JACC: CARDIOVASCULAR IMAGING
VOL. 9, NO. 6, 2016

ISSN 1936-878X/$36.00
PUBLISHED BY ELSEVIER
http://dx.doi.org/10.1016/j.jcmg.2016.02.022

# Follow-Up of Pulmonary Hypertension With Echocardiography

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**CME Objective for This Article:** After reading this article the reader should be able to: 1) review the current recommended echocardiographic techniques for follow up assessment of patients with pulmonary arterial hypertension; 2) understand the strengths and limitations of our current techniques, and physiological settings which may hamper accuracy, and lead to mismanagement; and 3) understand new techniques, and how to incorporate novel markers of RV function (RV free wall strain) into our current follow up.

**CME Editor Disclosure:** *JACC: Cardiovascular Imaging* CME Editor Ragavendra R. Baliga, MD, has reported that he has no relationships to disclose.

**Author Disclosure:** Dr. Dwyer has served on the advisory boards of Actelion, GlaxoSmithKline, and Bayer. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

**Medium of Participation:** Print (article only); online (article and quiz).

### CME Term of Approval

Issue Date: June 2016
Expiration Date: May 31, 2017

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Manuscript received December 21, 2015; revised manuscript received February 8, 2016, accepted February 11, 2016.

JACC: CARDIOVASCULAR IMAGING, VOL. 9, NO. 6, 2016
JUNE 2016:733–46

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## ABSTRACT

Individual patient response to effective therapies for pulmonary hypertension (PAH) is variable and difficult to quantify. Consequently, management decisions regarding initiation and continuation of therapy are highly dependent on the results of investigations. Registry data show that changes in cardiac index, mean right atrial pressure, and mean pulmonary artery pressure have the greatest influence on survival. It is recognized that pulmonary artery pressure (PASP) responses to PAH-specific drugs are heterogeneous. However, follow-up testing is strongly focused on assessing changes in PASP and functional status (6-min walk). The goals of therapy, which should be highlighted in follow-up imaging, include not only reduction of PASP, decrease in pulmonary vascular resistance, and improvements in right ventricular function, cardiac output, and tricuspid regurgitation. This paper reviews the echocardiographic follow-up of pulmonary hypertension, and especially focuses on right ventricular function—a major determinant of outcome, for which reliable echocardiographic assessment has become more feasible. (J Am Coll Cardiol Img 2016;9:733–46) © 2016 by the American College of Cardiology Foundation.

In patients with pulmonary arterial hypertension (PAH) treated with pulmonary vasodilators, continuation of therapy is conditional on the demonstration of treatment effect. Although assessment of response in early clinical trials of pulmonary vasodilator therapy was based on pulmonary artery (PA) systolic pressure (PASP), these trials were limited by small sample size, with often limited follow up (1). The assumption that pressure changes reflect mortality has been disputed by clinical trials and systematic reviews, and it is recognized that PASP responses to PAH-specific drugs are heterogeneous, with differences related to sex, race and disease patterns (2). Mortality rates in PAH trials have improved, with an average 5 year survival of 61%, increasing to 75% in those aged under 50 years (3). Hence, the endpoints of PAH trials are under review, with recommendations that invasive hemodynamics should only be used as a secondary endpoint (4).

As the risk and cost of performing right heart catheterization (RHC) make this test unattractive for 6-month follow-up, Doppler is used to document changes in cardiac function, and the 6-min walk (6MW) test is used to assess functional performance. The use of these methods is based on Level of Evidence: C, supported by expert opinion and small retrospective studies and registries (5). Moreover, there are important differences between echocardiographic and invasive measurements (5). Clinical trials have established a number of correlates of mortality in PAH patients. These range from measures such as baseline New York Heart Association functional class and heart rate, 6MW, echocardiographic measures and invasive measures of mixed venous $O_2$ saturation and prostacyclin. The National Institutes of Health registry shows that changes in cardiac index, mean right atrial pressure (mRAP), and mean pulmonary artery pressure (mPAP) have the greatest influence on survival. However, right ventricular (RV) impairment has important prognostic implications, and the presence of RV myocardial disease cannot be distinguished from RV impairment due to pressure overload unless RV afterload can be estimated.

## HEMODYNAMIC ASSESSMENT FOR DIAGNOSIS AND FOLLOW-UP

**1. RESTING HEMODYNAMICS.** The use of RHC to document a mean PASP >25 mm Hg is considered a necessity for PAH diagnosis (**Table 1**), although this is also supported by only Level of Evidence: C (5).

Complete hemodynamic assessment includes measurements of right atrial (RA), RV and pulmonary capillary wedge pressures, cardiac output (Fick or thermodilution), and mixed venous $O_2$. The normal pulmonary circulation is characterized by low pulmonary vascular resistance (PVR), with compliant vessels. The PVR (in dynes or Wood units [WU]) is calculated from the ratio of diastolic pulmonary gradient or transpulmonary gradient and cardiac output, and this is especially important for diagnosis of left heart-derived pulmonary hypertension.