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Arrhythmia characterization in catecholaminergic polymorphic ventricular tachycardia and CPVT-related phenotypes in children – a single centre experience

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Introduction:
Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare, inherited disorder associated with emotion/exercise-related syncope and sudden cardiac death. It is characterized by adrenergically induced ventricular and supraventricular arrhythmias in structurally normal hearts.

Objective:
Electrocardiographic characteristics of resting ECG and exercise- or isoproterenol-induced arrhythmias were investigated in our patients with CPVT and CPVT-related phenotypes. Demographic data and clinical presentation were also summarized in association with ECG findings.

Method:
Patients were diagnosed with CPVT or CPVT-related phenotypes according to exercise-induced or isoproterenol-provoked polymorphic or bidirectional ventricular tachycardia (VT) in 2004-2011 at our centre.

Results:
9 patients (5/4 male/female) were identified at the age of 2-15 years (median: 11.2). Syncope and sudden cardiac arrest (SCA) and other manifestations, such arrhythmic heart sounds or atypical spells, as presentation showed equal distribution among our patients. Baseline ECG showed sinus bradycardia and U waves in most cases, but ventricular ectopy appeared, also spontaneously in 2 cases. Mean QTc was 422.8ms (380-480ms; STD: 36.8ms). VT was provoked by exercise in 4 patients and by isoproterenol in 3 (small or disabled) patients, respectively. In 2 cases with Andersen-Tawil syndrome non-sustained VT occurred spontaneously, but not on exercise tests. Ventricular ectopy started to arise at a mean heart rate of 116.8/min (STD: 11.6/min). Non-sustained VT was bidirectional in 55% and polymorphic in 45%. The initiating beat of VT was late-coupled (mean coupling interval: 446ms, STD: 87ms) and wide (mean QRS duration: 126ms, STD: 14ms), and QRS morphology suggested an outflow tract origin in 66%. Multiple episodes of VT in 1 patient were not common, so reproducibility of VT morphology could not be investigated. The more serious presentation of SCA was not associated with a distinct type of VT or initiating ventricular beat, or with earlier age of presentation. Supraventricular arrhythmia was detected only in 1 patient during the median follow up of 3.9 years (IQR:1.3-5.6).

Conclusion:
In our patient population with CPVT ventricular tachycardia was often polymorphic, late-coupled, and initiated from the outflow tract.
Resting ECG was usually unremarkable with sinus bradycardia and prominent U waves.
Appearance of supraventricular arrhythmia was not common in childhood.