

Specialty area: The Forgotten Right Ventricle

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Highlights

- Right ventricular (RV) structure and function are important in health and disease.
- In the general population, RV hypertrophy is associated with a more than a two-fold increase in the risk of congestive heart failure and death.
- MRI allows for the visualization of structural and functional RV changes in a variety of pathologic conditions, including arrhythmogenic right ventricular cardiomyopathy (ARVC), RV infarction, and congenital heart disease.

Title: Clinical Applications of MRI in Evaluating the Right Ventricle

Target audience: Cardiologists, cardiac MRI radiologists

Objectives: To understand the role of cardiac MR imaging of the RV in health and disease.

Background: The RV plays an important role in both health and disease. Cardiac MRI is ideally suited to define the RV's complex anatomy and accurately assess RV volumes, RV mass, and RV function.

The RV in the General Population: The Multi-Ethnic Study of Atherosclerosis Right Ventricle Study (MESA-RV), a multicenter prospective multi-ethnic cohort study in subjects without clinical cardiovascular disease, has shown that men have greater RV mass and RV volumes but lower RV ejection fraction (RVEF) than women, and that African Americans have lower and Hispanics higher RV mass than Caucasians. These gender and ethnic differences in the general population must be considered when evaluating the RV clinically. RV hypertrophy is associated with a greater than two-fold increase in congestive heart failure or death after multivariable adjustment, suggesting that RV mass may play an important prognostic role. Future studies evaluating temporal changes in RV structure and function on outcomes are ongoing and may lend important insight into the relationship between the RV and cardiovascular health.

The RV in Disease: Many disease states can lead to changes in the RV that are clinically important, including Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), RV infarction, and congenital heart disease (CHD).

ARVC: ARVC is a rare inherited myocardial disease characterized by fibrofatty replacement of the myocardium predominantly in the RV, cardiac arrhythmias, and sudden cardiac death. ARVC is more common in men than in women and becomes clinically evident in the second to fourth decade of life. In 2004 with revisions in 2010, the Task Force of the Working Group of Myocardial and Pericardial Disease published criteria for the diagnosis of ARVC which included family history, arrhythmic, electrocardiographic, pathologic, and structure and functional abnormality criteria. A definite diagnosis of ARVC consists of 2 major criteria, 1 major and 2 minor, or 4 minor criteria. The structural and functional criteria are divided into major and minor criteria defined by MRI as: 1) Major: Regional RV akinesia or dyskinesia or dyssynchronous RV contraction **PLUS** either RV end-diastolic volume to BSA $\geq 110\text{mL/m}^2$ (men) or $\geq 100\text{mL/m}^2$ (women) or RVEF $\leq 40\%$; 2) Minor: Regional RV akinesia or dyskinesia or dyssynchronous RV

contraction **PLUS** either RV end-diastolic volume to BSA ≥ 100 to < 110 mL/m² (men) or ≥ 90 to < 100 mL/m² (women) or RVEF $> 40\%$ to $\leq 45\%$. MRI can also detect fatty infiltration of the RV but is less reproducible and lacks specificity compared to detection of RV dysfunction and regional RV enlargement. In ARVC patients who meet Task Force criteria, 75% will have intra-myocardial fat detected on black-blood imaging, most commonly in the RV inflow and outflow regions. Other findings associated with ARVC include focal wall thinning, RV hypertrophy, trabecular hypertrophy and disarray, moderator band enlargement, localized aneurysms, and RVOT enlargement.

RV Infarction: RV involvement in acute left ventricular (LV) inferior and anterior myocardial infarction (MI) has an estimated incidence of 50-80% in post-mortem studies however many RV infarctions, especially in association with anterior MI, may be clinically silent. MRI with delayed enhancement imaging can detect RV infarction and provides important clinical and prognostic information. RV infarction is associated with lower RVEF, lower LV ejection fraction, larger LV infarct size, and poorer prognosis than LV infarcts without RV involvement.

Congenital Heart Disease (CHD): Up to 10 in 1,000 births are complicated by congenital cardiac abnormalities. These can vary in clinical severity from small ventricular septal defects that may spontaneously close to more complex congenital abnormalities including Tetralogy of Fallot, Transposition of the Great Arteries, and Ebstein's anomaly. Due to advances in surgical and medical therapy, many of these patients survive into adulthood but need to be monitored and treated for the hemodynamic and structural consequences of their corrective or palliative surgery or sequelae of their original defect.

Tetralogy of Fallot (TOF): TOF, defined by an overriding aorta, pulmonary atresia/stenosis, RV hypertrophy, and a ventricular septal defect, accounts for 5-6% of all CHD patients. Total corrective surgery has been performed since the 1950's, however overall life expectancy is shortened for these patients compared to age-matched controls. Common issues after initial correction of TOF include pulmonary regurgitation and pulmonary stenosis, RV aneurysms, and RV dysfunction. MRI can visualize the RVOT as well as RV patch material, accurately assess RV volumes, RVEF, and RV mass, as well as qualitatively and quantitatively assess tricuspid and pulmonic regurgitation.

Transposition of the Great Arteries (TGA): Transposition of the Great Arteries (TGA) can occur as a congenitally corrected (L-transposition) form as well as a discordant ventriculoarterial connected (D-transposition) form. L-TGA is associated with other cardiac anomalies including tricuspid valve abnormalities, ventricular septal defects, and pulmonary stenosis. Progressive tricuspid regurgitation and RV failure is a concern for L-TGA and can be evaluated with MRI. D-TGA patients have a right atrium which connects to a morphologic RV which then connects to the aorta. The left atrium connects to the LV and empties into the pulmonary circulation. Without a concomitant cardiac shunt (ventricular septal defect, patent ductus arteriosus, or atrial septal defect), these patients have significant cyanosis and high mortality. Two types of surgical repair, atrial switch (performed before 1975) and the arterial switch procedures (performed after 1975) are performed in D-TGA. In the atrial switch procedure, a baffle is constructed within the atria

redirecting systemic venous blood into the LV and pulmonary venous return into the RV using either synthetic/pericardial tissue (Mustard procedure) or atrial tissue (Senning procedure). Because the RV must now function as systemic pump, it is prone to failure. Tricuspid regurgitation, baffle obstruction, and venous obstruction can also occur after this procedure. In the arterial switch procedure, the aorta is attached to the LV and the pulmonary artery is attached to the RV. RVOT obstruction or supra-ventricular pulmonary stenosis can occur, leading to RV dilation and failure. In both types of repair, cardiac MRI can play an integral role in evaluating RV structure and function.

Ebstein's Anomaly: Ebstein's anomaly is a congenital defect in which the septal leaflet of the tricuspid valve is apically displaced with "atrialization" of the basal portion of the RV. Ebstein's anomaly accounts for <1% of all cardiac defects. In severe cases, patients with Ebstein's may present with cyanosis, right-sided heart failure, arrhythmias, and sudden cardiac death. Short-axis and four chamber cine images can demonstrate the apical displacement of the valve, right ventricular volumes and function, as well as any tricuspid regurgitation.

Summary: If clinicians and radiologists recognize the ethnic and gender differences in RV structure and function in health, the imaging characteristics of ARVC, the prognostic importance of RV infarction in anterior and inferior LV MI, and the potential RV-related pathology that may occur in long-term follow-up for adults with CHD, MRI can add incremental and important information that will affect patient care.

References:

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