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PRIMARY PULMONARY HYPERTENSION

What is primary pulmonary hypertension?

Primary, or unexplained, pulmonary hypertension (PPH) is a rare lung disorder in which the blood pressure in the pulmonary artery rises far above normal levels for no apparent reason. The pulmonary artery is the blood vessel carrying oxygen-poor blood from the right ventricle, one of the pumping chambers of the heart, to the lungs. In the lungs, the blood picks up oxygen and then flows to the left side of the heart, where it is pumped by the left ventricle to the rest of the body through the aorta.

Hypertension is the medical term for an abnormally high blood pressure. Normal mean pulmonary-artery pressure is approximately 14 mmHg at rest. In the PPH patient, the mean blood pressure in the pulmonary artery is greater than 25 mmHg at rest and 30 mmHg during exercise. This abnormally high pressure (pulmonary hypertension) is associated with changes in the small blood vessels in the lungs, resulting in an increased resistance to blood flowing through the vessels.

This increased resistance, in turn, places a strain on the right ventricle, which now has to work harder than usual against the resistance to move adequate amounts of blood through the lungs.

People at risk are women between 20 and 40 years old, although the disease can affect any sex and age. The incidence is 8 out of 100,000 people. It is rare in children. More common is pulmonary hypertension in infants with persistent fetal circulation.

What are the symptoms of primary pulmonary hypertension?

progressive shortness of breath with activity

hyperventilation

chest pain under the sternum

weakness

fatigue

fainting

lightheadedness

How is primary pulmonary hypertension diagnosed?

Your doctor will take your history and perform a physical exam. He/she will look

for enlargement of the veins in the neck, enlargement of the liver, and edema (fluid retention).

Certain diagnostic tests and/or imaging procedures can help diagnose primary pulmonary hypertension. They include:

ECG

chest X-ray

echocardiogram

cardiac catheterization

lung scan

pulmonary arteriogram

How is primary pulmonary hypertension treated?

There is no known cure. The goal of treatment is control of the symptoms. Some people respond to treatment with vasodilators. Other medications used to relieve symptoms include diuretics and calcium channel blockers. As the disease progresses, oxygen may be needed for shortness of breath. Heart-lung transplantation for suitable candidates with available donors has been successful.