Awaji criteria for ALS diagnosis: limits and advantages compared to previous criteria.

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Introduction
The diagnosis of Amyotrophic Lateral Sclerosis (ALS) requires the combination of upper and lower motor neuronal signs. There is no established diagnostic biomarker of ALS in the early stage.

In the last two decades three different sets of diagnostic criteria for ALS have been developed; the first (El Escorial criteria, EEC, of 1994) were based only on clinical ground, whereas their revised version of 1998 (Airlie House Criteria, AHC) and the latest set of criteria (Awaji criteria, AC, 2006) included also EMG findings as indicator of lower motor neuronal (LMN) involvement.

The sensitivity of the AC criteria is however debated; a recent meta-analysis found that despite the overall superiority of AC in detecting EMG signs of LMN involvement, they are not always more sensitive than AHC in increasing the diagnostic certainty level.

Objective
To compare the sensitivity of the three set of criteria in a cohort of ALS patients at the time of the diagnosis.

Materials and Methods
We enrolled in the study 68 consecutive patients (M 39, F 29) with a suspected diagnosis of ALS attending our tertiary ALS unit during the 2013-2014 year.

All patients were clinically evaluated and underwent EMG.

A careful diagnostic workup excluded other potential diagnoses.

Results
According to EEC, 71% (n= 48/68) of patients were classifiable as “definite + probable” ALS, 13% (n=9) as “possible” and 16% (n=11) as “suspect” ALS.

The percentage of definite + probable ALS did not significantly change using AHC (75%; n=51) and AC (76%; n=52).

AC were however more sensitive in finding EMG signs of LMN involvement; indeed, EMG signs of LMN damage were observed in 2 or more regions in 81% of patients using AC in comparison to 60% with AHC (p < 0.0001).

This difference was more pronounced in bulbar onset ALS; 41% of patients showed three affected regions according to AC, compared to the 18% with AHC (p = 0.02).

Discussions and Conclusions
Our results are in agreement with a recent meta-analysis and confirm that the new set of criteria do not significantly increase the level of diagnostic certainty of ALS.

AC are however more sensitive than AHC in detecting EMG signs of lower motor neuron damage (with regarding fasciculation potentials as evidence of acute denervation), especially in in ALS patients with bulbar onset.

Bibliography