SUPPLEMENTAL MATERIAL

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is characterized by fibrofatty infiltration of the right ventricular (RV) myocardium and clinically presents with ventricular arrhythmias, heart failure, syncope, and sudden death The diagnosis of ARVC/D relies on the demonstration of structural, functional, and electrophysiological abnormalities that are caused by or reflect the underlying histological changes. In 1994, an International Task Force proposed criteria for the clinical diagnosis of ARVC/D which facilitated recognition and interpretation of the frequently non-specific clinical features of ARVC/D. The 1994 criteria were highly specific but they lacked sensitivity for early and familial disease. The criteria have been modified to incorporate new knowledge and technology to improve diagnostic sensitivity, but with the important requisite of maintaining diagnostic specificity. Revision of the diagnostic criteria provides guidance on the role of emerging diagnostic modalities and advances in the genetics of ARVC/D. Electrocardiographic measurements and arrhythmias have been refined. Technical advances in MRI and 2D echo have improved the capability to image the RV with reproducible measurements of volume and systolic function which permits classification of severity and differentiation from normality. Multiple genes, mostly those encoding desmosomal proteins, have been identified as causative in ARVC. In this modification of the Task Force Criteria, quantitative criteria are proposed and abnormalities are defined based on comparison with normal subject data. These modified criteria, in addition to genetic analysis, should assist the clinician to make an accurate diagnosis of ARVC/D and avoid the consequences of misdiagnosis.