Is Magnetic Resonance Imaging of Right Ventricular Volume Useful Clinically for Evaluation of Pulmonary Arterial Hypertension?

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Pulmonary hypertension (PH) is a rare but life-threatening cardiovascular disease characterized by a pathological increase in resistance in the pulmonary circulation. Pulmonary arterial hypertension (PAH) is a subtype of PH that is progressive and debilitating. The United States national prospective registry for primary PH reports the median survival for the idiopathic form of PAH to be only 2.8 years without specific treatment. The 1-, 3-, and 5-year survival rates are 68%, 48%, and 34%, respectively.

Our understanding of right ventricle (RV) performance in PAH has been hindered by a lack of techniques that give a reliable picture of RV morphology and function. Nevertheless, there have been recent major advances in our understanding of the mechanism of disease development in PAH and of diagnosis and treatment. Treatment options include modification of 3 different pathways by using prostacyclin analogs, endothelin-receptor antagonists, and phosphodiesterase type 5 inhibitors. Therapeutic advances in management have reinforced the requirement for noninvasive, accurate, and reproducible methods of assessment to yield endpoints for measuring the effects of treatment. New diagnostic and therapeutic targets for heart failure must therefore be established quickly.

Over the past 2 decades, improvements in the spatial and temporal resolution of magnetic resonance imaging (MRI) and in post-acquisition analytic algorithms have led to increased incorporation of MRI as a key element in numerous aspects of cardiovascular assessment and treatment. Cardiac magnetic resolution (CMR) imaging is an attractive modality for studying the complex geometry of the RV. This highly dynamic, rapidly evolving family of technologies has affected the recognition and management of PAH, and CMR is now regarded as the reference standard in the assessment of RV structure and function via the measurement of RV volumes and ejection fractions (EFs). CMR is likely to supplant invasive or radiation-based techniques for evaluating myocardial function. The advantages of CMR are its noninvasiveness, safety (it is non-ionizing), excellent spatial and temporal resolution, and repeatability. Moreover, CMR imaging enables a unique combination of morphological and functional assessment of the RV and pulmonary circulation. CMR is more reproducible than echocardiography for measuring RV function.

PAH affects the right side of the heart and results in RV hypertrophy and dilation and right atrial enlargement. The functional capacity of the RV is a major prognostic determinant in PAH. It is unknown why some patients with markedly elevated pulmonary artery pressure maintain well-preserved cardiac function for several years, whereas others with equal or less severe PAH suffer rapidly progressive right-sided heart failure. One factor that has hindered the understanding of RV performance in patients with PAH has been the lack of techniques that give a reliable picture of RV morphological and functional change in the face of increasing outflow obstruction.

In this issue of the Journal, Yamada et al review the CMR results from 121 consecutive examinations of patients who either had, or were suspected of having, PH. Of the 121 patients, 42 were diagnosed with idiopathic PAH and served as the study group. The RV end-diastolic volume index was an independent predictor of both hospitalization and mortality. The study focused on patients with idiopathic PAH, because survival curves are not comparable across all subgroups of category-1 PAH.

Which method is the best for evaluating RV function? In the clinical setting, echocardiography, radionuclide ventriculography, and MRI are available, but all have both merits and demerits in terms of invasiveness and quantitative capability. Van Wolferen et al found that a triad of prognostic signs (large RV volume, low stroke volume, and reduced left ventricle (LV) volume) were independent predictors of poor prognosis in patients with PAH. They suggested that increased RV end-diastolic volume may be the most appropriate marker of progressive RV failure in follow-up. However, the prognostic significance of LV shrinkage suggests that interventions aimed at better LV filling, such as atrial septostomy, should not be neglected. The possibility that RV stroke volume changes may be more relevant than changes in the cardiac index is quite interesting. Unlike healthy subjects, most patients with PAH...
fail to increase their RV stroke volume during exercise. In the light of this, the results of CMR are more reproducible than those of echocardiography when measuring RV function.

RV structural and functional assessment plays a central role in both diagnosis and serial follow-up of patients with PAH. However, the geometry of the RV is complex, and it is difficult to evaluate its contractile motion and functional parameters given the limitations of traditional 2-dimensional echocardiography. Unlike the LV, which can be reasonably modeled as a symmetric ellipsoid, the RV is crescent-shaped in cross-section and triangular when viewed laterally. When taking these factors into consideration, CMR may become the technique of choice to investigate RV morphology and function in patients with PAH.

The majority of the proposed methods of CMR assessment of RV function are based on volumetric approximations of the RV. Such approaches have inherent limitations, because volume-related measures such as EF are load-dependent. Right arterial pressure or RV end-diastolic pressure is an index of preload. Pulmonary vascular resistance is the most commonly used index of RV afterload. CMR imaging provides a direct evaluation of RV size, morphology, and function, and it allows non-invasive assessment of blood flow, including such parameters as stroke volume, cardiac output, pulmonary artery distensibility, and RV mass. CMR data can be used to evaluate right heart hemodynamics, particularly for follow-up. There is no doubt that RV function is a remarkably useful factor for predicting prognosis in patients with PAH and for considering the pathophysiology of PAH. Taken together, CMR might be one of the most powerful candidate tools for assessing the influence of preload and afterload on RV function.

A number of plasma biomarkers and non-invasive prognostic markers derived from exercise testing, when assessed at baseline, correlate with outcome in PAH. Echocardiography and computed tomography are also valuable investigations to perform in the assessment of PAH patients. CMR imaging allows a unique combination of morphological and functional assessment of the RV. In the future, because CMR assessment will become increasingly important as a prognostic tool, it may be listed in future guidelines as the routine method of assessment. Potentially, this powerful diagnostic tool offers many more options than other methods of assessment of RV function and its relationship with the pulmonary arterial system. We anticipate that CMR imaging will increasingly be used as the primary modality for combined anatomic and functional assessments that enable more complete and efficient evaluation of PAH patients. We believe that the day will come soon when the clinical characteristics and CMR findings of Japanese patients with PAH will be investigated using this modality. The study by Yamada and coworkers is a step in this right direction.

References