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Arrhythmic faces and risk stratifications of hypertrophic cardiomyopathy in children; Single center experience from Turkey

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Background: Hypertrophic cardiomyopathy (HCM) is the most common cause of sudden cardiac death(SCD) in the young adults. Although there are large series about arrhythmic characteristics of HCM in adults, it is limited in childhood. We evaluated pediatric HCM patients followed at our center from different angles.
Patients and Methods: We identified 120 pediatric patients with HCM between January 2010 to August 2017. Demographic characteristics, non invasive and invasive diagnostic tests findings, treatment and follow-up results of the patients were evaluated. Patients with ICD and poor prognosis were scanned backwards and their distinctive and prominent features were searched. The contributor factors, which could help to selection SCD candidate, were tried to be found.
Results: The mean age of patients was 8.2± 5.9 years (0.05-21) and 64.2 % were males. Electrocardiographically, the most common finding was ventricular preexcitation (n = 12, 10%) except for left ventricular hypertrophy. Echocardiographically, there were concentric hypertrophy in the majority (51.2%) and obstruction (LVOTO) in 26 (22%). Implantable cardioverter-defibrillator (ICD) implantation was performed in 24 patients(14 transvenous, 10 epicardial). Two of them were inserted for secondary prevention; whereas the remaining for primary prevention. ICD dependent complications were seen in 7 (29 %) patients. During follow-up period (mean 23.0± 22.5 months); 3(2.5%) patients has died (2patient with Danon's disease, due to heart failure and uncontrolled VF episodes, 1 patient with Noonan Syndrome in postoperative period) and none of them had an ICD. There was a significant difference between good and poor prognostic groups in terms of QT prolongation, left atrial dimensions, presence of preexcitation, and left ventricular outflow tract obstruction.
Conclusion: HCM is still an important disease, as it is common in the community. In order to avoid both SCD and ICD related complications, the right patient selection for ICD is important. New factors that may contribute to conventional risk factors are being investigated. In our own group, we showed that some of these modifier factors are effective on the prognosis.