

Guidelines first to focus on children with pulmonary hypertension

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For the first time, guidelines have been developed for children with pulmonary hypertension, a sometimes fatal heart and lung disease that affects nearly two of every 1,000 babies born each year. The joint American Heart Association/American Thoracic Society guidelines are published in the American Heart Association's journal *Circulation*.

"These children suffer with health issues throughout their lives or die prematurely—particularly if they're not properly diagnosed and managed. But with the proper diagnosis and treatment at a specialized center for [pulmonary hypertension](#), the prognosis for many of these children is excellent." said Stephen L. Archer, M.D., the head of medicine at Queens University, Kingston, Ontario, Canada and co-chair of the guideline's writing committee.

Children with pulmonary hypertension typically have blockages in the blood vessels of their lungs (the [pulmonary arteries](#)), which makes it harder for the heart's right ventricle to pump blood through the lungs. Symptoms in children with pulmonary hypertension include shortness of breath, passing out and appearing blue (cyanosis).

Some children have pulmonary hypertension as an isolated condition caused by circulatory problems in their lungs, however, many patients have pulmonary hypertension as a complication of other serious heart or lung health issues, such as [congenital heart disease](#), bronchopulmonary dysplasia (a form of chronic [lung disease](#)) and [congenital diaphragmatic hernia](#). Children with diaphragmatic hernias are born with a hole in the

diaphragm that allow abdominal organs to shift in the chest, which prevents the lungs from developing properly.

Untreated, pulmonary hypertension can lead to disability or death from heart failure.

Although there have been guidelines for the diagnosis and treatment of pulmonary hypertension in adults for many years, they are not directly applicable to children since the underlying causes of the disease differ markedly in children compared to adults. The new guidelines are a collaboration between the American Heart Association and American Thoracic Society. Twenty-seven physicians from different disciplines, such as pediatric cardiology, pulmonary, critical care, neonatology, surgery and others, worked together distilling information from more than 600 studies to develop a foundation for the best possible care for pediatric pulmonary hypertension.

The new guidelines provide doctors with practical advice about:

- how to classify the many types of pulmonary hypertension (a key step in determining treatment);
- proven and emerging medical and surgical therapies;
- which treatments are approved for children with pulmonary hypertension and which dosages should be used;
- optimal comprehensive care of the patient with pulmonary hypertension-including advice on supportive care, social aspects of care and pragmatic advice on exercise and travel restrictions; and
- advice on the role of accessing pulmonary hypertension centers that specialize in treating the disease.

The guidelines also point to gaps in knowledge about pediatric pulmonary hypertension and offer advice about issues that parents and healthcare providers struggle with, including whether children should receive blood thinners, how to determine whether a child with pulmonary hypertension can safely engage in exercise or travel on a plane, and how high altitude can cause or worsen pulmonary hypertension.

"It's important to note that although these guidelines provide a foundation for taking care of children with pulmonary hypertension, we still have a huge need for more specific data and research to further improve outcomes of children with pulmonary hypertension," said Steven H. Abman, M.D., the guidelines committee co-chair, a pediatric pulmonologist, and professor of pediatrics at the University of Colorado School of Medicine and Children's Hospital in Denver, Colorado.

When children are diagnosed with pulmonary hypertension, parents often feel hopeless. However, Archer said, it's important that parents seek doctors and centers that see these children on a regular basis and can offer them access to new molecular diagnostics, new drug therapies and new devices as well as surgeries that have recently been developed.

Provided by American Heart Association

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