



Letter to the Editor

Somatosensory evoked potentials and Hirayama disease

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Quick Response Code:



Dear Sir,

Hirayama disease (HD) is a rare sporadic condition which is defined by a slowly progressive, asymmetric distal amyotrophy in the upper limbs. This condition usually presents in the juvenile years and is often clinically detected by muscle weakness and atrophy in the arms. Patients can also endorse symptoms of cold paresis which is the aggravation of muscle weakness on exposure to the cold. While the etiology of HD is currently unclear, many theories exist. One theory is that there is forward displacement of the posterior wall of the lower cervical dural canal in neck flexion causing compression and flattening of the spinal cord. Given this, somatosensory evoked potentials (SSEPs) have been discussed as a potential electrophysiologic measure. A recent study by Fustes *et al.* presented a series of eight cases with HD for whom the SSEP test did not turn out to be an electrophysiologic marker.^[1]

We report a series of six patients who were diagnosed with HD between 2004 and 2012. The average age of our cohort at diagnosis was 32.8, with all six being males. These patients all underwent ulnar SSEP studies which were normal, providing additional data that the SSEP test may not be an electrophysiologic marker for patients with HD.

Our studies were not done with patients in the flexed neck position. However, a study by Misra *et al.* found that there was no significant change in SSEPs and F-wave parameters in HD patients when their neck was flexed versus at baseline.^[2] Other small studies have shown that there may be some difference in SSEPs when compared to controls.^[3,4] The usual protocol for upper extremity SSEP is to use the median sensory nerve, which will not interrogate the lower cervical segment as well as the ulnar nerve SSEP.

Further large studies are needed to determine the utility of SSEP in this patient population. Given the variance of data published, it is also possible that there are different phenotypes of the disease which may present differently, furthering the need for large studies.

Sincerely,

Adeel S. Zubair, MD

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Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Conflicts of interest

There are no conflicts of interest.

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