# The role of muscle MRI in the diagnosis of Amyotrophic Lateral Sclerosis (ALS)

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## *Background and Purpose*

Muscle has been always considered a secondary target in ALS, due to the degeneration of lower motor neuron. Some authors [1-2] demonstrated in transgenic mice that the muscular damage can be primary. Muscle MRI is commonly used for non-invasive diagnosis of inherited dystrophies and acquired myopathies, where T1-weighted images can show chronic modifications such as atrophy and fatty degeneration. Few and heterogeneous data are published in literature about the role of muscle MRI in ALS patients.

## *Materials and Methods*

We enrolled 10 patients (n=8 spinal-onset, n=2 bulbar-onset), newly diagnosed as ALS according to El Escorial criteria (mean ALSFRSr score, 40,3±4.64), and 9 age-matched healthy controls (mean age patients, 68,7±8,57 years, range 57,53-86,16; mean age controls, 68,7±5,44 years, range 58,36-74,59). All subjects underwent muscle MRI (T1-weighted images) of hands, paraspinal muscles, legs (68 muscles globally), and EMG exam with quantitative motor unit action potential (MUAP) analysis for each muscle (first dorsal interosseus, paravertebral thoracic, anterior tibial). We utilized the Mercuri score for the evaluation of MRI images, that were normal (grade=0) or altered (grade>0). An arbitrary grading was used for the analysis of MUAP duration.

## *Results*

Muscle involvement was more frequent in ALS patients than controls (respectively 69.02% and 43.3%; p<0,0001, fig. 1a), and in spinal-onset patients than bulbar-onset patients (73.21% vs 54.41%; p<0,0001, fig. 2a). Furthermore, muscle damage was more frequent in bulbar-onset patients than controls (54.41% vs 43.3%; p=0,0197, fig. 2b). The presence of atrophy was also significantly different between patients and controls (22.46% vs 0,72%; p<0,0001, fig. 1b). No difference was detected stratifying patients for the I motor neuron damage (p=0,2698, fig. 2c). We correlated MRI score and EMG grading in the sum of selected muscles, finding a positive and significant correlation (r=0,7133, p=0,0229, fig. 3a). A negative correlation between MRI score and ALSFRSr score for right upper limb (r=-0,7217, p=0,0110, fig. 3b) was also present.

## *Conclusions*

Muscle MRI, in particular T1-weighted images (fig. 4-5), can distinguish ALS patients from controls, and spinal-onset from bulbar-onset patients. Therefore, muscle MRI provides a feasible diagnostic tool for ALS, that can integrate clinical and electrophysiological data. Moreover, we confirm that the involvement of the I motor neuron is independent from the muscular damage. On the basis of the multifactorial pathogenesis of ALS, we can speculate that the mechanism of involvement is different between the I and II motor neuron.

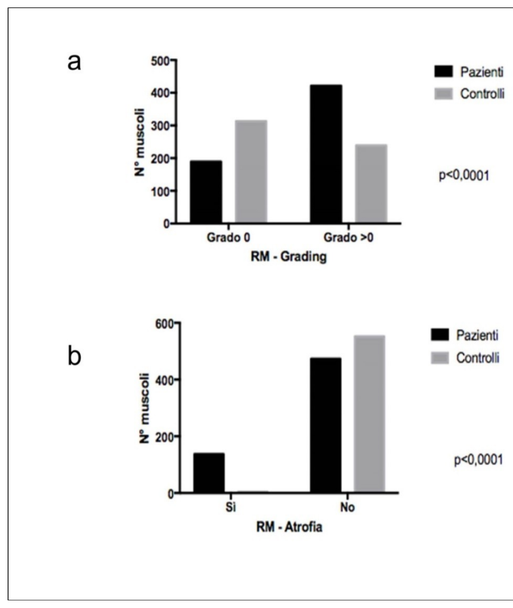


Fig. 1 – Fatty substitution (a) and atrophy (b) in patients and controls.

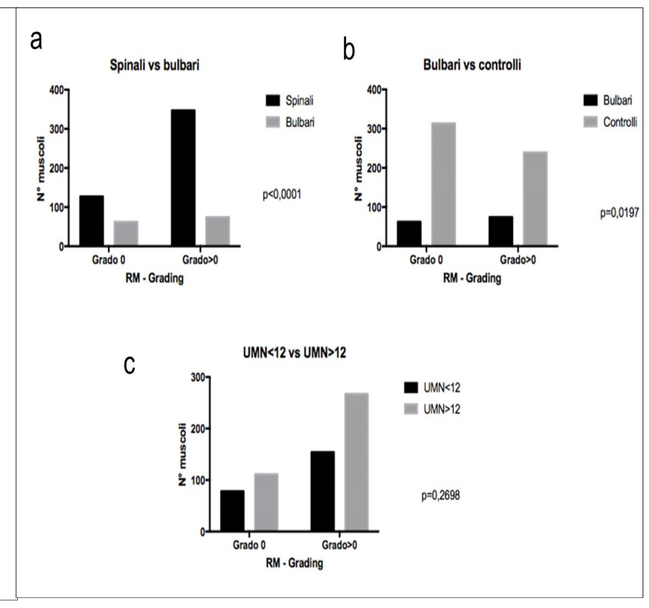


Fig. 2 – Fatty substitution in spinal and bulbar ALS (a), bulbar ALS and healthy controls (b), patients with UMN score <12 o >12.

