Subthalamic nucleus deep brain stimulation for severe idiopathic dystonia: impact on severity, neuropsychological status, and quality of life

Galit Kleiner-Fisman, M.D.,1 Grace S. Lin Liang, M.D.,2 Paul J. Moberg, Ph.D.,1 Anthony C. Ruocco, M.S.,3 Howard I. Hurtig, M.D.,4 Gordon H. Baltuch, M.D., Ph.D.,5 Jurg L. Jaggi, Ph.D.,5 and Matthew B. Stern, M.D.1,4

1Philadelphia Parkinson’s Disease Research Education and Clinical Center, Philadelphia Veterans Administration Hospital; 4Department of Psychology, Drexel University; Departments of 4Neurology and 1Neurosurgery, Pennsylvania Hospital and University of Pennsylvania, Philadelphia, Pennsylvania; and 5Parkinson’s Institute, Sunnyvale, California

Object. Medically refractory dystonia has recently been treated using deep brain stimulation (DBS) targeting the globus pallidus internus (GPI). Outcomes have varied depending on the features of the dystonia. There has been limited literature regarding outcomes for refractory dystonia following DBS of the subthalamic nucleus (STN).

Methods. Four patients with medically refractory, predominantly cervical dystonia underwent STN DBS. Intraoperative assessments with the patients in a state of general anesthesia were performed to determine the extent of fixed deformities that might predict outcome. Patients were rated using the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) and the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) preoperatively and 3 and 12 months following surgery by a rater blinded to the study. Mean changes and standard errors of the mean in scores were calculated for each subscore of the two scales. Scores were also analyzed using analysis of variance and probability values were generated. Neuropsychological assessments and quality of life ratings using the 36-Item Short Form Health Survey (SF-36) were evaluated longitudinally.

Results. Significant improvements were seen in motor (p = 0.04), disability (p = 0.02), and total TWSTRS scores (p = 0.03). Better outcomes were seen in those patients who did not have fixed deformities. There was marked improvement in the mental component score of the SF-36. Neuropsychological function was not definitively impacted as a result of the surgery.

Conclusions. Deep brain stimulation of the STN is a novel target for dystonia and may be an alternative to GPI DBS. Further studies need to be performed to confirm these conclusions and to determine optimal candidates and stimulation parameters. (DOI: 10.3171/JNS-07/07/0029)

Key Words • deep brain stimulation • dystonia • quality of life • subthalamic nucleus

DYSTONIA is a clinical syndrome characterized by sustained muscle contractions causing twisting and repetitive movements or abnormal postures.7 The pathophysiology of dystonia is still being elucidated, but it has been postulated that a lack of appropriate surround inhibition within the basal ganglia, normally mediated by the GPI and STN, may contribute to its pathophysiology.15 Authors of multiple studies have shown that pallidal DBS is effective in treating refractory primary dystonia.1,4,13,21 Other authors have reported improvement in some types of secondary dystonia2,10,13,23 but generally to a lesser extent.

Deep brain stimulation of the STN has also been effective in reducing the off-medications dystonia in patients with Parkinson disease,18 but experience in targeting the STN in other types of dystonia is limited. In the study of Sun et al.,21 eight patients with idiopathic generalized dystonia underwent DBS: four received STN DBS and four received GPI DBS. All four patients who underwent STN DBS experienced significant and immediate improvement, whereas only two of the patients undergoing GPI DBS improved and only after months of treatment.21 Authors of another case report described a patient with medically refractory cervical dystonia and tremor who experienced marked improvement in dystonia following bilateral STN DBS.3 In contrast, a case series of four patients with generalized dystonia (three secondary to pantothenate kinase–associated neurodegeneration and one primary dystonia) underwent STN DBS without benefit.9

We present four patients with idiopathic, medically refractory dystonia who were prospectively evaluated in a blinded protocol to determine the effects of bilateral STN
DBS on severity of dystonia, disability, and pain impact on neuropsychological function, and QOL.

