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**Cover Image**
Charles Dickens' character Joe from *The Posthumous Papers of the Pickwick Club* is the source of the term Pickwickian Syndrome, or obesity hypoventilation syndrome. He is depicted on the cover in an illustration by David Zwierz.

## Guest Editor's Memo

# Translating Evidence into Clinical Practice

Pulmonary hypertension (PH) associated with lung disease and hypoxia (WHO Group III) is one of the most common forms of pulmonary hypertension. It also encompasses a very diverse group of diseases where the PH is an extremely important determinant of functional limitation and prognosis. In this group of diseases, PH is common in advanced disease, however its prevalence in milder disease is not known. None of the non-invasive screening methods have been found to be accurate and the best screening tool, Doppler echocardiography, is especially prone to error. Treating these patients can be a frustrating experience because of the lack of prospective trials and the associated underlying lung disease. Several questions remain unanswered including the populations to be screened, best diagnostic approach, and treatment options. In the current issue of *Advances*, all these questions have been addressed by some of the key thought-leaders in this field.

In our article on PH in Obstructive Sleep Apnea (OSA), Dr Chua and I have tried to summarize the clinically relevant literature on pulmonary hemodynamics in patients with OSA. We point out that PH in OSA is a multi-factorial process and may have important implications in terms of functional capacity and prognosis in these patients. CPAP therapy may be helpful in patients with mild PH, however more studies are needed to better define the role of PH-specific therapy in patients with more significant PH.

The articles by Dr Klinger on PH in interstitial lung diseases, Dr Girgis on PH in sarcoidosis, and Dr Preston on PH in COPD all provide an exhaustive yet focused review of the literature. These authors make several recommendations that readers can utilize in their clinical practice in terms of diagnostic evaluation and when faced with various treatment dilemmas.

The roundtable is hosted by me and discussants are Drs Nicholas Hill and Steve Nathan, who bring a wealth of experience and wisdom to the discussion. During the

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## Editor's Memo

It is with great excitement that I introduce the Fall 2009 issue of *Advances*, devoted to pulmonary hypertension in the setting of lung or respiratory disease (or WHO Diagnostic Group III). Dr Omar Minai has done an outstanding job of coordinating the issue and contributing one of the articles.

The topic of pulmonary hypertension in patients with underlying pulmonary disorders is critical. What are the mechanisms involved in the development of PH in these patients? How common is PH in this group? How severe? When can we attribute PH in an individual patient to the underlying pulmonary disease and when is the PH "out of proportion"? And importantly, can we treat any of these patients with PAH-specific therapies?

This last question is, indeed, a charged topic. In reality, PAH-specific drugs are already being used in patients with underlying pulmonary disease. Does this represent inappropriate, off-label use of costly therapies, or thoughtful, albeit empiric, application of targeted treatment to patients who may share many of the histopathologic features with more "typical" PAH patients? The truth, of course, is probably somewhere in the middle, but what are the data?

As someone who lectures widely on all aspects of pulmonary hypertension, I am asked after virtually every talk whether the current PAH therapies are ever indicated in patients with PH and underlying lung or respiratory disease, especially COPD. This is not a surprising question. After all, these are the patients that physicians see. A community-based pulmonologist rarely ever sees a 30-year-old woman with idiopathic pulmonary arterial hypertension. What he or she sees regularly is a 65-year-old overweight patient with some underlying sleep apnea, hypertension, and COPD. Some

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**Program Description**

The mission of *Advances in Pulmonary Hypertension* is to serve as the premiere forum for state of the art information regarding diagnosis, pathophysiology, and treatment of pulmonary hypertension. The 2008 Dana Point revision of the World Health Organization Classification serves as a guide to categories of pulmonary hypertension addressed in *Advances in Pulmonary Hypertension*. While focusing on WHO Group I PAH, the other categories (Group II, pulmonary venous hypertension; Group III, Associated with chronic lung disease and/or hypoxemia; Group IV, pulmonary embolic hypertension; Group V, Miscellaneous) are also addressed. This mission is achieved by a combination of invited review articles, Roundtable discussions with panels consisting of international experts in PH, and original contributions. In addition, a special section in selected issues entitled "Profiles in Pulmonary Hypertension" recognizes major contributors to the field and serves as an inspiring reminder of the rich and collegial history of dedication to advancing the field.

**Objectives**

- Provide up-to-date information regarding diagnosis, pathophysiology, and treatment of pulmonary hypertension.
- Serve as a forum for presentation and discussion of important issues in the field, including new paradigms of disease understanding and investigational trial design.
- Recognize and preserve the rich history of individuals who have made major contributions to the field via dedication to patient care, innovative research, and furthering the mission of the PH community to cure pulmonary hypertension.



*The Mission of the Scientific Leadership Council is to provide medical and scientific guidance and support to the PHA by:*

- Developing and disseminating knowledge for diagnosing and treating pulmonary hypertension
- Advocating for patients with pulmonary hypertension
- Increasing involvement of basic and clinical researchers and practitioners

More information on PHA's Scientific Leadership Council and associated committees can be found at: www.PHAssociation.org/SLC/