Making Strides for Pediatric Pulmonary Arterial Hypertension

The evaluation and treatment of pulmonary arterial hypertension (PAH) in children represents distinct challenges. In this issue of *Advances*, Drs Dunbar Ivy and Usha Krishnan have done a fantastic job guest editing in a fashion that complements recently published guidelines for this important population.[1] The *Advances* issue, along with the pediatric guidelines, offers a more comprehensive review than is typically available in more conventional efforts.[2]

Amazingly, no FDA-approved pharmacological therapies are available for PAH in children. Nonetheless, dedicated clinicians and scientists have forged ahead relying on those medications approved for use in adults. For perspective, they offer an update on the status of 5 children who were featured in a 2011 issue on pediatric PAH who have managed to control their disease with expert management. In addition to the core articles, please take time to read the roundtable discussion that provides background of guideline development and the role in clinical practice; the review on dental care in children with PAH from our PH Professional Network colleague; and PH Grand Rounds that highlights one of the important mimics of chronic pulmonary thromboembolic disease. Lastly, I want to thank Dr. Slack for his careful review of a prior issue and need to provide corrective information.

One more important note: this issue hosts a tribute to Rino Aldrighetti, who has retired as President of the Pulmonary Hypertension Association. His dedication to the launch and evolution of *Advances* has been key to the success of the journal.

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References

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2. Galie N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). *Eur Heart J.* 2016;37(1):67-119.

Diagnosis and treatment of children with pulmonary hypertension (PH) remain challenging. Advances in therapy have followed the approval of adult therapies; however, there still is no FDA-approved medication for the treatment of pulmonary arterial hypertension (PAH) in children. Further, recognition of PH associated with chronic lung disease continues to grow, increasing the number of infants treated. To aid the clinician struggling to treat these children, a group of PH clinicians recently published "Pediatric Pulmonary Hypertension: Guidelines from the American Heart Association and American Thoracic Society" (*Circulation* 2015; 132(21):2037-99). In this issue of *Advances in Pulmonary Hypertension*, we address various aspects of pediatric PH that are discussed in the guidelines. We thank all the authors who contributed to this effort and to the experts who participated in the roundtable.

Nearly every 5 years, we have an issue of *Advances* dedicated to pediatric PH. The first pediatric PH issue, in 2006, featured photographs of 5 children diagnosed with PAH on the front page. In a 2011 issue we showed the same 5 children--older and doing well on targeted therapies. We are happy to showcase the same 5 children-- now grown into 3 young adults and 2 teenagers—in a photo montage on pages 118-119 of this issue. Two are still on prostanoids and the rest are on dual-targeted therapies, and all are WHO functional class I-II. One is now an RN, another is working- doing talent shows, 3 are in high school and college and participate in sports activities under supervision. They are all very productive individuals and are actively involved in PAH advocacy. Each one of them is grateful to the advances in PAH care and therapeutics and are very aware of the need for continued research toward better palliation and perhaps a cure for the disease.

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