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Commentary

Arrhythmogenic Right Ventricular Cardiomyopathy: How to Predict the Progression

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DESCRIPTION

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a heritable cardiomyopathy characterized by fibro fatty replacement of primarily the Right Ventricular (RV) myocardium, providing a substrate for potentially life-threatening Ventricular Arrhythmias (VAs).

Echocardiography has emerged as an ideal modality because it is available and safe for use In Cardioverter-Defibrillator (ICD) patients, as is common in ARVC.

Conventional echocardiographic measurements as described in the 2010 diagnostic Task Force Criteria (TFC), lack sensitivity for detection of early structural disease substrates in ARVC. Echocardiographic myocardial strain has emerged as a principal method for the quantification of subclinical Left Ventricular (LV) dysfunction in a variety of clinical settings.

Over the past decade, echocardiographic deformation imaging has emerged as a valuable tool for both early detection and prognosis in ARVC. A recent study has characterized the role of traditional and novel measures of RV dysfunction as they relate to ARVC disease progression. They suggested that echocardiographic right ventricular strain and strain rate at baseline are associated with the rate of future progression of structural abnormalities in patients with ARVC. Patients with suspected ARVC and abnormal baseline right ventricular strain or strain rate could be at higher risk of disease progression and may warrant closer observation.

Structural progression was independently associated with ventricular arrhythmic events during follow-up. In this cohort, study of 85 patients with 6 years of follow-up, one-third of the patients with ARVC showed significant progressive structural dysfunction, with marked interpatient variability in the rate of progression. Another interesting observation was the association between first appropriate ICD therapy and significant structural RV progression. Although the numbers are small, this association demonstrates the clinical importance of assessing structural progression and its association with electrical progression in ARVD/C. Accurate and reliable prediction of electrical progression is an important area of clinical interest, as presenting events can be fatal.

Several studies have also examined ECG evidence of progression. The largest of these, by Saguner et al., reported that, in 77 patients with ARVD/C, depolarization abnormalities increased during follow-up. In another study, low precordial voltages especially in right-sided leads and Inferior T-wave inversions showed consistent correlation with echocardiographic progression of disease, with the latter also predicting higher risk of VT. Finally, more pronounced right ventricular end-diastolic area appeared to predict risk of VT with higher risk of ICD shocks in those with history of syncope or sudden cardiac death.

ICD implantation is the most accepted therapeutic strategy for ARVC patients, because the natural history of this pathology is characterized mainly by the risk of arrhythmic death, and only secondarily by contractile dysfunction that leads to progressive heart failure.

The concept that ARVC is a progressive disorder could not only influence treatment plans but also explain the unfavorable outcomes. The high rate of adverse events related to the ICD leads parameters can be explained by the peculiar pathophysiology of ARVC that leads to fibrous and fatty replacement that can both generate difficulties in locating a suitable place to implant the leads, and affect the thresholds and sensitivity during clinical follow-up. The R-wave amplitude is crucial to the performance of ICDs. A fall in the R-wave amplitude (electrical progression) and poor sensing may prevent optimal defibrillator therapy. The concern in this regard assumes an even greater significance in the treatment of patients with ARVC. Previous studies have reported a remarkable decrease in the R-wave amplitude in patients with ARVC with ICDs during long follow-up periods.

Recently, another study underscore the predictive role of echocardiographic markers in determining electrical progression over time in patients with ARVC. They assessed lead parameters in patients with an ICD, at multiple points during the follow-up period. This study which is first study to assess the value of RV 2D and 3D deformation parameters in predicting electrical

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progression in patients with ARVC, confirms and extends prior observations demonstrating that ARVC is a progressive disease. These findings suggest that echocardiography has a pivotal role in predicting patients at high risk for electrical progression.

In conclusion, based on the results of the studies, ARVC is a progressive cardiomyopathy with marked interpatient variability in the rate of progression. Further multicenter studies with prospective data collection can help elucidate the predictors of adverse outcomes and ultimately reduce disease progression in patients with ARVC.