

Pulmonary hypertension

Definition

Pulmonary hypertension is abnormally high blood pressure in the arteries of the lungs. It makes the right side of the heart need to work harder than normal.

Alternative Names

Pulmonary arterial hypertension; Sporadic primary pulmonary hypertension; Familial primary pulmonary hypertension; Idiopathic pulmonary arterial hypertension; Primary pulmonary hypertension; PPH; Secondary pulmonary hypertension

Causes

The right side of the heart pumps blood through the lungs, where it picks up oxygen. Then, the blood returns to the left side of the heart, where it is pumped to the whole body.

When the small arteries (blood vessels) of the lung become narrowed, they cannot carry as much blood. When this happens, pressure builds up. This is called pulmonary hypertension.

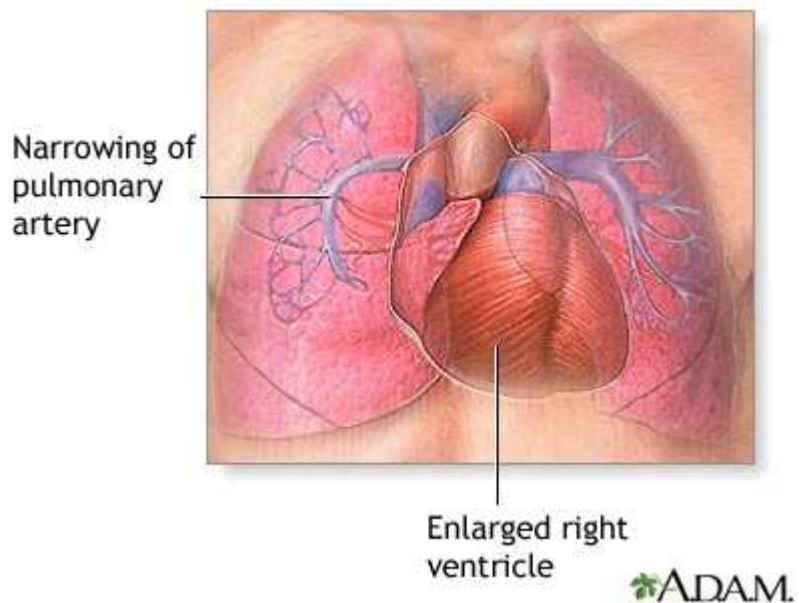
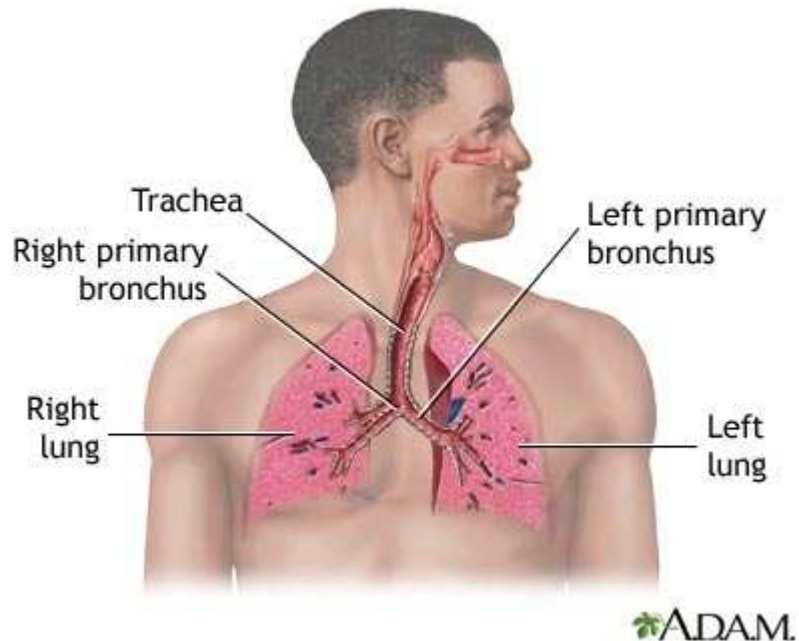
The heart needs to work harder to force the blood through the vessels against this pressure. Over time, this causes the right side of the heart to become larger. Not enough blood flows to the lungs to pick up oxygen.

