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Letter to the Editor

Duchenne muscular dxstrophy requires close cardio-pulmonary monitoring for therapeutic decision making

[1] Josef Finsterer, [2] Claudia Stollberger
[1] Neurology & Neurophysiology Center, Vienna, Austria
[2] Second Medical Department, Klinik Landstrasse, Vienna, Austria

Corresponding Author: **Josef Finsterer**

We read with interest the article by Yoon *et al*. about a retrospective analysis of data from the Korean Health Insurance database on the cardiac and pulmonary management of patients with Duchenne muscular dystrophy (DMD) [1]. Among 479 DMD patients, 41% underwent cardiologic investigations and 60% underwent pulmonary function tests [1]. Angiotensin-converting enzyme inhibitors (ACEI) were prescribed to 30% of patients undergoing cardiac evaluation and pulmonary rehabilitation to 42% [1]. Ventilatory support was provided to 17% [1]. Frequency of cardiac and pulmonary examination increased with progression of the disease. The study is appealing but raises concerns that should be discussed.

Because DMD patients frequently develop ventricular arrhythmias and because the study aimed at describing the cardio-therapeutic management of DMD patients, we should know how many of the included patients had received a pacemaker, and implantable cardioverter defibrillator (ICD), or a cardiac resynchronisation (CRT) system. It is also crucial to know how many of the patients died from sudden cardiac death.

Because 26 patients of the author's institute were under a treatment with steroids [1], we should know if the outcome measures (survival, time until respirator, heart failure), varied between those receiving steroids plus cardiac therapy and those who only received cardiac therapy without steroids. Furthermore, we should know the percentage of patients of the entire cohort who took steroids.

Because cardiac involvement in DMD can manifest with dilative cardiomyopathy, which can be complicated by heart failure [2], it would be interesting to know how many of the included patients underwent determination of pro-brain natriuretic peptide (pro-BNP). We should also know if elevated proBNP correlated with systolic function as assessed by fractional shortening or ejection fraction on echocardiography.

Because DMD patients can also present with left ventricular hypertrabeculation (LVHT), also known as non-compaction [3, 4], it would be interesting to know how many of the included patients had LVHT on echocardiography. Because LVHT can be complicated by heart failure, ventricular arrhythmias and intraventricular thrombus formation, it is crucial to identify these patients, as their risk of cardio-embolism, sudden cardiac death, or acute heart failure may be increased compared to the remainder of patients.

Missing is the information in how many of the included patients the diagnosis DMD was genetically confirmed and in how many patients the diagnosis was based on immune-histology.

An explanation should be provided why only 41% of patients underwent echocardiography [1]. Was this due to limited access to appropriate facilities or due to non-compliance? Because cardiac involvement should be recognised early and because of associated risks, it should be offered to every DMD patient and carried out in as many patients as possible.

Overall, the interesting study has some limitations that call the results and their interpretation into question. Clarifying these weaknesses would strengthen the conclusions and could improve the study. DMD patients should undergo extensive cardiac and pulmonary evaluation at regular intervals as the therapeutic management strongly depends on the results of these examinations.

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