Abstract: P521

Echocardiographic deformation imaging improves assessment of structural disease progression in arrhythmogenic cardiomyopathy

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Background: Arrhythmogenic cardiomyopathy (AC) is an inherited cardiomyopathy characterized by both electrical and structural cardiac disease. Remarkably, progression of structural disease is uncommon during the early stages of AC, while progression of electrical disease is seen frequently. We hypothesize that this discrepancy is caused by the inability of conventional imaging methods to detect subtle changes over time. Echocardiographic deformation imaging may be more sensitive than conventional imaging techniques for detection of structural disease progression in AC.

Objective: To study the incremental value of serial echocardiographic deformation imaging over conventional imaging for detection of structural disease progression in AC.

Methods: We included patients who fulfilled criteria for AC diagnosis (i.e. definite AC) and their mutation-carrying relatives who did not fulfill criteria for definite diagnosis (i.e. early AC). All study participants underwent a baseline and follow-up cardiac evaluation, including conventional echocardiography and deformation imaging. Global right ventricular strain (RV-GLPS) and regional analysis in the right ventricular (RV) subtricuspid region. The deformation pattern in this region was scored as type I (normal deformation), type II (delayed onset, decreased systolic peak, and post-systolic shortening), or type III (systolic stretching and large post-systolic shortening). All measurements were compared between baseline and follow-up to assess disease progression.

Results: 81 subjects were included (50 definite AC, 31 early AC). The mean follow-up duration was 6.7±3.0 years. In definite AC, measurements by conventional echocardiography and deformation imaging both indicated disease progression (p<0.001). In early AC, conventional functional measurements did not reveal disease progression: left ventricular ejection fraction went from 57.5% [IQR 5.5] at baseline to 57.0% [IQR 4.7] after follow-up (p=0.146), and RV fractional area change went from 47.6 ± 6.6% to 47.1 ± 7.2% (p=0.279). However, with deformation imaging, a small but significant decrease in RV global strain was seen in early AC (from 24.9 ± 3.9% to 23.3 ± 3.8%, p=0.035). More pronounced was the deterioration of regional strain patterns, 14 subjects (45%) with early AC showed a change of the deformation pattern in the subtricuspid region (p<0.001), of which 12 subjects progressed to type II and two progressed to type III.

Conclusion: While conventional imaging approaches indeed lack sensitivity to detect disease progression in early AC, echocardiographic deformation imaging reveals progressive mechanical dysfunction in the RV in half off our study population. Serial evaluation by echocardiographic deformation imaging may therefore be helpful in risk stratification of patients during the earliest stages of AC. The prognostic implications of this finding warrants further investigation.