Unilateral pallidal deep brain stimulation in a patient with cervical dystonia and tremor

Case report

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Bilateral deep brain stimulation of the globus pallidus pars interna (GPi) is the favored neuromodulation procedure in cases of cervical dystonia. The authors report on a case of unilateral GPi implantation that resulted in sustained benefit with marked improvement in pain and dystonia. (DOI: 10.3171/2010.4.JNS091722)

Key Words • cervical dystonia • deep brain stimulation • neuromodulation

Cervical dystonia is the most frequent dystonic movement disorder. Bilateral pallidal DBS is becoming the favored surgical procedure for cases that do not respond to medical treatment, particularly BTX injections.

Although the pathophysiology of cervical dystonia is still largely unknown, there is evidence of bilateral basal ganglia involvement and bilateral electromyographic muscle activation regardless of the clinical pattern and laterality. Transcranial magnetic stimulation studies have demonstrated that there are some bilateral corticomotor projections to the SCM muscle. Patients with severe CD unresponsive to BTX often have a very complex pattern of bilateral muscle hyperactivity, and thus, it has been presumed that unilateral DBS will be ineffective in such cases. Indeed, most surgical centers seem to prefer bilateral surgery. Little information is therefore available on the utility of unilateral stimulation and whether it is sufficient to help bilateral dystonic symptoms.

We describe a patient with disabling CD resistant to medical and BTX treatment, who was satisfactorily treated with unilateral stimulation of the GPi.

Case Report

History. This 76-year-old, right-handed woman with no family history of movement disorders or neurological disease was referred to our center for surgical opinion. Her medical and surgical history was positive for appendectomy at the age of 14 years, nephrectomy for renal cancer at the age of 46 years, and hysterectomy. No evidence of a cancer relapse was found at the time of the consultation.

Her dystonia symptoms started at the age of 16 years when she was noted to have head tilting to the left side with titubation. These symptoms slowly progressed, and dystonic tremor was diagnosed when she was 69 years old and was initially treated using benzodiazepines with poor benefit. The neck dystonia had worsened significantly in the past 2 years, and severe pain developed and in fact was her most disabling symptom at the time of consultation.

Further medical treatment with propranolol and tetrabenazine proved unsatisfactory. Type A BTX—3 trials—did not provide significant relief of the dystonia and pain, but she did experience some side effects of treatment, including dysphagia, neck pain, and difficulty turning her neck following the injections. She was taking 1 mg of lorazepam every day at the time of consultation.

Examination. On examination, her neck was rotated to the right side and her head was tilted to the left. She...
was able to move her head past midline but not to the extreme opposite positions. Significant hypertrophy of the left SCM muscle was also present. Sensory tricks provided little or no relief of her neck spasms. Her TWSTRS scores for disease severity, disability, and pain were 22, 15, and 14, respectively.

Magnetic resonance imaging of the brain demonstrated no significant structural abnormalities; and imaging of the cervical spine, moderate spinal stenosis from C-4/C-5 to C-6/C-7, with no evidence of myelopathy.

Her neuropsychiatric and neuropsychological assessments were unremarkable. Because of her age and to address patient comfort, a staged approach to bilateral pallidal DBS was chosen.

Operation. Deep brain stimulation involved implantation of the electrode on the side contralateral to the contracted SCM muscle. The right DBS electrode (3387, Medtronic, Inc.) was implanted using a microelectrode guidance technique, as previously described. Intraoperative microelectrode recordings revealed neuronal activity typical of the GPi, and correct localization was further confirmed by microstimulation-evoked visual percepts immediately below the GPi consistent with optic tract activation.

Postoperative Course. The patient experienced a remarkable early benefit from a microlesioning effect: a reduction in both her stiffness and the tremor in her neck. Postoperative MR imaging showed the electrode to be in a satisfactory position and no evidence of complications (Fig. 1). She had mild, transient confusion postoperatively.

Stimulation was begun 1 month after surgery. Programming of GPI stimulation was done according to the algorithm used for bilateral stimulation, as previously described.

After 3 months of continuous stimulation, there was a 75% reduction in the dystonic tremor, almost complete pain relief, and a 60% improvement in her neck dystonia. The TWSTRS severity scores decreased from 22 preoperatively to 13 at 3 months after surgery, disability scores went from 15 to 4, and pain scores went from 14 to 3. Initial settings of the stimulation parameters were 2.5 V, 130 Hz, 60 μsec, contacts 1–case+. Given her improvement, GPI DBS was not performed on the contralateral side. Scores remained satisfactory at 10 months postsurgery (TWSTRS severity score of 14).

Discussion

For patients with incapacitating CD refractory to medical treatment, especially BTX injections, bilateral GPI DBS is currently the surgical procedure preferred by many movement disorders centers. Most published studies are case reports or small series, reporting 43–76% improvement in the TWSTRS scores following bilateral GPI DBS.

