Deep brain stimulation for the treatment of childhood dystonic cerebral palsy

Clinical article

JOSEPH R. KEEN, D.O.,1 ALLISON PRZEKOP, D.O.,2 JOFFRE E. OLAYA, M.D.,3 ALEXANDER ZOUROS, M.D.,1 AND FRANK P. K. HSU, M.D., PH.D.4

Departments of 1Neurosurgery and 2Pediatric Neurology, Loma Linda University Medical Center, Loma Linda; 3Department of Neurosurgery, University of California at Irvine, California; and 4Division of Neurosurgery, Seattle Children’s Hospital, Seattle, Washington

Object. Deep brain stimulation (DBS) for dystonic cerebral palsy (CP) has rarely been reported, and its efficacy, though modest when compared with that for primary dystonia, remains unclear, especially in the pediatric population. The authors present a small series of children with dystonic CP who underwent bilateral pallidal DBS, to evaluate the treatment’s efficacy and safety in the pediatric dystonic CP population.

Methods. The authors conducted a retrospective review of patients (under the age of 18 years) with dystonic CP who had undergone DBS of the bilateral globus pallidus internus between 2010 and 2012. Two of the authors independently assessed outcomes using the Barry-Albright Dystonia Scale (BADS) and the Burke-Fahn-Marsden Dystonia Rating Scale–movement (BFMDRS-M).

Results. Five children were diagnosed with dystonic CP due to insults occurring before the age of 1 year. Mean age at surgery was 11 years (range 8–17 years), and the mean follow-up was 26.6 months (range 2–42 months). The mean target position was 20.6 mm lateral to the midcommissural point. The mean preoperative and postoperative BADS scores were 23.8 ± 4.9 (range 18.5–29.0) and 20.0 ± 5.5 (range 14.5–28.0), respectively, with a mean overall percent improvement of 16.0% (p = 0.14). The mean preoperative and postoperative BFMDRS-M scores were 73.3 ± 26.6 (range 38.5–102.0) and 52.4 ± 21.5 (range 34.0–80.0), respectively, with a mean overall percent improvement of 28.5% (p = 0.10). Those stimulated at least 23 months (4 patients) improved 18.3% (p = 0.14) on the BADS and 30.5% (p = 0.07) on the BFMDRS-M. The percentage improvement per body region yielded conflicting results between rating scales; however, BFMDRS-M scores for speech showed some of the greatest improvements. Two patients required hardware removal (1 complete system, 1 unilateral electrode) within 4 months after implantation because of infections that resolved with antibiotics.

Conclusions. All postoperative dystonia rating scale scores improved with pallidal stimulation, and the greatest improvements occurred in those stimulated the longest. The results were modest but comparable to findings in other similar series. Deep brain stimulation remains a viable treatment option for childhood dystonic CP, although young children may have an increased risk of infection. Of particular note, improvements in the BFMDRS-M subscores for speech were comparable to those for other muscle groups, a finding not previously reported.

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Key Words • deep brain stimulation • dystonia • cerebral palsy • pediatrics • functional neurosurgery

Cerebral palsy (CP) is a heterogeneous group of neuromotor disorders that result from an insult to the developing brain.4,33 It is the leading cause of physical disability in childhood17 with an overall prevalence of approximately 2 cases per 1000 live births.28,33 According to the Executive Committee for the Definition and Classification of Cerebral Palsy, the disorder can be classified into spastic, dystonic, and ataxic groups depending on the dominant type of tone or movement abnormality.4 The spastic type is the most common, accounting for approximately 80% of cases,2 followed by the dystonic group with a prevalence ranging from 3% to 17%1,13–15 which appears to be increasing since the 1990s.13–15 The dystonic group is dominated by “involuntary, uncontrolled, recurring, occasionally stereotyped movements”33 and can be further divided into dystonic and choreoathetotic subtypes.33 The dystonic subtype predominates, composing 70% of a dystonic CP cohort.4
and is commonly characterized by truncal hypotonia, spastic hypertonia of the legs, and dystonic hypertonia of the upper extremities, neck, and face.\textsuperscript{23} Children with dyskinetic CP, especially those with dystonia, tend to have more severe motor impairment and an increased likelihood of learning disability, epilepsy, and visual and hearing impairments.\textsuperscript{15,20}

