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Becker and Duchenne Muscular Dystrophy (BMD, DMD) are associated with myocardial fibrosis and abnormal cardiac energetics even in the presence of normal left ventricular ejection fraction

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Introduction

BMD and DMD are X-linked abnormalities of dystrophin associated with a high rate of cardiomyopathy and eventual cardiac death. Previous magnetic resonance spectroscopy (MRS) studies have shown impaired energetics that did not correlate with echocardiographic abnormalities. Early detection of myocardial disease is clinically important in commencing heart failure therapy.

Purpose

Therefore this prospective study was performed to assess cardiac function, fibrosis and energy metabolism in patients with BMD and DMD with normal left ventricular function.

Methods

Patients with BMD or DMD (n = 10) (age 39 ± 12 yrs) and normal left ventricular ejection fraction (LVEF 65.5 ± 4.3%; mean ± one standard deviation), and 10 matched healthy volunteers (age 40 ± 16 yrs) (LVEF 69.3 ± 5.3%) were scanned using a Siemens Tim Trio 3 T (Erlangen, Germany). 31P MRS was used to measure the cardiac phosphocreatine to adenosine triphosphate ratio (PCr/ATP) in the mid ventricular septum[1]. Anatomical and functional imaging was performed in all subjects and late gadolinium enhancement (LGE) imaging was performed in the patient group only.

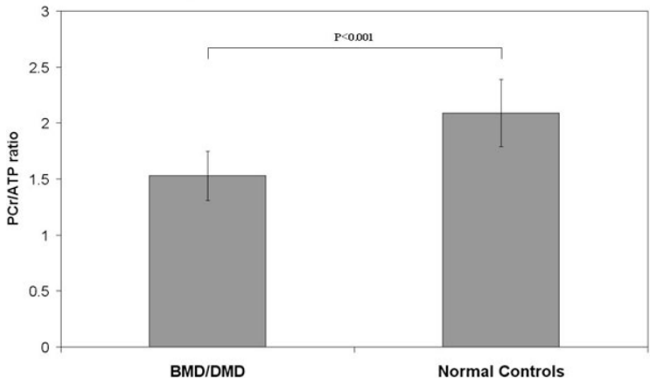

Figure 1
PCr/ATP ratio in BMD/DMD vs. Normal Controls (Error bars 1 stadard deviation).

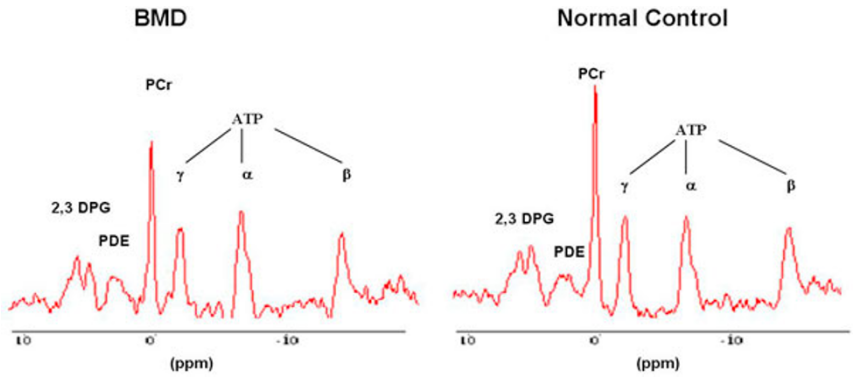

Figure 2
Typical 31P MR Acquisition in one patient with BMD and one normal health control showing the PCr, 2,3 disposoglyverate (DPG), phosphodiesters (PDE) and three ATP peaks (α, β, γ). The PCr/ATP ratio is reduced in the BMD patient.

Results

The cardiac PCr/ATP ratio in the BMD/DMD patients was significantly lower than the age matched controls (BMD/DMD 1.53 ± 0.13 vs. normal controls 2.09 ± 0.13; $p < 0.001$, Figures 1 and 2). Myocardial fibrosis as detected by LGE was detected in all BMD/DMD patients, with preponderance for the basal inferolateral wall only (4/10), lateral wall only (3/10), and basal inferolateral and lateral walls (3/10). LGE was quantified using QMASS 7.0 (Medis, Inc.) software with a threshold of 4 standard deviations above unaffected myocardium (LGE/total mass 16.4 ± 8.9%).

Conclusion

BMD and DMD are characterised by myocardial fibrosis and abnormal cardiac energetics even in the absence of left ventricular systolic dysfunction. These findings suggest incipient cardiomyopathy is more prevalent in this patient population than previously thought.

References

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