

Top Ten Things To Know American Heart Association and American Thoracic Society Joint Guidelines for Pediatric Pulmonary Hypertension

1. Few studies specifically address the safety and efficacy of therapies for pulmonary arterial hypertension (PAH) in children, and there are no treatment guidelines. Care guidelines have not been previously developed for specific use in children.
2. Epidemiology and statistics:
 - PPHN is rarely familial and few genetic causes have been identified.
 - PPHN often complicates developmental lung diseases, including Down Syndrome, alveolar capillary dysplasia (ACD), genetic abnormalities of surfactant function and lung hypoplasia in diverse settings.
 - Delivery prior to 39 weeks of gestation is now appreciated as being strongly associated with worse neonatal outcomes, in large part because of an elevated incidence of neonatal respiratory failure.
 - Evidence suggests that up to 2% of all preterm infants will have early acute PPHN, particularly following prolonged preterm rupture of membranes or oligohydramnios.
 - The prevalence of PPHN has been estimated as 1.9/1000 live births.
 - Even with precise diagnosis, the availability of specific pulmonary vasodilators such as iNO and extracorporeal support, early mortality for PPHN remains 8-10%.
 - Long term outcomes for PPHN include high rates of neurodevelopmental impairment at 18 months of age, as defined by mental developmental index < 70, cerebral palsy, deafness and blindness.
3. These joint guidelines represent consensus from experts in the field addressing current approaches to the diagnosis, evaluation and treatment of pulmonary hypertension in children.
4. The focus is on childhood disorders of PH due to pulmonary vascular disease (PVD), and includes PH related to cardiac, lung and systemic diseases, as well as idiopathic pulmonary artery hypertension (IPAH).
5. PH and related PVD cause significant morbidity and mortality in diverse childhood diseases. Despite availability of new drug therapies, long-term outcomes for children with severe pulmonary arterial hypertension (PAH) remains poor.
6. Unlike adult PH, pediatric PH is intrinsically linked to issues of lung growth and development including many prenatal and early postnatal influences
7. Adult and pediatric PH differ in vascular function and structure, genetics, natural history, response of the right ventricle and responsiveness to PAH-specific therapies. Importantly, there are many more conditions associated with PH in children than in adults.
8. Therapeutic strategies for adult PAH have not been sufficiently studied in children to allow definition of potential toxicities or optimal dosing. Clinical research in pediatric PH suffers from lack of age-appropriate clinical endpoints.
9. These guidelines offer recommendations for diagnosis, assessment, monitoring PH. Specific topics include a discussion of the genetics involved, conditions and events such as persistent PH of the newborn, congenital diaphragmatic hernia, bronchopulmonary dysplasia, idiopathic PAH, pediatric heart disease, PH crises/acute RV failure, hypobaric hypoxia, lung diseases, systemic disease, and outpatient care, and pharmacotherapy for PH.
10. There is clearly a need to better define the natural history and course of pediatric PAH; to develop new strategies to identify patients at-risk for development of PAH, and to establish novel approaches to diagnose, monitor disease progression and treat children with PAH.

Abman SH, et al; on behalf of the American Heart Association Council on Cardiopulmonary, Critical Care, Perioperative and Resuscitation, Council on Clinical Cardiology, Council on Cardiovascular Disease in the Young, Council on Cardiovascular Radiology and Intervention, Council on Cardiovascular Surgery and Anesthesia, and the American Thoracic Society. [Pediatric pulmonary hypertension: guidelines from the American Heart Association and American Thoracic Society](#) [published online ahead of print November 3, 2015]. *Circulation*. doi: 10.1161/CIR.0000000000000329.