Impaired Limb Proprioception in Adults With Spasmodic Dysphonia

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Summary: Objectives. Focal dystonia of the head and neck are associated with a loss of kinesthetic acuity at muscles distant from the dystonic sites. That is, while the motor deficits in focal dystonia are confined, the associated somatosensory deficits are generalized. This is the first systematic study to examine, if patients diagnosed with spasmodic dystonia (SD) show somatosensory impairments similar in scope to other forms of focal dystonia.

Methods. Proprioceptive acuity (ability to discriminate between two stimuli) for forearm position and motion sense was assessed in 14 spasmodic dystonia subjects and 28 age-matched controls using a passive motion apparatus. Psychophysical thresholds, uncertainty area (UA), and a proprioceptive acuity index (AI) were computed based on the subjects' verbal responses.

Results. The main findings are as follows: first, the SD group showed significantly elevated thresholds and UAs for forearm position sense compared with the control group. Second, 9 of 14 dystonia subjects (64%) exhibited an AI for position sense above the control group maximum. Three SD subjects had a motion sense AI above the control group maximum.

Conclusions. The results indicate that impaired limb proprioception is a common feature of SD. Like other forms of focal dystonia, spasmodic dystonia does affect the somatosensation of nondystonic muscle systems. That is, SD is associated with a generalized somatosensory deficit.

Key Words: Basal ganglia–Focal dystonia–Human–Kinesthesia–Somatosensation.

INTRODUCTION

Spasmodic dysphonia (SD) is a chronic voice disorder characterized by involuntary random movement of laryngeal muscles causing disruption of fluent speech with strained-strangled voice quality. Vocal interruptions or spasms, periods of no sound, and periods of near normal voice all co-occur in SD. SD signs and symptoms are task specific, occurring during speech but not during other phonatory (eg, prolonging vowels) or nonphonatory tasks (eg, breathing). Current therapeutic options are limited. SD does not respond to current forms of behavioral speech therapy and is treated primarily with Botulinum toxin injections (botox) to provide temporary symptom relief. There is no cure and misdiagnosis with other idiopathic voice disorders collectively termed *muscle-tension dysphonia* is not uncommon and can lead to inappropriate treatment.

Although its exact etiology is unknown, SD has been considered a form of focal dystonia. SD shares several abnormal neurologic signs with focal dystonia of the head, neck, and hand. For example, abnormal blink reflexes were observed in SD, torticollis, and blepharospasm[1–4] and abnormal long-latency responses to peripheral nerve stimulation have been observed in SD,[5] blepharospasm, and oromandibular dystonia.[6]

Substantial evidence indicates that basal ganglia-related diseases such as Parkinson's disease and certain forms of dystonia are associated with somatosensory and specifically proprioceptive abnormalities which are closely linked to the observed motor deficits[7–17] (for reviews, refer the studies by Patel et al, Konczak et al, and Konczak and Abbruzzese[18–20]). With respect to focal dystonia, the kinesthetic impairments are generalized and not restricted to the dystonic musculature.[17] Given that muscle spindles in dystonic muscles are intact[21] and focal dystonia is not associated with known proprioceptive receptor or peripheral nervous system damage, the most plausible explanation is that the observed kinesthetic deficits in focal dystonia are of central origin and not solely caused by the abnormal tone in the affected musculature or abnormal reflex circuitry in the brain stem. This assessment is corroborated by recordings of somatosensory-evoked potentials (SEPs) and transcranial magnetic stimulation documenting that abnormal processing of somatosensory information in focal dystonia is associated with abnormally enhanced cortical excitability and decreased intracortical inhibition,[22,23] which recently has also been confirmed for SD.[24]

The purpose of this study was to determine whether the proprioceptive acuity of nonspeech motor systems is altered in patients with the voice disorder SD. Acuity refers to one's ability to perceive the smallest, just noticeable difference (JND) between two detectable stimuli.[25] We measured proprioceptive acuity by determining (a) the psychophysical threshold and (b) the uncertainty around the threshold. Specifically, we examined the acuity of the position and passive motion sense of the forearm in SD patients and compared the results to the perceptual performance of a healthy control group. Controlled passive motion of the forearm, not requiring any active muscular contractions, was induced by a custom-built apparatus described in detail previously.[10] The systematic testing of two proprioceptive senses important for motor control allowed us to obtain a proprioceptive profile of each participant. Showing that SD

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patients have reduced proprioceptive acuity similar to patients with other forms of focal dystonia would provide additional evidence for classifying SD as a form of focal dystonia. More importantly, it could provide a justification for creating behavioral treatments that aim to alter proprioceptive inputs to the laryngeal muscles to overcome the SD symptoms. This would be especially relevant for patients who do not respond well to botox injections and who currently have no viable treatment option.