Clinical Material and Methods

Patient Population

All patients were men and had clinically diagnosed dystonia causing severe functional impairment. In all patients, cervical dystonia was the most prominent feature. Patients having additional disabilities was not the main goal. In some patients, cervical dystonia was the most prominent feature. These patients were included in a retrospective analysis of elderly patients (mean age 69 years) from the University of Pennsylvania, with a mean duration of 10 years (range 10-30 years). All patients had dystonia that was refractory to multiple medication trials and chemo-denervation with botulinum toxins A and B.

Baseline characteristics of the four patients prior to surgery are shown in Table 1. The mean age at time of surgery was 50.2 years (range 41-56 years) and the mean duration of illness prior to surgery was 24.5 years (range 10-39 years). All patients had disability from dystonia that was refractory to multiple medication trials and chemo-denervation with botulinum toxins A and B.

Case 1. At 41 years of age, this patient began experiencing symptoms of torticollis to the left and head tremor that had progressively worsened over the ensuing years. At the preoperative evaluation, there was severe retrocollis, right torsion, and left laterocollis with irregular, high-amplitude head tremor, as well as marked blepharospasm, lip pursing, severe dysarthria, and moderate left arm and mild truncal dystonia. Range of neck motion after the induction of general anesthesia revealed a significant fixed deformity (Table 1).

Case 2. When this patient was 40 years old, he experienced torticollis with onset of right head turn. Over the course of 10 years the torticollis worsened, and severe neck and shoulder pain was exacerbated by working at a computer for extended periods. Pain was the main source of disability. Baseline examination showed severe right torticollis, mild left tilt, left shoulder elevation, and moderate irregular, jerky, left hand and head elevation. He was unable to maintain a neutral position for more than a few minutes, and a sensory trick using the hand to push the chin to midline was only partially effective.

Case 3. This patient began experiencing head turning toward the right at 15 years of age. His brother had spasmodic dysphonia. Genetic tests were not performed because of patient's wishes. Pharmacological treatment for head tremor and neck pain was initiated at age 44 years. At age 48 years, a fall from a ladder resulted in fractures of the face, rib, and left leg resulting in fractures of the face, rib, and neck. The patient was admitted at age 44 years. At age 48 years, a fall from a ladder resulted in fractures of the face, rib, and neck pain was admitted. The patient was in a state of general anesthesia.

Case 4. At 14 years of age, this patient began experiencing symptoms of torticollis to the left and head tremor that had progressively worsened over the ensuing years. At the preoperative evaluation, there was severe retrocollis, left laterocollis, and severe deviation of the head to the left. Baseline examination showed irregular, high-amplitude head and neck elevation, and moderate left arm and mild truncal dystonia. Range of neck motion after the induction of general anesthesia revealed a significant fixed deformity (Table 1).

TABLE 1
Patient baseline characteristics

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Duration of Disease (yrs)</th>
<th>Family History</th>
<th>Dystonia Type</th>
<th>Sites of Involvement</th>
<th>Medications at Time of Op*</th>
<th>Previously Failed Medications</th>
<th>Neck ROM†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>41</td>
<td>56</td>
<td>15</td>
<td>no</td>
<td>segmental</td>
<td>face, neck, trunk, lt arm</td>
<td>30 mg diazepam, 12–16 mg benzotropine, 25,000 U botulinum toxin B to neck</td>
<td>botulinum toxin A</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>40</td>
<td>50</td>
<td>10</td>
<td>no</td>
<td>segmental</td>
<td>neck, shoulder</td>
<td>1.5 mg clonazepam, 5 mg trihexyphenidyl, 10 mg cyclobenzapine as needed, 100 mg amitriptyline, 1000 mg naproxen</td>
<td>tizanidine, baclofen, dilantin, botulinum toxins A &amp; B</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>15</td>
<td>54</td>
<td>39</td>
<td>yes</td>
<td>segmental</td>
<td>face, neck, trunk</td>
<td>none</td>
<td>clonazepam, trihexyphenidyl, benzotropine, diazepam, baclofen, phenol nerve blocks, botulinum toxins A &amp; B</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>7</td>
<td>41</td>
<td>34</td>
<td>no</td>
<td>generalized</td>
<td>speech, neck, shoulder, upper &amp; lower limbs</td>
<td>45 mg trihexyphenidyl, 4 mg tizanidine, 225 mg benadryl</td>
<td>levodopa, diazepam, botulinum toxins A &amp; B</td>
</tr>
</tbody>
</table>

