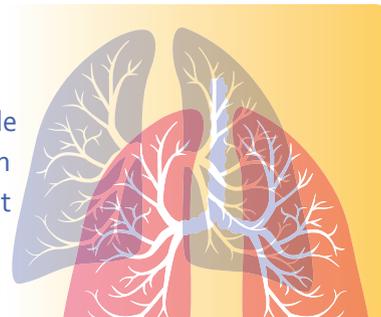


Pulmonary Hypertension in Children

Pulmonary hypertension is a condition in which the pressure in the pulmonary artery and lungs is too high. This puts stress on the right side of the heart because the muscles on the right side are not used to pushing blood out to the lungs against such high pressures. Over time, the right side of the heart is strained and begins to fail. This is a rare but serious condition that can develop in children or adults at any age. This fact sheet talks about Pulmonary Hypertension in children. For more information about Pulmonary Hypertension in adults, see the ATS Patient Information Series at www.thoracic.org/patients.



In a structurally normal heart, oxygen-poor blood flows to the right side of the heart from the body. The blood then flows into the right atrium and finally into the right ventricle. The right ventricle pumps this blood through the pulmonary artery into the lungs. In the lungs, the blood becomes enriched with oxygen. This oxygen-rich blood then flows to the left side of the heart so the body's organs can get enough oxygen to function properly.

In pulmonary hypertension (PH), there is high pressure in the blood vessels of the lungs. This makes the right ventricle work harder. The harder the right ventricle has to work with this strain, the higher the risk for failure.

Classes and Causes of Pediatric Pulmonary Hypertension

There are various causes of pulmonary hypertension in children. It can occur without any obvious reason which is called idiopathic (or primary) pulmonary hypertension or pulmonary arterial hypertension (PAH). PAH differs from other forms of PH in that the blood vessel walls remodel and become narrow. Some children can have PH that is inherited or familial (passed down or occurring in multiple family members). PH can also occur in association with another condition which is called secondary PH. These conditions include various forms of congenital heart disease, some types of lung disease, clotting disorders, autoimmune diseases, some genetic or developmental disorders, or as a side effect of certain medicines.

Changes in the Body Due to Pulmonary Arterial Hypertension

As the pressure increases in the lungs, the right ventricle can begin to struggle. Blood then starts backing up into the right atrium and finally, into the veins of the head, neck and belly. The child may develop bulging neck veins, belly pain, nausea with vomiting and/or liver abnormalities. Blood flow to the brain and kidneys can be affected also, causing them to not function as well. The child may start fainting or have decreased urine output.

What are the Signs and Symptoms of Pulmonary Hypertension?

Some of the symptoms of PH are common to other less severe

childhood conditions, which may result in delay of diagnosis.

These conditions often include asthma, behavioral concerns, or seizures. Symptoms can progress over time and may differ based on the age of the child and other underlying conditions. The most common symptoms are related to difficulty with breathing. It is important to tell your healthcare provider of any new or worsening symptoms. Symptoms include:

- Fatigue (tiring easily) which may worsen with activity
- Shortness of breath especially with activity
- Blue discoloration of the lips, hands and/or feet (cyanosis)
- Dizziness or lightheadedness
- Heart racing or pounding (palpitations)
- Fainting spells (syncope)
- Coughing up blood

On physical exam, people with pulmonary hypertension may show swelling (edema) of the lower legs and feet, enlarged liver and/or distension of the neck veins. A heart murmur or abnormal heart sounds may be heard with a stethoscope.

How is Pulmonary Hypertension Diagnosed?

Pulmonary Hypertension is a rare disease that can be difficult to diagnose and therefore it is important to be evaluated by a team that has expertise in PH. Many tests will be ordered to determine the severity of disease and the type of pulmonary hypertension the child has.

- One of the first tests performed is often an ultrasound of the heart called an **echocardiogram (echo)**. The echo will show if there is strain on the right side of the heart and can give an estimate of the pressures on the right side of the heart and pulmonary arteries. Echocardiograms are used to assist in diagnosis and to follow disease progression.
- A **chest x-ray** may show enlargement of the size of the heart and/or pulmonary arteries and evaluate for lung disease.
- An **electrocardiogram (EKG)** may be performed to evaluate for enlargement of the right side of the heart or changes in cardiac muscle function.

