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Echocardiographic deformation imaging improves detection of arrhythmogenic right ventricular cardiomyopathy; a head-to-head comparison of deformation imaging and conventional assessment

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Topic(s):
Imaging: Myocardial Disease

Citation:

Background
Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is an inherited cardiomyopathy diagnosed by a complex set of tests defined in the 2010 Task Force Criteria (TFC). For echocardiography, right ventricular (RV) dilatation and function are combined with visual wall motion assessment to obtain diagnostic criteria. However, subtle wall motion abnormalities can be missed by visual assessment, thereby limiting detection of disease. Recent studies have shown that echocardiographic deformation imaging has high sensitivity for detection of wall motion abnormalities. However, the performance of deformation imaging within the 2010 TFC for ARVC diagnosis remains unknown.

Objectives
To perform a head-to-head comparison of the diagnostic value of visual wall motion assessment versus deformation imaging in a real-world cohort of consecutive patients evaluated for ARVC.

Methods
We included a consecutive cohort of 163 patients who were referred for ARVC evaluation between 2009-2011, of whom 59 patients underwent an echocardiogram with images available for deformation analysis. Patients were diagnosed by consensus of 3 independent ARVC experts with access to all patient data including a median follow-up of 5.9 years IQR[2.7-7.6 yrs]. The original clinical assessment of RV outflow tract (RVOT) dimensions, fractional area change and wall motion was used. In addition, deformation patterns of the subtricuspid area were scored as normal (type I) or abnormal (type II/III), according to the presence of regional mechanical dysfunction (see figure). We evaluated the effect of replacing visual wall motion assessment with deformation imaging on the sensitivity, specificity and balanced accuracy of the echocardiographic TFC.

Results
Of 59 patients (age 38 ± 17 yrs, 49% male), the expert panel diagnosed 15 (25%) with ARVC. Conventional TFC, either minor or major, were observed in 10 patients; replacing visual wall motion assessment with deformation imaging led to 5 additional detections of ARVC patients, whereas 0 were lost. Consequently, deformation imaging increased sensitivity from 67% to 100%, whereas specificity decreased from 89% to 73%. The balanced accuracy increased from 0.78 to 0.86. Of the 12 patients with false positive TFC by deformation imaging, half were asymptomatic mutation carriers at risk for developing ARVC. Of the other 6 false positives, 3 were diagnosed with ventricular arrhythmia from the RVOT. There were no false negative diagnoses using deformation imaging.

Conclusion
All definite ARVC patients were detected when deformation imaging patterns were used to evaluate wall motion abnormalities. This increased sensitivity was accompanied by a slight decrease in specificity. Deformation imaging on its own was not able to reliably distinguish ARVC from other RV related disease. Since no ARVC diagnoses were missed, echocardiographic deformation imaging could be of great value to exclude ARVC in patients referred for ARVC evaluation.
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EMI = electro-mechanical interval, PVC = pulmonic valve closure, TFC = task force criteria