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Letter to the Editor

Cardiac Disease in Dystrophinopathies Requires Longitudinal MRI Studies

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Dear Editor,
We read with interest the article by Girija *et al*. on the prevalence of cardiac magnetic resonance imaging abnormalities such as late gadolinium enhancement (LGE), regional wall motion abnormalities (rWMA) and reduced left ventricular ejection fraction (LVEF) in 38 Indian patients with Duchenne muscular dystrophy (DMD) and eight patients with Becker muscular dystrophy (BMD) [1]. Only age and disease duration correlated with LGE, and most cMRI parameters differed between DMD and BMD [1]. The study is impressive, but some points should be discussed.
The first point is that cardiac involvement in DMD/BMD is not limited to systolic dysfunction due to dilated cardiomyopathy and myocardial fibrosis, but also includes supra- and ventricular arrhythmias [2], hypertrophic cardiomyopathy [3], left ventricular hypertrabeculation, also known as noncompaction (LVNC) [4], and cardiac disease due to pulmonary involvement. Detection of these other morphologic and functional cardiac abnormalities on cMRI or long-term ECG is critical, as ventricular arrhythmias, for example, can be complicated by sudden cardiac death [5]. Since LVNC is often complicated by cardiac embolism, heart failure and malignant ventricular arrhythmias, it is also important not to overlook LVNC in DMD/BMD patients.
Secondly, there is no mention of how many of the included patients had scoliosis [1]. Scoliosis is known to strongly influence systolic and pulmonary function, so it is important to assess this orthopedic complication in the analysis. In how many of the patients did scoliosis contribute to systolic and pulmonary dysfunction? How many of the included patients had undergone surgical stabilization of the spine and to what extent did this affect cardiac function?
Thirdly, how many of the patients had cardiac impairment secondary to pulmonary dysfunction was not investigated [6]. Pulmonary dysfunction in DMD/BMD can lead to right ventricular hypertrophy, right ventricular systolic dysfunction, tricuspid regurgitation and pulmonary hypertension.
The fourth issue is that there was no mention of how many of the included patients were on nocturnal or all-day oxygen and how many were on continuous positive airway pressure (CPAP) and how many were tracheostomized for intermittent CPAP ventilation. Knowing this number of patients is crucial as it can strongly influence cardiac function and thus cMRI results.
The fifth point is that cMRI was not repeated after a certain time to assess the progression of cardiac impairment. Since cardiomyopathy progresses in DMD/BMD, it would have been essential to study the cohort of interest not only cross-sectionally but also longitudinally. Tracking the progression of heart disease is essential to be able to intervene in case of malignant arrhythmias, severe heart failure or the development of LVNC.
The sixth point is that it is not clear how underage patients could give their consent to the examinations [1]. Do the authors mean that in patients under 18 years of age, their caregivers agreed or refused the examinations?
Since glucocorticoids may also have a positive effect on myocardial function [7], we should know how many patients were taking glucocorticoids regularly and whether patients taking glucocorticoids differed from those not taking glucocorticoids in terms of cardiac function on cMRI.
Overall, this interesting study has limitations that put the results and their interpretation into perspective. Addressing these limitations could strengthen the conclusions and support the message of the study. Cardiac disease in dystrophinopathies requires longitudinal studies with cMRI to avoid overlooking the progression of cardiomyopathy.

Declarations
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Author contribution: JF was responsible for the design and conception, discussed available data with coauthors, wrote the first draft, and gave final approval. CS and FS: Contributed to literature search, discussion, correction, and final approval.

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