Table of Contents

Guest Editors for this issue:

Erika Berman Rosenzweig, MD Assistant Professor of Pediatrics Department of Pediatrics Columbia College of Physicians and Surgeons New York, New York

Kristin Highland, MD

Assistant Professor
Division of Pulmonary and
Critical Care
Director, Pulmonary Hypertension
Clinic
Medical University of South Carolina
Charleston, South Carolina

- 108 Profiles in PH: Joseph K. Perloff, MD
- 121 Pulmonary Arterial Hypertension in Adults with Congenital Heart Disease: General Overview of Disease Mechanisms
- 126 Evaluating Operability in Adults with Congenital Heart Disease and the Role of Pretreatment with Targeted Pulmonary Arterial Hypertension Therapy
- 136 Emerging Therapy of Congenital Heart Disease Associated with Pulmonary Hypertension
- 142 Pulmonary Hypertension Roundtable

Publisher

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

© 2007 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: 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

Diagnostic tools used to evaluate and define shown perimembranous ventricular septal defect (center). From top left, clockwise: 2-D echocardiogram and continuous wave Doppler assessment of tricuspid regurgitant velocity, electrocardiogram, contrast ventriculography, and simultaneous RV and LV pressure tracing.

Editor's Memo

Another Milestone for PHA Mission to Promote Awareness



With this third issue of *Advances in Pulmonary Hypertension* in 2007, I must first recognize the enormous success of the Pulmonary Hypertension Resource Network Symposium held in Crystal City, Virginia this October. Attendance was a record 400+ (up from 60 in 2003), and the representation was far-reaching, with attendees and participants from Oregon to Washington, DC, and Canada to Texas. This clearly reflects the tremendous interest in this growing field and speaks to the success of PHA

in its mission to promote pulmonary hypertension awareness.

Associate Editor Erika Berman Rosenzweig, MD, has taken the role of Lead Editor of this issue and with Editorial Board member Kristin Highland, MD, has put together a comprehensive review of pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD), with three key pieces focused on CHD. Dr Ingram Schulze-Neick provides an overview of pulmonary vascular disease in CHD, while Dr Michael Landzberg covers emerging medical therapies, and Drs Konstantinos Dimopoulos and Michael Gatzoulis teach us about how to evaluate operability in adults with CHD, as well as the role of pretreatment with targeted PAH therapy. A specialist roundtable discussion is a highlight of this issue, as it covers the practical aspects of treating patients with CHD and associated PAH. Dr Berman Rosenzweig and her contributors are to be congratulated for their efforts in producing such a wonderful issue of *Advances*.

Ronald J. Oudiz, MD Editor-in-Chief

Guest Editor's Commentary

Focusing on Pulmonary Arterial Hypertension in Adults with Congenital Heart Disease



This issue of *Advances in Pulmonary Hypertension* focuses on the growing population of adults with congenital heart disease (CHD) and associated pulmonary arterial hypertension (PAH). Although progress in diagnostics and surgical intervention has dramatically decreased the incidence of classic Eisenmenger syndrome in the Western world, these advances have led to a rapidly growing number of adults with CHD and associated PAH.

In this journal edition, the authors highlight some of the considerations and challenges specific to the adult with PAH associated with CHD. Admittedly, there are still more questions than answers for these patients; the authors hope to bring to light some of the important issues that require further study.

Erika Berman Rosenzweig, MD, Guest Editor

Assistant Professor of Pediatrics (in Medicine)
Division of Pediatric Cardiology
Columbia University College of Physicians and Surgeons
New York, New York