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Cover Image

Diagnostic modalities in pulmonary hypertension. Clockwise, starting at the top: chest X ray shows an implantable device for continuous hemodynamic monitoring; cardiac magnetic resonance evaluates the performance of a severely dilated right ventricle (RV); RV pressure overload creates a D-shaped septum on a short-axis echocardiogram image; mediastinal window image of a thoracic computed tomography shows severe enlargement of right-heart chambers; chest X ray shows a dilated RV and confirms the correct position of a pulmonary artery catheter tip. Background: hemodynamic tracings showing simultaneous left ventricular pressure (red) and pulmonary capillary wedge pressure (blue) waveforms.

Guest Editor's Memo

A Definitive Guide to New Applications in Diagnosis



This issue of the journal focuses on current diagnostic modalities used to diagnose and manage patients with pulmonary hypertension (PH) and especially the more complex and severe form, pulmonary arterial hypertension (PAH). Thanks to the impressive evolution of this field in recent years, physicians who take care of patients with PH are being faced with a very diverse population in whom PH etiology is frequently multifactorial. While potentially life saving, the cost and complexity of some of the treatments available for PAH mandate that clinicians perform a comprehensive evaluation of patients to confirm or exclude its presence. Likewise, given the high risk for rapid deterioration—especially in patients with PAH—we must use a variety of tests to closely monitor treatment response.

In the first half of this issue, the authors discuss new applications and selective information obtained from the more established diagnostic tests such as echocardiography and right heart catheterization. Potential applications of newer diagnostic tests such as magnetic resonance of the heart and continuous hemodynamic monitoring through implantable devices are also discussed. Some areas of this issue will also shed light on complex case-scenarios that clinicians who treat PH currently face: exercise-induced PH and pulmonary vascular response in patients with nonsystolic heart failure (ie, diastolic dysfunction).

Given the amount of unanswered questions in this field and the significant complexity of the many cases we evaluate, correctly diagnosing and managing PH has truly become an art. The authors who participated in this issue sincerely hope that the information contained in these pages will give the PH-treating community additional tools to establish a more accurate diagnosis and promptly identify signs of disease progression that require therapeutic interventions.

Francisco J. Soto, MD, MS

Guest Editor

Editor's Memo



In September, 2007 the 3rd Pulmonary Hypertension (PH) Resource Network Symposium held in Crystal City, Virginia was a fantastic success, with a huge (>350) attendance of health professionals including nurses, nurse practitioners, and respiratory therapists convening to teach, learn and network with other allied health professionals. This year, Pulmonary Hypertension Resource Network hopes to outdo it self again. The program, titled "Leading Progress, Creating Partnerships: Empowering the Interdisciplinary PH Team" will again be held in Crystal City, VA, September 24-26, 2009.

In addition to the PH Resource Network symposium, several additional PHA programs have been in development this past year, all focused on improving education for PH practitioners and patients. These programs have been enabled by the recently created PHA Medical Education Fund, with \$2 million in unrestricted educational industry grants, and include the PH Preceptorship program, PHA Online University, and the 30 City Program and PHA on the Road Educational programs for PH patients. Undeniably, this "explosion" of PH programs in medical education reflects the rapidly growing interest in this field and speaks to the overwhelming success of PHA in its mission to find ways to prevent and cure PH, and to provide hope for the PH community through support, education, advocacy, and awareness.

In addition to these medical education programs, we have seen rapid advances in therapies for PAH which have enabled practitioners to choose from several targeted therapies for their patients. During this time, we have also gained a better understanding and appreciation for the multitude of diagnostic and prognostic modalities that have resulted from improvements in biotechnology, and we can now better appreciate the physiological aberrations caused by pulmonary vascular disease in our patients.

There are many scientists and clinicians with refined expertise in echocardiography and in MRI as well as in invasive hemodynamics and exercise physiology, not to mention those who avidly study a host of clinically relevant biomarkers. Each expert can readily

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Advances in Pulmonary Hypertension

Author Guidelines 2008

Scope of Manuscripts

Advances in Pulmonary Hypertension considers the following types of manuscripts for publication:

- Reviews that summarize and synthesize peer-reviewed literature to date on relevant topics in a scholarly fashion and format
- Letters to the Editor
- Clinical Case Studies

Manuscript Submission

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Manuscript Preparation

Length: Full-length manuscripts should not exceed 4,000 words, including references. Please limit the reference list to 50 citations. Manuscripts should be accompanied by figures and/or tables. Generally, 4 to 5 figures and 2 to 3 tables are preferred for each manuscript. Please include a brief description to accompany these items, as well as a key for all abbreviated words.

Spacing: One space after commas and periods. Manuscripts should be double spaced. Manuscripts should not contain an abstract but an introduction is recommended.

References: All submissions should include numbered references that are referred to in the text by superscripts and that conform to AMA style. Example: Lewczuk J, Piszko P, Jagas J, et al. Prognostic factors in medically treated patients with chronic pulmonary embolism. *Chest*. 2001;119:818-823.

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(continued from inside front cover)

demonstrate the many attributes that their specialized modality offers, and can provide a unique clinicopathologic perspective on pulmonary vascular disease.

Dr Francisco Soto served as the Guest Editor for this issue, which focuses on several diagnostic modalities used to study PAH patients, and provides a view from the experts that have studied these modalities in great detail. From this information, it is clear that several markers of PAH severity can be considered valuable tools that enhance our understanding of PAH pathophysiology, but no single test can be accepted as a gold standard. Indeed, as

evidenced by the Expert Roundtable in this issue, some of the issues surrounding the utility and performance of these tests have been somewhat controversial.

It is my hope that the contents of this issue will not only help your understanding of the diagnostic challenges facing PAH clinicians, but also stimulate new growth in this field and catalyze additional research initiatives to further our knowledge base in pulmonary vascular disease.

Ronald J. Oudiz, MD
Editor-in-Chief