

Table of Contents

Guest Editors for this issue:

Robert Schilz, DO, PhD

Medical Director of Lung
Transplantation and Pulmonary
Vascular Disease

University Hospital of Cleveland
Case Western Reserve University
Cleveland, Ohio

Francisco J. Soto, MD, MS

Assistant Professor
Director, Pulmonary Hypertension
Program

Medical College of Wisconsin
Milwaukee, Wisconsin

- 160 Profiles in Pulmonary Hypertension: Sir Magdi Yacoub
- 168 Pulmonary Venous Hypertension: A Diagnostic and Therapeutic Dilemma
- 176 The Vasoreactive Patient: Diagnosis, Treatment, and Follow-up
- 182 Chronic Thromboembolic Disease: Underdiagnosis and Nonsurgical Options
- 186 Lung Transplantation and Idiopathic PAH: A Case Study
- 190 Case of Catastrophic Antiphospholipid Syndrome: Roundtable Discussion

Publisher

Pulmonary Hypertension Association
Michael D. McGoon, MD, *Chair of the Board*
Rino Aldrighetti, *President*
Rachel Pokorney, *Associate Director of Medical Services*

PHA Office

Pulmonary Hypertension Association
801 Roeder Rd. Suite 400
Silver Spring, MD 20910-4496
301-565-3004, 301-565-3994 (fax)
www.phassociation.org

© 2008 by Pulmonary Hypertension Association. All rights reserved. None of the contents may be reproduced in any form whatsoever without the written permission of PHA.

ISSN: 1933-088X (print); 1933-0898 (online)

Editorial Offices

Advances in Pulmonary Hypertension, DataMedica,
P.O. Box 1688, Westhampton Beach, NY 11978
Tel (631) 288-7733 Fax (631) 288-7744
E-mail: sbelsenchapman@aol.com

Advances in Pulmonary Hypertension is circulated to cardiologists, pulmonologists, rheumatologists, and other selected physicians by the Pulmonary Hypertension Association. The contents are independently determined by the Editor and the Editorial Advisory Board. All past issues of the journal are available at:
www.PHAssociation.org/Medical/Advances_in_PH/

Cover Image

Unofficially called "The Case Discussion" by PHA editorial staff, this painting by Jamil Aboulhosen, MD, is one of several for *Advances in Pulmonary Hypertension*. Dr. Aboulhosen is an assistant professor of medicine (cardiology) at the UCLA School of Medicine. His clinical interests include pulmonary hypertension and adult congenital heart disease as well as general cardiology. He is a member of the Ahmanson/UCLA Adult Congenital Heart Disease Center. Dr. Aboulhosen paints "for fun," pursuing topics from medicine to landscapes and figures. His medical illustrations have been published in journals and textbooks, including *Circulation* and the latest *Hurst's the Heart* (12th ed).

Editor's Memo

Marking a Milestone for the Journal and Previewing PHA's Upcoming Conference and Scientific Sessions



This issue of *Advances in Pulmonary Hypertension* is a milestone in the journal's history: it is the first to produce a series of clinical cases and expert commentaries since the journal's inception in 2002. The Editorial Board and I were excited when the idea was conceived. Now that the "cases" issue is in print, I am happy to see that we were right to be energized about this issue. Publishing these cases is one of several components introducing new dimensions in PHA's tireless efforts in educating the world about PAH:

1) Formal case presentations presenting practical approaches to PAH diagnosis and management, 2) CD-ROM-based case presentations, with interactive multimedia (coming soon), and 3) On-line "live" case discussions via Pulmonary Hypertension Clinicians and Researchers (you must be a PHCR member to view and participate in these discussions).

The many contributors to this issue deserve a great deal of thanks for their efforts in helping to produce it. These include our Editorial Board members Drs Francisco J. Soto, Robert Schilz, Charles D. Burger, Srinivas Murali, E. Clinton Lawrence, and Micah R. Fisher.

This issue also coincides with the opening of registration for PHA's 8th International Pulmonary Hypertension Conference and Scientific Sessions, which will be held on June 20-22 in Houston, Texas. The theme of the 2008 Scientific Sessions, a daylong educational program specifically for medical professionals who work in the field of pulmonary hypertension, is *Determinants of RV Function on Molecular, Pharmacogenomic, and Metabolic Levels*. PHA's biennial conference is the largest meeting of its kind and is unique in bringing together pulmonary hypertension patients, caregivers, and medical professionals from around the world. For more information on this exciting event, be sure to see the ad in this issue of *Advances*. As someone who has attended past PHA Conferences and worked to develop their programming, as well as the 2008 Conference program, I can assure you that this will be an exceptional weekend.

Ronald J. Oudiz, MD

Editor-in-Chief

Guest Editor's Commentary

Pulmonary Hypertension: Challenges and Controversies in Diagnosis and Management



The case presentations and discussions in this issue of *Advances* illustrate both common and uncommon challenges in the diagnosis and management of patients with pulmonary hypertension. Although significant guidelines and evidence-based recommendations exist for patient evaluation and care, practical application of these guidelines and managing uncertainty in areas where evidence may be sparse are key elements in the daily activities of specialized pulmonary hypertension clinics.

The contributors present case-based analyses exploring important concepts and controversies in pulmonary venous hypertension, transplantation, vasoreactivity, chronic thromboembolic disease, and catastrophic antiphospholipid syndrome. Two of the cases not only illustrate important concepts but are examples of notable

(continued on page 179)