Performance of sudden cardiac death prediction strategies before and after Septal Reduction Strategies in patients with Hypertrophic Cardiomyopathy

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Background: Actual sudden death (SD) risk stratification strategies for Hypertrophic Cardiomyopathy have not been validated for patients after a septal reduction treatment (SRT). Therefore, it is largely unresolved the quantification of the ventricular arrhythmias (VA) burden after an SRT and how risk stratification should be performed in such patients.

Purpose: To evaluate the performance of ESC SD Risk stratification model and AHA ACC Risk Factors for SD in the identification of patients with lethal ventricular arrhythmias after SRT.

Methods: Data from 11 high-volume HCM specialty centers from the international SHaRe Registry were used to describe the natural history of patients with SRT. Patients were followed from SRT until last follow-up or meeting the composite outcome of ventricular arrhythmias (sudden cardiac death (SCD), resuscitated cardiac arrest, or appropriate implantable cardioverter-defibrillator (ICD) therapy). Cox proportional hazards models were used to identify predictors of prognosis and incident development. Calculation of SD score was performed before the SRT and at 1 year from the procedure.

Results: Of the 10385 patients part of the ShaRe Registry, 1835 (18%, 968 (53%) males) underwent a SRT during a median follow-up of 6.8 (IQR 3.4-9.8). A total of 455 (25%) had Alcohol Septal Ablation (ASA) and 1377 (75%) had myectomy. Patients who underwent myectomy were younger (49 ± 18 vs 56 ± 15 years, p < 0.01), more frequent with a sarcomeric mutation (396 (29%) vs 111 (24%), p < 0.01). Over 6.8 years after the procedure, 77 (4%) died because of HCM (0.6%/year) and 87 (5%) presented a composite VA outcome (0.7%/year). Development of the composite VA event was associated with younger age at procedure (38 ± 20 vs 54 ± 18 years, p < 0.01), the development of a LV EF < 50% (29 (33%) vs 351 (20%), p < 0.01), a more severely dilated LA (47 ± 7 vs 43 ± 11 years, p < 0.01) and greater LV maximal wall thickness (24 ± 6 vs 21 ± 6 years, p < 0.01). Significant predictor of the composite VA outcome was the presence of a LA diameter ≥ 45 mm (HR, 2.6 [95% CI, 1.7–3.5]). Patients who presented the VA outcome were classified according to ESC SCD Risk pre and post SRT as: 40 (46%) as high risk, 31 (36%) as intermediate risk, 16 (18%) as low risk pre intervention and 26 (46%) as high risk, 41 (47%) as intermediate risk, 20 (18%) as low risk post procedure. AHA ACC SCD risk factors pre and post SRT classified patients as: 56 (64%) as class IIA 25 (29%) as Clas IIB, 6 (7%) as Class III and post SRT 42 (48%) as Class IIA, 31(36%) as Class IIb, 14 (16%) as Class III.

Conclusions: Current stratification strategies potentially underestimate the risk of SD pre and post SRT. Specifically, ESC SD risk strategies underperformed in the identification of intermediate high risk patient, whereas AHA ACC were capable of identifying 84% of patients who had a malignant VA.