Progress in the Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia by Cardiac Magnetic Resonance Imaging Using Feature Tracking

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Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a condition which continues to pose diagnostic challenges for noninvasive imaging. The 2010 ARVC/D Task Force criteria was a major step forward for diagnosis. These Task Force criteria emphasized the role of quantitative imaging for both echocardiography and cardiac magnetic resonance imaging (CMR). In particular, multicenter study data were used to generate sex-specific cut-off values that distinguished borderline and definite ARVC for volumetric CMR data. The Task Force criteria were established to achieve high specificity of an imaging feature (eg, right ventricular [RV] volume or ejection fraction) at approximately the 95th percentile. That relatively high specificity resulted in lower sensitivity (68% to 76%) for CMR criteria in the original North American ARVC/D Registry. Major CMR criteria include RV ejection fraction ≤40% or RV end-diastolic index ≥110 mL/m² for men or 100 mL/m² for women. The rationale for choosing stringent criteria for any one diagnostic test, such as CMR, is that the combination of several criteria is used for the final diagnosis of disease. The use of several diagnostic criteria (structural changes, tissue characterization, arrhythmia, repolarization abnormalities, and family history) is the current standard of care for diagnosis of ARVC/D. The task force criteria for CMR require a regional wall motion abnormality to be present (in addition to RV dilatation or decreased ejection fraction). ARVC/D is believed to begin initially as a regional disease that eventually can involve major portions of the RV in addition to the left ventricle. For this reason, regional akinesia or dyskinesia must be present. However, because the normal RV is asymmetrical in shape, assessment of regional wall motion abnormality is considerably more difficult than evaluation of the more symmetrical left ventricle. Visual recognition of abnormal wall motion of the RV is extremely challenging even by experienced reviewers: in one study of 29 healthy subjects, regional dyskinesia was thought to be present in 22/29 subjects. Thus, a major weakness to date has been the ability to quantitatively evaluate regional wall motion abnormalities of the RV.

The traditional method of assessing regional wall motion abnormalities by CMR is tissue tagging. Unfortunately, the RV wall is thin, so that tag lines are poorly visualized and tracked in the RV. Low resolution of tag lines can be resolved in part using the displacement encoding with stimulated echoes technique. However, in ARVC/D, the RV wall may be replaced by fibrofatty tissue. This results in shorter T1 times in the diseased ventricle—so that CMR tag lines relax more quickly when disease is present, further weakening the tissue tagging approach.

In this issue of Circulation: Cardiovascular Imaging, Prati et al have adapted methods originally used for echocardiography to track motion of the RV on CMR. Feature tracking uses the inherent irregular nature of the endocardial and epicardial surfaces of the RV to follow the location of these features during the cardiac cycle. The motion of points on the surfaces of the RV can be used to calculate surrogates for myocardial strain, defined as the percentage change in length between 2 features relative to the baseline distance between the features. As opposed to CMR tissue tagging, the concept of tissue tracking is much better suited to identifying regional motion of the thin-walled RV.

The results of Prati et al are particularly encouraging. The authors evaluated 29 patients with definite ARVD/C and compared these subjects to 32 patients with RV outflow tract tachycardia and normal controls. Patients with RV outflow tract tachycardia are thought to have a benign course without structural abnormalities of the RV. Importantly, feature tracking analysis was able to be performed in all study subjects. Reader reproducibility for global longitudinal strain was excellent (coefficient of variation 4%–6%). The regional differences between patient groups and normal subjects were the greatest at the base of the RV: basal longitudinal strain was −35% in normal subjects and RV outflow tract patients but was −22% in ARVC/D patients. Similar differences were present at the mid and apical levels, but to a lesser degree. Strain abnormalities were also present in the circumferential direction at the base (−25%, −22%, and −14% for normal subjects, RV outflow tract patients, and ARVC/D patients). Again, differences were less marked at the mid and apical levels. Strain abnormalities that are most prominent at the base of the RV are consistent with current paradigms of the...
spectrum of disease of ARVC/D. Further, the results of Prati et al using feature tracking is consistent with the results of other authors. Thus, the work by Prati et al continues to strengthen the argument that feature tracking of the RV will eventually have a role in the diagnosis of ARVC/D using CMR methods.

There are several caveats before adaptation of these methods for analysis of CMR data. First, these results represent single center data, whereas the Task Force criteria were validated in a multicenter study. Feature tracking is relatively new, and multicenter data in ARVC/D remains rare. The incremental value of feature tracking for the diagnosis of the difficult patient remains to be determined. In particular, a challenging situation is the patient who is genetic (or presumed to be so) based on genetic or family history. In these individuals, we seek to find early phenotypic expression of disease. Regional wall motion abnormalities are likely to precede global alterations in volumes and ejection fraction. Can we use feature tracking methods to assess changes in regional wall motion over time? Even at its relatively early stage of development, the intra- and interreader variability of feature tracking of the RV seems to be relatively low. However, to track change, we also need to know the scan–rescan reproducibility of feature tracking techniques.

In conclusion, CMR feature tracking for ARVC/D seems to be a promising method for improving the quantitative assessment of this challenging group of patients. Prati et al are to be congratulated for providing excellent insight that may help in noninvasive diagnosis of ARVC/D. Because the feature tracking technique can be applied rapidly for the analysis of multiple cardiac chambers, we should expect to see a proliferation of studies determining the appropriate use of feature tracking for CMR.

Disclosures

The opinions expressed in this editorial are those of the author, and do not necessarily reflect the opinions of NIBIB, the NIH Clinical Center, the National Institutes of Health, or the Department of Health and Human Services. Dr Bluemke discloses that he is a full-time employee of the intramural program at the NIH Clinical Center.

References


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