

Spinocerebellar Ataxia Type 2.

Spinocerebellar Ataxia Type 3.

Hashimoto et al., investigated the effects of deep brain stimulation (DBS) or lesions of the ventral intermediate nucleus (Vim) of the thalamus for spinocerebellar ataxia (SCA) and examined the pathophysiological role of neuronal activity of the Vim underlying ataxia.

METHODS: Five patients with SCA with cortical atrophy (ages 60-69 years; 2 sporadic and three familial SCA) and five patients with essential tremor (ET) (ages 57-71 years) were treated with Vim surgery. Intraoperatively, we recorded neuronal activity from single neurons in the Vim thalamus while patients were at rest and compared the physiological properties of those neurons between patients with SCA and those with ET.

RESULTS: Postsurgery mean scores for the Fahn-Tolosa-Marin Tremor Scale were improved from 78 to 44 in SCA patients and from 54 to 21 in ET patients. Stronger stimulation was necessary to optimize outcomes in SCA as compared to ET patients. We analyzed 68 Vim neurons in SCA and 60 Vim neurons in ET. Mean discharge rates, burst characteristics, and oscillatory activity were similar for both patient groups, however, we observed that the ratio of cells responding to passive manipulation was significantly smaller ($P = 0.0001$) in SCA (22%) than in ET (71%).

INTERPRETATION: Thalamic surgery led to a significant improvement in tremor in SCA patients. One potential mechanism underlying ataxia in SCA may be disruption of cerebellar sensory feedback, which modulates motor commands in the cerebello-thalamo-cortical network ¹⁾.

¹⁾

Hashimoto T, Muralidharan A, Yoshida K, Goto T, Yako T, Baker KB, Vitek JL. Neuronal activity and outcomes from thalamic surgery for spinocerebellar ataxia. *Ann Clin Transl Neurol.* 2017 Dec 10;5(1):52-63. doi: 10.1002/acn3.508. eCollection 2018 Jan. PubMed PMID: 29376092; PubMed Central PMCID: PMC5771317.

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