Hybrid CMR- and FDG-PET-imaging gives new insights into the relationship of myocardial metabolic activity and fibrosis in patients with Becker muscular dystrophy

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Background: Cardiac involvement in patients with Becker muscular dystrophy (BMD) is an important predictor of mortality. The cardiac phenotype of BMD patients is characterized by slowly progressive myocardial fibrosis that starts in the left ventricular (LV) free wall segments and extends into the septal wall during the disease course.

Purpose: Since the reason for this characteristic cardiac phenotype is unknown and comprehensive approaches using e.g. hybrid imaging combining cardiovascular magnetic resonance (CMR) with 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET) are limited, the present study addressed this issue by a comprehensive non-invasive imaging approach.

Methods: Hybrid CMR- and FDG-PET-imaging was performed in N=14 patients with BMD on a whole-body Biograph mMR system. The CMR protocol comprised cine- and late-gadolinium-enhancement (LGE)-imaging. Metabolism was assessed with FDG-PET after oral glucose loading to effect myocardial carbohydrate uptake. PET was acquired for 65 minutes starting with tracer injection. Uptake values from 60 to 65 minutes p.i. were divided by the area under the blood activity curve and reported as percentages relative to the segment with maximal myocardial FDG uptake.

Results: In the total study group, mean left ventricular (LV) ejection fraction (EF) was 55±5% and there were 6/14 patients with a reduced LV-EF $<$55%. A characteristic pattern of LGE in LV lateral wall was observed in 13/14 patients whereas an additional septal LGE pattern was documented in 6/14 patients only. Segmental FDG uptake was 88±6% in the LV lateral wall vs. 77±10% in the septal wall ($p<0.001$). There was a rather inverse relationship between segmental FDG activity compared to segmental LGE extent ($r=-0.33$, $p=0.089$). There were N=6 patients (= $\triangle$FDG-high) with a segmental difference in FDG uptake of $>$15% in the LV lateral wall compared to the septal wall (lateral FDG = 91±3% vs. septal FDG = 69±8%; $p<0.001$) while the remaining N=8 patients (= $\triangle$FDG-low) showed a segmental difference in FDG uptake of $\leq$15% (lateral FDG = 85±7% vs. septal FDG = 83±5%; $p=0.37$). Patients in the $\triangle$FDG-high group showed a similar LV lateral wall vs. septal wall LGE extent of 12±6% vs. 7±15%, respectively, while those patients in the $\triangle$FDG-low group demonstrated a large difference in LV lateral wall vs. septal wall LGE extent of 33±24% vs. 4±6%, respectively.

Conclusions: Segmental FDG uptake – reflecting myocardial metabolic activity – is in principal higher in the LV free wall of BMD patients – possibly due to a higher segmental work load. However, segmental metabolic activity seems to be dependent on and limited by the respective segmental extent of myocardial fibrosis as depicted by LGE-imaging.