treatment (see below) would be helpful or not. Other tests are performed to screen for associated diseases, including blood tests, EKG, chest X-ray, pulmonary function testing, and a test for lung blood clots called a ventilation/perfusion scan (V/Q scan). A 6 minute walk time is often performed to assess a person's exercise capacity.

What is the Treatment?

Treatment of PH is directed towards improving symptoms, exercise capacity, and when possible, survival. The therapy used for PH depends upon its underlying cause. PH can improve with treatment of associated heart or lung disease. Important non-medication treatment includes: oxygen therapy, giving up smoking, removal of deleterious drugs and low level exercise . When PH develops into heart failure, diuretics, digitalis, and sodium restriction can be helpful. Because people with idiopathic PAH are at higher risk for pulmonary blood clots, the blood thinner warfarin is used to prevent blood clots.

People whose pulmonary blood pressure "responds" to intravenous vasodilator treatment during RHC are candidates for calcium channel blocker therapy (CCB). People with idiopathic, familial, or connective tissue associated PH who do not improve with CCB treatment or do not respond to vasdilator challenge during RHC are candidates for oral medications. These medications include: phosphodiesterase-5 (PDE-5) inhibitors (sildenafil, tadalafil) or endothelin receptor antagonists (ERA's) (bosentan, ambrisentan, sitaxsentan). PDE-5 inhibitors and ERA's can improve exercise capacity, and sometimes, slow the progression of the disease. It is unclear if these drugs provide benefit to people with PH associated with lung or heart disease.

Medications called prostanoids may improve symptoms, and sometimes, survival. Prostanoids can be given by nasal inhalation (iloprost), injection (treprostinil), or via IV infusion (epoprostenol). While the prostanoids can provide significant improvement, they have many side effects. They should be managed at PH centers experienced with

their use. Surgical removal of blood clots can benefit people with chronic thromboembolic pulmonary hypertension (CTEPH). Finally, lung or combined heart/lung transplantation is appropriate in a select group of people who have progressive disease and have failed traditional therapy.

What do we do at National Jewish Health?

We provide comprehensive cardiology evaluation and consultation and non-invasive cardiac testing. We evaluate and treat heart problems such as coronary artery disease, high blood pressure, high cholesterol, heart valve problems and heart failure. In addition to traditional heart problems, we offer expertise in many other focus areas, including evaluation of patients with shortness of breath with exercise, sarcoid of the heart, diastolic dysfunction and secondary pulmonary hypertension.

Why National Jewish Health?

At National Jewish Health, we treat the whole person, not just the disease. Our cardiology team works with healthcare providers from all areas of the medical center, including rehabilitation therapists, dietitians and clinical researchers.

Note: This information is provided to you as an educational service of **LUNG LINE® (1-800-222-LUNG)**. It is not meant to be a substitute for consulting with your own physician.

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