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Prognostic value of decremental responses to repetitive nerve stimulation in ALS patients

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Reply from the Authors:

We thank Dr. Kaires for her interesting comment. Decremental responses recorded in patients with ALS after repetitive nerve stimulation probably are not related exclusively to a neuromuscular junction problem restricted to pre- and postsynaptic levels. Therefore, we preferred to speak about impairment of the neuromuscular transmission, which implies prejunctional, perijunctional, and postjunctional levels. A prejunctional level of failure is characteristic of immature motor axon branches with incomplete myelinization, particularly at early stages of reinnervation. This prejunctional mechanism, which does not exclude an axonal ion channel disturbance, may be an explanation for decremental motor responses after repetitive nerve stimulation in patients with ALS, as mentioned in our article. 1 Our methodology does not allow us to determine which prejunctional mechanism, immature myelinization, or axonal ion channel dysfunction is prominent in the impairment of the neuromuscular transmission observed in patients with ALS.

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