Electrodiagnostic Criteria for diagnosis of ALS

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ALS is primarily a clinical diagnosis, though a definite diagnosis requires additional diagnostic testing to help exclude other causes for a patient's clinical examination. For example, typical findings of mixed upper and lower motor neuron (UMN, LMN) findings in a single limb do not meet established criteria for a definite diagnosis of ALS. Electrodiagnostic studies are, therefore, helpful, as they can extent clinical findings by suggesting LMN findings in muscles or other body regions that appear to be clinically unaffected. At the time of this review, the El Escorial criteria had been used to help establish a diagnosis of ALS based on combined clinical and electrophysiological data. However, there were some inherent limitations to these criteria, from an electrodiagnostic standpoint, that enhanced the risk for a delayed diagnosis of ALS. Specifically, limbs affected by ALS were required to show evidence for both ongoing denervation (fibrillations, or fibs, and/or positive sharp waves, or SWs) and chronic re-innervation changes (large, prolonged, unstable MUPs), though many muscles do not show fibs or SWs. This could lead to insufficient data, on the part of the clinician, and a delayed diagnosis of ALS. For all of the above, the International Federation of Clinical Neurophysiology met in Awaji Island Japan in 2006 to review the El Escorial criteria for a diagnosis of ALS, to determine if modifications were needed. They additionally reviewed other, emerging techniques that might be of clinical utility. The goal of this review was to summarize their findings and consensus criteria.

Overall Conclusions of the Consensus Group: There were three main conclusions from the conference regarding modified EMG criteria for ALS. These are summarized below.

1) The study group modified the El Escorial criteria for ALS, as in **Table 2**. They re-affirmed the need for full NCS and EMG studies for the purpose of assisting in an ALS diagnosis, and they re-affirmed that an abnormal EMG finding should be interpreted in the context of the clinical exam.

2) The study group suggested that abnormalities required for a diagnosis of ALS in a given limb may be obtained by either neurophysiologic OR clinical criteria. This re-enforced that an EMG abnormality in a single limb is of equal importance to a clinical finding in that same limb.

3) The consensus group proposed that evidence for ongoing denervation changes on EMG could be fulfilled by the presence of **fasciculation potentials (FPs),** and that FPs should have equal significance as both fibrillations (fibs) and positive sharp waves (SWs). This would obviate the need for fibs/SWs, alone, as being indicative of ongoing denervation, as these can be difficult to find (especially in bulbar musculature), even in someone with clinically evident ALS.

In addition to the above, the authors also reviewed relevant criteria and concepts related to needle EMG and NCS in ALS. These are further noted below.

<u>1) Benign vs. Neurogenic FPs</u>: A major modification to the El Escorial criteria was the addition of fasciculation potentials (FPs), if found on EMG, as an indicator of on-going neurogenic disease. The authors also further characterize FPs, as FPs can be a benign variant. Specifically, FPs associated with ALS are complex in morphology and are unstable (increased jitter with a high band pass filter). Benign FPs do not have these features and are also in the absence of associated motor unit potential (MUP) changes. Unstable FPs can also be seen in other neurogenic conditions as well (i.e., neuropathies), so they must be interpreted along with the remainder of the clinical exam and in relation to one's clinical suspicion for ALS.

<u>2) FP equivalence to Fibs and SWs</u>: FPs are often evident in distally in muscles, and are suggestive of damaged axons, as are fibs and SWs. All results as a consequence of enhanced Na+ and reduced K+ conductance in ALS patients, due to axonal damage within the peripheral nerve. The authors do highlight that FPs on EMG can also occur as a result of other neurological conditions, such as a peripheral neuropathy, so the finding of FPs should be interpreted in the setting of the clinical exam.

3) Unstable MUPs: Chronic reinnervation changes on EMG are suggested by the presence of abnormal MUP morphology, though the consensus committee also highlighted that unstable MUPs serve as indicators of chronic reinnervation changes. Unstable MUPs (**Fig. 1**) are seen when using a trigger delay line and a 500Hz low band pass filter, and appear as single units dropping in and out of the recording. Unstable MUPs were not considered as a diagnostic requirement, but were still highlighted as being useful indicators of chronic changes.

4) Choice of Muscle for EMG: To search for the involvement of additional body regions in ALS, good muscles to sample include bulbar, facial, tongue, masticatory, thoracic paraspinals, and rectus abdominus muscles.

5) Nerve Conduction Studies in ALS: The consensus criteria group re-affirmed the importance of NCS in a diagnosis of ALS (**Table 3**). On NCS, findings in ALS include normal SNAP amplitude and conduction velocities (CVs, if no other super-imposed peripheral neuropathy), motor CVs >75% of the lower limit of normal, distal CMAP latencies <150% of normal, and the absence of temporal dispersion and conduction blocks (drop in CMAP by >50% on proximal vs. distal stimulation). The presence of pathological temporal dispersion is often due to a demyelinating condition.

6) Electrophysiological Evaluation of UMN Disease: EMG readily defines LMN but not UMN pathology. Identification of UMN disease via electrodiagnostic testing is often difficult, but important for confirmation of clinical findings of UMN involvement. Continued research is underway investigating Transcranial Magnetic Stimulation techniques (i.e., looking for increased central motor conduction time or abnormal results on triple stimulation techniques) and or F-wave study techniques (assessing the F/M ratio) for this purpose. In addition, MRI-based imaging studies may be able to identify UMN tract abnormalities in ALS.

Conclusions: As noted above, conclusions from this consensus meeting reviewed and revised electrodiagnostic + clinical criteria for a diagnosis of ALS, leading to the updated Awaji-Shima Consensus Recommendations and/or Revised El Escorial Criteria. This helped to further clarify the diagnostic criteria that are now often used today in the diagnosis of ALS via electrodiagnostic data.

Summary created by Elaine Sinclair, D.O.