Sustained ventricular tachycardia of left, right or both bundle branch block morphology in patients with Arrhythmogenic Cardiomyopathy

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Aims: In arrhythmogenic cardiomyopathy (ACM) sustained monomorphic ventricular tachycardia (VT) typically displays left bundle branch block (LBBB) morphology. Sustained VT with right bundle branch block (RBBB) morphology is very rare despite the frequent left ventricular involvement. The present study sought to assess the prevalence of spontaneous sustained LBBB-VT, RBBB-VT or both as well as clinical and genetic differences associated with these VT types.

Methods and results: Twenty-six centers from 11 European countries provided information on 952 patients with ACM and >1 episode of sustained VT observed during the patients' clinical course. VT was classified as: LBBB-VT; RBBB-VT or LBBB+RBBB-VT.

Among 952 patients, 881 (92.5%) had LBBB-VT alone, 71 (7.5%) had RBBB-VT [alone in 42 (4.4%) patients or with LBBB-VT in 29 (3.0%) patients]. Male prevalence was 90.5%, 79.2% and 55.9% in the RBBB-VT, LBBB-VT and LBBB+RBBB-VT groups, respectively (P=0.001). Patients' age at first VT did not differ amongst the 3 VT groups. ICD implantation was more frequent for the RBBB-VT and the LBBB+RBBB groups (≈90% each) vs. 67.9% for the LBBB-VT group (P=0.001). Death incidence (9.5%–17.2%) was not significantly different between the 3 groups (P=0.425). Plakophylin-2 mutations predominated in the LBBB-VT and LBBB-VT+RBBB-VT groups (47.2% and 27.3%, respectively) and Desmoplakin mutations in the RBBB-VT group (36.7%).

Conclusion: This large European survey demonstrates: 1) Sustained RBBB-VT is documented in 7.5% patients with ACM; 2) Males markedly predominate in the RBBB-VT and LBBB-VT groups but not in the LBBB+RBBB VT group; 3) Distribution of desmosomal mutations appears to be different in the 3 VT groups.