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Structural progression increases the risk of ventricular arrhythmias in patients with arrhythmogenic cardiomyopathy

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Topic(s):
Myocardial Disease – Clinical: Arrhythmogenic Right Ventricular Cardiomyopathy

Citation:
Background: Arrhythmogenic cardiomyopathy (AC) is an inheritable cardiomyopathy with incomplete penetrance, variable phenotype severity and poorly described disease progression. It is characterized by high risk of life-threatening ventricular arrhythmias and sudden cardiac death in young individuals. Risk stratification and selection of patients presenting without history of life-threatening arrhythmic events for cardioverter-defibrillator implantation in primary prevention remains challenging.

Purpose: We aimed to assess the impact of disease progression on arrhythmic outcomes in AC patients.

Methods: We included consecutive AC probands and mutation-positive family members with at least one complete follow-up evaluation. Echocardiographic and electrical parameters were defined according to the 2010 Revised Task Force criteria at inclusion and at last follow-up. Structural progression was defined as development of new echocardiographic diagnostic criteria. Electrical progression was defined as the development of new diagnostic depolarization, repolarization and/or premature ventricular complex count criteria during follow-up. Non-sustained ventricular tachycardia or ventricular tachycardia occurring during follow-up defined incident ventricular arrhythmic events.

Results: We included a total of 144 patients (48% female, 47% probands, 40±16 years old). At inclusion, 54 patients (37%) had a history of arrhythmic events, 30 patients (21%) had overt structural disease and 114 (79%) had no or minor structural disease. During 7.0 (IQR: 4.5 to 9.4) years of follow-up, 49 patients (43%) with no or minor structural disease at inclusion developed new structural criteria being defined as progressors. Among 80 participants with no or minor structural disease and no arrhythmic history at inclusion, a first arrhythmic event occurred in 14 (17%). The incidence of arrhythmic events was higher in progressors (11/27, 41%) than in non-progressors (3/53, 6%) (p<0.001) (Figure). Structural progression was associated with higher risk of first arrhythmic events during follow-up when adjusted for sex, age at inclusion and follow-up duration, independent of electrical progression (7.6, 95% CI [1.5, 37.2], P=0.01).

Conclusion: Almost half of patients without overt structural cardiac disease at genetic diagnosis develop new structural criteria during 7 years follow-up and 17% experienced their first ventricular arrhythmic event. Structural progression was independently associated with ventricular arrhythmic events during follow-up. These findings highlight the increased risk of arrhythmias when structural abnormalities are detected. Their finding may initiate the evaluation for primary prevention cardioverter-defibrillator implantation.
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