Pulmonary Hypertension

Definition
Pulmonary hypertension is the narrowing of the pulmonary arterioles within the lung. It is caused by changes in the cells that line the pulmonary arteries. These changes cause extra tissue to form, eventually narrowing or completely blocking the blood vessels, making the arteries stiff and narrow. This makes it harder for blood to flow, raising the blood pressure in the pulmonary arteries. This creates resistance and an increased work load for the heart. The heart then enlarges from pumping blood against the resistance and raises pressure within the arteries or lungs. This results in the heart muscle to weaken and eventually completely fail. Since there is no cure other than transplantation, the goal of treatment is control of the symptoms, although the disease usually develops into congestive heart failure.

Although anyone can develop either type of pulmonary hypertension, older adults are more likely to have secondary pulmonary hypertension, and young people are more likely to have idiopathic pulmonary hypertension. Idiopathic pulmonary hypertension is also more common in women than it is in men.

Idiopathic pulmonary hypertension
When an underlying cause for high blood pressure in the lungs can't be found, the condition is called idiopathic pulmonary hypertension (IPH). Some people with IPH may have a gene that's a risk factor for developing pulmonary hypertension. But in most people there is no known cause. IPAH is rare.

Secondary pulmonary hypertension
Pulmonary hypertension that's caused by another medical problem is called secondary pulmonary hypertension and is more common than idiopathic pulmonary hypertension. Causes of secondary pulmonary hypertension include:
• Blood clots in the lungs (pulmonary emboli)
• Chronic obstructive pulmonary diseases, such as emphysema
• Connective tissue disorders, such as scleroderma or lupus
• Sleep apnea and other sleep disorders
• Congenital heart disease
• Sickle cell anemia
• Chronic liver disease (cirrhosis)
• AIDS
• Lung diseases such as pulmonary fibrosis
• Left-sided heart failure
• Living at altitudes higher than 8,000 feet (2,438 meters)
• Climbing or hiking to altitudes higher than 8,000 feet without acclimating first
• Use of certain stimulant drugs, such as cocaine

Symptoms
Shortness of breath or light-headedness during activity is often the first symptom. Tachycardia may be present. Over time, symptoms occur with less activity or even while at rest. The signs and symptoms of pulmonary hypertension in its early stages may not be noticeable for months or even years. Symptoms in this phase include: chest pain, weakness, shortness of breath, and fatigue. As the disease progresses, symptoms become worse. Other symptoms include:
• Ankle and leg edema
• Ascites
• Bluish color of the lips or skin (cyanosis)
• Chest pain or pressure, usually in the front of the chest
• Dizziness or fainting spells
• Palpitations

Complications
Pulmonary hypertension can lead to a number of complications, including:
Right-sided heart failure (Cor Pulmonale). In Cor Pulmonale, the heart's right ventricle becomes enlarged and has to pump harder than usual to move blood through narrowed or blocked pulmonary arteries. At first, the heart tries to compensate by thickening its walls and expanding the chamber of the right ventricle to increase the amount of blood it can hold. This thickening and enlarging works temporarily, and eventually the right ventricle fails from the extra strain.
Blood clots. Clots in a vein that breaks off and travel to the lungs can lead to a form of pulmonary hypertension that is reversible with time and treatment. Having pulmonary hypertension makes it more likely that clots will develop in the small arteries in the lungs, which is dangerous if there is already narrowed or blocked blood vessels.
Arrhythmia. Irregular heartbeats from the upper or lower chambers of the heart are complications of pulmonary hypertension. These can lead to palpitations, dizziness or fainting and can be fatal.
Bleeding. Pulmonary hypertension can lead to bleeding into the lungs and coughing up blood. This is another potentially fatal complication.
Tests and diagnosis

Pulmonary hypertension is hard to diagnose early because it's not often detected in a routine physical exam. Even when the disease is more advanced, the signs and symptoms are similar to other heart and lung conditions. The first tests done to diagnose pulmonary hypertension include:

**Chest X-ray.** This test may be able to check for pulmonary hypertension if your pulmonary arteries or the right ventricle of your heart is enlarged. The X-ray will appear normal in about one-third of people who have pulmonary hypertension.

**Echocardiogram.** These images show how the heart is functioning and records pictures to provide measurements of the size and thickness of the heart muscle. An exercise echocardiogram will help determine how well the heart works under stress.

**Transesophageal echocardiogram.** In this procedure, a flexible tube containing a transducer is guided down into the esophagus using after numbing the back of the throat with a spray. From here, the transducer can get detailed images of your heart. It is used when the echocardiogram is not diagnostic.

**Right heart catheterization.** After an echocardiogram, if pulmonary hypertension is suspected, a right heart catheterization is done and is the most reliable way of diagnosing pulmonary hypertension. The catheter is threaded into the right ventricle and pulmonary artery. This allows direct measurement of pressures in the main pulmonary arteries and right ventricle. It's also used to see what effect different medications may have on your pulmonary hypertension.

Additional tests may include:

**Pulmonary function test.** This noninvasive test measures how much air the lungs can hold, and the airflow in and out of the lungs.