- **A six-minute walk test** is used to assess exercise capacity, or endurance. This is performed in children who are old enough to follow commands and to walk as directed. During the test, children will walk the measured course at their own pace while wearing a pulse oximeter to measure heart rate and oxygen levels. The distance walked is measured and compared to previous six-minute walk tests. Six-minute walk testing is also used to follow response to therapy and for clinical staging of disease severity.
- **Pulmonary function tests (PFTs)** are typically performed in children older than 5 years of age because of the need to follow directions and to forcefully exhale for at least four to six seconds. A trained expert will guide the child to take a deep breath and force air out as hard and long as possible. PFTs are done to evaluate air flows and lung volumes; abnormalities may suggest lung disease. For more information on Pulmonary Function Testing in Children, go to www.thoracic.org/patients.
- Sleep study or **polysomnography** may be performed if there is concern for sleep disordered breathing as a contributing factor or cause of PH.
- **Blood tests** may be done to look for the cause of PH and to check the strain on the right side of the heart. Blood tests can also be very important in monitoring progression of disease.
- **Cardiac catheterization** is the gold standard in diagnosis of PH. During cardiac catheterization, the pressures in the right side of the heart and blood vessels in the lungs will be measured using specialized catheters. Through this procedure, the degree of PH can be measured, response to therapy (oxygen/medications) can be tested and anatomy of the heart and blood vessels can be studied. Cardiac catheterization can help guide therapy as well as help monitor disease progression.

How is Pulmonary Hypertension Treated?

There are many treatments available which may relieve some symptoms and may slow the progression of the disease. Early treatment is important because advanced disease may be less responsive to therapy. Oxygen is given with sleep or all the time if oxygen levels are low (see also ATS Patient information series "Oxygen Therapy in Children" at www.thoracic.org/patients). Treatment usually includes use of specific PH disease modifying medications that can be swallowed (oral medications), inhaled, or given continuously either directly into the vein or under the skin (subcutaneous) with a pump. Other medications include diuretics and possibly blood thinners (anticoagulants). These PH medications are given to relax the blood vessels in the lungs, improve blood flow through the lungs, and reduce strain on the right heart.

Children with PH should be referred to a pulmonary hypertension specialist or pulmonary hypertension center that has a comprehensive approach to treating PH. This includes physical therapy, nutrition and social work services. To find a Pediatric PH specialist in the U.S., you can go to the Pulmonary Hypertension Association website (<https://phassociation.org/pha-pediatrics/> and <https://www.phassociation.org/PHCareCenters/Patients/AccreditedCenters/Peds>).

People being treated for PH need to have regular appointments with their specialist. They often need regular testing including blood tests, echocardiograms and cardiac catheterization to determine how well their medications are working. The child's caregiver needs to contact the PH specialist right away with any

changes in symptoms. Children should avoid contact sports and only take part in activities that allow them to rest when tired. If medications fail, treatment options may include surgical measures to decompress the heart or lung transplantation. These two advanced procedures are only done in highly specialized pediatric centers after careful consideration. (For more information on Lung Transplantation in Children, go to www.thoracic.org/patients).

What is the Prognosis for Pediatric Pulmonary Hypertension?

The prognosis is generally better for children with PH than for adults with PH. However, the prognosis depends on the type and severity of PH (such as whether it is the only problem or is associated with another condition). For PH which occurs with another condition, the prognosis is often tied to the prognosis of the underlying disease or condition.

In contrast, PH that occurs without an obvious cause (i.e. PAH) is a progressive disease, which is life-shortening. There is no cure. However, with early and aggressive treatment, improved survival has been reported. Advances in research continue to provide hope for the future treatment of PH.

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This information series piece reflects standards defined in the American Thoracic Society/American Heart Association Clinical Practice Guidelines (2015) <http://circ.ahajournals.org/content/early/2015/10/29/CIR.0000000000000329>

Rx Action Steps

- ✓ If your child has trouble breathing with exercise or fainting spells, that are not improving with treatment or getting worse, ask your healthcare provider about checking for pulmonary hypertension.
- ✓ If your child has been diagnosed with Pulmonary hypertension, seek care from a PH specialist.
- ✓ Take all medications as prescribed and call right away with any change in symptoms.
- ✓ Talk with your child's PH specialist about what activities your child can safely do.

Doctor's Office Telephone:

Resources:

Pulmonary Hypertension Association (PHA)

<https://www.phassociation.org/Parents>

Pulmonary Hypertension Association – Europe

<http://www.phaeurope.org/disease-information/children-pha/>

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