At this point, heart failure involves the right side of the heart. This is called cor pulmonale.

Pulmonary hypertension may be caused by:

- Autoimmune diseases that damage the lungs, such as scleroderma and rheumatoid arthritis
- Birth defects of the heart
- Blood clots in the lung (pulmonary embolism)
- Congestive heart failure
- Heart valve disease
- HIV infection
- Low oxygen levels in the blood for a long time (chronic)
- Lung disease, such as COPD or pulmonary fibrosis
- Medicines (for example, certain diet drugs)
- Obstructive sleep apnea

In many cases the cause is unknown. In this case, the condition is called idiopathic pulmonary arterial hypertension (IPAH). It used to be called primary pulmonary hypertension (PPH).



IPAH is rare. It affects more women than men.

If pulmonary hypertension is caused by a known medicine or medical condition, it is called secondary pulmonary hypertension.

Symptoms

Shortness of breath or light-headedness during activity is often the first symptom. Fast heart rate (palpitations) may be present. Over time, symptoms occur with lighter activity or even while at rest.

Other symptoms include:

- Ankle and leg swelling
- Bluish color of the lips or skin (cyanosis)
- Chest pain or pressure, usually in the front of the chest
- Dizziness or fainting spells
- Fatigue
- Weakness

People with pulmonary hypertension often have symptoms that come and go. They report good days and bad days.

Exams and Tests

A physical examination may show:

- Abnormal heart sounds
- Feeling of a pulse over the breastbone
- Heart murmur on the right side of the heart
- Larger-than-normal veins in the neck
- Leg swelling
- Liver and spleen swelling
- Normal breathing sounds

In the early stages of the disease, the exam may be normal or almost normal. The condition may take several months to diagnose. Asthma and other diseases may cause similar symptoms and must be ruled out.

Tests may include:

- Blood tests
- Cardiac catheterization
- Chest x-ray
- CT scan of the chest
- Echocardiogram
- ECG
- Lung function tests
- Nuclear lung scan
- Pulmonary arteriogram
- Six-minute walk test
- Sleep study

Treatment

There is no known cure for pulmonary hypertension. 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.

Many new treatment options for pulmonary arterial hypertension (IPAH) and other forms of pulmonary arterial hypertension are becoming available. Medicines used to treat pulmonary hypertension include:

- Ambrisentan (Letairis)
- Bosentan (Tracleer)
- Calcium channel blockers
- Diuretics
- Prostacyclin or similar medicines
- Sildenafil and similar medicines

Your doctor will decide which medicine is best for you. You will be closely monitored during treatment to watch for side effects and to see how well you are responding to the medication. Never stop taking medicines without talking to your doctor.

Some patients are put on blood thinners to reduce the risk of blood clots in leg veins and lung arteries.

People with low oxygen levels in the blood may need oxygen therapy at home.

As the illness gets worse, you will need to make changes in your home and get more help around the house.

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

If treatment with medicine does not work, a lung or heart-lung transplant may help some people.

Outlook (Prognosis)

The long-term outlook has been poor, but new treatments may lead to better results. Some people with this condition may have heart failure that could lead to death.

It is not a good idea to get pregnant if you have this condition.

When to Contact a Medical Professional

Call your health care provider if:

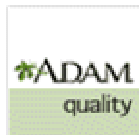
- You begin to develop shortness of breath when you are active
- Shortness of breath worsens
- You develop chest pain
- You develop other symptoms

Most patients with pulmonary arterial hypertension are treated at centers that specialize in the care of these patients.

References

McLaughlin VV, Archer SL, Badesch DB, Barst RJ, Farber HW, Lindner JR, et al: American College of Cardiology Foundation Task Force on Expert Consensus Documents; American Heart Association; American College of Chest Physicians; American Thoracic Society, Inc; Pulmonary Hypertension Association. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association developed in collaboration with the American College of Chest Physicians; American Thoracic Society, Inc; and the Pulmonary Hypertension Association. *J Am Coll Cardiol*. 2009;53:1573-1619.

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