Guidelines for Managing Pulmonary Hypertension: What the Pediatrician Needs to Know

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Abstract

Pediatric pulmonary vascular disease contributes to morbidities and death in diverse clinical settings, ranging from idiopathic or heritable forms of pediatric arterial hypertension, congenital heart disease, developmental lung disorders, chronic lung disease, left heart disease, sickle cell disease, oncologic disease and systemic disorders. Despite its impact on the clinical courses in so many diseases, information is limited on how to best approach the diagnosis and evaluation of pediatric pulmonary hypertension. This review highlights a few key topics underlying guidelines for the care of children with pulmonary hypertension that are especially important for the practicing pediatrician and others, and presents some of the major recommendations from the published guidelines report. Overall, the author emphasizes that these guidelines are based on the best current evidence and clinical experience of experts in the field, yet much more clinical research is needed to improve long term outcomes in pediatric pulmonary hypertension. Over the past few decades, there has been a growing awareness of the contributions of pulmonary hypertension (PH) and related pulmonary vascular diseases (PVD) to poor outcomes in children with diverse diseases, associated with heart, lung, blood and systemic conditions. Despite advances in our understanding of its pathobiology and the growing availability of drug therapies, PH and related PVD continue to cause significant morbidity and mortality. Although PH is a relatively rare condition, it is surprising how often abnormalities of the pulmonary circulation are found to contribute to the pathophysiology and clinical course in acute and chronic settings. PH is present in such settings as the extremely rare form of idiopathic or isolated pulmonary artery hypertension (IPAH) to more common conditions such as acute respiratory failure, chronic lung diseases and hematologic disorders and others. In fact, over the past several years, PH-related hospitalizations of children have markedly increased, which likely reflects improved recognition of the important role of PH in these diverse clinical settings. Despite increased understanding regarding the adverse impact of PH in many settings, pediatric PVD has been understudied and relatively little is known about basic disease mechanisms, natural history, long-term outcomes, age-appropriate clinical endpoints and optimal therapeutic strategies for neonates, infants and children with PAH. Studies are often complicated by the marked heterogeneity of conditions and co-morbidities associated with pediatric PVD, the relatively small number of patients at each center, the dependence on anecdotal experience or on studies of adult subjects, and other critical factors. While similarities exist regarding the etiology and disease pathogenesis of some forms of pediatric and adult PH, many cardiopulmonary and systemic diseases associated with PH are unique to children.

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