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Motor neuron disease: a multimodal longitudinal follow up

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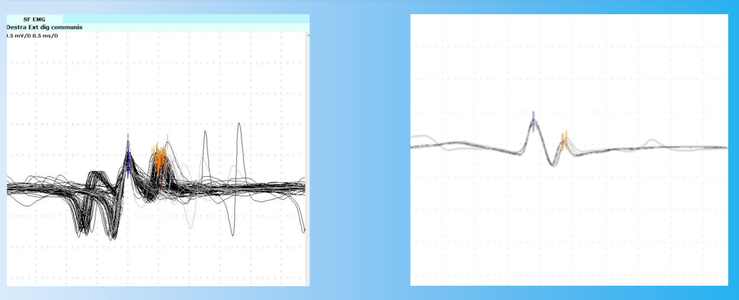
Objective: The aim of our study is to evaluate the reliability and usefulness of different neurophysiological tests in the assessment of clinical evolution in motor neuron disease (MND), through a six months longitudinal follow up.

Materials and methods: we have evaluated 14 subjects with MND at baseline, after three and six months; each patient undergo clinical evaluation with Amiotrofic lateral Sclerosis Functional rating scale (ALSFRS) Neuromuscular Symptoms Score (NSS) and Extended Muscular Research Council Status Score (EMRCSS). We also performed a single fiber electromyography with concentric needle on extensor digitorum communis (EDB), MUNIX and MUSIX on abduttor pollicis brevis (ABP), quantitative motor unit potential evaluation on biceps brachii (BP) and vastum medialis (VM). We evaluated the differences from the baseline with *Student's T-test*.

Figure 1. Single fiber EMG potentials
A abnormal jitter **B** normal Jitter



A B

Results: we enrolled 14 patients, 10 with progressive muscular atrophy (PMA) and 4 Amyotrophic lateral sclerosis (ALS) clinically definite for El Escorial criteria. All patients reached the follow up at three months and 9 patients undergo the sixth month visit. For the nine patients who completed the follow up, the mean ALSFR score was 32,2±3,2 at baseline, 28,7±4,0 at three months and 26,1±5,2 at sixth month. The mean NSS at baseline was 44,3±4,6 , at three months 39,5±9,9 and at six months 32,3±10,3. MUNIX value on ABP was 43,5±39,2 at baseline, 44,6±41,4 after three months and 38,7±47,4 at sixth month. The mean of all mean Jitter values was 54,3±20,7 μs at baseline, 51,4±15,6 μs and 65,6±18,9 μs after three and six months respectively. The mean of patological jitter percentage was 59,4±28,2 at baseline, 66,1±29,3% at third month and 76,6±27,1% at the last follow up.

Figure 2. Results for 9 patients with 6 months follow up

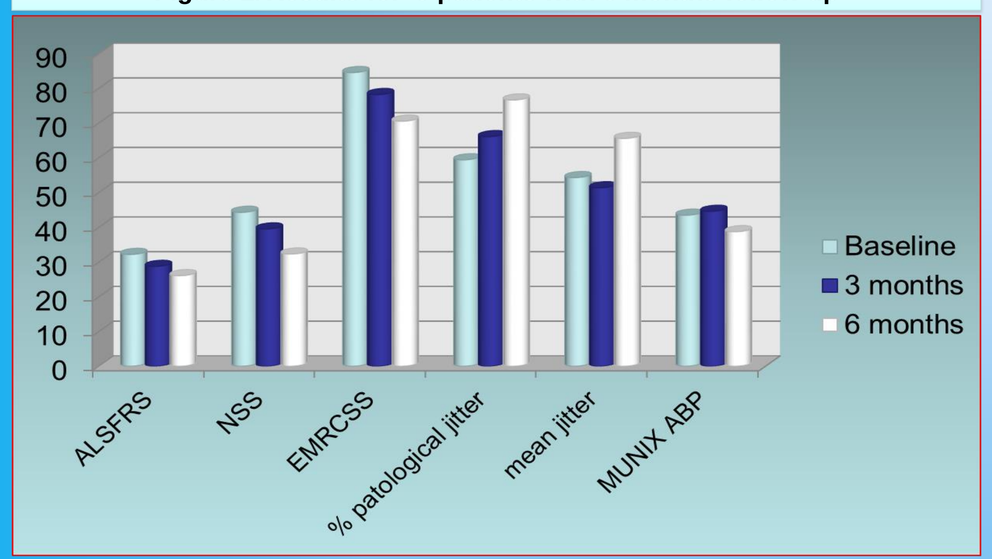


Table 1. Characteristics of 14 patients.

Age at disease onset (mean±sd)	60,9 ±9,2
Sex(M/F)	9/5
Disease duration at baseline (months)	18,2±13,2
Kind of onset	1 D-UL; 1 P-UL; 6 D-LL; 3 P-LL; 2 A/B

D-UL= distal upper lims; P- UL= proximal upper lims; D-LL= distal lower limbs;
P-LL= proximal lower limbs; A/B= axial/bulbar

Table 2. Results at baseline and after three months for all 14 patients

MEAN±SD	Basal	3 months follow up	P value
ALSFRS	32,21±2,99	29,5±3,61	*0,006*
NSS	45,78±6,42	41,71±9,81	*0,037*
EMRCSS	84,78±12,83	80,25±14,9	*0,019*
% patological jitter	63,57±29,17	73,92±26,39	*0,035*
Mean jitter (μs)	58,98±22,00	62,99±23,09	*0,354*

Discussion: we evaluated different neurophysiological approaches in order to find a possible method to quantify the progression of MND. All 14 patients had a statistically significant worsening of all the three clinical scales applied at third month, like the 9 patients who completed the follow up at sixth month. For all the other neurophysiological test applied there is a trend of worsening, but not significant, except for the jitter % at third (p=0,034) and sixth month (p= 0,039). Single fiber emg is a helpful diagnostic tool in MND; now we emphasized a possible role in the assessment of clinical course evolution. Anyway we find a trend in all the other tests, suggesting that a larger study could be helpful to investigate their usefulness.

• **Lower motor neuron involvement in ALS assessed by motor unit number index (MUNIX): Long-term changes and reproducibility** *Davood Fathi , Bahram Mohammadi, Reinhard Dengler, Sebastian Böselt, Susanne Petri , Katja Kollewe* , Clinical Neurophysiology 127 (2016) 1984–1988

• **Quantitating Changes in Jitter and Spike Number Using Concentric Needle Electrodes in Amyotrophic Lateral Sclerosis Patients** *Ming-Sheng Liu, Jing-Wen Niu, Yi Li, Yu-Zhou Guan, and Li-Ying Cui*, Chin Med J (Engl). 2016 May 5; 129(9): 1036–1040.

• **Single fiber electromyography in 78 patients with amyotrophic lateral sclerosis.** *Cui LY[1], Liu MS, Tang XF.* Chin Med J (Engl). 2004 Dec;117(12):1830-3

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