Fixed dystonia unresponsive to pallidal stimulation improved by motor cortex stimulation

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Fixed dystonia is a rare condition in which immobile dystonic postures do not return to a neutral position at rest. It is typically focal or segmental and painful in about 50% of cases. Corticectomies, pleotomies, and peripheral denervation are often tried, but responses are typically short-lived. Immobility and anomalous postures do not return to a neutral position at rest. It is typically focal or segmental and painful in about 50% of cases. Corticectomies, pleotomies, and peripheral denervation are often tried, but responses are typically short-lived. Immobility and anomalous postures do not return to a neutral position at rest.

**Case report.** A right-handed woman with no remarkable personal or family history developed severe painful elevation of the left shoulder in 1990 at age 31. Left anterior scalene myotomy in 1990 was unsuccessful. By 1998 she had progressed to severe segmental dystonia, with fixed elevation and anterorotation of the left shoulder, abduction of the upper limb, severe trunk involvement, and fixed kyphoscoliosis (figure, A). Videotaping (video 1 [on www.neurology.org]). Gestes antagonistes and overflow dystonia were not present. Attempts to stand or walk were thwarted by unbearable pain in the affected shoulder and arm, whereas at rest there was little pain. Benzedrinate, baclofen, and trihexyphenidyl produced no benefit. DY17 and DY75 gene mutations were absent. Nothing indicated somatoform or psychogenic disorder.

Brain MRI was normal. Under deep sedation, left shoulder and arm posture became almost normal, and the continuous electromyographic (EMG) activity of the left trapezius and pectoralis major disappeared. Repeated botulinum toxin (Dysport) injections to the left superior trapezius (up to 500 U), levator scapulae (up to 75 U), and pectoralis major (up to 300 U) did not relieve pain or dystonia. At age 44, internal global pallidus (GPi) stimulators were implanted bilaterally, but no improvement in dystonia or pain occurred over 12 months.

A four-plate Medtronic Restrue electrode array was placed epidurally over the right primary motor cortex parallel to the central sulcus (figure, C) under local anesthesia and MRI control. The GPi electrodes and subclavian generators were left in place. The right generator was connected to the plate, and the left was switched off. Stimulation started the day after implant (3.8 V, 60 Hz, 60 microseconds, contacts 0 +1 -2) and remained unchanged thereafter.

Gradual recovery became evident at 4 months, and by 6 months the pain and dystonic postures of the shoulder and trunk had almost resolved, although the fist remained clenched (figure, B; video 2). This marked improvement persisted at 22 months.

Two resting [(18)F]fluorodeoxyglucose ([18]FDG) PET scans were performed (6 months before and 6 months after cortical implant), using a multitracking tomograph. The patient’s parametric images of [(18)F]FDG distribution were compared with those in 21 healthy subjects using voxel based SPM99 procedures (Wellcome Department of Cognitive Neurology, London, UK). Differences were considered significant at p < 0.001. PET during GPi stimulation showed significantly increased glucose consumption in the sensorimotor cortex (more extensive on the left) and supplementary motor cortex and anterior cingulate gyrus bilaterally. PET under cortical stimulation showed significant hypometabolism in the cerebellum, more pronounced on the right, and no increase in cortical metabolism (figure, D).

**References**


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Discussion. Mechanisms of fixed dystonia have not been elucidated but may differ from those of primary torsion dystonia. Our patient did not improve with GPi stimulation, which is often effective in primary and nonprimary dystonia, suggesting that the modulation of data flow within the GPi during stimulation was insufficient to improve the fixed dystonia symptoms. The improved movements and fixed dystonia during cortical stimulation were not due to reduced pain because before cortical implant, fixed posturing at rest persisted, although there was little pain. A PubMed search uncovered no reports of fixed dystonia treated by epidural cortical stimulation, but found a report that stroke-related hand dystonia and pain improved with epidural cortical stimulation. It remains unclear why motor cortex stimulation is effective in these conditions. Low-frequency motor cortical stimulation is thought to activate neurons within the cortex, which results in modulation of the corticopontocerebellar and the corticopallidothalamocortical loops.

The significant bilateral reduction of cerebellar glucose metabolism during cortical stimulation (figure, D) suggests a modulating effect on cerebellar function. Interestingly, selective elimination of cerebellar output improves dystonia in experimental animals. Alternatively, modified cerebellar outflow to the motor cortices may have caused plastic reorganization of the cortical representation of movements, to produce the improved motor function.

Both basal ganglia and cerebellum—key structures in motor control—may be involved in the various manifestations of dystonia. The current case suggests that motor cortex stimulation may be a useful treatment option for fixed dystonia.

References