Well-known etiological factors are hypoxic-ischemic encephalopathy (HIE), infection, and hyperbilirubinemia,\textsuperscript{15} which preferentially affect the basal ganglia and thalamus. These areas are particularly vulnerable given the high metabolic demands of the peri- and neonatal developmental periods.\textsuperscript{3,13,18} In a study to investigate the MRI correlates of CP, the European Cerebral Palsy Study found that basal ganglia and thalamus damage was associated with dystonic CP in 75.6% (34 of 45 cases) of the study cohort.\textsuperscript{3} The posteroventrolateral globus pallidus internus (GPI) has become a well-established target for deep brain stimulation (DBS) in both primary and secondary dystonia.\textsuperscript{1,18,23,24} While primary dystonia has been shown to respond robustly to pallidal stimulation in both adults and children,\textsuperscript{1,2,6,12,21,29,39} secondary dystonia, especially that related to CP, appears to be less responsive.\textsuperscript{1,2,9,21,23,27,30} The inherent difficulty in evaluating the effect of DBS within this heterogeneous population is that the literature is sparse and fraught with case series that include incomplete data, involve the inconsistent use of dystonia rating scales that may not be sensitive to the subtleties of secondary dystonia, and consist of patients of different age groups suffering from various forms of secondary dystonia.\textsuperscript{1,18,23,24}

Recently, however, better controlled and more homogeneous studies have been published.\textsuperscript{16,22,24,27,36} Vidaillhet et al. demonstrated sustained mean improvement of 24.4% in motor symptoms at the 1-year follow-up, with additional improvements in functional disability, pain, and mental health–related quality of life.\textsuperscript{26} For children specifically, Marks et al. observed significant improvements in Burke-Fahn-Marsden Dystonia Rating Scale–movement (BFMDRS-M)\textsuperscript{3} and disability (BFMDRS-D) scores as well as Barry-Albright Dystonia Scale (BADs)\textsuperscript{3} scores at 6 months, and when directly comparing children with DYT1 primary dystonia and those with dystonic CP, the latter group responded more favorably (7% vs 24% reduction in overall BFMDRS-rated motor impairment, respectively).\textsuperscript{1,2,23} Furthermore, Marks et al. advocated for early intervention in the CP group.\textsuperscript{24} A recent meta-analysis of DBS for dyskinetic CP revealed moderate but significant improvement in BFMDRS movement (23.6%) and disability (9.2%) scores as well as a negative correlation between severity of dystonia and clinical outcome.\textsuperscript{18}

The purpose of the present study was to report the results of bilateral pallidal stimulation in a uniform cohort of children with dystonic CP.

**Methods**

**Data Collection**

This was an institutional review board–approved study in which the authors retrospectively identified children diagnosed with dystonic CP who had undergone implantation of DBS electrodes into the bilateral GPI in the period between 2010 and 2012. Dystonic CP was diagnosed by a pediatric neurologist (A.P.) specialized in movement disorders, and surgery was performed by one neurosurgeon (F.P.K.H.) at Loma Linda University Medical Center. Inclusion criteria included disabling and generalized CP-related dystonia refractory to medical and rehabilitative therapies and an age less than 21 years. Videotaped assessments were obtained starting 1 month prior to surgery and at each monthly postoperative appointment. In addition to making a general assessment of the patients’ resting posture, muscle tone, and voluntary and involuntary movements, we asked patients to perform a standard set of tasks, such as reaching for objects, finger and toe tapping, fist clenching, reciting the alphabet and/or counting, and walking (with assistance), to assess the degree of dystonia as well as the provoking factors in the following body regions: eyes, mouth, neck, trunk, extremities, and speech. In a nonblinded fashion, two authors (J.R.K. and A.P.) independently evaluated these sessions and scored performance using the BADs\textsuperscript{3} and the BFMDRS-M.\textsuperscript{7} The BADs scores were calculated monthly for the first 6 months, then every 3 months for the following 6 months, and finally every 6 months thereafter. Only preoperative and latest postoperative BFMDRS-M scores were recorded.

The BADs is a modification of the BFMDRS and is the preferred rating scale for CP-related dystonia.\textsuperscript{4} It rates dystonia severity in 8 body regions, including eyes, mouth, neck, trunk, and extremities, ranging from 0 (no dystonia) to 4 (severe dystonia present more than 50% of the time), with a maximum total score of 32. In contrast, the BFMDRS-M rates dystonia severity as well as provoking factors in 9 body regions, including eyes, mouth, speech and swallowing, neck, trunk, and extremities. Each region is scored using a 4-point scale, and a score that ranges from 0 (no dystonia at rest or with action) to 4 (dystonia present at rest) is generated for “provoking factors,” with intervening scores for dystonia triggered by other body actions. There is also a “severity factor” score that ranges from 0 (no dystonia) to 4 (severe prolonged dystonia). The scores for eyes, mouth, and neck are multiplied by 0.5 to down-weight them before the final calculation, which is the sum of the products of the provoking, severity, and weighting factors. The maximum total score is 120. Rarely a full rating scale could not be completed because of poor patient cooperation or a body action not captured on video. In these instances, the particular body region was assigned the previous month’s score. Although the BFMDRS-M may be limited in evaluating secondary dystonia,\textsuperscript{3,5,10,22,24,27} scores were included to allow for comparison with other published literature.

