Clinical Spectrum of Primary Cardiomyopathy Associated Ventricular Arrhythmia in Children: 5-year Single Center Experience

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Objectives
The aim of our study was to evaluate the characteristics of ventricular arrhythmias in children with primary cardiomyopathies (CMPs).

Methods
Patients with cardiomyopathy who had visited pediatric cardiology department from January 2010 to December 2014 were retrospectively reviewed. Patients with secondary cause of cardiomyopathy were excluded.

Results
A total of 163 patients with cardiomyopathy was enrolled and grouped according to the echocardiography and MRI findings as follows; Hypertrophic cardiomyopathy (HCMP, n=71, 43.6%), dilated cardiomyopathy (DCMP, n=50, 30.7%), isolated noncompaction cardiomyopathy (IVNC, n=32, 10.6%), restrictive cardiomyopathy (RCMP, n=7, 4.3%), arrhythmogenic right ventricular cardiomyopathy (ARVC, n=3, 1.8%).

Twenty-five(15.3%) of 163 patients had premature ventricular contractions (PVCs) and/or ventricular tachycardia (VT) (17 male, 68%; 8 female, 32%). Frequency of ventricular arrhythmia with cardiomyopathies was as follows; HCMP (n=8, 11.3%), DCMP (n=9, 18%), IVNC (n=4, 12.5%), RCMP (n=1, 14.3%) and ARVC (n=3, 100%). The mean patient age and the mean follow-up period were 5.3 ± 11.5 years and 12 ± 12.3 months, respectively.

Exercise testing was performed in 4 (16%) patients and sustained/nonsustained VT was not induced in any patient. Atrial fibrillation/flutter was developed in 2 patients with ARVC. PVCs and VT characteristics of patients were mild monomorphic PVCs (n=7), mild polymorphic PVCs (n=2), moderate monomorphic PVCs (n=1), moderate polymorphic PVCs (n=1), frequent monomorphic PVCs (n=1), sustained monomorphic VT (n=1), sustained polymorphic VT (n=3), nonsustained monomorphic VT (n=4), nonsustained polymorphic VT (n=5).

Antiarrhythmic treatment was instituted in 20 cases (80%). ICDs were implanted in 10 patients( 2 cardiac arrest, 1 sustain VT, 7 primary). One patient died suddenly, 2 with ARVC, 1 with IVNC and 1 with DCMP patients died for incessant VT, and 1 with ARVD died for multiorgan dysfunction after implantation of biventricular assist device.

Conclusion
VT is a potentially life-threatening arrhythmia because it leads to VF and sudden cardiac arrest especially in cardiomyopathic patients. Consequently, these patients should be monitored closely and carefully for ventricular arrhythmia.