



POSTER PRESENTATION

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Cardiac involvement of *myotonic dystrophy type II* in patients with preserved ejection fraction - Detection by CMR

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Background

Myotonic dystrophy type II (MD2) is a genetic multisystemic disorder characterized by skeletal muscle (SM) symptoms, metabolic changes as well as arrhythmias¹. Histopathologic changes of the SM may include fibrosis and fatty degeneration². The aim of this study is to evaluate myocardial structure in preserved ejection fraction (EF).

Methods

We prospectively enrolled 32 subjects with a genetically confirmed diagnosis of MD2. Exclusion criteria were known cardiac diseases and contraindication for CMR. We assessed left-ventricular (LV) volumes, mass and function applying state of the art cine imaging using a 1.5 T Scanner. Late enhancement imaging (LGE; slice thickness (sth) 7 mm) was performed to detect myocardial fibrosis 10 minutes after injection of gadoteridol (0.2 mmol/kgbw). We applied T1 Mapping based on MOLLI (TI native 211 ms, TI post-contrast 281 ms, TE 1.08 ms, sth 6 mm) before and 15 minutes after contrast application and assessed resultant extracellular volume fraction (ECV). Fat-water-separated imaging³ (GRE, TR 944.80 ms, TE 1.53-8.22 ms, sth 6 mm) was performed to identify myocardial fat deposits. Furthermore, we used ¹H magnetic resonance spectroscopy (MRS) (TR 1600 ms, TE 35 ms, septal voxel 20 x 15 x 6 mm³) to quantify myocardial Triglycerides (MTG). Data were analyzed using cvi⁴² and standard line-fitting procedure.

Results

26 data sets were totally completed (age 53.8 ± 11 y, LVEF 65 ± 0.6 %, 19 women). None of the patients had wall motion abnormalities. LGE was detectable in 6 of 28 subjects (LGE+; 3 women); the location was mostly subepicardial inferolateral basal (Figure 1). In case of LGE+, the T1-values of the enhanced regions were significantly different to remote myocardium in both the native (p = 0.03) and the post-contrast maps (p = 0.03). ECV and T1 values of the remote myocardium were not different between LGE+ and LGE negative (LGE-) groups besides to the inferolateral located ECV (25.6 % vs. 34.3 %, p = 0.03) (Figure 2). Fat deposits were noticeable in 6 of 30 subjects (all women, one with LGE+) in the apical portion of the interventricular septum. The content of MTG in LGE+ and LGE- subjects was not significant different (0.34 % vs. 0.74 %, p = 0.47).

Conclusions

Despite preserved LVEF we could detect myocardial injury in patients with myotonic dystrophy type II. Already native T1-values were increased in case of subepicardial fibrosis compared to remote myocardium. To conclude, this is the first study constituted that CMR is feasible to detect sub-clinical myocardial manifestations in MD2.

