Catecholaminergic polymorphic ventricular tachycardia is an uncommon, potentially lethal, ion channelopathy. Standard therapies have high failure rates and little is known about treatment in children. Newer options such as flecainide and left cardiac sympathetic denervation are not well validated. We sought to define treatment outcomes in children with catecholaminergic polymorphic ventricular tachycardia. This is a Pediatric and Congenital Electrophysiology Society multicenter, retrospective cohort study of catecholaminergic polymorphic ventricular tachycardia patients diagnosed before 19 years of age. The cohort included 226 patients, including 170 probands and 56 relatives. Symptomatic presentation was reported in 176 (78%). Symptom onset occurred at 10.8 (interquartile range, 6.8-13.2) years with a delay to
diagnosis of 0.5 (0-2.6) years. Syncope (P<0.001), cardiac arrest (P<0.001), and treatment failure (P=0.008) occurred more often in probands. β-Blockers were prescribed in 205 of 211 patients (97%) on medication, and 25% experienced at least 1 treatment failure event. Implantable cardioverter defibrillators were placed in 121 (54%) and was associated with electrical storm in 22 (18%). Flecainide was used in 24% and left cardiac sympathetic denervation in 8%. Six deaths (3%) occurred during a cumulative follow-up of 788 patient-years. This study demonstrates a malignant phenotype and lengthy delay to diagnosis in catecholaminergic polymorphic ventricular tachycardia. Probands were typically severely affected. β-Blockers were almost universally initiated; however, treatment failure, noncompliance and subtherapeutic dosing were often reported. Implantable cardioverter defibrillators were common despite numerous device-related complications. Treatment failure was rare in the quarter of patients on flecainide. Left cardiac sympathetic denervation was not uncommon although the indication was variable.