Right ventricular arrhythmia: emergence of magnetic resonance imaging as an investigative tool

There is increasing evidence that previously unrecognised abnormalities of the right ventricle may be associated with a variety of ventricular arrhythmias originating from the right ventricular outflow tract that have been well characterized for some years, but considered idiopathic in origin. The possibility of detecting minor abnormalities of the right ventricle is mainly due to the advent of magnetic resonance imaging. This safe, non-invasive and highly dimensionally accurate technique for cardiac imaging can evaluate both the anatomy and function of the right ventricle, and also detect abnormalities of shape, wall motion, and tissue composition. Perhaps not surprisingly therefore significant new information in the field has been obtained by using this technique.

Carlson et al. found that structural and functional abnormalities can be seen by magnetic resonance imaging in as many as 95% of patients with right ventricular outflow tract ventricular tachycardia. Furthermore, the abnormalities detected by magnetic resonance imaging were found to be closely related to the earliest site of ventricular electrical activation during ventricular tachycardia. Structural abnormalities of the right ventricle included focal wall thinning with or without excavations, decreased wall thickening, and abnormal systolic wall motion. Whilst anatomical and functional abnormalities were seen in various parts of the right ventricle, they were most frequently seen in the right ventricular outflow tract. In addition, as the paper from Proclemer et al. in this issue shows, patients with frequent premature ventricular beats of right ventricular outflow tract origin commonly show enlargement of the right ventricular outflow tract, as well as decreased right ventricular wall thickness and decreased systolic wall motion confined to the right ventricular outflow tract. These findings lead to two overall conclusions, namely that there may be a common substrate for patients with ventricular premature beats and those with ventricular tachycardia of right ventricular outflow tract origin, and that magnetic resonance imaging is a valuable technique for identifying previously unsuspected right ventricle abnormalities.

It is interesting to note that the abnormalities described in the patients above by magnetic resonance imaging are not dissimilar to those found in mild forms of arrhythmogenic right ventricular dysplasia. The abnormalities found by magnetic resonance imaging in arrhythmogenic right ventricular dysplasia patients include akinetic segments of the right ventricle (with about a quarter located in the right ventricular outflow tract), and increased right ventricular volume and right ventricular outflow tract diameters. A pathognomonic sign is fatty infiltration of the right ventricle, usually in the free wall, which is seen as intense signal by magnetic resonance imaging compared to the surrounding myocardium, particularly if clearly remote from the normal distribution of adipose tissue surrounding the coronary arteries. Fat infiltration is, however, seen less commonly than other morphofunctional changes and typically in more advanced cases who may present with sustained ventricular tachycardia and are at risk of sudden death.

Arrhythmogenic right ventricular dysplasia probably includes a spectrum of different conditions rather than a single entity, with different pathological processes manifesting in a common manner. Since arrhythmogenic right ventricular dysplasia is a disease that seems to have a temporal progression the disease may manifest differently according to the time of presentation. It is therefore not unreasonable to speculate that patients with right ventricular outflow tract arrhythmias may have a mild, early, attenuated or a more localized form of arrhythmogenic right ventricular dysplasia, and the differentiation between the two entities may not always be possible unless adipose deposition is seen. A large follow-up study in patients with right ventricular outflow tract-related arrhythmia using magnetic resonance imaging to show morphofunctional changes would be particularly helpful in order to gain more information on any link between right ventricular outflow tract arrhythmias and arrhythmogenic right ventricular dysplasia. Some answers may come from a registry for
arrhythmogenic right ventricular dysplasia patients that has been established in the U.S.A.\[8\] to monitor diagnostic and prognostic parameters over time, which should help in the understanding of the progression of the disease, and diagnostic criteria. Other attempts to resolve some of the issues have been made by the Task Force of the Working Group on Myocardial and Pericardial disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology, which suggests that the diagnosis of arrhythmogenic right ventricular dysplasia should rely on a combination of both clinical and morphofunctional criteria\[7]\.

The right ventricle has received only limited attention in the past, which may reflect the difficulty in imaging it adequately. Arrhythmogenic right ventricular dysplasia in its more advanced form may be demonstrated by other imaging techniques such as echocardiography\[8]\, but magnetic resonance imaging appears to be more sensitive\[9]\ and has the specific advantage of clearly characterizing adipose infiltration\[10]\.