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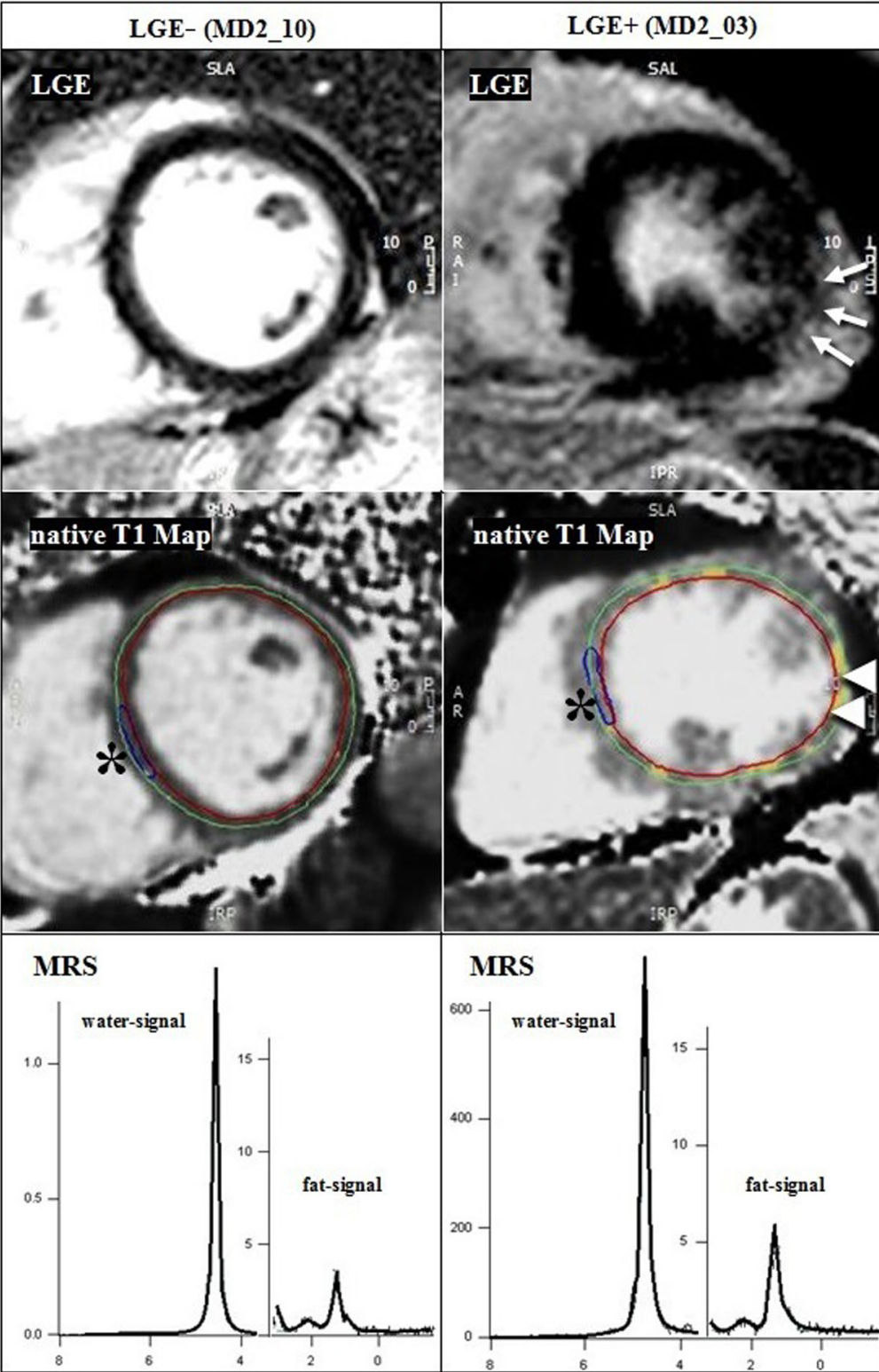


Figure 1 Image findings in LGE negative (LGE-, on the left) and positive (LGE+, on the right) patients. On top the LGE images, followed by native T1 Maps and MRS spectra. Arrows: LGE+; colored voxels (arrowheads): T1 values + 2 standard deviations to the reference myocardium (stars)

		LGE- (n = 22)	LGE+ (n = 6)	p-value (Mann-Whitney)	correlation to LGE+ (Spearman)
sex		18 women (82%)	3 women (50%)	0.12	-0.30
age (years)		52.3 ± 10	62.8 ± 11	0.15	0.28
Body-Mass-Index (kg/m ²)		23.6 ± 3	25.5 ± 2	0.28	0.21
LV Ejection Fraction (%)		0.66 ± 0.07	0.62 ± 0.05	0.14	-0.28
LV-EDV-Index (ml/cm)		0.8 ± 0.1	0.8 ± 0.1	0.23	0.23
LV-Mass-Index (g/cm)		0.6 ± 0.1	0.7 ± 0.2	0.10	0.33
MTG (%)		0.34 ± 0.20 (0.03 – 0.78)	0.74 ± 0.96 (0.01 – 2.36)	0.47	0.15
apical fat in fat/water separated imaging		5 (all women)	1 (woman)	0.72	-0.07
T1 value (ms) native Maps	septal	1024 ± 28 (980 – 1085)	1042 ± 49 (994 – 1119)	0.82	0.04
	inferolateral	1027 ± 41 (965 – 1111)	1087 ± 93 (1012 – 1249)	0.05	0.37
T1 values (ms) post-contrast Maps	septal	469 ± 38 (385 – 535)	471 ± 33 (430 – 517)	0.86	0.03
	inferolateral	463 ± 35 (396 – 511)	426 ± 44 (371 – 488)	0.06	-0.37
ECV (%)	septal	24.8 ± 3	27.7 ± 4	0.15	0.30
	inferolateral	25.6 ± 3	34.3 ± 8	0.003	0.62*

Figure 2 Comparison of LGE negative (LGE-) and LGE positive (LGE+) subjects. The inferolateral extracellular volume fraction (ECV) was significantly different in these two groups. MTG: Content of myocardial triglycerides measured by magnetic resonance spectroscopy. * The correlation to LGE+ was significant (p < 0.01)

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