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Prognosis and evaluation of the risk markers of arrhythmia in a large population of Brugada syndrome patients

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Introduction: Brugada syndrome (BrS) is an inherited arrhythmia syndrome with an increased risk of sudden cardiac death (SCD) despite a structurally normal heart. Many parameters have been suggested to be associated with the risk of ventricular arrhythmias, but only previous symptoms and spontaneous ECG pattern have been consistently associated with the risk of ventricular arrhythmia occurrence.

Objective: The aim of this study was to evaluate the association of these parameters with arrhythmic events in the largest cohort of BrS patients ever described.

Methods: Consecutive patients affected with BrS were recruited in a multicentric prospective registry in France (15 centers) between 1994 and 2016. Data were prospectively collected with an average follow-up of 6.5±4.7 years. ECGs were reviewed by 2 physicians blinded to clinical status.

Results: In this study, we enrolled a total of 1613 patients (mean age 45±15 years; 1119 males, 69%). At baseline, 462 patients (29%) were symptomatic (51 (3%) aborted SCD, 257 (16%) syncope). A spontaneous type 1 ECG pattern was present in 505 patients (31%). Implantable cardiac defibrillator was implanted in 477 patients (30%).

During the follow-up, 91 patients (6%) underwent arrhythmic events (16 SCD (10%), 48 appropriate ICD therapy (3%) and 27 ventricular arrhythmias (2%). Thirty-six patients (2%) died of non-arrhythmic causes. Mean age at the first event was 44±15 years.

In our cohort, event predictors were SCD (HR: 18.3; 95% CI: 11.2–29.8; $p<0.0001$), syncope (HR: 2.9; 95% CI: 1.8–4.9; $p<0.0001$), age >60 years (HR: 0.11; 95% CI: 0.032–0.377; p=0,0004), gender (HR: 2.96; 95% CI: 1.6–5.4; p=0.0005), spontaneous type 1 (HR: 2.14; 95% CI: 1.42–3.23; p=0.0003), type 1 ST elevation in peripheral ECG lead (HR: 3.6; 95% CI: 1.9–7.1; p=0,0001), fragmented QRS (HR: 3.37; 95% CI: 1.37–8.32; p=0,008), AvR sign (HR: 2.2; 95% CI: 1.4–3.8; p=0,0007), QRS >120ms in D2 lead (HR: 2.2; 95% CI: 1.4–3.6; p=0,001) and QRS >90ms in V6 (HR: 2.1; 95% CI: 1.3–3.3; p=0,001). All the others parameters including early repolarization pattern (ERP) and EPS were not predictor of events.

Conclusion: In the largest cohort of BrS patients ever described, we confirmed that symptoms, age, gender, spontaneous type 1, type 1 ST elevation in peripheral ECG lead, fragmented QRS, AvR sign, QRS >120ms in D2 and QRS >90ms in V6 are associated with arrhythmic events whereas ERP and EPS were not.