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Impact of the Revision of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia Task Force Criteria on Its Prevalence by CMR Criteria

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OBJECTIVES The purpose of our study was to assess the impact of revised versus original criteria on the prevalence of arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) criteria in cardiac magnetic resonance (CMR) studies.

BACKGROUND Recently, the ARVC/D task force criteria have been revised, aiming for a better diagnostic sensitivity. The implications of this revision on clinical decision making are unknown.

METHODS We retrospectively evaluated the CMR scans of 294 patients referred for ARVC/D between 2005 and 2010, and determined the presence or absence of major and minor CMR criteria using the original and the revised task force criteria. Previously, major and minor abnormalities were identified by the presence of right ventricle dilation (global or segmental), right ventricle microaneurysm, or regional hypokinesis. The revised criteria require the combination of severe regional wall motion abnormalities (akinesis or dyskinesis or dyssynchrony) with global right ventricle dilation or dysfunction (quantitative assessment).

RESULTS Applying the original criteria, 69 patients (23.5%) had major original criteria, versus 19 patients (6.5%) with the revised criteria. Forty-three patients (62.3%) with major original criteria did not meet any of the revised criteria. Using the original criteria, 172 patients (58.5%) had at least 1 minor criterion versus 12 patients (4%) with the revised task force criteria; 167 patients (97%) with minor original criteria did not meet any of the revised criteria. In the subgroup of 134 patients with complete diagnostic work-up of ARVC, 10 patients met the diagnosis of proven ARVC/D without counting imaging criteria. Only 4 of 10 met major criteria according to the revised CMR criteria; none met minor criteria. However, 112 of 124 patients without ARVC/D were correctly classified as negative by major and minor criteria (specificity 94% and 96%, respectively).

CONCLUSIONS In our experience, the revision of the ARVC/D task force imaging criteria significantly reduced the overall prevalence of major and minor criteria. The revision, although maintaining a high specificity, may not have improved the sensitivity for identifying patients with ARVC/D. Larger studies including follow-up are required. (J Am Coll Cardiol Img 2011;4:282–7) © 2011 by the American College of Cardiology Foundation

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Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is an inherited cardiomyopathy characterized by structural and functional abnormalities due to a progressive replacement of predominantly right ventricular myocardium by fibrofatty tissue (1–4). ARVC/D predisposes patients to complex ventricular arrhythmias and sudden cardiac death, typically among young subjects.

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Establishing the clinical diagnosis of ARVC/D remains challenging because of the lack of a single test to establish a definite diagnosis. Even endomyocardial biopsy, sometimes considered to be the gold standard for ARVC/D, is limited because the interventricular septum as a typical sampling site is less commonly involved (5). In 1994, the Task Force of the European Society of Cardiology and the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology proposed a set of criteria (6). These criteria were based on medical history, as well as on morphological, functional, and structural abnormalities, including right ventricular dilation, regional dysfunction, (fibro)fatty replacement of the right ventricle (RV), electrocardiographic (ECG) changes, arrhythmias, and a family history of sudden cardiac death. These original task force criteria (TFC) were universally used to identify patients with ARVC/D. They were considered highly specific, but some authors have suggested that they may lack sensitivity, especially for early and familial disease (7,8). Moreover, most of these criteria (global and structural abnormalities) were based on qualitative rather than quantitative information and were defined based on the experience before the wide availability of cardiac magnetic resonance (CMR).

Over the past 15 years, CMR has emerged as the noninvasive diagnostic tool of choice for assessing RV anatomy, structure, and function (9,10). High-resolution cine imaging with state-of-the-art steady-state free-precession techniques is widely considered the gold standard for the assessment of ventricular volumes, myocardial mass, and systolic function; and CMR demonstrates high intraobserver and interobserver agreement and accuracy (11,12). The high spatial and temporal resolution enables a detailed assessment of the RV for regions of severely reduced wall thickness and wall motion abnormalities.

The CMR evidence of intramyocardial fat and fibrosis in the RV has been used as supportive diagnostic information; however, because of the limited specificity of these findings and technical limitations of CMR in visualizing the thin RV myocardium, the diagnostic utility of intramyocardial fat and fibrosis as diagnostic targets in ARVC/D remains controversial (13).

Recently, a revision of the TFC has been proposed, incorporating quantitative assessment of RV size and RV function (14). On the basis of as yet unpublished pilot data from a partially genetically defined cohort of patients, the authors propose imaging criteria using a combination of RV dilation and severe regional wall motion abnormalities to establish evidence for ARVC/D. The main differences to the previous set of criteria include the removal of RV microaneurysms (focal akinesis with early diastolic bulging) (Fig. 1) and segmental RV dilation, and the use of a different and more detailed quantitative definition of RV dilation.

The purpose of our study was to assess the impact of revised versus original criteria on the prevalence of ARVC/D criteria in CMR studies.

ABBREVIATIONS AND ACRONYMS

ARVC/D = arrhythmogenic right ventricular cardiomyopathy/dysplasia

CMR = cardiac magnetic resonance

ECG = electrocardiographic

EF = ejection fraction

LV = left ventricle

RV = right ventricle

TFC = task force criteria

METHODS

Study population. We performed a retrospective analysis of patients referred to our CMR center for ARVC/D between 2005 and 2010. To minimize inappropriate indications, we included only patients re-

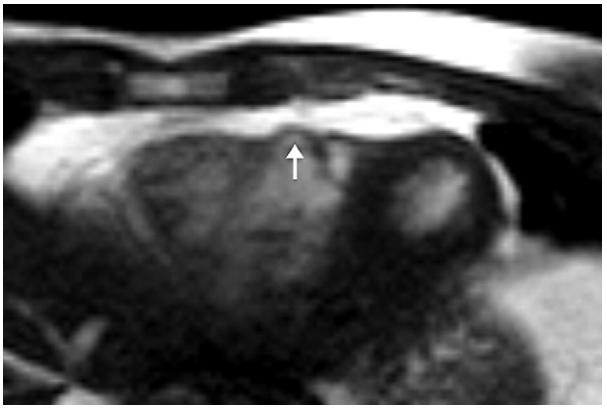

Figure 1. Microaneurysm in a Patient With Suspected ARVC

There is a focal outpouching of the free RV wall in early diastole ("early diastolic bulging," **arrow**). According to the original criteria, this finding itself was considered a major criterion while the revised criteria require a combination of regional akinesis, dyskinesis, or dyssynchronous contraction with global RV dysfunction or dilation. See Online Video 1. ARVC = arrhythmogenic right ventricular cardiomyopathy; RV = right ventricle.