

Is Cardiac Magnetic Resonance Imaging Underutilized in the Diagnosis of Pulmonary Hypertension?

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Cardiac magnetic resonance imaging (CMR) provides an important and complementary role to conventional imaging in the evaluation of patients with pulmonary hypertension (PH). Echo-cardiography remains vital given its ability to quickly assess cardiac morphology, function, and hemodynamics. It is also portable, readily available, and relatively inexpensive. However, certain limitations of echocardiography do occur in PH patients such as inability to fully or accurately characterize the right ventricle (RV), which remains crucial for therapeutic decision making and prognostic determination. Given the geometric complexity of the RV as well as patient-specific factors such as obesity, echocardiography may fail to adequately depict the RV. Conversely, CMR is well suited for PH imaging for various reasons, not least of which is its ability to fully characterize RV morphology and function. In addition, CMR provides various components

during a PH examination, including assessment of pulmonary artery (PA) flow and stiffness, ventricular function and strain, shunt, emboli, and tissue characterization.

Generally, echocardiography provides a detailed and accurate assessment of the RV in many PH patients.¹ However, in those instances where accuracy or visualization is limited, or where more precise determination is required, CMR should be strongly considered as it remains the reference standard for RV assessment.^{2,3} On a typical CMR examination, a series or stack of short- and long-axis cine images are performed and post-processed using commercially available software. Left and right ventricles are traced from systolic and diastolic still frames providing volumetric data, allowing for calculations of end-diastolic and systolic volumes, ejection fraction, mass, and stroke volume. As the severity of PH progresses, the RV

thickens, enlarges (Figure 1), and will subsequently fail demonstrated by a progressive decline in ejection fraction. These changes to RV size and function are tightly coupled with mortality.⁴ Also, as pulmonary pressures rise, there is a characteristic flattening of the interventricular septum leftward. The degree of flattening can be quantified by comparing the curvatures of the interventricular septum to left ventricular free wall, a value that is strongly correlated to the degree of PH.⁵ Evaluation of myocardial strain using conventional tagging or other novel sequences may also accurately characterize the RV, but at this point is largely relegated to research protocols.

The ability to accurately assess PA stiffness presents an exciting new avenue for CMR. The development of adverse pulmonary vascular remodeling is the hallmark feature, which begins the cascade of pulmonary arterial hypertension (PAH). With the use of specific flow sequences, CMR can accurately measure cross-sectional phasic changes to the PA, thereby providing a means

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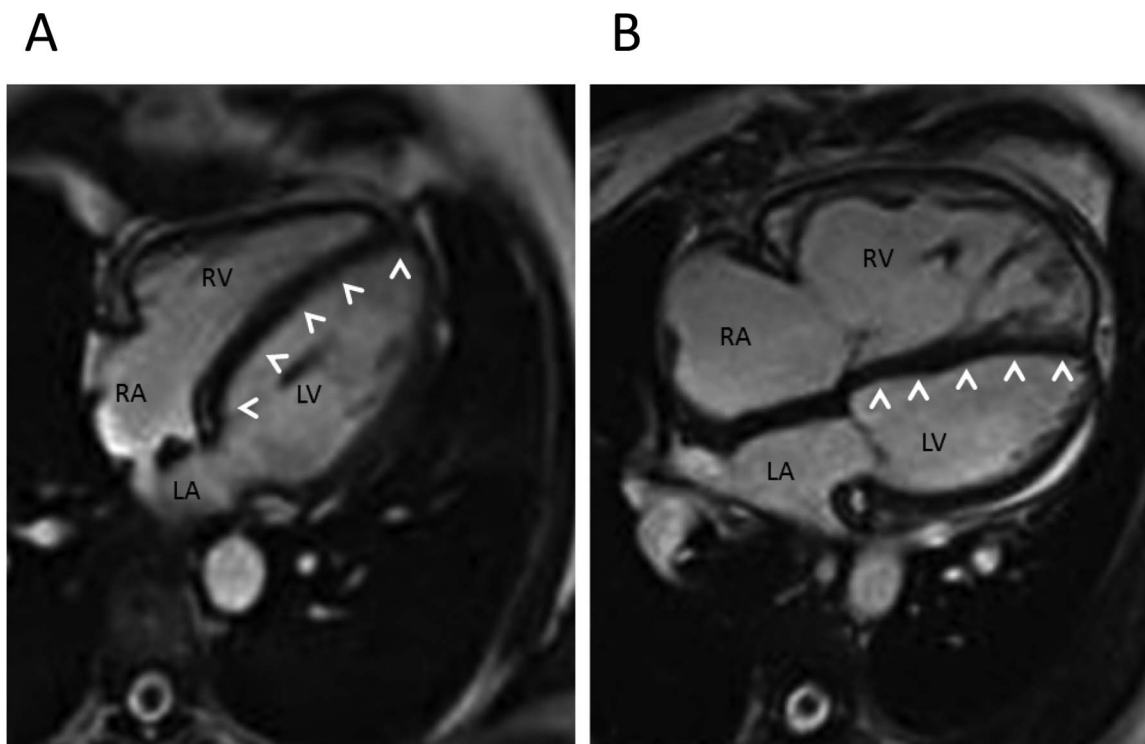