Previous experience with thalamotomies and pallidotomies for incapacitating axial or bilateral dystonia have shown that bilateral procedures yield better results than unilateral procedures. However, the risk of severe permanent adverse events has discouraged the adoption of bilateral pallidotomies at times in favor of more peripheral procedures, such as posterior ramisectomy.

In severe CD several deep muscles are often involved, and it may be difficult to reach them for BTX treatment even when using electromyographic guidance. This factor might explain why some patients with CD fail to get relief from BTX treatment. Patients with disabling CD often have bilateral neck muscle hyperactivity, and therefore, bilateral DBS is presumed to have an important role in treatment. The necessity of bilateral stimulation or the role of staging has not been formally tested, however. The patient in our case, who presented with idiopathic CD, showed significant improvement after unilateral stimulation of the GPI contralateral to the contracting SCM muscle.

There is more experience with unilateral procedures for Parkinson disease. Whereas unilateral thalamotomy mainly produces tremor reduction in the contralateral hemibody, unilateral pallidotomy can improve the severity of bilateral bradykinesia and rigidity, as measured by standard clinical rating scales. Since bilateral surgery results in modest benefits but is associated with an increased risk of complications, bilateral pallidotomy is usually considered only as a staged procedure. Unilateral GPI and subthalamic nucleus DBS for Parkinson disease can lead to improvement in both contralateral and ipsilateral motor function, although in general the motor benefit is less effective than with bilateral stimulation.

If a second procedure is required, staged DBS can have a surgical outcome and adverse effects similar to those of the simultaneous bilateral procedure. A persistent bilateral effect from unilateral interventions in patients with tremor is less common, however.

Experience with other circuit disorder conditions, such as Gilles de la Tourette syndrome, has yielded positive results after unilateral procedures. In Tourette syndrome, a mix of bilateral motor and complex tics, obsessive-compulsive and attention deficit disorders, and limbic, associative, and motor circuits of the basal ganglia seem to be involved. Unilateral capsulotomy and DBS in the nucleus accumbens have demonstrated a bilateral reduction in symptoms similar to the effect produced by...
bilateral interventions. For dystonia, however, a few studies show different degrees of improvement in axial dystonic symptoms with unilateral pallidal DBS (Table 1). 

There have been 4 reports of unilateral DBS in dystonia. Foote et al. implanted a unilateral pallidal DBS system in a patient with craniofacial dystonia and blepharospasm. The patient initially experienced complete resolution of his bilateral symptoms after surgery, and a 45–50% improvement in his Unified Dystonia Rating Scale scores on stimulation. After 6 months, the patient underwent contralateral electrode implantation. With bilateral DBS, the patient had a 75% improvement. İşlek et al. reported a case of posttraumatic cervical dystonia due to an ipsilateral focal basal ganglia lesion. This patient experienced significant improvement following unilateral globus pallidus stimulation contralateral to the contracted SCM muscle. Escamilla-Sevilla et al. documented a case of segmental cervical and truncal dystonia treated with bilateral GPi DBS surgery. Despite some benefit from the bilateral stimulation, the patient derived the maximum clinical improvement from unilateral stimulation ipsilateral to the contracting SCM muscle. This level of benefit was maximal at 3 months after surgery and was maintained for at least 18 months.

Based on these studies, the efficacy of unilateral DBS for bilateral motor symptoms in dystonia suggests that staged procedures could be preferred in selected patients. However, selecting the side for stimulation may be difficult in dystonia. Indeed, the anatomical pathways underlying the bilateral effects remain uncertain. Previous studies have shown that lesioning the ipsilateral hemisphere yields ipsilateral SCM muscle weakness, but there have been a few cases in which lesions in the contralateral GPi have been involved in posttraumatic cervical dystonia. In our case, we decided on the side of treatment by stimulating the side contralateral to the contracted left SCM muscle.

A staged pallidal DBS can be a good strategy in selected patients with CD, since they could benefit from unilateral procedures and thus obviate the need for a second surgery. Unilateral GPi DBS can induce a remarkable improvement in a subset of patients and decrease the risk of complications related to a simultaneous bilateral surgery.

The mechanism of action of DBS has not been elucidated, but it is possible that DBS has a more significant bilateral effect than lesioning procedures. Probably a complex pattern of interactions in basal ganglia circuits is involved in the clinical effects of unilateral pallidal DBS. However, it is also possible that the unilateral GPi stimulation has given our patient only a temporary benefit and that bilateral stimulation may in fact lead to a better longer-term outcome. Further studies are needed to elucidate the mechanism of amelioration of CD and perhaps other dystonia types following pallidal DBS.

### References

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