Published in final edited form as:

Mov Disord. 2018 July ; 33(7): 1190. doi:10.1002/mds.27430.

Reappraisal of Cortical Myoclonus. Electrophysiology is the gold standard

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Having a long interest in cortical myoclonus,¹ I was attracted to the recent article from Latorre and colleagues reappraising the entity.² The authors performed physiological analysis of “44 patients diagnosed according to clinical criteria (i.e. distal, stimulus sensitive, negative myoclonus) and etiology”. (I presume positive myoclonus was actually included as well as negative myoclonus.) The diagnoses were heterogeneous. As the authors note, electrophysiology is the “gold standard” and the patients are only suspected as having cortical myoclonus (although they do not reveal their suspicion until the end). They were able to “prove” cortical myoclonus in 47% of the cases, finding that many patients were too difficult to study and some did not have positive findings. They conclude that further improvements in clinical neurophysiological testing is needed. While I agree with the conclusion, readers might get some wrong impressions from the article.

The title is misleading. They are analyzing patients with a clinical syndrome of myoclonus, not patients with known cortical myoclonus. Indeed, some of the entities studied here have been evaluated previously and, it is already known that they do not have the classic findings of cortical myoclonus. For example, Thompson et al. studied 14 patients with corticobasal syndrome, found only one large somatosensory evoked potential and no jerk-locked backaverage.³ While cortical origin is suspected, it is not proven.

While C-reflexes can be seen in cortical myoclonus, it is well known not to be present in all cases, and, on the other hand, C-reflexes can be found in other forms of myoclonus as well (e.g., reticular myoclonus⁴). Hence, the presence of a C-reflex cannot be used as evidence for a cortical origin.

The article has value, but what it really does is appraise how often current standard clinical neurophysiological testing reveals a cortical origin in patients with the clinical syndrome of “distal myoclonus”. The answer is less than half the time, but the reason is a mixture of technical and biological. Clinical neurophysiological testing does provide the gold standard, and advances in methodology will hopefully help in allowing more patients to be properly diagnosed. However, considering all comers with myoclonus, as was done in another recent report, it is very clear that even with current techniques, physiology can aid in diagnosis and subtyping.⁵

Relevant conflicts of interest/financial disclosures: Nothing to report.

Full financial disclosures and author roles may be found in the online version of this article.

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