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Functional electrical stimulation for the treatment of lower extremity dystonia

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Introduction

Primarily used as an alternative to ankle foot orthosis (AFO) for central foot drop secondary to stroke or other CNS disease, lower extremity functional electrical stimulation (FES) works by electrical stimulation of the common peroneal nerve during the swing phase of gait.[1] This results in contraction of the anterior tibialis muscle and lateral compartment muscles with resultant dorsiflexion and eversion of the foot. Dystonia, which subsumes a range of involuntary hyperkinetic movements, may manifest as an action-induced stereotyped movement. We present a case in which the use of radiofrequency controlled FES was associated with improvement in right lower extremity dystonia and gait.

Case Presentation

A 62-year-old right-handed woman presented with abnormal right lower extremity movements while walking. Her first symptoms, involuntary right toe flexion and plantarflexion during walking, began in the setting of a rigorous exercise routine six years prior to presentation. Two years after symptom onset she fell and fractured her femur. Her symptoms worsened during her recovery. In order to walk, she raised her right leg higher, so her toes would not drag on the floor. She perceived her right leg was longer than her left. At

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Conflicts of Interest. MO - Scientific Advisory Board, Victhom, Inc. (paid), periodic speaker for Bioness, Inc. (unpaid), director of clinical trial for NDI Inc (electrical stimulation for shoulder pain in stroke), director of clinical trial for BioNess, Inc (L300 trial in stroke). The other authors report no other relevant conflicts of interest.

Beth Israel Medical Center does not require the approval of an Institutional Review Board for case reports.

The patient provided written consent to be videotaped and for the “images to be used for professional and scientific purposes and for medical education, including their publication in professional journals and medical books.”

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that time, she was diagnosed with focal dystonia. In addition to her right leg symptoms, she reported a right hand tremor for twenty years with two years of worsening handwriting. Prolonged writing was associated with increasing loss of control of her right hand. She had no history of depression, apathy, or anxiety. She had never taken a neuroleptic drug. She had no other significant past medical history. Her father was diagnosed with an essential tremor. She otherwise had no family history of dystonia or Parkinson's disease.

Her neurological exam revealed normal muscle tone, strength, reflexes, and sensation throughout. With writing she had hyperactive contraction of muscles of her right hand and wrist. With walking, she had dystonic plantarflexion of her right foot accompanied by exaggerated right hip flexion, giving the impression of a steppage gait. These abnormal limb contractions were not present at rest or during heel-walking, toe-walking, or walking backwards. Two independent movement disorders specialists diagnosed her abnormal leg movements as dystonia (SB, SF). There were no other neurological findings.

Testing for the DYT1 GAG deletion was negative. An MRI of her brain revealed a small venous angioma in her left posterior cingulate gyrus, but was otherwise normal. MRIs of her cervical and thoracic spine were unremarkable. She had a normal EMG of her right lower extremity. Three treatments with onabotulinum toxin A, up to 300U total in her right tibialis posterior and medial and lateral gastrocnemius, provided no benefit. A trial of levodopa up to 300 mg a day was also not effective. Her concern about side effects made her reluctant to try other oral medications. To compensate for her dystonia, she wore an AFO, which reduced her falls, but was cumbersome and not aesthetically pleasing.

As her gait resembled that accompanying foot drop, she was fitted with a radio frequency-controlled FES device (NESS L300, Bioness Ltd).[2] After a six-week adaptation period, she experienced significant improvement in her gait. Wearing the device without electrical stimulation was not sufficient to result in gait improvement. Over time she required less stimulation to achieve the same effect, and there was mild gait improvement when FES was turned off. (See video.) Benefit with the device has been sustained for 18 months, although her response has fluctuated over time.

Selected clinical parameters were available at 3 and 15 months after initiating the device. Her distance on the 6 minute walk test, a basic measure of endurance, without the L300 increased 20% from 765 feet to 947 feet over that 12 month period. There was little change in distance with the device in place. Also, single leg stance time on the affected leg with eyes closed and the device off improved from 8.7 seconds to 22.3 seconds. Despite apparent improvement in balance and endurance, her Timed Up and GO (TUG) time increased from 6.7 to 8.2 seconds.

Discussion

We present the application of FES for the treatment of task-specific leg dystonia. Although radio-frequency controlled FES was designed for foot drop after spinal cord or brain injury, our patient's dystonic gait was biomechanically similar. The rationale behind this approach was that electrical stimulation of the peroneal nerve during the swing phase of gait would drive dorsiflexion and eversion of the foot and overcome the patient's dystonic plantarflexion.

Potential pathogenic mechanisms underlying the development of dystonia are exuberant synaptic plasticity and altered sensorimotor organization.[3] Just as maladaptive sensorimotor plasticity contributes to the development of dystonia, sensory and motor retraining techniques have been proposed as strategies to beneficially alter sensorimotor organization.[4] Via costimulation of the sensory afferent and motor afferent components

and activation of motor afferents and proprioceptive inputs during dorsiflexion, FES has the capability of affecting central sensorimotor pathways and modifying central sensorimotor maps. However, after eighteen months of using FES daily, our patient continued to have dystonia without the device and her sensory distortion persisted.

Without more formal gait parameters assessing stride count and symmetry, it is difficult to determine why our patient's endurance and balance improved over time. Possible explanations include muscular strengthening of the affected leg secondary to increased activity or increased motor power and control as a result of stimulated contractions. A cardiopulmonary conditioning effect as a result of increased ambulation may account for the improved endurance.

Our patient's dystonia, with coexistent task-specific right leg and hand dystonia, could be characterized as bi-focal dystonia or mild hemidystonia. Since the majority of hemidystonia cases are associated with a structural lesion,[5] the left posterior cingulate venous angioma may be involved in the pathogenesis of our patient's symptoms. Previous imaging studies found aberrant activation of the cingulate cortex in patients with dystonia,[6, 7] but isolated cingulate lesions have not been associated with dystonia.

FES is a novel symptomatic treatment for the abnormal contractions present during dystonia. The device used in our patient was perfectly suited for dystonic plantarflexion and would not necessarily work for other dystonic movements of the lower limb. Our patient's dystonia was task-specific, present only during walking, and may have been more amenable to treatment. Despite these shortcomings, in an appropriately selected patient who has failed botulinum toxin and cannot tolerate oral medications, FES can improve motor control and function in leg dystonia.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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