

Table of Contents

Guest Editor for this issue:

Omar A. Minai, MD

Department of Pulmonary,
Allergy and Critical Care Medicine
Cleveland Clinic
Cleveland, Ohio

- 117 Profiles in Pulmonary Hypertension:
Emmanuel Weitzenblum, MD
- 118 Ask the Expert
- 120 Article Reviews
- 129 Advances in Pulmonary Hypertension CME Section
- 131 Pulmonary Hypertension in Obstructive Sleep Apnea Syndrome
- 141 Diagnosis and Management of Pulmonary Hypertension Associated with Pulmonary Fibrosis
- 148 Pulmonary Hypertension Associated with Sarcoidosis
- 156 Pulmonary Hypertension in Chronic Obstructive Pulmonary Disease: Diagnosis and Management
- 161 CME Self-Assessment Examination
- 163 Pulmonary Hypertension Roundtable: PAH in Patients with Lung Disease and Hypoxia
- 176 Clinical Trials Update
- 178 PHRN Corner

Publisher

Pulmonary Hypertension Association
Carl Hicks, *Board Chair*
Rino Aldrighetti, *President*

Publishing Staff

Managing Editor
Deborah L. McBride
McBride Strategic Services
mcbridedeb@aol.com
Design Director
Michael McClain

PHA Office

Pulmonary Hypertension Association
801 Roeder Road., Ste 400
Silver Spring, MD 20910
301-565-3004, 301-565-3994 (fax)
www.PHAAssociation.org

© 2009 by Pulmonary Hypertension Association. All rights reserved. None of the contents may be reproduced in any form whatsoever without the written permission of PHA.

Advances in Pulmonary Hypertension is circulated to cardiologists, pulmonologists, rheumatologists and other selected physicians by the Pulmonary Hypertension Association. The contents of the articles are independently determined by the Editor and the Editorial Advisory Board.

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
(continued on page 160)

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
(continued on page 160)

Guest Editor's Memo

(continued from inside front cover)

roundtable discussion, we addressed several clinically relevant questions that many of us are faced with everyday including the elusive questions on “disproportionate PH” and the art of choosing who to treat and with what.

I have really enjoyed planning this issue with the expert review of Dr Charles Burger and hope that all of you will come away with important information that will prove useful in caring for your patients.

Omar A. Minai, MD
Guest Editor

Editor's Memo

(continued from inside front cover)

of these patients develop pulmonary hypertension, sometimes with severe right ventricular failure.

The practitioners want to know from us, the experts, what to do to help these patients. We need to provide a framework by

which clinicians can approach and evaluate such a patient when he or she develops PH. I am hopeful that this issue of *Advances* will provide, to the practicing physician, at least parts of that framework.

Richard N. Channick, MD
Editor-in-Chief

Upcoming Events for Medical Professionals Include:

Pulmonary Hypertension Symposium: Optimizing Patient Centered Care in Clinical Practice

February 27, 2010 Coral Gables, Fla.
University of Miami
Miller School of Medicine
Contact info@excelcme.com

The Alfred P. Fishman Conference: Right Ventricular Structure and Function in Health and Disease

April 9 - 10, 2010 Philadelphia, Penn.
University of Pennsylvania
School of Medicine
Contact aspen@upenn.edu

4th Annual Pulmonary Hypertension Symposium: Updates on Classification and Therapies

June 3, 2010 East Haddam, Conn.
Yale School of Medicine
Visit http://cme.yale.edu/conferences/conference_schedule.asp



Building Medical Education in PH

A Partnership Initiative to Advance Medical Understanding of Pulmonary Hypertension

What did this physician hope to achieve by hosting a pulmonary hypertension continuing education event?

“Our PH Symposium is aimed primarily at educating and updating providers. We try to be as inclusive as possible by inviting providers from all over New England to serve on the faculty, and inviting nurses, other health care professionals and patients in addition to physicians. We had an excellent turnout in 2008 and very enthusiastic feedback. PHA has been a very important component for the patient attendees.”

Nicholas Hill, M.D.
6th Annual Update in Pulmonary Hypertension
Tufts Medical Center

Building Medical Education in PH events are designed to foster partnerships between PHA and PH Centers to promote continuing education in the field of pulmonary hypertension through CME educational events.

To partner with PHA in *Building Medical Education in PH* for your upcoming CME event, please contact Emily Koenig, Medical Education Program Associate, at 301-565-3004 X776 or Emily@PHAssociation.org.