Pulmonary Hypertension Associated with Congenital Heart Disease: It's Not All the Same

With improvements in medical and surgical therapeutics over the past two decades, the number of adults living with congenital heart disease now exceeds the number of children. Whether as a result of excessive pulmonary blood flow in childhood, or related to post-capillary obstruction, many of these adults have associated pulmonary hypertension (APAH-CHD) and require advanced management strategies. The evaluation of adults with APAH-CHD, which is often accompanied by complex cardiac lesions including single ventricle anatomy, can be extremely challenging. Presently, with the emergence of novel targeted PAH agents, medical-surgical approaches to APAH-CHD patients are rapidly evolving. In this edition of *Advances*, Guest Editor Dr Rich Krasuski calls upon authors to highlight the latest advances in the management of PAH in adults with structural heart disease. From the basics on anatomy for the non-congenital heart expert, to imaging, novel medical and interventional therapeutics, and the importance of transition programs, experts cover it all in this issue.

On a personal note, this edition of *Advances* represents the final journal published during my term as Editor-in-Chief. I want to extend a tremendous thanks to the editorial board and to Deb McBride for their dedication and assistance during my term. It is with great pleasure that I am able to hand off the position to a close colleague and friend, Dr Myung Park. Dr Park's expertise in the field, and enthusiasm for helping the PH community, will undoubtedly serve her well in this new position as Editor-in-Chief. Congratulations, Myung!

Finally, I want to express my deepest gratitude for the years of mentorship by Dr Robyn J. Barst who recently lost her own battle with illness, but won so many for the PH community. While many of us will miss her dearly, I am certain that Dr Robyn Barst's legacy will continue to impact the field for many years to come.

Signing off,

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Guest Editor's Memo

This issue of *Advances in Pulmonary Hypertension* focuses on the management of patients with congenital heart disease and associated pulmonary hypertension. More than a million adults in the United States have congenital heart defects, and adults now outnumber children with congenital heart defects. Many of these patients present complex cases with unique anatomical defects and very complicated interplay between pulmonary blood flow and pulmonary vascular resistance. Up to 40% of congenital heart patients are at risk for developing pulmonary hypertension and up to 10% actually develop it. Half of these can progress to Eisenmenger syndrome, a condition resulting in profound cyanosis from venous to systemic blood flow, when shunt lesions go unrecognized and untreated.

The goal of the following articles is to provide a broad overview of the congenital heart lesions most likely to result in pulmonary vascular disease, so the pulmonary hypertension specialist can become aware of the presenting features and the unique management strategies required. A multitude of treatments are now available for these patients, including medical therapies targeting the pulmonary vasculature, percutaneous devices that can be used to close abnormal intracardiac and vascular communications, balloons and stents that can be used to increase blood flow when the circulation is compromised, and even catheter-based valve prostheses that can be implanted without requiring surgery. A variety of surgical procedures also exist that can target the many different heart defects and valve abnormalities and dramatically alter the natural history of these disorders. Often what is utilized is a hybrid technique with several different specialists working together to improve the quantity and quality of life for this challenging patient population. It is indeed an exciting time to provide care for this constantly expanding group of patients!

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