**Operative Technique**

All patients underwent the implantation of DBS electrodes into the bilateral posteroventrolateral GPI. To prevent intraoperative injury as a result of severe dystonic movements and to ensure the most accurate placement of the DBS electrodes, all implantations were performed with the patient under general anesthesia. Once the patient had been intubated and general anesthesia induced,
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a Cosman-Roberts-Wells head frame was affixed to the patient’s head, followed immediately by a preoperative Stealth-assisted MRI study. The appropriate sequences were selected, and the imaging data set was transferred to a Medtronic StealthStation, where the images were fused. Using the Framelink software, we chose the desired posteroverentralateral GPi target (2–3 mm superolateral to the optic tract and 19–21 mm lateral to the midcommissural line) and trajectory. The patient was secured to the operating table in a semi-sitting position, and the implantation was performed according to established protocol. No microelectrode recording was performed, and all implantable pulse generators (IPGs) were implanted at the same surgery in a subclavicular location. Either bilateral or unilateral IPGs were used and were accompanied by approximately 1 g of vancomycin powder sprinkled into the subcutaneous pocket prior to skin closure.

Postoperative Care

Postoperatively, the patients were briefly monitored in the pediatric intensive care unit and either brain MRI or head CT was performed to confirm electrode placement. Two weeks postoperatively, the DBS systems were activated by the pediatric movement disorder specialist (A.P.), who tested each contact and programmed each electrode according to the clinical response. Patients returned monthly for adjustments, if needed, and videotaped clinical assessments, which were scored using the BDS and BFMDRS-M.

Statistical Analysis

Descriptive statistics were generated using Prism 6 software, and a 1-tailed paired t-test was used to evaluate changes in dystonia rating scales. Statistical significance was set at p < 0.05.

Results

Patient Data

From 2010 through 2012, 5 children, all with an age ≤ 17 years (mean 11 years, range 8–17 years) and diagnosed with dystonic CP, underwent implantation of DBS electrodes into the bilateral posteroverentralateral GPi performed by one neurosurgeon. All patients had been younger than 1 year at the time of brain insult from various causes: HIE (2 patients), intracerebral hemorrhage (1 patient), hyperbilirubinemia (1 patient), and meningitis (1 patient). Magnetic resonance imaging revealed basal ganglia and thalamic damage in 4 of the 5 children, whereas the 1 patient with meningitis had cortical-subcortical damage (Table 1). The mean follow-up was 26.6 months (range 2–42 months).

Lead Location and Stimulation Parameters

Relative to the midcommissural line, the mean left and right lateral electrode positions were 20.6 mm (range 19.7–21.8 mm) and 20.5 mm (range 19.4–21.6 mm), respectively, with the overall lateral position of 20.6 mm (Table 2). This is consistent with the 20 mm reported by Air et al. but more lateral than the values reported in two studies by Marks et al., whose leads were 14.5 and 15.8 mm.

Regarding stimulation parameters, voltage was started at 0.5 V and increased by 0.5-V increments up to 2.0–2.5 V and then by 0.2- or 0.3-V increments up to an overall maximum of 4.0 V, according to the clinical effect. Pulse width and frequency remained stable and similar for the entire cohort at 120 μsec and 160–180 Hz, respectively. The children in this cohort did not tolerate large voltage increases beyond 2.0–2.5 V and fared better at 180 Hz compared with 160 Hz. Ideal parameters achieved the best clinical effect with the least side effects (Table 2). These settings are within the ranges identified in a recent meta-analysis, which reported a mean amplitude of 3.2 ± 1.0 V (range 0.8–6.5 V), mean frequency of 111.8 ± 40.1 Hz (range 30–180 Hz), and mean pulse width of 167.6 ± 56.6 μsec (range 90–450 μsec). No specific parameters have been associated with improvements in dystonia. In contrast to primary dystonia, dystonic CP appears to need higher frequencies, probably because of anatomical derangements due to an initial injury that may have affected targeting accuracy and diffusion of the electrode current.