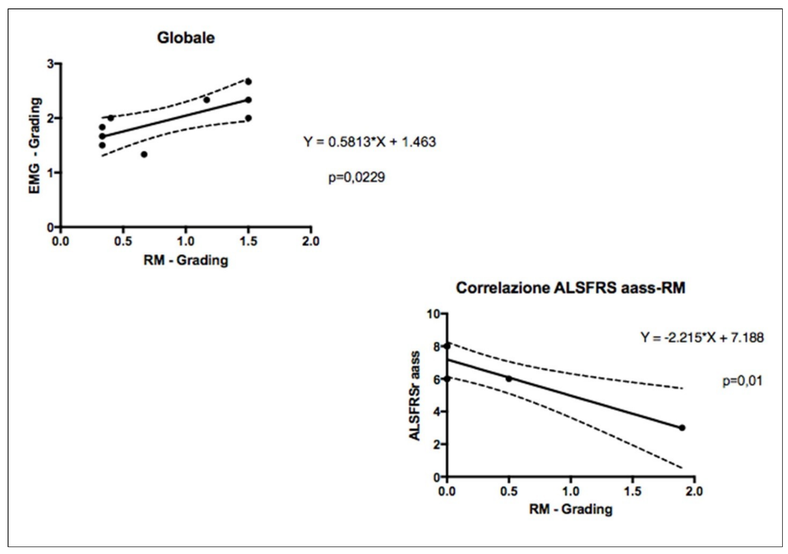


Fig. 3 – MRI-EMG correlation and ALSFRSr (upper limbs)-MRI correlation.

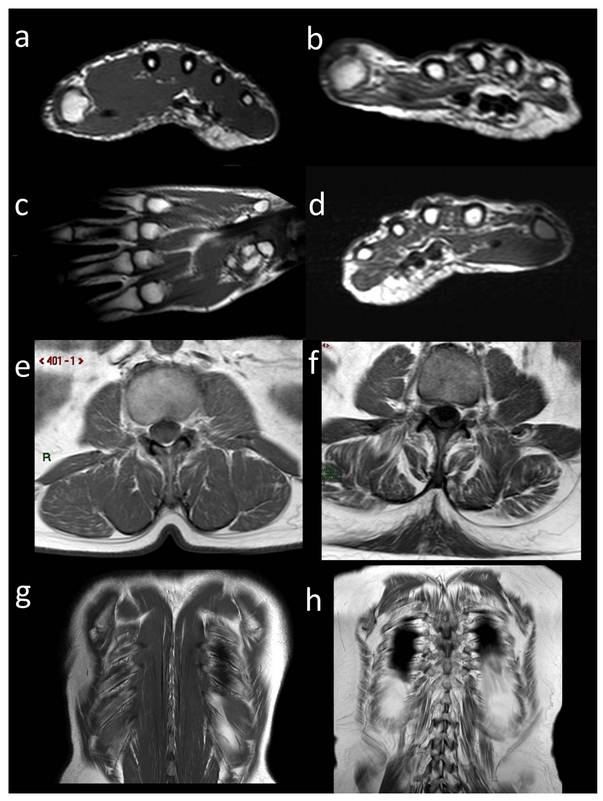


Fig. 4 – Muscle MRI, controls (a, c, e, g) and patients (b, d, f, h): left hand axial T1 (a, b, d), right hand coronal T1 (c), paraspinal axial T1 (e, f) and paraspinal coronal T1 (g, h).

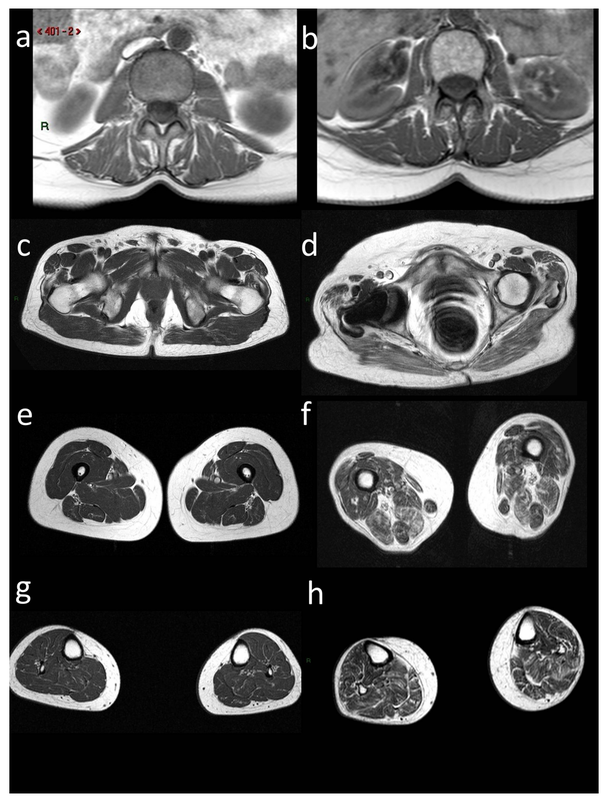


Fig. 5 – Muscle MRI, controls (a, c, e, g) and patients (b, d, f, h): iliopsoas axial T1 (a, b), gluteus maximus axial T1 (c, d), thigh axial T1 (e, f), calf axial T1 (g, h).

## *References*

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