**Perfusion lung scan.** This test uses small amounts of injected radioisotopes to study perfusion in the lungs. Immediately afterward, a gamma camera takes pictures of blood flow in the lungs' blood vessels. A lung scan is then used to determine whether blood clots are causing symptoms of pulmonary hypertension. A perfusion lung scan is usually performed with a ventilation scan. In this test, a small amount of radioactive substance is inhaled while a gamma camera records the movement of air into the lungs. The two-test combination is known as a ventilation-perfusion (V/Q) scan.

**Computerized tomography (CT) scan.** A CT scan provides organs in two-dimensional "slices."

**Magnetic resonance imaging (MRI).** This test is sometimes used to get images of the blood vessels in the lungs. A computer creates tissue "slices" from data generated by magnetic field and radio waves. An MRI can't measure artery pressure.

**Open-lung biopsy.** In rare situations an open-lung biopsy may be necessary.

**Genetic tests.** If a family member has had pulmonary hypertension, genetic testing may be done to screen for pulmonary hypertension. First the individual with Pulmonary Hypertension is
tested to identify the gene and then other family members are screened for that particular gene.

**Pulmonary Hypertension classifications**
Pulmonary hypertension is classified using guidelines developed by the New York Heart Association.

- **Class I** - Although diagnosed, patient is asymptomatic.
- **Class II** - Asymptomatic at rest, but fatigue, shortness of breath or chest pain with normal activity.
- **Class III** - Comfortable at rest but symptoms when physically active.
- **Class IV** - Symptomatic at rest.

**Medications**
There is no known cure for pulmonary hypertension, so the goal of treatment is to control symptoms and prevent more lung damage. It is important to treat medical disorders that cause pulmonary hypertension, such as obstructive sleep apnea, lung conditions, and heart valve disorders. Medications used to treat pulmonary hypertension include:

- **Vasodilators.** Vasodilators open narrowed blood vessels. One of the most commonly prescribed vasodilators for pulmonary hypertension is epoprostenol (Flolan). The drawback to epoprostenol is that its effects last only a few minutes. This drug is given as a continuous infusion. Potential side effects include jaw pain, nausea, diarrhea, leg cramps, as well as pain and infection at the IV site. Another form of the drug, iloprost (Ventavis), avoids many of these problems. Iloprost can be inhaled every three hours through a nebulizer, a machine that vaporizes your medication, making it far more convenient and less painful to use. And because it’s inhaled, it goes directly to the lungs. Side effects associated with iloprost include chest pain — often accompanied by headache and nausea — and breathlessness.

- **Endothelin receptor antagonists.** These medications reverse the effect of endothelin, a substance in the walls of blood vessels that causes them to narrow. One of these medications, bosentan (Tracleer), may improve energy level and symptoms. This drug is contraindicated in pregnant women.

- **Sildenafil.** Revatio (sildenafil) is sometimes used to treat pulmonary hypertension. It works by opening the blood vessels in the lungs to allow blood to flow through more easily. Side effects include dizziness and vision problems.

- **High-dose calcium channel blockers.** These drugs help relax the muscles in the walls of your blood vessels. They include medications such as amlodipine (Norvasc), diltiazem (Cardizem, Tiazac) and nifedipine (Adalat, Procardia). Although calcium channel blockers can be effective, only a small number of people with pulmonary hypertension respond to them.
• **Ambrisentan.** Ambrisentan (Letairis) is another medication that stops the narrowing of the blood vessels. This drug can cause serious liver damage if not taken appropriately, and is contraindicated in pregnant women.

• **Anticoagulants.** Anticoagulant help prevent the formation of blood clots within the small pulmonary arteries. Because anticoagulants prevent normal blood coagulation, they increase the risk of bleeding complications.

• **Diuretics.** These medications help eliminate excess fluid and reduce the amount of work the heart has to do. They also may be used to limit fluid buildup in your lungs.

• **Oxygen.** May be used to help treat pulmonary hypertension, especially for those who live at a high altitude or have sleep apnea. Some people with pulmonary hypertension eventually require constant oxygen therapy.

Other Instructions/Treatments
Other important tips to follow:
• Avoid pregnancy
• Avoid heavy physical activities and lifting
• Avoid traveling to high altitudes
• Keep up-to-date with yearly flu and pneumococcal vaccines
• Stop smoking
• Get plenty of rest to reduce fatigue
• Stay as active as possible. Even the mildest forms of activity may be too exhausting for some people with pulmonary hypertension. For others, moderate exercise such as walking may be beneficial, and using oxygen during exercise may be especially helpful.
• Avoid situations that can excessively lower blood pressure. These include sitting in a hot tub or sauna or taking long hot baths or showers. These activities lower blood pressure and cause fainting or even death.
• Avoid activities that cause prolonged straining, such as lifting heavy objects or weights.
• Find ways to reduce stress. Many people with pulmonary hypertension find that simply reducing stress can greatly improve the quality of their lives.
• Follow a nutritious diet and stay at a healthy weight.

Surgical Procedures
**Atrial septostomy.** If medications don’t control pulmonary hypertension, this open-heart surgery may be an option. In an atrial septostomy, an opening between the left and right chambers of the heart is created to relieve the pressure on the right side of your heart. Atrial septostomy can have serious complications, including arrhythmias.

**Transplantation.** In some cases, a lung or heart-lung transplant may be an option, especially for younger people who have idiopathic pulmonary hypertension.
References


