**Program Overview:** Pulmonary arterial hypertension (PAH), an incurable disease, is characterized by medial hypertrophy, intimal fibrosis, and in situ thrombi in small muscular pulmonary arteries. PAH was considered a rapidly fatal illness with a median survival of 2.8 years in the 1980s when no evidence-based therapies were available. Since then the treatment of this disease has made tremendous advances, and in the last 10 years the discovery of new medications have positively influenced the prognosis and survival of patients with PAH.

This self-study activity is based on 4 articles that review the management of patients with congenital heart disease and pulmonary hypertension.

This activity is jointly sponsored by Washington University School of Medicine and the Pulmonary Hypertension Association.

**Target Audience:** This self-study activity is appropriate for cardiologists, pulmonologists, rheumatologists, and other physicians who treat patients with PH.

**Learning Objectives:** Upon completion of this activity, participants will be able to:

1. Describe the anatomy of the most common congenital heart defects (and their repairs) that are associated with the development of PAH.
2. Demonstrate the importance of simple and advanced imaging technique in adults with congenital heart disease and PH as diagnostic and risk stratifying tools.
3. Understand the complex interplay between pulmonary blood flow and pulmonary vascular resistance and the techniques (medicinal, catheter-based, and surgical) that are used to modify it.
4. Discuss the clinical studies that have established the basis for pharmacology therapy, and explore the new therapeutic frontiers in patients with congenital heart disease and PH.

**Self-Assessment Examination:** See pages 196 and 197 for self-assessment questions, answer key, and evaluation form.

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**Accreditation Statement:** This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint sponsorship of Washington University School of Medicine and the Pulmonary Hypertension Association. Washington University School of Medicine is accredited by the ACCME to provide continuing medical education to physicians.

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**Instructions for Earning Credit:** This activity is a self-study program; a self-assessment examination is included on page 196 to help physicians review important points. A form is also included on page 197 for physicians to evaluate the CME activity. Completion of this activity involves reading the journal and completing the self-assessment examination and evaluation form with a passing grade of 70% or higher, which may take up to 2 hours. Credits for this self-study program are available from May 31, 2013 through April 30, 2014. There is no fee for this program. Please note that this self-study program may also be viewed online at https://cme-online.wustl.edu/pha.

**Accreditation Statement:** Department of Continuing Medical Education, Washington University School of Medicine, Campus Box 8063, 660 South Euclid Ave., St. Louis, MO 63110

**Disclosures:** The Accreditation Council for Continuing Medical Education and the Association of American Colleges have standards and guidelines to ensure that individuals participating in CME activities are aware of relationships between authors and commercial companies that could potentially affect the information pre-

*(Continued on page 196)*

Washington University in St. Louis
School of Medicine

## Self-Assessment **Examination**

See answer key on next page

1. **Aggressive monitoring for the development of PAH should occur in Fontan patients because:**
   a. They often have a mean pulmonary pressure >25 mm Hg at right heart catherization
   b. An Eisenmenger physiology is associated in most cases
   c. A conduit obstruction may lead to cardiac decompensation
   d. Even slight increase in PVR may have significant hemodynamic consequences
2. **Which of the following forms of congenital heart disease is most likely to lead to the development of PAH?**
   a. Partial anomalous pulmonary venous return without repair
   b. Secundum atrial septal defect without repair
   c. An unrestricted ventricular septal defect without prior repair
   d. Patent foramen ovale without repair
3. **A 30-year-old woman is diagnosed with a membranous VSD. She undergoes echocardiographic imaging evaluation and an invasive hemodynamic study. The hemodynamic study demonstrates pulmonary artery pressures of 110/50 mm Hg. Her central aortic pressure is 100/60 mm Hg. A pulmonary venous oximetry sample has a saturation of 95% and her femoral artery oximetry sample has a saturation of 86%. There is severe pulmonary regurgitation, as well. Which of the following is correct regarding treatment?**
   a. Closure of the VSD is indicated
   b. PAH vasodilator therapy only is indicated
   c. Heart transplantation should be considered
   d. She should have pulmonary valve replacement
4. **Which of the following is the most common subtype of ASD?**
   a. Muscular
   b. Outlet
   c. Secundum
   d. Sinus venosus
   e. Membranous
5. **Echocardiographic indicators of poor outcomes in patients with Eisenmenger syndrome are:**
   a. Pericardial effusion + low TAPSE
   b. Low TAPSE + shortened RV filling time + increased RA area/LA area
   c. Pericardial effusion + low TAPSE + shortened RV filling time
   d. Bi-atrial enlargement
6. **In Eisenmenger patients, the presence of RV late gadolinium enhancement at cardiac MRI:**
   a. Is a pathologic finding in any cases
   b. Is usually located at the apex
   c. Is a normal feature typically evident at the insertion points
   d. When present at the insertion points is an indicator of poor outcome
7. **Catheter-based interventions are available for all of the following lesions except:**
   a. Secundum atrial septal defect
   b. Muscular ventricular septal defect
   c. Ostium primum atrial septal defect
   d. Patent ductus arteriosus
8. **In a 4-month-old infant with APAH-CHD due to a nonrestrictive VSD, what would you most expect hemodynamics to resemble on cardiac catheterization?**
   a. Elevated pulmonary artery pressure, normal wedge pressure, elevated pulmonary blood flow, normal pulmonary vascular resistance
   b. Elevated pulmonary artery pressure, elevated wedge pressure, normal pulmonary blood flow, normal pulmonary vascular resistance
   c. Normal pulmonary artery pressure, normal wedge pressure, normal pulmonary blood flow, normal pulmonary vascular resistance
   d. Elevated pulmonary artery pressure, normal wedge pressure, normal pulmonary blood flow, elevated pulmonary vascular resistance
9. **The BREATHE-5 trial involving which endothelin receptor antagonist was the first randomized, double-blind, placebo-controlled drug trail conducted solely on Eisenmenger patients?**
   a. Sildenafil
   b. Bosentan
   c. Ambrisentan
   d. Treprostinil

Disclosures

*(continued from page 165)*

sented. To be disclosed to participants are all personal financial relationships with a com-mercial interest whose products are relevant to the content of this CME activity. It is the policy of Washington University School of Medicine, Continuing Medical Education, to ensure balance, independence, objectivity, and scientific rigor in all its educational activities. All faculty participating in this activity are expected to disclose to the audience any financial interest or other potential conflict. Each author was asked to complete a disclosure information form for this activity. Disclosures are reported below:

Dr Aboulhosn has no financial relationships to disclose. Dr Babu-Narayan has no financial relationships to disclose. Dr Bashore has no financial relationships to disclose. Dr Gatzoulis has no financial relationships to disclose. Dr Kiefer has no financial relationships to disclose. Dr Li has no financial relationships to disclose. Dr Rubens has no financial relationships to disclose. Dr Scognamiglio has no financial relationships to disclose. Dr Zuckerman has no financial relationships to disclose.

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