Combined IDEAL and Diffusion imaging to characterize Limb-girdle muscular dystrophy

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Introduction: Limb-girdle muscular dystrophies (LGMD) are a group of autosomal dominantly or recessively inherited muscular dystrophies that also present with primary proximal (limb-girdle) muscle weakness. This type of dystrophy involves the shoulder and pelvic girdles, distinct phenotypic or clinical characteristics are recognized. LGM dystrophies affect posterior thigh muscle compartment, predominantly gracilis and sartorius muscles. In the thigh, muscles at the back are affected, with a tendency to preserve the tibialis anterior and gastrocnemius. With standard histological techniques, biopsy specimens demonstrate variation in fiber size, increased numbers of central nuclei, and endomysial fibrosis, changes that are common to all dystrophic muscle [1]. The aim of this study was to compare quantitative MRI measurements from IDEAL-based imaging and DW imaging in the thigh muscles of adults with LGMDs and healthy volunteers.

Figure 1. Experimental setup showing coil

Material and Methods: Six women (three patients and three healthy volunteers) were examined as follows: all subjects were scanned while in the supine position with thighs relaxed and parallel to the magnet magnetic field direction. Imaging experiments were conducted on a 1.5T GE scanner (General Electric Health Care, Milwaukee, USA), using a combination of two eight-channel coil array as shown in Figure 1. T1 IDEAL 2D images were acquired with the following parameters TR/TE=700/62.5, slice number=18, slice thickness=8mm, slice spacing=8mm, FOV = 42x42cm^2, A/P, matrix=320x192 and NEX=2, in phase and out-phase. High-resolution T1 images were acquired using Fast Spin-Echo with the following parameters: TR/TE=600/23ms, slice thickness 8mm, matrix=256x256 and NEX=2. Diffusion Images were acquired with the following parameters TR/TE=5500/67.8, slice number=18, slice thickness=8mm, slice spacing=8mm, FOV = 42x42cm^2, A/P, matrix=128x128 and NEX=7. *Data Analysis:* Fat percentage values and ADC values were computed according to eqs (1) and (2), respectively: [5]:

$$Fat\% = \frac{S_{inphase} - S_{outphase}}{2S_{inphase}} \quad (1) \qquad \ln(S/S_0) = -\sum_{i=1}^{3}\sum_{j=1}^{3} b_{ij} D_{ij} \quad (2)$$

Where b is the effective difussion weighting and D is the diffusion tensor. Osirix [4] together with eqs (1) and (2) were used to digitally process all the images for the regions: around maximus gluteus, lateral vastus (VL), vastus intermedius (VI), vastus medial (VM), sartorius (S), gracilis (G), adductor longus (AL), adductor (AM), semi membranous (SM), semi-tendinous (ST), biceps femoris to form volumes of interest (VOI). Mean signal intensities were then measured for each VOI from the fat and water images to provide a percentage fat value [1,6].

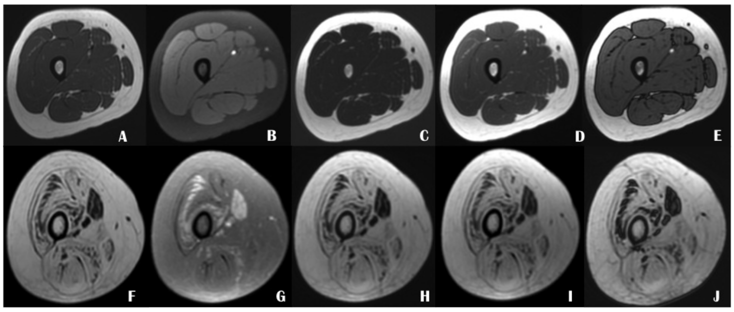

Figura 2. Comparison of water/fat images of healthy (top row) control and patient with LGMDs (bottom row): a) FSE-T1 weighted, b) and c) IDEAL fat/water and, d) and e) in/out phase IDEAL. Note the important fatty infiltration muscle planes.

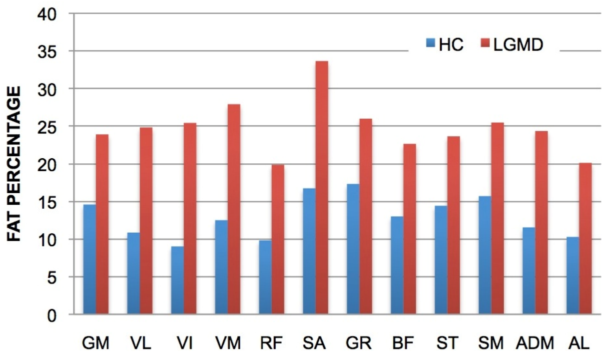

Figure 3. LGMDs (red) and HC group (blue) average percentage fat.

Results: Figures 2-3 show the mean values for fat percentage and ADC for LGMDs and HC. *Fat percentage:* A consistently higher average fat content was observed for the LGMDs compared to de HC group. *ADC:* The mean ADC values for the LGMDs was consistently lower than the HC group values for all muscles evaluated, except for the gracilis and sartorius muscles.

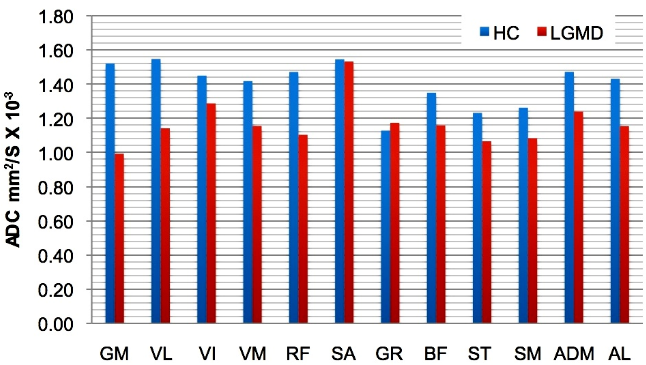

Figure 4. LGMDs (red) and HC group (blue) average ADC

Discussion and conclusion: The increased intramuscular fat percentage in subjects with LGMDs as quantified by the in phase-out phase-IDEAL method is consistent with the qualitative findings obtained with the Mercuri classification. ADC values were lower in patients with LGMDs, indicating restricted diffusion and infiltration of connective tissue. Our results demonstrated that the use of noninvasive MRI techniques may provide the means to characterize the muscle through quantitative methods to determine the percentage of fat and ADC values.

References: [1] Wattjes et al. Eur Radiol (20), 2010; [2] Mercuri et al. Curr Opin Neurol (18): 2005; [3] Quan D. Rheum Dis Clin NA (37), 2011; [4] Rosset et al. J Digit Imaging (17), 2004; [5] Hussain et al. Radiology (237), 2005; [6] Kim et al. Radiology (255): 2010.