# Sympathetic skin response in Kennedy disease patients

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**Background.** Spinal and bulbar muscular atrophy (SBMA), also known as Kennedy disease, is a rare X-linked neuromuscular disease, caused by a CAG repeat expansion >35CAGs in the exon 1 of the androgen receptor gene. SBMA classically manifests with lower-motor neuron symptoms. Patients commonly present with muscle cramps, tremor, leg weakness, dysarthria and dysphagia. Autonomic nervous system involvement in SBMA is not well studied nor fully known. Sympathetic skin response (SSR) is used in diagnosing the impairment of non-myelinated sympathetic fibres in peripheral neuropathies.

**Aim.** The aim of the study was to investigate the autonomic nervous system's involvement in SBMA by using sympathetic skin response test.

**Methods.** All Kennedy disease patients in Latvia (n=5) took part in this study – five unrelated Caucasian male patients. We deeply phenotyped all patients and carried out sympathetic skin response test as a non-invasive approach to investigate the sympathetic system, as well as nerve conduction studies.

**Results.** The study included all patients with Kennedy disease in Latvia (n=5) that were 34 to 68 years old. Neurological examination revealed typical manifestations of lower-motor neuron damage. Motor and sensory nerve conduction velocities and compound muscle action potentials and sensory action potential amplitudes were abnormal for two out of five patients indicating peripheral large nerve fibre neuropathy. All patients had complaints about sweating disturbances – 3 patients complained about increased sweating and 2 patients about decreased sweating. Sympathetic skin response test results indicated that three of five patients had peripheral sympathetic nervous system dysfunctions. All of those three patients demonstrated an increased latency and two of them also had a decreased amplitude during examination.

**Conclusion.** Our data supports the evidence that Kennedy disease is multisystemic disease with peripheral nervous system involvement, including sympathetic nervous system. However, our study group size was limited and bigger SBMA patient cohorts should be evaluated for autonomic nervous system involvement.

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