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Dilated cardiomyopathy - course and outcome in children

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Introduction: Dilated cardiomyopathy (DCM) is a congenital myocardial disorder resulting end-stage heart failure usually with rapid progression. The aim of study was to provide a detailed description of the etiology, course, and outcome of the DCM in children.
Methods: Retrospective analysis of 61 children enrolled in the Hungarian Pediatric DCM Registry between 2005-2010 was performed. Inclusion criteria were defined as decreased left ventricular function (FS<30%) and/or increased left ventricular end-diastolic diameter (LVEDD>2SD), which were not caused by any other structural abnormality, or arrhythmia. End points were defined as death, heart transplantation (htx) or implantation of ventricular assist device (VAD). Patient's demographic data, time elapsed from diagnosis to the end points, echocardiographic parameters measured at the time of diagnosis, and left ventricle trabeculation (non-compaction) were investigated.
Results: 55 patients were enrolled. Sex 6/5 (m/f); median age at diagnosis was 5.38 years. Non-compaction cardiomyopathy was diagnosed in 15 cases. Familiar inheritance could be proven in 16 cases (29%) involving 13 families. Htx was performed in 11 cases, VAD implantation in 4 cases. The mean time elapsing from diagnosis to htx or VAD application was 3.7 years. In lack of htx 13 patients died. Their average follow-up time from diagnosis to death was 3.39 years. Known etiology was proven only in 1 case (Alström syndrome).
Conclusions: 1. Etiology of DCM was not found in most cases. 2. Pediatric DCM has a rapid progression and high need for htx. 3. Familiar cases were found in high percentage of the cases. 4. Due to the high number of idiopathic cases, molecular diagnostic tests are recommended to support early recognition in uncertain cases.