METHODS

Participants

The study was approved by the Institutional Review Boards of the University of Minnesota, and Chang Gung Memorial Hospital, Taiwan. A total of 43 subjects participated, including 15 SD patients (mean age 55.4 ± 10.6 years; six male and nine female), and 28 healthy controls (mean age 61.1 ± 11.6 years; 13 male and 15 female). All SD patients and 13 control subjects were recruited in Minnesota. Fifteen additional controls were recruited in Taiwan. All patients were seen at the end of their Botox cycle when voice symptoms were most pronounced and the effect of medication was minimized. Before testing, clinical status was assessed through a questionnaire and clinical speech evaluation.

Clinical evaluation was based on audio recordings of vowel prolongation and connected speech with the latter using the 10 "adductor" sensitive sentences.[26] Sentence audio recordings were used for rating dysphonia severity and for identifying voice breaks. Two judges, blinded to the diagnosis, were given access to the secured audio files and listened to them online. Both were certified speech-language pathologists who work exclusively with voice-disordered patients. Judges used a visual-analog scale to rate dysphonia severity. The scale was a line of 100 mm in length, where the 0 mm mark represented no voice impairment and the most severely impaired status was by the 100 mm mark. Based on the same recordings, acoustic analysis was performed to determine the presence of voice breaks using PRAAT software.[27] Voice (phonation) breaks were identified for a voiced component (usually a vowel) within a word and only when the break was greater than 50 milliseconds following established guidelines.[28] A voice break was identified as an absence of voicing occurring during a voiced segment, and lasting 50 milliseconds or more. Nasoendoscopic examinations were available for 10 patients to confirm the diagnosis of SD. The remaining five patients were referrals and the original diagnosis had been made elsewhere. Clinically, they were treated as SD and did respond to botox. Table 1 lists the patient characteristics in detail. Data sets of one SD patient and five control subjects were incomplete (subjects did not perform both tests). For the analysis of the position sense acuity, the data of 14 SD and 25 controls were considered. For the analysis of motion sense acuity, the data of all 15 SD patients and 26 controls were included.

Instrumentation

A passive motion apparatus was used to test motion and position sense acuity (Figure 1A). Subjects placed their forearm on a rotatable aluminum splint that was moved passively by a direct current five-phase stepping motor (precision: 5466 steps per $1^\circ = 0.00018^\circ$ per step; Nyden Inc., San Jose). The splint was padded with 16-cm-thick foam to attenuate possible vibration effects of the DC motor. Control of the apparatus was realized through customized software routines coded in MATLAB Technical Programming Language.

TABLE 1.
Clinical Characteristics and Basic Demographics of Spasmodic Dysphonia Patients

ID	Age (y)	Gender	Reported Symptom Duration (y)	Duration Since Official Diagnosis (y)	Voice Symptom Severity (0–100)	Number of Voice Breaks
SD01	40	F	15	8	40	0
SD06	68	M	47	27		
SD10	50	F	1.9	0.2	35	0
SD09	67	F	37	22	34	1
SD16	48	M	3.5	2	55	2
SD21	47	F	2.5	0.75	61.0	5
SD27	67	F	1.5	1.2	47.7	0
SD25	60	M	15	8	61.3	5
SD28	57	M	9	1.5	72.3	10
SD29	49	M	22	2	21.3	2
SD34	46	F	3.5	3	62.7	0
SD36	45	M	3	2.5	92.7	0
SD37	70	F	1.5	1	61.7	12
SD43	70	F	11	9	38.0	0
SD44	47	F	1.4	0.4	64.0	0

Notes: For SD06, no audio/video data could be obtained and thus no data on voice quality could be derived. Phonation (voice) breaks were identified for a voiced component (usually a vowel) within a word and only when the break was greater than 50 milliseconds.