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Letter to the Editor

Cardiac and Neuromuscular Findings Are Required to Interpret Systolic Function in Duchenne Muscular Dystrophy

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Letter to the Editor

In a recent article, Cirino et al. investigated 40 young male patients with Duchenne Muscular Dystrophy (DMD) and normal left ventricular ejection fraction [1]. They found in 50% of the patients an abnormal global longitudinal strain (GLS >-18) indicating subtle disturbances in the longitudinal contraction of the myocardium. We have the following questions and concerns:

GLS has been shown to be subject to "Physiologic Variations" and loading conditions depending on patient demographics (age, gender, race), clinical factors (heart rate, blood pressure, weight or body surface area) [2]. Thus, it would be interesting to know the ethnicity of the included patients, their heart rate, blood pressure, volume status, and body surface area, and if there were any associations of these parameters with GLS.

Measurement of GLS has been shown to have a considerable intra- and interobserver variability [3]. Thus, it would be of interest if any studies about reproducibility of the measurements in the investigated cohort of patients were carried out.

Scoliosis is a frequent problem in DMD patients, especially when they become wheelchair-bound. Scoliosis may affect cardiac function. Thus, it would be of interest to know how many patients suffered from scoliosis and if they differed regarding GLS from patients without scoliosis.

Among electrocardiographic findings, only the rate of left ventricular hypertrophy is reported. There are, however, other electrocardiographic abnormalities which might indicate cardiac involvement and risk for sudden cardiac death like sinustachycardia, ST-elevation, broadening of the QRS complex or QT-prolongation. How many patients suffered from supraventricular or ventricular tachycardia? Was 24-hour Holter monitoring carried out to look for arrhythmias?

Among the echocardiographic findings, only left ventricular ejection fraction is reported. It would be of interest to know the thickness of the myocardium, the morphology and function of the cardiac valves and if there were any cases of left ventricular noncompaction [4].

Cardiac involvement may also be detected by laboratory findings like levels of brain natriuretic peptides or troponin. Were these investigations carried out in the included patients?

Regarding pharmacotherapy, only corticosteroid is mentioned. Did any of the included patients receive any other medication which may affect the cardiovascular system? How many of the patients received angiotensin-converting enzyme inhibitors, angiotensin II receptor-blockers, beta-blockers, or diuretics?

Cardiac function may strongly depend on pulmonary functions in DMD patients. How many of the patients suffered from muscular respiratory dysfunction and how many patients were on intermittent or permanent non-invasive positive pressure ventilation? To which degree did GLS depend on respiratory functions? Cardiac functions may also depend on the degree of limb muscle involvement [5]. Did GLS or systolic function correlate with any of the scores which assess the degree of limb muscle involvement?

In conclusion, more data about cardiac and neuromuscular findings should be provided in order to explain the observed disturbances in the longitudinal contraction of the myocardium and to assess if measurement of GLS is a useful tool for detection of beginning cardiac involvement in DMD patients.

Declarations

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.' None of the authors has any conflict of interest to disclose.

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