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Left ventricular volumes and function is affected by the cardiac fibrosis in patients with Becker and Duchenne muscular dystrophies in CMR.

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Introduction
Duchene Muscular Dystrophy (DMD) and Becker Muscular Dystrophy (BMD) are chromosome X-linked dystrophinopathies affecting skeletal, cardiac and respiratory muscles. Cardiac dysfunction is among leading causes of morbidity and mortality in this group of patients. Despite Cardiovascular Magnetic Resonance (CMR) is considered a useful tool for evaluation of cardiac function and fibrosis, in DMD and BMD patients the data is still scarce.

Methods
Of 79 patients with genetically confirmed diagnosis, 41 (aged 12.0 ± 3.1 years, DMD 88%, n=36, BMD 12%, n=5) were qualified and successfully examined using CMR. Disqualification criteria was age < 6 years, autism or metal implants. CMR protocol included LV dimensions, stroke volume (LVSV), ejection fraction (LVEF) measurement in short axis, and late gadolinium enhancement (LGE; 10–15 minutes after contrast injection) to provide fibrosis assessment. The obtained values were indexed to BSA and normalized (z-score) according to reference data published by Kawel-Boehm. Data is presented as mean ± standard deviation or median (range) dependently on the distribution. Chi-square test, Pearson and Spearman correlations were employed.

Results
Left Ventricle End Diastolic Volume index (LVEDVi) was 63.6 ± 17.4 ml/m2 and was abnormal in 24% (n=10). Left Ventricle End Systolic Volume index (LVESVi) was 30.0 ± 9.0 ml/m2, abnormally high in 12% (n=50) and abnormally low in 2% (n=1). Left Ventricle Mass index (LVMi) was 54.0 ± 12.2 g/m2 and normal in 93% of patients (n=37). LGE was assessed in 39 patients and was positive in 38% (n=15), most often in mid-anterolateral (38%, n=15), basal-anterolateral (36%, n=14), basal-inferolateral (31%, n=12), mid-inferolateral (26%, n=10) and apical-lateral segments (18%, n=7). LVSVi was 37.0 ± 10.8 ml/m2, abnormally low in 39% of cases (n=16), and LVEF was 58% ± 6.4%, low in 44%, n=18. Older patients had significantly lower LVEDVi-z (r=-0.41, p=0.008) and LVSVi-z (r=-0.50, p<0.001 respectively). LGE is significantly more prevalent in older patients (p<0.001). Patients with positive LGE had significantly lower LVSVi-z (p=0.022) and LVEF (p<0.001).

Conclusions
Fibrosis advances with age and DMD/BMD progression, causing worsening of cardiac function by limiting LVEDV and LVSV. The effect of pharmacotherapy is subject of a separate study.