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Resting ECG and Signal-Averaged ECG can Detect Depolarization Abnormalities in Pediatric Catecholaminergic Polymorphic Ventricular Tachycardia

Tofighi T. (1), (2), Hamilton R.M. (2)
Faculty of Medicine, University of Toronto, Toronto, CA (1); The Hospital for Sick Children, Toronto, CA (2)

INTRODUCTION: Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) is characterized by ventricular arrhythmias (VA) in response to physical activity or emotional stress, resulting in presentations of cardiac syncope or sudden cardiac arrest (SCA) with an untreated mortality of 50%. Despite limitations in non-ambulatory patients, diagnosis currently relies on the presence of VA during exercise stress testing, although a negative result does not exclude a diagnosis of CPVT. We sought to assess the diagnostic utility of resting ECGs and Signal-averaged ECGs (SAECGs) in children subsequently diagnosed with CPVT.

METHODS: We conducted a retrospective analysis of baseline ECG and SAECG data in 33 patients diagnosed with CPVT between March 1997- March 2016. A series of 192 waveform measurements across 12 leads were used for ECG comparisons, and for SAECG we extracted: 1) unfiltered (QRSDu); 2) filtered (QRSDf) QRS durations; 3) duration of high frequency (HFLA) signals; 4) terminal root-mean-square (RMS) voltage. A 1:2 age and gender match was used to compare ECG parameters from initial and most recent ECGs in 29 CPVT patients with a cohort of 88 normal school children using a 2-sample t-test assuming equal variance. Data from this cohort of 150 control patients was used to derive BSA and gender corrected Z-score from regression equations.

RESULTS: Baseline ECG data in 29 CPVT patients as compared to controls revealed statistically significant bradycardia (p=0.016) and QRS prolongation (p=0.005). Raw QT was prolonged (p=<0.001) but rate-corrected QT was not. Baseline SAECGs were performed in 17 (52%) of patients. For SAECG, the QRSDu was abnormal in 9 patients with an abnormal Z-score >2 (53%). Abnormalities of QRSDf, HFLA and RMS were less common. QRSDu was not normally distributed, with patients distributed throughout Z values of -1.0 to 4.5 (Figure 1).

CONCLUSION: ECG and SAECG are abnormal in a significant proportion of children with CPVT. SAECG abnormalities, particularly prolongation of unfiltered QRS duration, may be helpful in identifying CPVT, particularly in non-ambulatory patients who cannot formally complete exercise testing. Recent studies of patch clamp recordings in CPVT iPSC cardiomyocytes identify a decrease in action potential upslope V_{max} that may explain QRS prolongation.

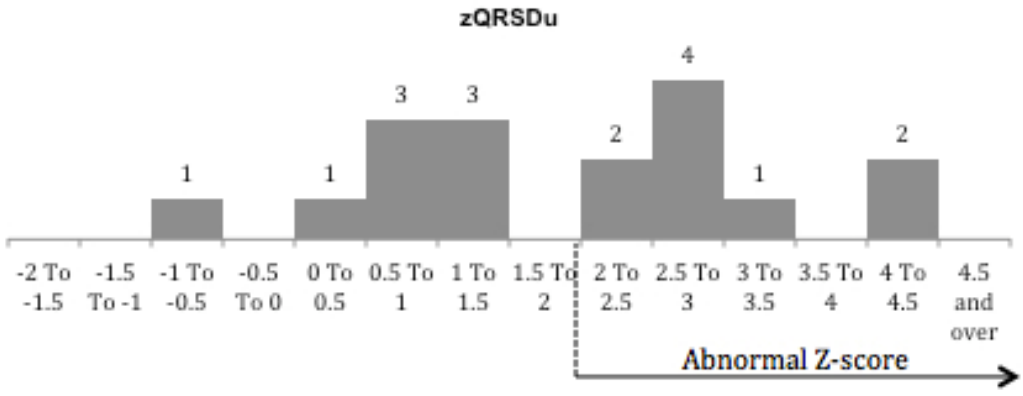

Figure 1