# **Table of Contents**

# Guest Editors for this issue:

### Robert Frantz, MD

Consultant in Cardiovascular Diseases and Internal Medicine Assistant Professor of Medicine Mayo Clinic College of Medicine Rochester, Minnesota

# Srinivas Murali, MD

Professor of Medicine
Drexel University College of Medicine
Director, Division of Cardiovascular
Medicine
Medical Director, Gerald McGinnis
Cardiovascular Institute
Allegheny General Hospital
Pittsburgh, Pennsylvania

- 4 Profiles in Pulmonary Hypertension: Victor F. Tapson, MD
- 13 Oral Therapies for PAH: State-ofthe-Art and Investigational Approaches
- 18 Combination Therapy for PAH: Current Rationale, Future Concepts
- 23 Advances in Prostanoid Therapy: New Studies, New Methods of Delivery
- 32 Pulmonary Hypertension Roundtable: Recapping 5 Years, Exploring Emerging Approaches

#### **Publisher**

Pulmonary Hypertension Association Michael D. McGoon, MD, Chair of the Board Rino Aldrighetti, President Justine Elliot, Director of Medical Services

### **Publishing Staff**

Stu Chapman, Executive Editor Susan Chapman, Managing Editor Heidi Green, Associate Editor Gloria Catalano, Production Director Michael McClain, Design Director

#### **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

© 2006 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) 288-7734 Fax (631) 288-7744 E-mail: sbelsonchapman@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 Pulmonary Hypertension are available at: www.phassociation.org/Medical/Advances\_in\_PH/

#### Cover image:

Targets for current or emerging therapies in pulmonary arterial hypertension. Three major pathways involved in abnormal proliferation and contraction of the smooth-muscle cells of the pulmonary artery in patients with pulmonary arterial hypertension are shown. (Reprinted with permission of *New England Journal of Medicine*; Humbert et al. 2004;351:1425-36.)

# Editor's Memo

# Reflecting on Progress in PH and Appreciating the Contributions of Our Physicians



The theme of this year-end issue is "emergence" as we survey the ever-expanding role of new therapies, from emerging combinations to new orally administered agents to advances in prostanoid treatment. As the acronyms of new trials have worked their way into our lexicon, we have grown accustomed to refer to new studies such as VISION, TRIUMPH, COMPASS, FREEDOM, and PHIRST, and await the latest results that will help guide

patient management choices in the future. Emergence is the theme of the contents, but it also applies to the role of physicians on our Editorial Boards as we welcome physicians who have stepped forward to help guide the journal in the years ahead and express appreciation to those who have contributed their time and energy.

Medical journals thrive on an infusion of new ideas reflected in the transition to new editors-in-chief and editorial advisory boards. By passing the torch to a new group of editors we promote an even greater exchange of ideas and keep the level of enthusiasm as high as it can be for producing the most comprehensive source of information on pulmonary hypertension available to practicing physicians. Somewhat like members of Congress but, thankfully, in a completely different arena, we also must observe term limits. In our case we have a self-imposed 2-year limit on the verge of expiring, and I wish to welcome physicians who will help guide this journal through the end of 2008.

I am pleased to welcome a new Editor-in-Chief, Ronald J. Oudiz, MD, beginning with the next issue of *Advances in Pulmonary Hypertension*. Dr Oudiz is an exemplary physician, a distinguished colleague, and a close personal friend who has demonstrated a tremendously strong commitment to the pulmonary hypertension community for many years. In turning over the job of leading the journal, I am confident that he will strive for the same editorial independence, unbiased perspective, and excellence that we have established over the first 5 years of the journal's existence and that characterize our approach to developing scientific content.

Dr Oudiz has already contributed in many ways to the selection of relevant and timely topics and to development of our content. His pivotal role in programs offered by the Pulmonary Hypertension Association (PHA) speaks volumes for the dedication he has shown to medical research, patient advocacy, and quality of care. I wish him well in this new endeavor during 2007 and 2008.

No journal can succeed without a supporting cast of physicians whose experience at the bench and bedside helps to create reference points from which we can select appropriate topics and a context for chronicling the evolution in care. During the last 2 years we have been graced with the contribution of three outstanding clinicians who served as Associate Editors: Ramona Doyle, MD, Karen Fagan, MD, and Olivier Sitbon, MD. We thank them for their service to the journal and helping us in the kind of peer review essential to the journal's integrity.

We are also pleased to welcome back to our Editorial Advisory Board Richard Channick, MD, as an Associate Editor and Editor-in-Chief Elect. His enthusiastic participation in Roundtable discussions over the years has given readers a clear and thoughtful perspective on important clinical issues. Similarly, we welcome (continued on page 39)

stress, you can have patients referred for at least a one-time visit. But we all approach patients a little differently and there are different thresholds for treating patients that we use. There is not one way to do it. I don't know how you can enforce any standards, really.

Dr McLaughlin: Vic, Rich, Ivan, thanks again for your participation. As always, it has been great working with you. ■

#### **Editor's Memo**

(continued from page 2)

Erika Berman Rosenzweig, MD, to the team of Associate Editors. Her focus and enthusiasm for the journal are refreshing and will help guide content in 2007.

We will benefit also from new input by physicians who are joining our Editorial Board, including Kristin Highland, MD, Ioana Preston, MD, Zeenat Safdar, MD, Rajan Saggar, MD, and Francisco Soto, MD. They will be taking over from physicians whose contribution as Editorial Board members is also much appreciated: Gregory Ahearn, MD, Jacques Benisty, MD, Raymond Benza, MD, and Jeffrey Edelman, MD.

We have seen significant progress in our effort to provide more hope to patients with pulmonary hypertension and I am honored to have been able to work with my colleagues and serve as Editor-in-Chief during the last 2 years. I also look forward to continued involvement with the journal and its outstanding educational program for more than 30,000 physicians engaged in pulmonary hypertension care.

I am sure I speak for all of our physicians and PHA staff in extending our best wishes for a joyous holiday season and a healthy and happy new year.

Vallerie V. McLaughlin, MD Editor-in-Chief