Figure 1: Four-chamber long axis cine images. A) Normal RV, normal curvature of the interventricular septum (arrows). B) Significant RV dilatation and hypertrophy as seen in severe PH. The curvature of the interventricular septum is lost (arrows) and presents as a “D”-shaped left ventricle. Also notice the presence of pericardial effusion and dilated right atrium: both are markers of poor prognosis in PH.

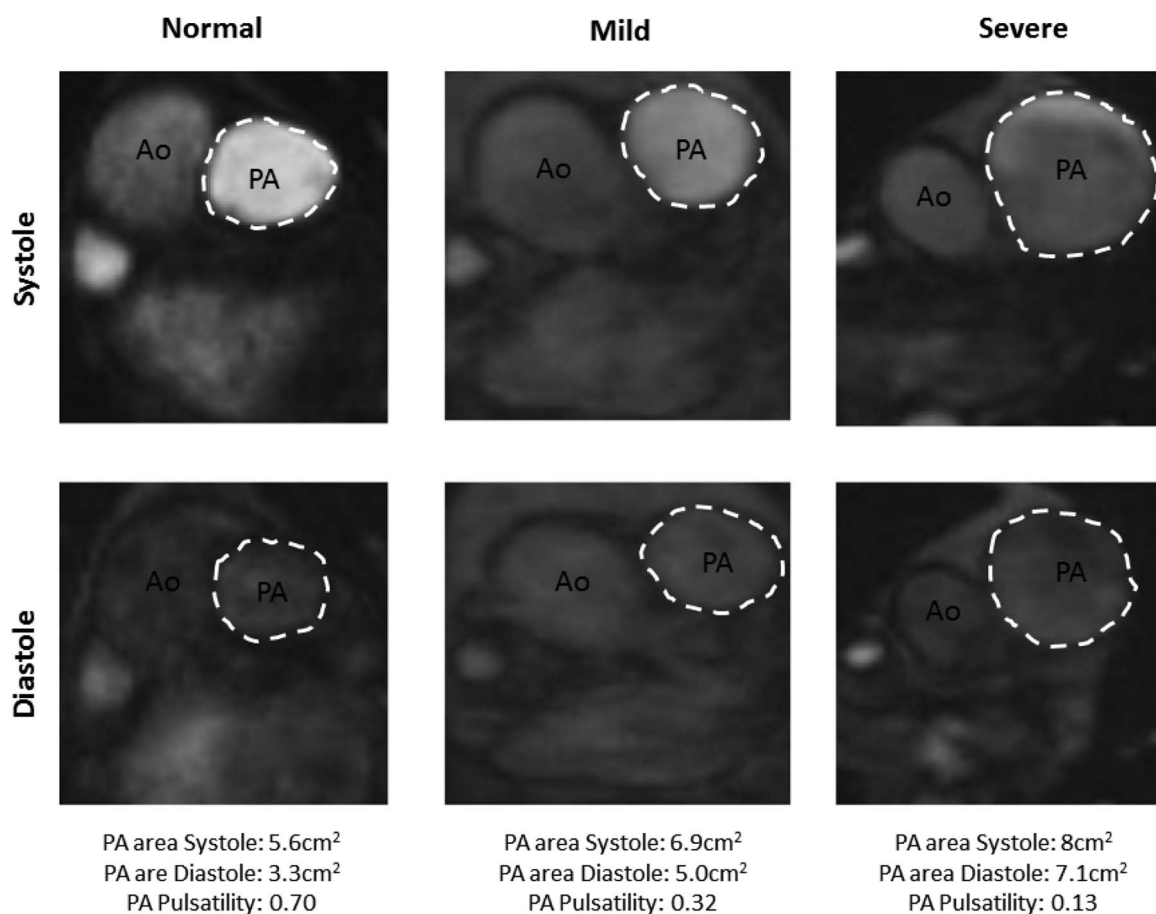


Figure 2: Short axis flow images of the pulmonary artery and aorta in normal, mild, and severe PH.

to calculate indices of PA stiffness including pulsatility $[(PA \text{ area}_{\max} - PA \text{ area}_{\min})/PA \text{ area}_{\min}]$ (Figure 2). As PH progresses, the PA typically dilates and the degree of pulsatility declines, reflecting loss of elasticity and worsening stiffness.^{6,7} This technique may provide a means to detect pulmonary vascular remodeling at an earlier stage of development, and may also ultimately be considered a target for PH-specific therapies. One potential role of CMR is detection in those patients who are “at risk” (ie, scleroderma patients) or have clinical findings suggestive of early pulmonary vascular remodeling, but exhibit negligible pulmonary pressure elevations above normal during right heart catheterization. Pulmonary arterial measurements from CMR can also be coupled with invasive catheterization, yielding additional characterization of compliance, capacitance, and distensibility and may hold promise in assessing ventricular-vascular coupling.^{8,9}

The use of magnetic resonance angiography (MRA) or pulmonary perfusion to assess for World Health Organization Group 4 chronic thromboembolic PH (CTEPH) and pulmonary blood flow is also a part of routine PH CMR protocols. While lacking the sensitivity of traditional testing such as ventilation perfusion lung scans or computed tomography angiography, MRA may be useful in selected cases to aid in the diagnosis. In addition to CTEPH, conventional cine and flow sequences may also identify other potential causes for PH that may not have been previously recognized such as atrial septal defects and shunts, patent ductus arteriosus, and anomalous pulmonary veins. Following perfusion imaging, delayed imaging post-contrast is performed to assess whether late gad-

