

EDITOR'S MEMO

Welcome back all. The summer of 2021 is commencing. As we all step into this new season with optimism and anticipation, it is a great time reflect back on this challenging last year. I would like to thank everyone at PHA, Allen Press, and every member of the editorial board, guest editor, author and panelist who have generously contributed their time, effort and expertise in creating outstanding issues of *Advances*, during a time when none of us *had* a lot of time. These issues have received some of the highest readerships we have seen. It is this enduring dedication by staff and faculty alike who make this unique

journal so well-loved in our community. Thank you to all.

I would like to congratulate Drs. Oksana Shlobin and Anjali Vaidya, the co-guest editors of this edition of *Advances*. The topic is Group 5 PH, an ever changing and ever challenging topic for us all. Drs. Shlobin and Vaidya and their faculty have met the challenge of organizing this difficult group of conditions into a beautifully written resource which touches on the most important subgroups in Group 5. The group focused the round table and PHGR section on Sarcoidosis, a major topic in the 6th WSPH. Readers will turn to these doc-

uments when looking for expert viewpoints on this controversial topic.

I know you will enjoy this issue of *Advances*. This issue will serve as a great resource in 2021 and beyond, as this topic will continue to ebb and flow with more experience and knowledge.

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GUEST EDITOR'S MEMO

Group 5 Pulmonary Hypertension (PH), i.e. the Miscellaneous Group, is a heterogeneous group that includes a myriad of conditions that differ in etiology and pathophysiology. One can argue that Group 5 PH is the most complex group in the current classification of PH, as, in addition to variability of the underlying conditions, the mechanisms of superimposed PH differ vastly, making a proper diagnosis and management more nuanced and challenging.

In this issue of *Advances*, we are very excited to bring to the readers an array of topics, covered by the experts in the field with particular clinical and research interests in specific forms of Group 5 PH.

Drs. Melendres-Groves and Bostwick's article *The Story of Group 5 PH* offers the readers an overview of the historical context of the classification system and how Group 5 has morphed into its current classification. The paper further succinctly describes major subgroups of Group 5 PH including hematologic (chronic hemolytic anemias, myeloproliferative disorders), systemic and metabolic disorders (Pulmonary Langerhans Cell Histiocytosis, Gaucher disease, Glycogen storage disease, Neurofibromatosis, and Sarcoidosis), congenital heart diseases that do not fall into Group 1 PAH, and also covers chronic renal failure and fibrosing mediastinitis.

The Myriad Presentations of Sickle Cell Disease-Related Pulmonary Hypertension

by Drs. Prohaska and Machado is a comprehensive review of the most common hemolytic anemia that fall in Group 5 PH: sickle cell disease. The authors offer an insight and guidance into the pathogenesis, diagnosis, prognosis and treatment of this complex group of patients.

In their article *Metabolic Disorders of Pulmonary Hypertension*, Dr. Oliveros and the guest co-editor of this issue Dr. Vaidya offer a deeper dive into the two metabolic diseases: Glycogen storage diseases and Gaucher's disease. In their paper, they also cover a topic of the complex relationships between PH and thyroid abnormalities.

Sarcoidosis associated pulmonary hypertension (SAPH) was a hot topic during the 6th World Symposium on Pulmonary Hypertension with a discussion centered on whether it should be "housed" in Group 3 rather than Group 5 category. While the supporters of the move made many persuading arguments that in majority of the cases, SAPH occurs in the setting of a significant parenchymal lung disease, at the end, it remained in Group 5, as there are a number of other etiologic processes that can lead to development of PH in this group of patients. Dr. Kristen Alman and authors offer the readers *An Interesting Case of Sarcoidosis Associated Pulmonary Hypertension* describing one of such situations in the PH grand Rounds Section. The topic of SAPH is further

discussed in the roundtable with great insights from Drs. Roxana Sulica, Stacy Mandras, and Bob Baughman, and facilitated by the co-editors of this issue.

We hope that the readers will find this issue thought provoking, educational and practical. It is important for us as a PH community to continue the ongoing discussions regarding this complex agglomerate of distinct conditions. It is our hope that with the increased awareness of Pulmonary Hypertension in general, this often under-recognized and under-appreciated group will be on the forefront of research, as more guidance is needed to help clinicians caring for these complicated patients.

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