Primary Pulmonary Hypertension

What is primary pulmonary hypertension (PPH)?
Pulmonary hypertension is a lung disorder in which the blood pressure in the pulmonary artery rises far above normal levels.

History of the disease:
The first reported case of primary pulmonary hypertension occurred in 1891 with a published description of an autopsy that showed thickening of the deceased's pulmonary artery, but no indications of heart or lung disease. In 1951, when 39 cases were reported in the United States, the illness received its name. Between 1967 and 1973, an unexplained increase in primary pulmonary hypertension was reported in central Europe. The increase in the number of cases was eventually attributed to aminorex fumarate, an amphetamine-like drug introduced in Europe in 1965 to control appetite. When aminorex was removed from the market, the incidence of primary pulmonary hypertension went down to normal levels.

More recently, in the United States and France, primary pulmonary hypertension has been associated with the appetite suppressants fenfluramine and dexfenfluramine, which were taken off the market in 1997. In the US, there are an estimated 300 new cases diagnosed each year. It is most common in women between the ages of 21 and 40.

What are causes of pulmonary hypertension?
Exact causes of primary pulmonary hypertension remain unknown. Researchers believe the blood vessels are particularly sensitive to certain internal or external factors, and constrict, or narrow, when exposed to these factors. There may be a genetic factor, an immune system factor, or sensitivity to drugs or other chemicals.

What are the symptoms of pulmonary hypertension?
The following are the most common symptoms for pulmonary hypertension. However, each individual may experience symptoms differently. Symptoms may include:

- fatigue
- difficulty in breathing (dyspnea)
- dizziness
- fainting spells (syncope)
- swelling in the ankles or legs (edema)
- bluish lips and skin (cyanosis)
- chest pain (angina)
- racing pulse
- trouble getting enough air
- palpitations, strong throbbing sensations brought on by increased heart rate

More severe symptoms indicate a more advanced disease. In advanced stages, the patient:

- is able to perform minimal activities.
- has symptoms even when resting.
- may become bedridden if the disease becomes worse.
The symptoms of primary pulmonary hypertension may resemble other conditions or medical problems. Consult your physician for a diagnosis.

How is primary pulmonary hypertension diagnosed?
Pulmonary hypertension is rarely discovered in a routine medical examination, and in its later stages, the signs of the disease can be confused with other conditions affecting the heart and lungs. Pulmonary hypertension is a diagnosis of exclusion. Diagnostic procedures may include:

- **electrocardiogram (EKG or ECG)** - a test that records the electrical activity of the heart, shows abnormal rhythms (arrhythmias or dysrhythmias), and detects heart muscle damage.
- **echocardiogram (echo)** - a procedure that evaluates the structure and function of the heart by using sound waves recorded on an electronic sensor that produce a moving picture of the heart and heart valves.
- **pulmonary function tests** - diagnostic tests that help to measure the lungs’ ability to exchange oxygen and carbon dioxide appropriately. The tests are usually performed with special machines that the person must breathe into.
- **perfusion lung scan** - a nuclear medicine procedure that can detect a blood clot in the artery leading to the lung. This procedure can also assess the function of the lungs.
- **cardiac catheterization** - a procedure that evaluates blood flow to the heart muscle, blockage of coronary arteries, congenital heart defects, functioning of the heart valves, and other heart structures. A small catheter is advanced from a blood vessel in the groin or arm through the aorta to the heart.

Treatment for pulmonary hypertension:
Specific treatment will be determined by your physician based on:

- your age, overall health, and medical history
- extent of the disease
- your tolerance for specific medications, procedures, or therapies
- expectations for the course of the disease
- your opinion or preference

Treatment may include one or more of the following:

- **medications**, including:
  - **anticoagulants** - to decrease the tendency of the blood to clot and permit blood to flow more freely
  - **diuretics** - to decrease the amount of fluid in the body and reduce the amount of work the heart has to do
  - **drugs** - to help lower blood pressure in the lungs and improve the performance of the heart in many patients
  - **calcium channel blocking drugs** - to improve the heart's ability to pump blood
- **supplemental oxygen**
  Some patients also require supplemental oxygen delivered through nasal prongs or a mask if breathing becomes difficult.
- **lung transplantation**