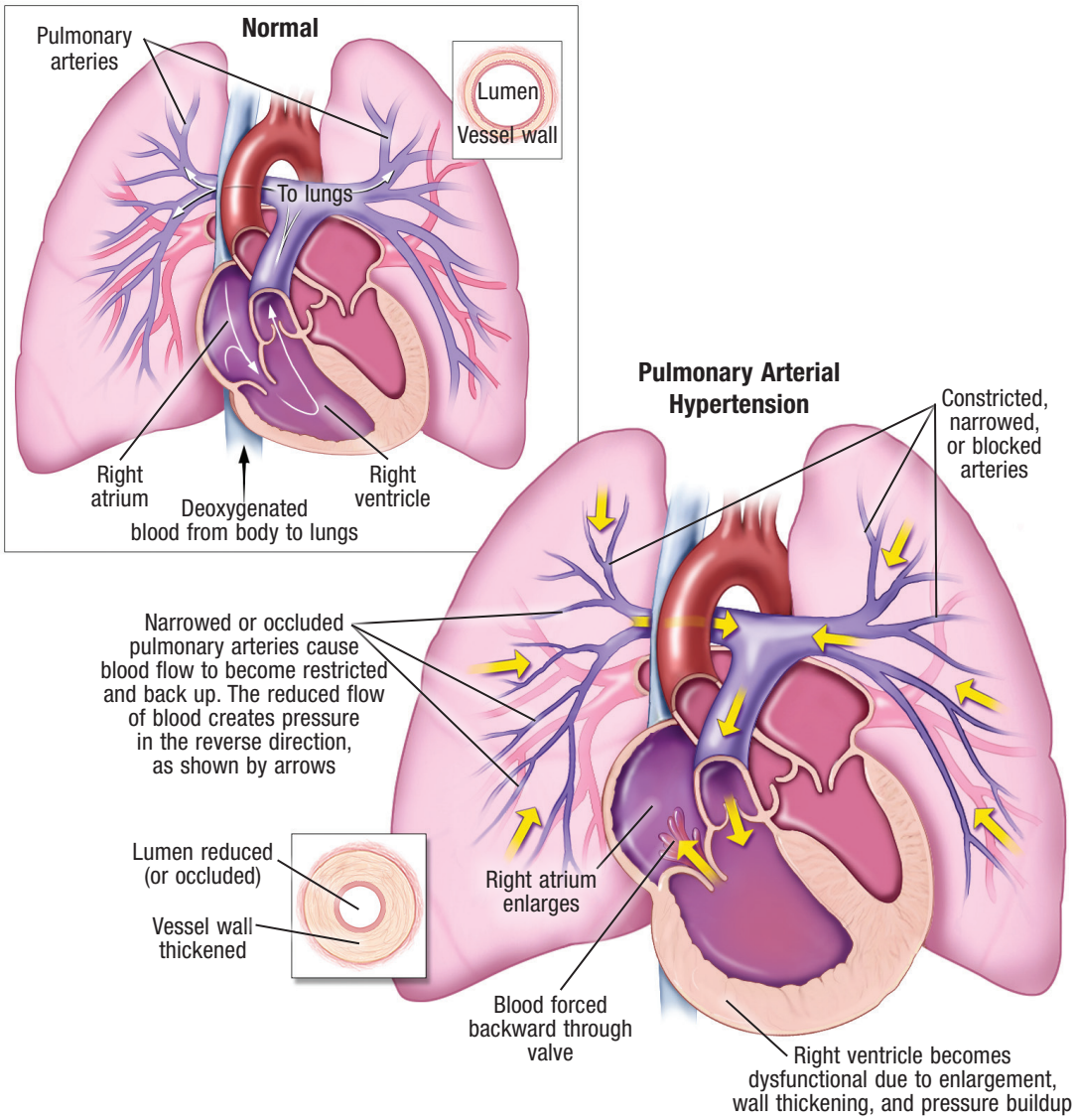
TEAR ALONG PERFORATION

Pulmonary Arterial Hypertension



High Blood Pressure in the Lungs

Pulmonary arterial hypertension, or PAH, occurs when the arteries in the lungs are narrowed, making it harder for the heart to pump blood through the lung circulation to become oxygenated. This results in damage to the heart as well as the lungs. PAH is a rare but serious condition that worsens over time. Although it can be genetically inherited, this condition is most often caused by other diseases or drug toxicity. The symptoms of PAH develop slowly and are similar to those of other heart or lung diseases, so specialized tests are needed to make the diagnosis.

PAH has no cure, but medications can help control symptoms, prevent complications, and improve the quality of life for patients. A healthy diet and exercise as tolerated are part of the treatment of PAH.

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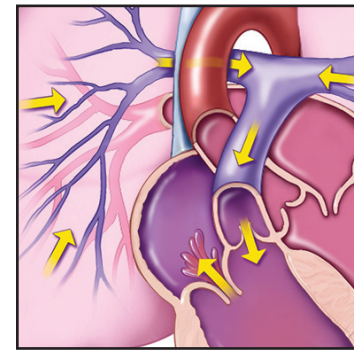
Symptoms May Be Similar to Asthma or Heart Disease



Narrowed or blocked pulmonary arteries cause blood flow to become restricted and back up.

Pulmonary arterial hypertension (PAH), or high blood pressure in the pulmonary arteries, is a rare disease that affects approximately 15 people per million population. Although PAH can occur at any age, it is most common in individuals aged 36 to 50 years.

Disease Development

PAH can develop for a number of reasons. The walls of the arteries may thicken, or they may constrict, making them more narrow. Small blood clots can form, blocking blood from flowing easily through the arteries. Regardless of the reason for the narrowing of the pulmonary arteries, the result is the same—the right ventricle must work harder to pump blood to the lung circulation. This leads to less effective oxygenation of the blood that passes through the lung tissue (causing shortness of breath) and enlargement of the right side of the heart (causing heart symptoms and fluid retention).

Symptoms

Symptoms of PAH develop slowly and are similar to those of other conditions, such as asthma or heart disease. The most common symptoms are shortness of breath, rapid heartbeat, dizziness, chest pain, and swollen feet and ankles. If left untreated, the condition may lead to right-sided heart failure and possibly death.

Diagnosing the Condition

Diagnosis is made using information from the patient's history and the physical examination. Some tests that may be ordered to confirm the diagnosis include chest x-ray, ECG, and echocardiogram. These tests often show the right side of the heart to be enlarged as a result of the increase in work required to pump blood into the narrowed pulmonary arteries. A right heart catheterization procedure measures the pressure in the right ventricle and pulmonary artery and determines how well the right heart is working. A lung perfusion scan shows the blood flow through the lungs. A pulmonary function test (PFT or breathing test) is used to measure lung function, and a skin sensor can reveal the amount of oxygen in the blood.

Treatment Options

Treatment of PAH is aimed at stopping the progression of the arterial narrowing and relieving symptoms by reducing the strain on the heart. Any disease causing PAH should be treated first. Activity levels are determined by the severity of symptoms. Oxygen supplementation can help relieve shortness of breath. When fluid retention is a symptom, diuretics (water pills) help remove excess fluid. Blood thinners can be used to prevent blood clots from forming in the pulmonary arteries.

Oral, inhaled, and IV drugs may be used to open the blood vessels in the lungs, reducing the strain on the heart. Digoxin is useful in some patients to strengthen a weakened heart. Surgical procedures such as a lung transplant or heart-lung transplant have also been successful in treating severe PAH.