Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

Proposed Modification of the Task Force Criteria

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Abstract

Background— In 1994, an International Task Force proposed criteria for the clinical diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) that facilitated recognition and interpretation of the frequently nonspecific clinical features of ARVC/D. This enabled confirmatory clinical diagnosis in index cases through exclusion of phenocopies and provided a standard on which clinical research and genetic studies could be based. Structural, histological, electrocardiographic, arrhythmic, and familial features of the disease were incorporated into the criteria, subdivided into major and minor categories according to the specificity of their association with ARVC/D. At that time, clinical experience with ARVC/D was dominated by symptomatic index cases and sudden cardiac death victims—the overt or severe end of the disease spectrum. Consequently, the 1994 criteria were highly specific but lacked sensitivity for early and familial disease.

Methods and Results— Revision of the diagnostic criteria provides guidance on the role of emerging diagnostic modalities and advances in the genetics of ARVC/D. The criteria have been modified to incorporate new knowledge and technology to improve diagnostic sensitivity, but with the important requisite of maintaining diagnostic specificity. The approach of classifying structural, histological, electrocardiographic, arrhythmic, and genetic features of the disease as major and minor criteria has been maintained. In this modification of the Task Force criteria, quantitative criteria are proposed and abnormalities are defined on the basis of comparison with normal subject data.

Conclusions— The present modifications of the Task Force Criteria represent a working framework to improve the diagnosis and management of this condition.

Clinical Trial Registration— URL: http://www.clinicaltrials.gov. Unique identifier: NCT00024505.

Key Words:
- arrhythmias, cardiac
- arrhythmogenic right ventricular cardiomyopathy/dysplasia
- death, sudden, cardiac
- diagnosis
- echocardiography
- electrocardiography
- magnetic resonance imaging