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 and Purpose

- 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.
- The purpose is to estimate the 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.

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 -Sqared 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.
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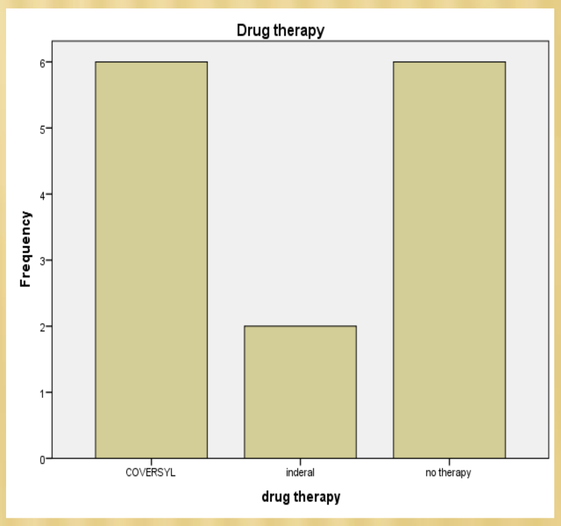
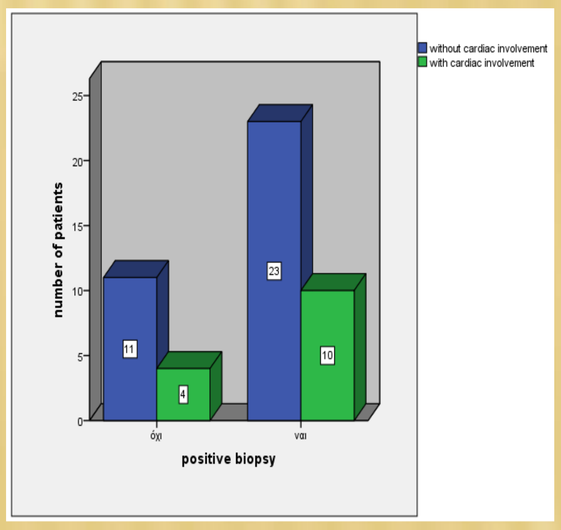
- No improvement of EF (>70%) were seen after the treatment. ECG changes were noticed in 71% of the patients with cardiac involvement and EF<70%.
- Muscular biopsy was positive in 10 out of 14 DMD patients with cardiac involvement and no significant correlation was found between these two parameters.





Conclusions

- The current recommendations of the DMD Considerations Working Group identified ACE-Is as first line therapy in patients with DMD with LV dysfunction.
- 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.

References

- A current approach to heart failure in Duchene muscular dystrophy. Domenico D´ Amario et al, http://heart.bmj.com on July 1st 2017.
- Cardiac Involvement Classification and Therapeutic Management in Patients with Duchenne Muscular Dystrophy. Abdallah Fayssoil et al, Journal of Neuromuscular Diseases 4 (2017) 17–23.
- Long-Term Pathological Follow-Up of Myocardium in a Carrier of Duchenne Muscular Dystrophy With Dilated Cardiomyopathy. Toru Kondo et al. *Circ Heart Fail*. February 2017.