Understanding Pulmonary Artery Hypertension

Pulmonary hypertension is a condition in which the blood pressure in the arteries of the lungs is unusually high. In a healthy person, the blood pressure in the arteries of the lungs is usually lower than the pressure of the blood in the rest of the body because the blood is pumped from the weaker right side of the heart (a shorter distance from the heart) into the lungs. By contrast, the stronger left side of the heart has to pump blood forcefully enough to circulate oxygen-rich blood through the entire body. While blood pressure in the body is normally about 120/80 millimeters (mm) of mercury (Hg), in the arteries of the lungs it is only 25/15 mm Hg.

When blood pressure rises in the arteries of the lung, damage can occur over time to the large and small arteries in the lungs. The walls of the blood vessels thicken and are less able to transfer oxygen and carbon dioxide to and from the blood. When less oxygen gets into the blood, pulmonary arteries become narrower. A spiral of increasing pressure on the lung's arteries begins.

Over time, damage to the heart occurs. The muscles of the lower right chamber (the right ventricle) become thicker and larger (a condition called cor pulmonale) and less able to pump efficiently. Heart failure can develop.

In some people, the body tries to compensate for the lower oxygen it is getting by producing more red blood cells in the bone marrow. The extra red blood cells make the blood thicker, stickier and even harder for the heart to pump. The extra cells also increase the risk of a person developing a pulmonary embolism because the thicker blood clots more easily.

There are often no symptoms until cor pulmonale is quite advanced. Then the symptoms are still those of pulmonary hypertension: shortness of breath upon exertion, lightheadedness, fatigue and chest pain. Symptoms of heart failure, such as swelling (edema) in the legs and progressively worse shortness of breath, develop.

Symptoms

The most common symptom that virtually everyone who develops pulmonary hypertension has is shortness of breath when they are active. Other symptoms include:

- Lightheadedness
- Tiring easily
- Chest pain like angina

If there is coughing and wheezing, it's usually caused by an underlying lung disease. If pulmonary hypertension has reached the stage of cor pulmonale, a person may have

swelling of the legs. People with primary pulmonary hypertension are prone to developing achy joints, sometimes years before they are diagnosed with pulmonary hypertension.

Some rheumatic disorders, such as scleroderma, may be associated with pulmonary hypertension. When a person has both scleroderma and hypertension, they may develop symptoms of Raynaud's phenomenon before the symptoms of pulmonary hypertension become noticed.

Causes

There are two kinds of pulmonary hypertension, primary pulmonary hypertension and secondary pulmonary hypertension.

Primary pulmonary hypertension is more rare and has no known cause. It most likely begins when the muscles the line the walls of the pulmonary arteries go into spasms (contractions). Women are twice as likely as men to have this kind of pulmonary hypertension. Half the people who are diagnosed with pulmonary hypertension are 35 or older when the diagnosis is made.

Secondary pulmonary hypertension occurs as a result of another disorder that affects how the lungs are structured or work. This can be any condition or disease that interferes with the flow of blood through the lungs or causes periods in which the blood does not receive enough oxygen. Some examples include chronic obstructive pulmonary disease, cystic fibrosis or occupational lung diseases. Other causes can include:

- Losing lung tissue from surgery
- Injury
- Heart failure
- Diseases, such as scleroderma
- Obesity, which impairs the ability to breathe
- Nerve disorders that involve the muscles used to breathe
- Chronic liver diseases
- HIV infection
- Diet drugs, such as dexfenfluramine-phentermine (fen-phen)

Diagnosis

A doctor can make a tentative diagnosis of pulmonary hypertension based on a person's symptoms, especially if he or she also has another lung condition. Specific tests that may be ordered to confirm the diagnosis include:

- A chest X-ray, which may show enlargement of the pulmonary arteries
- An electrocardiogram and echocardiogram to check for problems, such as thickening of the right ventricle or backflow of blood through the tricuspid valve

between the right atrium and ventricle. These signs may be present even before cor pulmonale develops
- Blood tests to check oxygen levels in the blood
- Pulmonary function to check for lung damage

The best way to confirm that pulmonary hypertension is present is to measure the blood pressure in the pulmonary artery. This is done by passing a tube (a wire catheter) through a vein in the neck, arm or leg into the right side of the heart to measure the blood pressure in the right ventricle and in the pulmonary artery.
Treatment

Treating secondary pulmonary hypertension usually focuses on treating the underlying lung disease

Drugs to make the blood vessels larger (such as calcium channel blockers, nitric oxide and prostacyclin) can be helpful when the hypertension is due to scleroderma, chronic liver disease or HIV infection. These drugs haven't been proven effective for people with secondary pulmonary hypertension due to a lung disease

Drugs to open up the blood vessels can improve the quality of life and increase survival rates for those who have primary pulmonary hypertension. A doctor will usually first do a test using cardiac catheterization to make sure that the drugs will be safe

If a person with pulmonary hypertension is not getting enough oxygen in the blood, he or she may need to continuously use oxygen through a device on the nose (a nasal canula) or an oxygen mask. The extra oxygen may also help lower blood pressure in the arteries of the lung while relieving shortness of breath.

Drugs to eliminate excess fluids from the body (diuretics) can be helpful in lowering the pressure on the arteries. Use of drugs to prevent the blood from clotting may be used to help prevent a pulmonary embolism.

Ultimately, single or double lung transplantation may be necessary when pulmonary hypertension gets severe enough and the underlying cause cannot be identified or treated.

Contact Us

For more information/question or comment about Pulmonary Artery Hypertension
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