

Cardiac involvement and therapeutic outcomes in pediatric patients with Duchene Muscular Dystrophy in the pediatric cardiology department of Ahepa in Thessaloniki during the period 2014-2015: a retrospective study.

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Introduction: Duchene muscular dystrophy(DMD) represents the most common and severe form of muscular dystrophy and it is due to mutations in the dystrophin gene on chromosome Xp21.1. Typical presentation of cardiac involvement are dilated cardiomyopathy affecting the left ventricle, chronic heart failure and heart rhythm disorders.

Materials and Methods: It is a retrospective survey that took place during the period 01/01/14-01/01/15 in Ahepa pediatric cardiology department. Children with muscular dystrophy diagnosed by muscular biopsy were enrolled in the study. The statistical methods that were used are the following: a) descriptive analysis of demographic data and cross-tabulation matrices, b) Pearson Chi -Squared Test. The statistical analysis was carried out with the use of statistical parcel IBM SPSS Statistics 22.

Results: Of the total of 99 children with muscular dystrophies, 48 had DMD. The mean age of diagnosis of DMD was 5 years and 2 months. Cardiac involvement was seen in 14 children (29, 2%). All of them had an LV ejection fraction (EF) <70% measured with M-Mode. Drug treatment in the means of angiotensin- converting enzyme inhibitors (ACE) and beta blockers (BBs), received 8 patients. No improvement of EF (>70%) were seen after the treatment. ECG changes were noticed in 71% of the patients with cardiac involvement and EF<70%. Muscular biopsy was positive in 10 out of 14 DMD patients with cardiac involvement and no significant correlation was found between these two parameters.

Conclusions: The current recommendations of the DMD Considerations Working Group identified ACE as first line therapy in patients with DMD with LV dysfunction. On the other hand, the use of BBs is controversial and limited in small-sized studies. According to our study, the use of drug treatment preserved rather than improved the LV function.