



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

D'Errico E. ^{1,2}, Zoccolella S. ¹, Introna A. ¹, Distaso E. ¹, Scarafino A. ¹, de Leo G. ¹, Tempesta I. ¹, Cortese R. ¹, Tortelli R. ³, Capozzo R. ³, Simone I. L. ¹, Logroscino

¹ Department of Basic Medical Sciences, Neurosciences and Sense Organs - University Aldo Moro – Bari

- ² Neurological Department, General Regional Hospital "F. Miulli" Acquaviva delle Fonti (BA)
- ³ Department of Clinical Research in Neurology at Pia Fondazione "Card. G. Panico", Tricase, LE University Aldo Moro Bari

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 (EI 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 sign 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 <u>definite + probable ALS</u> 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.



Bibliografia

1. Jang JS, Bae JS. AWAJI criteria are not always superior to the previous criteria: A meta-analysis. Muscle Nerve. 2015 Jun;51(6):822-9. 2. Costa J, Swash M, de Carvalho M. Awaji criteria for the diagnosis of amyotrophic lateral sclerosis: a systematic review. Arch Neurol. 2012 Nov;69(11):1410-6.

3. Noto Y, Misawa S, Kanai K, Shibuya K, Isose S, Nasu S, Sekiguchi Y, Fujimaki Y, Nakagawa M, Kuwabara S. Awaji ALS criteria increase the

diagnostic sensitivity in patients with bulbar onset. Clin Neurophysiol. 2012 Feb;123(2):382-5.