

Title : Right Ventricle and Arrhythmogenic Right Ventricular Dysplasia
By Harikrishna Tandri MD (Thursday May 11, 2011)

Background. Right Ventricular function is an important determinant of prognosis both in congenital and acquired forms of right ventricular myopathies [1]. As such an accurate way to assess RV function is important both for diagnosis and prognosis. MRI is currently the modality of choice to evaluate the right ventricle due to its multiplanar imaging capabilities and the non-invasive nature of the exam [1]. In conditions such as right ventricular dysplasia, MRI has the unique ability to provide tissue characterization and depict the histopathologic hall marks of the disease obviating the need for invasive biopsy. Primary RV involvement with minimal or no LV involvement can be seen in genetic conditions such as ARVD, inflammatory granulomatous disease such as Sarcoidosis and rarely myocarditis. RV involvement in ARVD should be differentiated from other arrhythmic disorders such as idiopathic ventricular arrhythmias, sarcoidosis and non-ischemic cardiomyopathy. Distinguishing these conditions is important for accurate diagnosis and management. Further, pattern of cardiac involvement varies with the underlying genetic mutations that result in ARVD. As such better understanding of genotype phenotype correlations are important for prognostic purposes and for screening family members.

MRI techniques and applications for RV cardiomyopathy evaluation. MRI for ARVD consists of several techniques that may be performed separately or in various combinations during a patient examination. Major techniques include:

a) *Cine-MRI is the using steady state free precession*, performed in the short axis four chamber and axial planes (including the pulmonary outflow tract). This series of scans provides accurate depiction of RV global morphology and function. For right sided disorders in particular, MRI offers superior depiction of wall motion abnormalities as well as better/ more accurate quantification of right heart function compared to echocardiography.

b) *Inversion-recovery prepared myocardial 10-20 min delayed enhancement sequences*, also acquired in the short and axial planes following 0.15-0.2 mmol/kg intravenous gadolinium administration. These sequences are identical to those used for evaluation of myocardial scar due to coronary artery disease. For evaluation of the right ventricle, inversion recovery times are typically shortened by 25 msec to obtain improved suppression of the right ventricle. Unlike scar from myocardial infarction, scar associated with nonischemic conditions may be intermingled with normal myocardium, so that relatively lower signal intensity (than dense scar) is present [3]. Our current standard is to use delayed gadolinium enhancement images for all patients referred for nonischemic cardiomyopathy.

c) *Double inversion recovery turbo/ fast spin echo imaging* [2]. These sequences are used primarily for arrhythmogenic right ventricular dysplasia (proton density weighted with/ without fat suppression) and hypertrophic cardiomyopathy (T2 weighted with fat suppression). Optimization of the sequence is difficult in order to achieve black blood, particularly for long axis views in diseased ventricles with low rates of blood flow. Imaging is optimized by synchronizing the image acquisition period to the rest period of the cardiac cycle.

Arrhythmogenic right ventricular dysplasia (ARVD). ARVD is characterized by enlargement, dysfunction and fibrofatty infiltration of the right ventricle (RV). It is recognized clinically by ventricular tachyarrhythmia, abnormal RV morphology and RV dysfunction. Although rare, it may be responsible for 5% of sudden cardiac death due to arrhythmias among young people in certain populations [4]. Fibrofatty tissue might have a role on the development of cardiac arrhythmias. Tandri et al. assessed 30 consecutive patients referred for diagnostic evaluation. Of the patients identified as having ARVC by RV biopsy, RV late gadolinium enhancement was observed in 100% [5]. Desai et al [6] found that the TI for myocardial signal suppression appears to be different between left and right ventricles. Potential mechanisms include partial volume averaging with fat or blood pool (related to increased trabeculation) in the RV. The primary diagnostic features of ARVD are a) enlargement and dysfunction of the right ventricle out of proportion to LV dysfunction, b) regional aneurysm formation or wall motion abnormalities. Fatty infiltration on MR imaging is poorly reproducible among observers and is therefore not considered a criterion for the disease. It can also occur in other circumstances such as steroid use and obesity [7] Small amounts of RV fat with normal RV function are seen in normal individuals, but individuals with large amounts of RV fatty infiltration and normal function may be seen [8], [9].

References

1. Boxt LM. Radiology of the right ventricle. *Radiol Clin North Am* 1999 ;37:379-400.
2. Simonetti OP, Finn JP, White RD, Laub G, Henry DA. "Black blood" T2-weighted inversion-recovery MR imaging of the heart. *Radiology* 1996; 199: 49-57.
3. McCrohon JA, Moon JC, Prasad SK, et al. Differentiation of heart failure related to dilated cardiomyopathy and coronary artery disease using gadolinium-enhanced cardiovascular magnetic resonance. *Circulation* **2003**;108:54-59
4. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes. *N Engl J Med* **1998**;339:364-369
5. Tandri H, Saranathan M, Rodriguez ER, et al. Noninvasive detection of myocardial fibrosis in arrhythmogenic right ventricular cardiomyopathy using delayed-enhancement magnetic resonance imaging. *J Am Coll Cardiol* **2005**;45:98-103
6. Desai MY, Gupta S, Bomma C, et al. The apparent inversion time for optimal delayed enhancement magnetic resonance imaging differs between the right and left ventricles. *J Cardiovasc Magn Reson* **2005**;7:475-479
7. Macedo R, Prakasa K, Tichnell C, et al. Marked lipomatous infiltration of the right ventricle: MRI findings in relation to arrhythmogenic right ventricular dysplasia. *AJR Am J Roentgenol* **2007**;188:W423-427
8. Fontaine G, Fontaliran F, Zenati O, et al. Fat in the heart. A feature unique to the human species? Observational reflections on an unsolved problem. *Acta Cardiol* **1999**;54:189-194
9. Burke AP, Farb A, Tashko G, Virmani R. Arrhythmogenic right ventricular cardiomyopathy and fatty replacement of the right ventricular myocardium: are they different diseases? *Circulation* **1998**;97:1571-1580