*Medications are listed in daily doses, except for botulinum toxin, which is indicated as the last dose administered prior to surgery.
†The neck ROM measurements were made while the patient was in a state of general anesthesia.
Treatment of idiopathic dystonia with STN DBS

Case 4. When this patient was 7 years of age, curling and left foot eversion were noticed. Shortly thereafter, right foot turning and plantar flexion developed, affecting balance and gait. Spasmodic dysphonia developed at age 10 years and resolved over the years, but stuttering of speech persisted into adulthood. Cervical dystonia with retrocollis and head tremor began at age 13 years. Due to progressive right-hand dystonia, writing was switched to the left hand. Medications had minimal benefit, although botulinum toxin A injections of the neck and legs initially provided approximately 75% symptomatic improvement; however, immunity eventually developed. A trial of botulinum toxin B produced intolerable side effects. Investigations including myelography and brain MR imaging were unremarkable. Genetic testing during childhood did not reveal any findings, and the patient refused further testing as an adult.

Baseline examination showed hypertrophy of the left sternocleidomastoid muscle, right head turn, left tilt and lateral shift, retrocollis, and intermittent head tremor, most prominent with neck flexion. Bilateral hand posturing was present intermittently at rest and with action. There was a jerky tremor of the left hand. Gait was severely affected from constant right foot intorsion and left ankle flexion making the patient unable to place his feet flat on the ground.

Surgical Procedure and Stimulator Programming

Permanent quadripolar DBS electrodes (model 3389, Medtronic, Inc.) were implanted in the STN bilaterally following stereotactic localization with MR imaging guidance after induction of general anesthesia as described in detail elsewhere. Prior to electrode placement but with the patient in a state of anesthesia, passive ROM of the neck was measured to determine to what extent there were fixed deformities. The patients underwent postoperative MR imaging to confirm optimal electrode location within the STN. The neurostimulators (Soletra, Medtronic, Inc.) were placed subcutaneously in the subclavicular region the same day. The stimulators were turned on and initially programmed 1 week following surgery. Medications and stimulator settings were adjusted at monthly intervals for the first 3 months and thereafter according to the patient’s clinical status (Table 2).

Clinical Evaluations

Clinical evaluations were done at a presurgical visit and at 3 and 12 months after surgery. Patients were examined and videotaped before and after surgery according to a standardized protocol incorporating elements of the validated BFMDRS and the TWSTRS. Intraoperative neck ROM evaluations are displayed in Table 1. During the post-surgical evaluations, patients underwent examinations in the stimulator-on and stimulator-off states; both the patient and the examiner were blinded to stimulation status. There was a 30-minute interval between adjusting the stimulators either on or off and the clinical evaluation. This length of time was chosen to balance the capture of a difference in stimulation states with minimization of patient discomfort when stimulation was turned off. Patient-scored QOL, functional, and disability scales and neuropsychological evaluations were all administered during the stimulation-on state. A movement disorders specialist, blinded to both the surgical status and the stimulation status of patients, independently rated the videotaped examinations using the TWSTRS for cervical dystonia and BFMDRS movement scale for segmental or generalized dystonia if there was involvement of additional areas. Mean changes and standard errors of the mean in scores were calculated for each subscore of the two scales. Analysis of variance was performed on the subscores of the TWSTRS and the BFMDRS to determine whether there was a significant difference in scores over time, and probability values were generated.

