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Predictors of Adverse Events in Children with Arrhythmogenic Right Ventricular Cardiomyopathy

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Introduction/Objective: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an increasingly recognized cause of sudden death in young adults and children. Children may be referred for symptoms, signs or a positive family history of ARVC or an ARVC-causing gene. We sought to identify predictors of adverse events among referred children meeting 2010 proposed modified Task Force criteria for probable or definite ARVC.

Methods: We reviewed all patients referred to a pediatric cardiovascular genetics/electrophysiology program for assessment of possible ARVC. Patients were routinely investigated with ECG, SAECG, Holter, Echo MRI and (after 2003) gene testing. If only partial criteria for ARVC were present, patients underwent catheterization for RV angio, EPS and biopsy. Adverse events were defined as sustained VT, appropriate ICD discharge or death. Univariate predictors of adverse events were assessed by unpaired T-test, Chi-square analysis or Fisher's Exact Test as appropriate.

Results: Of 423 patients referred for assessment, 77 met inclusion criteria (48 ARVC, 29 probable ARVC) with a mean age of 14.9 ± 3.3 (range 5 to 18) years at last follow-up. No adverse events were identified in 71, whereas 6 had adverse events (including 1 death, 3 appropriate ICD discharges and 3 sustained VT). Predictors of adverse events included presentation as the index case v. relative (6/35 v. 0/35, p=0.026), extra-systoles, (4/21 v.1/45, p=0.049) prolonged S duration in V1 (73 v. 53 ms, p=0.006), major criteria for global or regional dysfunction (6/32 v. 0/33, p=0.027, documented LBBB VT (4/13 v 2/57, p=0.020), any minor arrhythmia criteria (5/24 v 1/44, p=0.031) and absence of any major family history criteria (6/34 v/ 0/36, p=0.018). There was a tendency for patients meeting ARVC criteria to have adverse events compared to those diagnosed with borderline criteria.

Conclusions: Adverse events can occasionally occur in children identified with probable or definite ARVC. Children presenting with symptoms, signs of right ventricular dysfunction or documented arrhythmias appear to be at highest risk of adverse events, compared to those referred with or identified to have a family history of ARVC. These findings should be validated in a larger cohort or multicenter study of children with ARVC.