

What is pulmonary hypertension?

Pulmonary hypertension is a disorder of the blood vessels that carry blood from the heart to the lungs. The vessels become narrowed. This causes the pressure in the arteries to rise above normal. The heart has to work harder and puts strain on the lungs. It can lead to serious problems.

What causes it?

It is most often caused by another health problem. In many cases, taking care of this health problem can help prevent or control pulmonary hypertension. Some of the most common causes are:

- Severe lung problems in newborns.
- Lung conditions like cystic fibrosis, interstitial lung disease, or pulmonary fibrosis.
- Heart disease or heart failure.
- Heart defects at birth.
- HIV, scleroderma, lupus, or sickle cell disease.

Sometimes there is no known cause.

What are the symptoms?

- Shortness of breath.
- Getting tired easily, especially when active.
- Chest pain or pressure.
- Fast heartbeat.
- Fainting or dizzy spells.
- Blue lips or fingernails. These are signs the body is not getting enough oxygen.
- Bloating or swelling in the legs or ankles.

How is it diagnosed?

A complete physical exam will be done. We will check heart and lungs, blood pressure and oxygen level. Some of these tests may be done:

- **Electrocardiogram (EKG).** Records the heart's electrical activity.
- **Chest X-ray.** Shows the inside of the chest, including the lungs and the heart.
- **Echocardiogram.** Looks at a moving picture of the heart.
- **Blood tests.** Measure certain body functions.
- **Cardiac catheterization.** Gives the doctor detailed information about the structure of the child's heart and pressures in the heart and lungs. A thin tube, called a catheter is put into a blood vessel in the groin and guided to the heart. This may need to be repeated.
- **Pulmonary function tests.** Measure breathing and lung capacity.
- **Six minute walk test.** Looks at how well a child does with exercise.

How is it treated?

Treatment depends on age, health, and the severity of the disease. The doctor may prescribe:

- Medicines to decrease the pressure of the blood vessels in the lungs.
- Medicines to thin blood and prevent clotting.
- Medicines to get rid of excess body water.
- Oxygen.

What are the long-term concerns?

If the health problem causing it can be treated, the outcome is often good. Rarely, a lung transplant is needed if the condition is long lasting and severe.

ALERT: Call your child's doctor, nurse, or clinic if you have any questions or concerns or if your child:

- Is short of breath or has chest pain.
- Gets dizzy or faints.
- Has a fever of 100.4°F or higher.
- Has any new symptoms
- Is getting worse.
- Has special health care needs that this sheet did not cover.

This sheet was created to help you care for your child or family member. It does not take the place of medical care. Talk with your healthcare provider for diagnosis, treatment and follow-up.