Quality of Life

To determine the impact of this procedure on QOL the SF-36, widely used and validated in multiple disease states and heterogeneous populations, was administered before surgery and 3 and 12 months following surgery. Higher scores represented better health states. Confidence intervals for the PCS and MCS scores, which have the greatest reliability among the SF-36 scores, were examined for changes between baseline and 3-month scores, and between baseline and 12-month scores.

Neuropsychological Assessment

Each patient was administered a comprehensive neuropsychological evaluation preoperatively and 3 months and 1 year after surgery (Table 3). Tests were administered by a board-certified clinical neuropsychologist (P.J.M.) according to a standardized protocol. To ensure comparability across tests, the raw neuropsychological test scores were rescaled to standard equivalents (Z-score transformation) using the means and standard deviations from published normative data corrected for age, sex, and education. A patient’s score for each neuropsychological domain was obtained by averaging the Z-scores for tests measuring that function. To gauge change in neuropsychological abilities with DBS implantation, the percentage of change from baseline relative to 1-year follow-up was calculated (Table 4).

Results

Postoperative MR imaging confirmed electrode placement in the anterior dorsolateral STN bilaterally in all patients. Table 2 lists the clinical results following surgery. All assessments are reported in the stimulator-on state. Overall, significant improvements in scores over time were noted, with a 5 ± 2.5–point improvement in the TWSTRS motor score (p = 0.04), a 7 ± 1.6–point benefit in the TWSTRS disability score (p = 0.02), and a 14.9 ± 5.9–point improvement in the TWSTRS total score (p = 0.03). There were no significant differences in the movement or disability subscores of the BFMDRS. Because no differences were found between stimulation-on or -off states (results not shown), reported scores are in the stimulation-on state.

Case 1

Following surgery, neck and shoulder pain and head tremor were reduced. However, the BFMDRS worsened slightly over the follow-up period, mainly in the areas of...
speech and walking. The TWSTRS scores modestly improved in all domains, especially in the pain subscale (60% improvement at 12 months). Medication doses were reduced by approximately half at the last follow-up visit.

**Case 2**

Immediately following bilateral stimulator placement the patient experienced marked improvement in the torticollis and head tremor, complete resolution of the hand tremor, and improvement in neck pain at 3 months. All medications for dystonia were discontinued. The patient was able to maintain a neutral position the majority of the time, and he no longer required the sensory trick to bring his head to midline. Following surgery he was well enough to return to work; however, due to constant computer use his neck pain recurred which was reflected again by higher pain scores at the time of evaluation. When the patient had discontinued computer use, his neck pain once again resolved completely (after study termination).

**Case 3**

This patient experienced immediate reduction in the severity of low-back pain and retrocollis/lordosis after initial programming. However, the BFMDRS and TWSTRS scores at the 3- and 12-month evaluations were similar or slightly worse than baseline, and he continued to be severely disabled from the dystonic postures, neck pain, and worsening depression.

**TABLE 2**

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Time of Evaluation</th>
<th>BFMDRS Score</th>
<th>TWSTRS Score</th>
<th>Stimulator Settings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Movement (max 120)</td>
<td>Disability (max 30)</td>
<td>Motor (max 35)</td>
<td>Disability (max 30)</td>
</tr>
<tr>
<td>case no.</td>
<td>preop</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>3 mos</td>
<td>36.5</td>
<td>5</td>
<td>31</td>
</tr>
<tr>
<td></td>
<td>12 mos</td>
<td>22.5</td>
<td>8</td>
<td>21</td>
</tr>
<tr>
<td>2</td>
<td>preop</td>
<td>ND</td>
<td>ND</td>
<td>21</td>
</tr>
<tr>
<td></td>
<td>3 mos</td>
<td>—</td>
<td>—</td>
<td>15</td>
</tr>
<tr>
<td>3</td>
<td>preop</td>
<td>53</td>
<td>14</td>
<td>26</td>
</tr>
<tr>
<td></td>
<td>3 mos</td>
<td>53</td>
<td>12</td>
<td>22</td>
</tr>
<tr>
<td>4</td>
<td>preop</td>
<td>43</td>
<td>5</td>
<td>19</td>
</tr>
<tr>
<td></td>
<td>3 mos</td>
<td>32</td>
<td>3</td>
<td>14</td>
</tr>
<tr>
<td></td>
<td>12 mos</td>
<td>12</td>
<td>3</td>
<td>14</td>
</tr>
</tbody>
</table>