olinium enhancement is present in the myocardium, a finding suggestive of cardiac pathology such as with fibrosis, scar, infarction, or infiltrative cardiomyopathy. In the case of PAH, late gadolinium enhancement is often pictured in the RV insertion sites, which is a finding suggestive of increased wall tension and strain. An appearance of scar in the interventricular septum RV insertion sites may reflect worsened PH and may be linked to poor prognosis.¹⁰

While it is highly unlikely that CMR would replace conventional echocardiography or right heart catheterization, there is little doubt regarding its value in selected patients with PH where the diagnosis or cause remains unclear or the RV is poorly characterized. Newer techniques and data will be necessary to determine the usefulness of other sequences that measure such things such as myocardial strain, pulmonary vascular remodeling, and tissue characterization. Certainly, CMR is not without its limitations such as cost, limited availability, expertise, and study time. Patient-related limitations include claustrophobia and potential contrast reactions. However, the superb tissue characterization combined with important morphologic and physiologic data afforded by CMR make it an extremely useful and promising means for assessment of pertinent changes to the pulmonary circulation and RV in the setting of PH. No other technique at present provides such a comprehensive means of assessment of important morphologic changes to the heart and vasculature in this patient population. While we cannot definitively state whether CMR is presently underutilized in PH, its value and appreciation coupled with new advancements and research will likely increase the proportion of CMR used in PH in the future.

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