Characteristics of pediatric Brugada Syndrome


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[Introduction] Brugada syndrome (BS) is a primary cardiac electrical disorder, which manifests as coved-type ST-segment elevation, either spontaneously after the administration of intravenous class I antiarrhythmic drugs. The syndrome predisposes to ventricular arrhythmias eventually leading to sudden cardiac death (SCD). However, diagnosis of BS is sometimes difficult in children.

[Methods] Children for the evaluation and diagnosis of BS, and definite diagnosis of BS were included in this study.

[Results] The consecutive 9 patients with BS (mean age 10.4 ± 5.6 years, M:F=8:1) were included in this study. The initial onset was syncope in 3 patients (1 was due to vasovagal syncope, and the other 2 were due to VT), heat screening in 3 patients, coincident recording of electrocardiogram in 2 patients, and familial study in 1 patient. In these patients, 5 of 6 patients had SCN5A mutation and 1 patient was under investigation of genetic anomaly. Electrophysiological Study (EPS) was performed in 4 patients, ventricular tachycardia (VT) was induced in 2 patients, and ventricular fibrillation (VF) in 1 patient. Associated arrhythmias were: sick sinus syndrome (SSS) in 4 patients, VT in 2 patients, and atrial tachycardia (AT) in 2 patients, and supraventricular tachycardia (SVT) in 1 patient. Implantable cardioverter defibrillator (ICD) was implanted in 2 patients. Catheter ablation for atrial tachycardia was performed in 2 patients. During mean follow up of 6.7 ± 5.0 years, 1 patient detected VT by remote monitoring of ICD, however no other patients developed VF, syncope or sudden death.

[Conclusions] In this study, none of the patient documented spontaneous syncope due to VF. Incidence of SCN5A anomaly is higher (5/6 patients, 83%) than adult patients. Supra ventricular arrhythmia (SSS, AT, SVT) was noted more than half of the patients. And also, monomorphic VT was noted 22% of the patients. In children, fibrosis of right ventricular outflow tract may not be profound, that may reduce the possibility of the occurrence of VF or polymorphic VT. Catheter ablation of AT, or SVT may be useful to prevent inappropriate discharge of ICD in children.