Incidence and Clinical Management of Atrial Arrhythmias in Patients with Catecholaminergic Polymorphic Ventricular Tachycardia

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Abstract

Background: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a genetic arrhythmia syndrome characterized by adrenergically-triggered ventricular arrhythmias, syncope, and sudden cardiac death. Several small studies suggest that atrial arrhythmias (AAs) are common in patients with CPVT. Objective: To determine the incidence and type of AAs observed within a large, single-center cohort of CPVT cases as well as the efficacy and durability of AA-directed management. Methods: In this retrospective study, the electronic medical record of 129 patients (52% female; average age at diagnosis 20.8 ? 15.3 years) with CPVT (95% with a putative CPVT1-causative RYR2 variant) between 01/2000 and 09/2019 were reviewed for electrocardiographic evidence of AAs. Clinical features and efficacy of pharmacologic and ablation therapy were assessed. Results: Overall, 10/129 (7.8%) CPVT patients, all RYR2 variant-positive, had evidence of an AA (atrial fibrillation/flutter in 6, atrial tachycardia in 3, and supraventricular tachycardia in 1). The median age at AA diagnosis was 23 (14.2-35.5) years. 8/10 of patients experienced symptoms attributed to their AA, including inappropriate shocks. All patients were trialed on anti-arrhythmics, including ?-blockers, and/or flecainide. Owing to drug failure (1/10), drug intolerance (1/10), or patient preference (2/10); 4/10 patients received an ablation. Over a median follow-up of 23.5 (4.5-63) months, no AA recurrences were observed. Conclusion: Compared to prior studies, the incidence of AAs in this large, single-center referral cohort of CPVT patients was substantially lower (7.8% vs. 26%-35%). Although larger multi-center studies are needed to confirm, this study suggests that ablation is efficacious and durable in CPVT-associated AAs.

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