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Letter to the Editor

Specificity of Decremental, Repetitive Median Nerve Stimulation for Diagnosing Myasthenia Remains Unproven

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In a recent article, Lee *et al*., reported about a retrospective investigation of repetitive nerve stimulation (RNS) of the median nerve in 95 patients with myasthenia gravis (MG), of whom 27 patients were seronegative, and other neuromuscular conditions.(Lee, T.H, & Li, Y. 2019)Among the 95 MG patients undergoing RNS of the median nerve 62 (59%) showed a decremental response. It was concluded that an abnormal decrement in median nerve RNS is highly specific for diagnosing MG, and has a diagnostic sensitivity for MG that is comparable to the spinal accessory nerve but less than that of the facial nerve(Lee, T.H, & Li, Y. (2019). It was proposed that median nerve RNS should be considered as a routine diagnostic study for work-up of patients with putative MG.(Lee, T.H, & Li, Y. 2019) We have the following comments and concerns.

The main shortcoming of the study is that only 74.4% were seropositive. Among the seronegative patients MG was diagnosed upon RNS in 17 patients, upon single fiber EMG in 5, upon the edrophponium test in 3, and upon the response to anti-myasthenic medication in 3 patients. Symptoms and signs indicative of MG alone do not justify the diagnosis of MG. Given the low number of seropositive patients, the probability of false positive diagnoses (no myasthenia) is high. Thus, we should know how many of the included 95 MG patients had a decremental RNS in any of the stimulated nerves, how many had thymoma or thymus hyperplasia, how many an increased jitter, how many a positive edrophonium test, and how many a favourable response to cholinergics/immunosuppressants. We also should know how many of the included patients had generalised myasthenia and how many had ocular myasthenia. If patients with ocular myasthenia were included, it is unlikely that RNS of any peripheral nerve showed a decremental response. Since acetylcholine-receptor antibodies (AchR abds) may become positive during the disease course,(Yamada, Y.,*et al*.,2015) we should know if AchR were repeatedly determined and in how many of the initially aeronegative patients the test became positive over time.

We do not agree that an abnormal response to RNS of the median nerve is highly specific for MG. A decremental response in a single peripheral nerve does not justify the diagnosis MG. MG should be diagnosed upon a bundle of tests, including clinical exam, edrophonium test, thoracic CT, AchR-abs or MUSK-abs titres, single´-fiber electromyography, and response to cholinergic medication.(Mantegazza, R.,& Cavalcante, P. (2019) An abnormal response to low-frequency RNS does not necessarily imply MG. Abnormal decremental responses to RNS may be false positive since it has been also reported in other neuromuscular conditions.(Finsterer, J.,*et al*.,2002) With respect to RNS of the median nerve, figures provided in the result section are misleading. In the second paragraph the authors mention that 62 MG had a decremental response to RNS of the median nerve. However, in table 4 only 33 MG patients with a decremental response to RNS of the median nerve are listed. This discrepancy should be clarified.

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Furthermore, sensitivity of RNS may not only depend on the location of the stimulation, but also on the disease duration, titres of Ach-R-abs, concomitant drugs, co-pathologies in investigated nerves, concomitant diseases, compliance of the patient, and dosage of the anti-myasthenic medication.

A further shortcoming of the study is that the drugs the 95 patients were taking at the time of NCS were not reported. We should know how many of the included patients were taking cholinergic drugs or other compounds influencing the severity and course of MG.

We do not agree with the figure provided for the specificity of the RNS of the median nerve. The authors themselves mention that RNS has been only infrequently carried out in MG patients and to our knowledge there are no systematic studies investigating specificity/sensitivity of RNA of the median nerve in MG patients. We should know how many of those with normal RNS of the median nerve had MG and how many of those with abnormal RNS did not have MG.

A decremental response to RNS of the facial nerve in only 57% of the patients is low compared to other studies.(Lin, Z.Z., *et al*., 2019) The authors should explain the low sensitivity of RNS of the facial nerve in their patients.

In summary, the presented study has a number of shortcomings, such as different investigators, insufficiently reported concomitant comorbidities of peripheral nerves or other organs, and insufficient data about time to diagnoses, disease duration, and current medication taken at the investigation. Before proposing RNS of the distal median nerve as a specific diagnostic tool for MG, prospective studies on large, homogenous cohorts of patients with seropositive MG are warranted.

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