

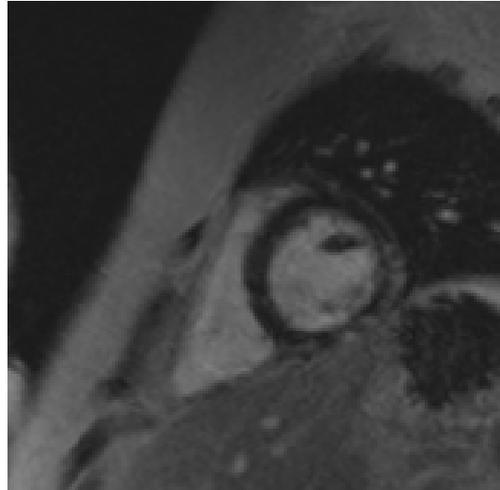
Cardiac involvement in Duchenne Muscular Dystrophy (DMD) – a detailed cardiac magnetic resonance imaging study in 39 patients

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Background: In the second decade of life about 70% of DMD patients develop initially asymptomatic cardiomyopathy with impairment of left ventricular function, which is one of the major reasons for subsequent morbidity and mortality in DMD. The value of cardiac resonance imaging (CMR) regarding sensitivity and significance in the diagnosis of cardiac involvement is to be evaluated.

Methods: CMR including late gadolinium enhancement (LGE) sequences as well as standard cardiological diagnostics and Tissue Doppler Imaging were performed in 39 male DMD patients (mean age 13 years, range 6-20 years). We here focus on MRI analysis.

Results: 16 patients (42%) had a limited left ventricular ejection fraction. Left ventricular mass was reduced without dilation; diminished end diastolic volume was present in 14 patients (37%), only one patient had a dilated left ventricle. CMR showed regional wall motion abnormalities in 47% of the patients. LGE was detectable in 89%, preferentially in the inferolateral and anterolateral segments of the left ventricle. The average mass of LGE was 7 g/m² (range 0-29 g/m²) with a mean fraction of 14% of left ventricular mass (0-51%). The youngest patient with evidence of LGE was five years old. LGE correlated with the wall motion abnormalities in CMR and Tissue Doppler Imaging.



Conclusions: In our group of 39 Duchenne patients, cardiomyopathy manifested with a left ventricular muscular atrophy without dilation. Localised LGE marks the extent of inflammation and fibrotic tissue transformation. Regional impairment of wall motion starts at an early stage. Further and ongoing investigations will show, if regional myocardial impairment and LGE in CMR analysis are reliable indicators for early medical treatment.