| mean change in score (± SEM) | 10.8 ± 10.8 | -1.5 ± 1.5 | 5 ± 2.5 | 7 ± 1.6 | 2.9 ± 2.4 | 14.9 ± 5.9 |
| p value† | 0.47 | 0.43 | 0.04 | 0.02 | 0.38 | 0.03 |

* Ratings obtained at the postsurgical evaluations were performed in the stimulation-on state; the maximum possible score for each scale is indicated (0 = no disability). Abbreviations: BP = bipolar; config = configuration; LS = left stimulator; MP = monopolar; ND = not done; RS = right stimulator; SEM = standard error of the mean; — = denotes worsening of scores.
† Calculated using analysis of variance.

**TABLE 3**

<table>
<thead>
<tr>
<th>Parameter Tested</th>
<th>Test Name</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neuropsychological Tests</strong></td>
<td></td>
</tr>
<tr>
<td>attention/working memory</td>
<td>WAIS-III Digit Span</td>
</tr>
<tr>
<td>executive function</td>
<td>Wisconsin Card Sorting Test</td>
</tr>
<tr>
<td>language</td>
<td>Boston Naming Test</td>
</tr>
<tr>
<td>visual memory</td>
<td>Hopkins Verbal Learning Test–Revised</td>
</tr>
<tr>
<td>motor processing speed</td>
<td>Brief Visuospatial Memory Test–Revised</td>
</tr>
<tr>
<td>visuospatial</td>
<td>Trail Making Test–Part A</td>
</tr>
<tr>
<td>affect/mood</td>
<td>Beck Depression Inventory–II</td>
</tr>
</tbody>
</table>

* BDAE = Boston Diagnostic Aphasia Examination; WAIS-III = Wechsler Adult Intelligence Scale–Third Edition.
Case 4

The patient’s foot posture improved, and his neck ROM increased in all directions 1 month following DBS placement and initiation of stimulation. At the 3-month evaluation, he exhibited improved dexterity and handwriting with no hand tremor or posturing. Pain and disability were markedly improved by 3 months. Despite mild residual foot posturing, he was now able to walk 10 blocks easily, which he had not been able to do prior to surgery. All medications for dystonia were discontinued. Examinations at 3 and 12 months showed moderate left laterocollis and shift, but no head tremor.

Quality of Life

Figure 1 represents the PCS scores for the four patients. None of the patients showed a significant difference between their baseline and 3-month scores. Only the patient in Case 2 showed a significant improvement between baseline and the 12-month visit. The MCS scores are shown in Fig. 2. Two patients (Cases 1 and 4) displayed significant improvement between the baseline and 3-month visits, and three of the four patients displayed significant improvements between baseline and 12 months.

The condition of the patient in Case 3 deteriorated between the preoperative and 3-month examinations. These deficits were still present 12 months after surgery. The condition of the patient in Case 4, the remaining three patients experienced improvement in their ratings of affective distress (that is, depression and anxiety) 1 year following surgery. The patient in Case 4 experienced a mild worsening in his anxiety and depression scores, although these were markedly higher at baseline than those in the other patients. The scores were not in the clinically relevant range either at baseline or at follow-up.

Neuropsychological Function

The Z-score profile on the neuropsychological assessment for each patient is presented in Fig. 3. The degree of decline and specific cognitive domains affected varied considerably between patients. On average, at 1 year, all four patients demonstrated a mild, insignificant decline in basic executive functions (~7.5%), although none of these scores fell in the clinically impaired range. Across patients, attention, processing speed, and visuospatial abilities did not significantly change (either better or worse) following surgery.

