## **GUEST EDITOR'S MEMO**

I am honored to serve as Guest Editor for this issue of Advances in Pulmonary Hypertension and thank Dr Harrison Farber (Hap) for the opportunity to serve in this role. In this issue, we focus on scleroderma. Scleroderma is a devastating disease that affects not only the skin, but also many of the internal organs, including the gastrointestinal tract, kidneys, heart, and lungs. It is exactly this multiorgan involvement that makes identifying and treating pulmonary vascular disease in patients with scleroderma so challenging. However, it is also this known risk of developing pulmonary hypertension (PH) that makes scleroderma an ideal paradigm in which to study the pathogenesis of PH and the interplay between autoimmunity and pulmonary vascular disease.

We have assembled a distinguished panel of experts in both scleroderma and pulmonary vascular disease to summarize the current knowledge at the intersection of these complex diseases. First, Professor Christopher Denton from University College London provides a comprehensive review of the clinical aspects of PH in scleroderma and contrasts these features with other

forms of connective tissue disease. Dr Maria Trojanowska from the Arthritis Center at Boston University offers insight into the pathogenesis of PH in the setting of scleroderma, highlighting the advances in our understanding of the pathobiology of these diseases and identifying potential molecular and cellular targets for therapy. Drs. Anklesaria, Rajeev Saggar, Derhovanessian, and Rajan Saggar contribute what, to date, is likely the most complete review of non-Group 1 PH in scleroderma. In this review, the authors not only summarize the epidemiology and impact of various forms of PH in scleroderma, but also proffer practical suggestions for the evaluation and management of patients with non-Group 1 pulmonary vascular disease. Importantly, Sophy Dedopoulos and Dr Sameer Verma focus on the often unseen individuals who are also impacted by scleroderma and PH, the patient caregivers, and emphasize the need to better understand the emotional and physical burden of these diseases on those persons closest to the patient. I have written a short review on a vexing problem in scleroderma, digital ulceration. I find this a particularly relevant

topic for this issue as some pulmonary arterial hypertension medications are often used off-label to treat this problem. Finally, we have a convened a roundtable of experts in both scleroderma and PH, including Dr Virginia Steen from Georgetown University, Dr Laura Hummers from Johns Hopkins University, and Dr Rajan Saggar from the University of California, Los Angeles, to discuss topics such as screening for PH in scleroderma, treatment of non-Group 1 PH in scleroderma, monitoring response to therapies, and novel therapeutic approaches in scleroderma-related PH.

I hope that you find these topics as interesting and the discussions as valuable as I do. Our expectation is that through exploration of these subjects as they pertain to scleroderma, we can better identify and treat the various manifestations of pulmonary vascular disease in the setting of this connective tissue disease.

## Stephen Mathai, MD, MHS

Associate Professor of Medicine Johns Hopkins University Baltimore, Maryland

## EDITOR'S MEMO

It is now Volume 16, Number 2 of *Advances*, and I have the honor of taking over as Editor-in-Chief for the next 8 issues. Will either of us survive this marriage? Well, like any good relationship, I hope that both of us will grow from this experience, expand our horizons, and be far better off at the end. I follow a line of illustrious Editors and hope to build on what they have created thus far. Over these two years, I hope to add some new

features to *Advances*, including basic research perspectives whenever appropriate for the topic of that issue. And, please, give me/us your feedback/suggestions as we incorporate these features.

For the first issue of my tenure, we will be discussing scleroderma and pulmonary hypertension, with Stephen Mathai as the Guest Editor. He has assembled an outstanding group of contributors for this issue, and, as promised,

we have included a review of the current basic science in this field.

So, time to get underway, have fun, and impart some knowledge. Let's rock and roll!

## Harrison (Hap) Farber, MD

Professor of Medicine Director, Pulmonary Hypertension Center Boston University/Boston Medical Center Boston, Massachusetts