Editor's memo

Tackling the Gray Zone

It is a great honor to take on the role of editor-in-chief of *Advances in Pulmonary Hypertension* from Dr Richard Channick and to welcome new experts to the editorial board. I'd first like to thank Rich Channick and Deb McBride, managing editor, for paving the way for a relatively smooth transition for me into this role. With this talented and dynamic group of editorial board experts, we intend to continue the *Advances* commitment to bringing the PH community the latest updates in the field of pulmonary hypertension.

When the editorial board approached me about devoting an issue of *Advances* to Group 2 pulmonary hypertension, and specifically to pulmonary hypertension (PH) associated with left ventricular diastolic dysfunction, I wondered about whether we would be able to fill the issue with quality content as, admittedly, what we know about managing diastolic dysfunction is less than what we don't know. However, the fact that many of the novel therapies for pulmonary arterial hypertension (PAH; Group 1 PH) can worsen symptoms for a patient with left-sided heart disease reinforced the need to cover this topic and provide clinicians with some tools to ably distinguish PAH (Group 1) patients from PH (Group 2) patients. In this issue, the editorial board and expert authors do an excellent job of highlighting this challenging topic that affects an increasing number of patients encountered within our clinical practices today.

As Dr Myung Park bravely took the lead as guest editor for this issue, she commissioned authors to cover some of the frequently-faced diagnostic dilemmas and management challenges that occur when caring for patients with Group 2 PH. Articles include Dr Francisco Soto's piece on the clinical features that should raise one's suspicion about Group 2 PH. Dr Paul Forfia and Dr Amresh Raina provide us with an eloquent article on non-invasive imaging strategies, and Dr Michael Mathier provides an overview of the hemodynamic nuts and bolts of evaluating patients for Group 2 PH. Both articles describe the diagnostic tools to help us to differentiate between Group 1 (PAH) and Group 2 PH. Finally, Dr Hunter Champion brings forth some general treatment strategies for this challenging group of patients, which underscore the need for further research in this arena.

In this issue, Dr Park and authors as well as section editors have done a terrific job at balancing what we know with what we don't know as it relates to Group 2 Pulmonary Hypertension and begin the dialogue of how we can advance the overall assessment and care of this growing group of PH patients.

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Guest editor's memo

Group 2 PH: Striving To Get A Handle On This Epidemic

Pulmonary hypertension (PH) associated with left heart disease is the single most common form of PH we encounter in clinical practice today. Divided into 3 categories – systolic dysfunction, diastolic dysfunction, valvular disease – the presence of PH is well recognized to portend worse outcome across the spectrum of cardiac disorders. PH associated with diastolic dysfunction presents unique challenges for clinicians, as at first glance it shares many similar features with pulmonary arterial hypertension (PAH). Furthermore, the incidence of PH with diastolic dysfunction is in sharp rise, in parallel with the aging population and increase in incidence of obesity. It is no wonder that I often hear my referring colleagues comment, "*All* of my patients have PH. It has become an epidemic!"

Thus, it has been a distinct pleasure to have the opportunity to bring together a leading group of expert clinicians in this rapidly evolving field to address specifically some of the most frequently asked questions in the areas of presentation, diagnosis, and treatment of PH associated with diastolic dysfunction (aka diastolic heart failure, pulmonary venous hypertension, and heart failure with preserved ejection fraction [HFpEF]). Dr Soto, representing the pulmonary group who are often asked to evaluate these patients for dyspnea of unclear etiology, gives a well-thought-out strategy to formulate pretest probability of HFpEF versus PAH by integrating the relevant clinical risk factors. How best to utilize echocardiography to differentiate between PAH and HFpEF is eloquently addressed by Drs Raina and Forfia in their detailed explanation of key findings that readily separate the two entities. They also give a step-wise, integrative approach to prioritize echo features to further add to the pretest probability of distinguishing diastolic dysfunction versus PAH – and why the pulmonary artery systolic pressure number may be

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sildenafil group consisted of flushing in 3 patients and headache in 2 patients.

The authors concluded that in this group of stable heart failure patients, long-term use of sildenafil was well tolerated. The results point to some improvement in the LV and diastolic function properties and cardiac geometry. The authors found that these patients improved their functional capacity and clinical status as well.

The benefits of PDE-5 inhibition have been demonstrated in patients with Group 1 PAH in multiple trials. However, the question of whether this class of drugs or other PAH-specific therapies may play a role in those patients who come to our clinics with PH associated with left heart disease needs to be evaluated in larger clinical trials. Currently, the use of these agents remains limited to those who have documented PAH by right heart catheterization. This study, along with others, explores the use of PDE-5 inhibitors in patients with Group 2 PH and may provide rationale for the development of future trials to evaluate patients with Group 2 PH.

Reference

1. Guazzi M, Vicenzi M, Arena R, Guazzi MD. PDE5 inhibition with sildenafil improves left ventricular diastolic function, cardiac geometry, and clinical status in patients with stable systolic heart failure: results of a 1-year prospective, randomized, placebo-controlled study. *Circ Heart Fail.* 2011; 4(1):8-17.

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the least important of all the findings that echocardiogram provides.

Right heart catheterization remains the ultimate arbiter and, like any tool, it is critical for the operator to know how to maneuver and interpret the information correctly. Dr Mathier provides the key do's and don'ts of performing right heart catheterization and how to avoid the pitfalls in interpreting the data. Furthermore, he explores the often asked question "What is PH 'out of proportion'?" Finally, Dr Champion walks us through the therapeutic realm for PH associated with left heart disease, most notably those therapies that have been studied and found to be ineffective. The promising aspects of phosphodiesterase-5 inhibitors are discussed with the need for randomized clinical trials to determine its potential usefulness in this population. The Roundtable discussion engages Drs Alvarez, De Marco, Robbins, and Semigran, who share their views on how HFpEF impacts their clinical practice and how they approach this entity.

Though we may not have a clear picture yet, we are making strides in improving our understanding and coming to appreciate PH associated with diastolic dysfunction. I believe this issue will prove to be a useful resource in managing this increasing group of PH patients.

Finally, I would like to thank Erika Berman Rosenzweig for her encouragement and guidance in completing this issue.

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