The conditions of three of the four patients were considered clinically impaired at baseline in the verbal memory domain; two of these conditions worsened further following surgery although only one (Case 3) worsened appreciably. All four patients were considered clinically impaired at baseline regarding visual memory; two of these patients worsened further after surgery. Language skills declined slightly for three patients when tested at follow-up, two of whom fell into the clinically impaired range.

With the exception of the patient in Case 4, the remaining three patients experienced improvement in their ratings of affective distress (that is, depression and anxiety) 1 year following surgery. The patient in Case 4 experienced a mild worsening in his anxiety and depression scores, although these were markedly higher at baseline than those in the other patients. The scores were not in the clinically relevant range either at baseline or at follow-up.

Discussion

In this series of adults with severe idiopathic dystonia refractory to medical therapy, bilateral STN DBS led to improvement of dystonia severity and reduced disability with 3 months that was also reflected in improvement in QOL scores. This was sustained for at least 12 months following surgery in three of the four patients. In these patients, medication doses for dystonia treatment were markedly reduced.

**Table 4**

Percentage of change from presurgery baseline to 1-year postsurgical follow-up for each patient across neuropsychological domains

<table>
<thead>
<tr>
<th>Domain</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>attention/working memory</td>
<td>9.52</td>
<td>7.87</td>
<td>1.79</td>
<td>11.11</td>
</tr>
<tr>
<td>executive function</td>
<td>-28.63</td>
<td>-6.92</td>
<td>-1.01</td>
<td>6.55</td>
</tr>
<tr>
<td>language</td>
<td>-28.79</td>
<td>7.73</td>
<td>-20.36</td>
<td>-9.50</td>
</tr>
<tr>
<td>verbal memory</td>
<td>12.50</td>
<td>4.19</td>
<td>-45.91</td>
<td>-12.73</td>
</tr>
<tr>
<td>visual memory</td>
<td>8.70</td>
<td>-39.36</td>
<td>-65.79</td>
<td>0.00</td>
</tr>
<tr>
<td>motor (dominant hand)</td>
<td>0.00</td>
<td>-32.43</td>
<td>0.00</td>
<td>-76.47</td>
</tr>
<tr>
<td>motor (nondominant hand)</td>
<td>-12.50</td>
<td>50.00</td>
<td>-19.35</td>
<td>67.86</td>
</tr>
<tr>
<td>processing speed</td>
<td>19.64</td>
<td>0.00</td>
<td>-10.00</td>
<td>-22.62</td>
</tr>
<tr>
<td>visuospatial</td>
<td>3.44</td>
<td>19.52</td>
<td>7.97</td>
<td>24.74</td>
</tr>
<tr>
<td>mood/affect</td>
<td>68.44</td>
<td>69.77</td>
<td>16.19</td>
<td>-38.60</td>
</tr>
</tbody>
</table>

* Negative numbers denote a decline from presurgical baseline levels and positive numbers indicate improvement.
Clinical Outcome

Although the BFMDRS and TWSTRS have been validated and used in previous clinical trials to measure the efficacy of dystonia treatments, dystonia is highly variable even within an individual in both the severity of movement and its duration, and both scales may lack sensitivity and may not accurately reflect the extent of clinical or functional improvement noted by the patient or the examiner. In these cases, tremor severity was much improved but was not reflected anywhere in the scoring as the scales do not measure tremor severity.