In patients with right ventricular outflow tract arrhythmias, however, magnetic resonance imaging is necessary, as shown by Proclemer et al., where all the patients had normal echocardiography. This supports the widely held belief that magnetic resonance imaging is the best technique for detailed study of the right ventricle. Even for magnetic resonance imaging, however, normal range limits for right ventricular volumes and regional function are from studies with small subject numbers, and considerable functional heterogeneity is likely to lead to equivocal results in some cases. In addition, several abnormalities reported to be specific for arrhythmogenic right ventricular dysplasia have also been found in other conditions, thus raising the possibility that different diseases might damage the right ventricle similarly, resulting in similar morphofunctional changes and clinical presentation. Newer magnetic resonance imaging techniques may, however, help in this regard, with myocardial tagging offering an objective quantified analysis of right ventricular mechanics\[11]\, and the widespread use of ultrafast sequences during a breath-hold continuing to improve image quality by eliminating respiratory artefacts, allowing full ventricular imaging in approximately 5 min\[12]\.

Myocardial characterization techniques are also improving with the use of breath-hold turbo-T2 weighted spin echo, fat suppression and STIR sequences.

Thus, magnetic resonance imaging offers the clinician a unique method with which to study the right ventricle, as it can detect signal abnormalities from the right ventricular myocardium (reflecting abnormal tissue composition), and wall motion abnormalities, as well as right ventricular volumes and shape. For these reasons, magnetic resonance imaging represents the best available technique with which to evaluate even small abnormalities of the right ventricle. Whilst many uncertainties remain as to whether patients with right ventricular outflow tract-related arrhythmias and no apparent underlying disease have a mild, early or localized form of arrhythmogenic right ventricular dysplasia, magnetic resonance imaging is changing our way of approaching these patients. Based on recommendations of the Task Force, magnetic resonance imaging can offer all the main morphofunctional criteria for diagnosis of arrhythmogenic right ventricular dysplasia within a single study. Thus so far this is the best imaging modality for patients with a clinical suspicion of arrhythmogenic right ventricular dysplasia or with arrhythmias originating from the right ventricular outflow tract.

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**References**


Impact of systolic and diastolic dysfunction on postoperative outcome in patients with aortic stenosis

See page 1977 for the article to which this Editorial refers

In most patients with aortic stenosis, long-term survival after aortic valve replacement is excellent. In the absence of coronary artery disease, operative mortality is less than 2–3% and 10-years survival is more than 85%. Early and late mortality is dependent on various factors such as age, clinical symptoms, severity of valve disease, left ventricular function, presence of coronary artery disease etc. In this issue Lund et al. evaluate several parameters, such as left ventricular systolic and diastolic dysfunction as predictors for postoperative outcome in patients with severe aortic stenosis. Left ventricular systolic dysfunction was defined as an ejection fraction of less than 61%, a peak ejection rate of less than 2.29 end-diastolic volumes per second and a prolonged time to peak ejection. Left ventricular diastolic dysfunction was defined as a peak filling rate of less than 2.86 end-diastolic volumes per second in patients younger than 49 years and of less than 2.00 in those older than 50 years. Furthermore, a diminished fast filling fraction of less than 69% in the younger and of less than 55% in the older patients, as well as a reduced late filling fraction and a prolonged time to peak filling, were evaluated as parameters for diastolic dysfunction. Lund et al. observed an adverse early and late outcome in patients with diastolic dysfunction either alone or in combination with systolic dysfunction. It is surprising that in patients with severe aortic stenosis the presence of systolic and diastolic dysfunction predicts an unfavourable postoperative outcome, since it is well known that most patients with this valve disorder already have left ventricular diastolic dysfunction at an early stage of the disease when left ventricular hypertrophy is present.

Left ventricular systolic dysfunction caused by afterload mismatch usually improves after aortic valve replacement, but recovery is delayed or improvement lacking when it results from myocardial dysfunction. Coronary artery disease is an important confounding factor which contributes to left ventricular dysfunction and which is associated with an enhanced risk for an adverse outcome after aortic valve replacement. Recently, the postoperative survival rate in patients with aortic stenosis with reduced left ventricular ejection performance has been shown to be similar to an age-matched control group. In contrast, the presence of coronary artery disease, especially a history of previous myocardial infarction, has been shown to be an independent risk factor for an adverse outcome and reduced survival after valve replacement. In the study of Lund et al. more than 40% of all patients had coronary artery disease and some of them had previous myocardial infarction. Thus, early and late mortality may have been influenced by this risk factor, since coronary artery disease influences left ventricular ejection performance significantly and is one of the major causes of left ventricular systolic dysfunction.

Left ventricular diastolic dysfunction has been found in 50 to 60% of all patients with aortic stenosis and has been considered a major cause for the development of congestive heart failure. The most common cause of diastolic dysfunction in patients with aortic stenosis is, however, left ventricular hypertrophy. In the study of Villari et al. diastolic dysfunction was defined either as abnormal relaxation, decreased diastolic filling or increased myocardial stiffness. Diastolic dysfunction was observed in approximately 50% of all patients with a normal systolic ejection performance, but was found in 95% of those with depressed systolic function. Villari and coworkers reported that diastolic stiffness increases