Cardiomyopathy in a pediatric population: Key results from 10 years


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Introduction and objectives: Cardiomyopathy is a common cause of heart failure and the main cause of heart transplantation in children. Objectives: (1) To elaborate a pediatric cardiomyopathy registry, (2) to describe the incidence, presentation and outcome of cardiomyopathy in a pediatric population, (3) to examine the medium and long-term course of functional status related to clinically important events.

Methods: Medical records of the patients under 15 years old affected by cardiomyopathy according to echocardiographic patterns, from January 2002 to December 2012, were reviewed. Patients with congenital heart defects and infants of diabetic mothers were excluded.

Results: 57 patients met inclusion criteria, 35 (61.4%) boys and 22 (38.5%) girls. Median age at diagnosis was 12 months. 30 (52.6%) patients were less than one year old and 3 patients were diagnosed during fetal life. The type of cardiomyopathy was: hypertrophy in 30 (52.6%), dilated in 18 (31.5%), left ventricular noncompaction in 7 (12.2%), restrictive in 1 and ventricular dysplasia in 1. In the hypertrophic cardiomyopathy group, 23 (76.6%) had a known cause: 7 with familial cardiomyopathy, 5 with malformation syndrome, 4 with neuromuscular disorder, 4 with arterial hypertension, 1 with mitochondrial disease, 1 with errors of metabolism, 1 with hyperinsulinism. In the dilated cardiomyopathy group, 12 (66.6%) had a known cause: 6 with familial cardiomyopathy, 2 with myocarditis, 1 with syndrome, 1 with anthracycline toxicity, 1 with arterial hypertension, 1 with mitochondrial disease. Mean follow up time was 3.7 years. 25 (43.8%) were admitted due to heart failure decompensation, 7 (12.2%) had an arrhythmogenic event and 2 (3.5%) had an aborted sudden death event. Surgical intervention was undertaken in 4 (7%), heart transplantation in 11 (19.3%), and implanted defibrillation device in 5 (8.8%). 6 (10%) patients dead, all of them were less than 1 year old.

Conclusions: More than 50% of children affected by cardiomyopathy have a known cause. The most frequent type of presentation is hypertrophic cardiomyopathy. Cardiac decompensation occurred in 44% of children affected by cardiomyopathy and heart transplant was undertaken in 19% of them. Younger age at presentation are associated with poor outcomes.