As electrode placement in the STN was optimized in all patients, the relative lack of benefit in the patients in Cases 1 and 3 may be related to other factors. The latter patient became increasingly depressed over the course of follow-up, and he had fixed orthopedic lower-limb and neck deformities resulting in severely restricted ROM (as demonstrated when he was in a state of general anesthesia) as well as other complicated medical problems that further limited his mobility and reduced his QOL. The lack of improvement in his scores partially reflected fixed defects. Fixed tonic postures or orthopedic changes may be common in adults with refractory dystonia and are an important consideration in the selection of candidates and the timing of surgery. On the other hand, patients with good neck ROM intraoperatively in a state of general anesthesia were most likely to exhibit improvement in mobility. In those patients with fixed deformities while in a state of anesthesia, disability was unchanged following surgery. Of note, even in these refractory patients, there was improvement in pain scores suggesting that DBS may modulate pain independently of its effects on movement. Despite these limitations and the small sample size, effectiveness of the procedure was reflected in significant improvements between patients over time in the TWSTRS motor, disability, and total scores. The lack of differences in scores between on- and off-stimulation states likely reflected an inadequate waiting interval between states as changes in dystonia may occur slowly. In dystonia, the minimal time interval between states necessary to reflect a change in clinical status is not known.
Treatment of idiopathic dystonia with STN DBS

Similarly, the ideal stimulation parameters may vary significantly according to the cause and type of dystonia: the experience with pallidal DBS suggests that higher stimulation amplitudes and pulse widths may be required to control dystonia than those used for Parkinson disease.26 Alternately, there are also cases of profound benefit with low-frequency stimulation (M. Tagliati, personal communication, 2005).

Quality of Life

The STN DBS resulted in minimal improvements in PCS. However, significant improvements on MCS were found in three of the four patients between baseline and 12 months. The MCS scores have been found to relate to changes in mental health, emotional health, and emotional well-being.24 Although there was no significant improvement in the PCS scores, the fact that the MCS scores improved dramatically suggests that other features of the condition aside from the motor aspects contribute to well-being. Even in those patients whose dystonic posture and disability did not improve, pain improved which is likely an important contributor to QOL.

Neuropsychological Function

Overall, there was no marked decline in cognitive function attributed to the procedure. Most patients demonstrated impairments in multiple neuropsychological domains at baseline testing, particularly verbal and visual memory and fine motor speed. Difficulties in fine motor speed at the baseline assessment are not surprising given dystonic involvement of the hands in two of four cases. Deficits in neuropsychological function have previously been identified in patients with primary idiopathic dystonia. Specifically, neuropsychological deficits have been reported in attention and executive function domains;15 however, there are also reports of no executive disturbance in these patients but instead deficits in category fluency and fine motor speed.16 Visuospatial deficits have also been observed in patients with primary idiopathic spasmic torticollis.12 Although none of the patients in this series presented with clinically impaired attention or executive function abilities at the baseline assessment, all patients had below-average attention and three of four had below-average executive function abilities prior to surgery. Patients were taking psychoactive medications to treat their symptoms, which may have contributed to the impaired baseline cognitive function. Importantly, decreased levels of depression and anxiety were observed in most patients.

Conclusions

To our knowledge, this is the first series of patients with idiopathic dystonia treated with STN DBS and evaluated in terms of clinical results, QOL, and neuropsychological outcomes. Although further study is required to identify patient characteristics that predict best surgical outcome, appropriate selection of target and stimulation settings, and long-term effects of the procedure, these results suggest that STN DBS can reduce dystonia severity, disability, and pain, and provide improvement in some aspects of QOL in patients with refractory segmental or generalized dystonia.

Acknowledgment

We thank Dr. David N. Fisman for assistance in statistical analysis and critical review of the manuscript.

References

Executive cognitive deficits in primary dystonia. Mov Disord 18: 539–550, 2003

Accepted January 3, 2007.
The Parkinson’s Disease Research, Education and Clinical Center provided a Seed Grant in support of this research.
Address reprint requests to: Galit Kleiner-Fisman, M.D., Morton and Gloria Shulman Movement Disorders Center, Toronto Western Hospital, University of Toronto, 399 Bathurst Street, McL-7, Toronto, Ontario, M5T 2S8 Canada. email: gkleinerfisman@yahoo.com.