LETTER TO EDITOR

Localized Right Precordial QRS Prolongation in Provocable Brugada Syndrome – A Risk Marker of Additional Electrical Disease in Typical Arrhythmogenic Cardiomyopathy

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Additional arrhythmogenic cardiomyopathy features contributes to a higher risk of life-threatening arrhythmias in Brugada syndrome (1). The measurement of the diameter of right ventricular outflow tract was the only parameter of arrhythmogenic cardiomyopathy features in this study.

If we take typical cases with arrhythmogenic cardiomyopathy by ECG, MRI features, right ventricular angiography, and echocardiographic measurement of the right ventricular size, diameter of the right ventricular ouflow tract and diameter of the right ventricular inferior wall with provocable Brugada ECG a single electrocardiographic risk marker predicts high risk of arrhythmias: localized right precordial QRS prolongation, as published elsethere (2).

Additionally, genetic analysis of plakophilin-2 can cause isolated Brugada syndrome by decreasing sodium current, is by far the dominant cause of isolated arrhythmogenic cardiomyopathy, and what is more important, a combination of arrhythmogenic cardiomyopathy and provocable Brugada ECG with a high risk of ventricular arrhythmias (3). The same is true for other mutations: desmoglein-2 (4), desmoplakin (5), and plakoglobin in animal models (6).

SCN5A is, indeed, the typical gene mutation for Brugada syndrome with atrioventricular conduction disturbance. SCN5A, too, can cause typical arrhythmogenic cardiomyopathy in a minor range of the disease with about 2% (7). Whether these patients have provocable Brugada ECG and high risk of ventricular arrhythmias is still unknown.

In conclusion, echocardiographic, electrocardiographic and genetic features predicts worse prognosis in typical Brugada syndrome with arrhythmogenic cardiomyopathy features and in typical arrhythmogenic cardiomyopathy with provocable Brugada ECG.

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