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Expert Opinion: International Task Force Criteria for the Clinical Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy

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Diagnostic criteria for arrhythmogenic right ventricular cardiomyopathy (ARVC) were first established qualitatively by an international task force in 1994 and subsequently updated in 2010 to include quantitative assessment. The updated criteria have resulted in improved specificity at the expense of decreased sensitivity. With this in mind, we asked 3 leading experts the following question:

“If you had the opportunity to update the 2010 International Task Force Criteria for the clinical diagnosis of ARVC, what would you change and why?”

The objective of the imaging revisions of the 2010 ARVC Task Force criteria was to move from completely qualitative criteria (e.g. “reduced” ejection fraction) to quantitative criteria (e.g. ejection fraction $\geq 40\%$). Unfortunately, the 2010 Task Force criteria still involve subjective analysis: a regional wall motion abnormality (akinesia, dyskinesia or

dyssynchrony) must be present. For the left ventricle, identification of a wall motion abnormality is thought to be reliable using visual evaluation. But even in the normal right ventricle, imaging physicians frequently describe “abnormal” regional function. Thus, a major improvement for future Task Force revisions would be to add quantitative measures of regional RV function. While speckle tracking seems feasible for echocardiography, the MRI solution to quantification of regional contraction remains under investigation.

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ARVC is an uncommon autosomal dominant disorder that can involve both the right and the left ventricles. Thus, I would include cardiac magnetic resonance imaging data regarding left ventricular systolic dysfunction as well as late gadolinium enhancement of both the left ventricle and right ventricular free wall in the diagnosis criteria for suspected ARVC patients with abnormal right cavity size and regional/global systolic function.

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The 2010 Task Force guidelines for the diagnosis of ARVC stipulate concomitant presence of both regional wall motion abnormality and global dilation/dysfunction of the right ventricle to fulfill a single criterion in the imaging group. Yet, local and global right ventricular dilation/dysfunction not infrequently occur independently of each other, warranting revision of the criteria to reflect this, as in the original 1994 version. Late gadolinium enhancement of the left ventricle in a subepicardial or midmyocardial pattern is another feature that merits incorporation, particularly if the criteria are extended to include biventricular and left-dominant subtypes of arrhythmogenic cardiomyopathy.

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No conflicts of interest to disclose.

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