Barry-Albright Dystonia Scale

All postoperative scores were improved compared with the preoperative scores, with the greatest improvements occurring around 6–9 months and plateauing thereafter (Fig. 1). The mean preoperative score for the entire cohort was 23.8 ± 4.9 (range 18.5–29.0) and postoperatively improved to 20.0 ± 5.5 (range 14.5–28.0), accounting for a mean overall percent improvement of 16.0% (p = 0.14). When selecting for patients stimulated for at least 23 months (4 patients), the mean overall percent improvement increased to 18.5% (p = 0.14; Table 3).

Burke-Fahn-Marsden Dystonia Rating Scale—Movement

All postoperative scores were improved compared with the preoperative scores. The mean preoperative score for the entire cohort was 73.3 ± 26.6 (range 38.5–102.0) and postoperatively improved to 52.4 ± 21.5 (range 34.0–80.0), with a mean overall percent improvement of 28.5% (p = 0.10). When selecting for patients stimulated for at least 23 months (4 patients), the mean overall percent improvement increased to 30.5% (p = 0.07; Table 3).

Improvement per Body Region

In an attempt to evaluate outcome per body region, the mean percentage improvement was calculated for each of the 8 body regions on the BDS and 9 regions on the BFMDRS-M. According to the BDS scores (Table 4), the axial muscle group (neck and trunk) showed greater improvement than the extremities (17.6% vs 16.4%) and the least improvement (10%) in the face group (eyes and mouth). The opposite was discovered for the BFMDRS-M scoring (Table 5), in which the face group showed the greatest improvement (59.6%), followed in descending order by the bilateral extremities (21.0%), speech (19.2%), and axial (13.4%) groups. There was no overall significant difference in the laterality of motor effects. However, the patient whose right electrode was removed showed less...
improvement in the contralateral side according to BFM-DRS-M scoring (left 0% vs right 14.2%).

Complications

Two patients developed infections, which required removal of the right electrode and the IPG in Case 4 and the entire DBS system in Case 5. The patient in Case 4 had early cerebritis that initially resolved with intravenous antibiotics, but he later developed a wound infection that tracked along the subcutaneous portion of the DBS electrode. The patient in Case 5 had a persistent subcutaneous fluid collection around the IPG that progressed to involve the right frontal scalp incision. Both patients were 11 years old or younger, and the infections occurred within 4 months of implantation despite the intraoperative use of vancomycin powder and routine preoperative intravenous antibiotics. Wound cultures were positive for Staphylococcus aureus (Case 4) and S hominis (Case 5); once the necessary hardware was removed and appropriate antibiotics were administered, the infections resolved without further complications. All removed electrodes have not been reimplanted because of parent refusal.

Illustrative Case

This 11-year-old female (Case 1) had dystonic CP caused by hyperbilirubinemia as an infant. She showed the greatest degree of improvement (47.5%) among all the patients. Preoperatively, when she was 8 years old, her dystonia was so severe that the exam had to be done while she was supine (Video 1). Postoperatively, at 11 months (Video 2) and 42 months (Video 3), she had marked improvement in the ability to sit upright unassisted, better control of her upper extremities, and improved verbal communication.

**Video 1.** Case 1. Video clip showing preoperative evaluation. Her baseline dystonia was so severe that the examination had to be conducted while the patients was supine. Copyright Joseph R. Keen. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

**Video 2.** Case 1. Video clip showing evaluation at 11 months postoperatively. Note the improved ability to sit upright without assistance and the smoother movements of her upper extremities. Copyright Joseph R. Keen. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

**Video 3.** Video clip showing evaluation at 42 months postoperatively. She has maintained her ability to sit upright without assistance and has improved in her ability to reach for and hold objects. She also demonstrates subtle improvement in verbal communication. Copyright Joseph R. Keen. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

Discussion

Dyskinetic CP accounts for up to 17% of the CP cases and, according to newer population studies, is increasing. Dyskinesia is more prevalent than the choreoathetotic subtype and commonly manifests as dystonic hypertonia of the upper extremities, neck, and face with concomitant

<table>
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<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age at Disease Onset (mos)</th>
<th>Age at Surgery (yrs)</th>
<th>Etiology</th>
<th>Abnormalities on T2W MRI</th>
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<tr>
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<td>bilat GPi</td>
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<tr>
<td>2</td>
<td>M</td>
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<td>M</td>
<td>10</td>
<td>14</td>
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<td>corpus callosum w/ bifrontal atrophy</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>NB</td>
<td>7</td>
<td>HIE</td>
<td>bilat thalamus, posterior lentiform nuclei</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>7</td>
<td>11</td>
<td>ICH</td>
<td>bilat GPi, thalamus w/ periatral leukomalacia</td>
</tr>
</tbody>
</table>

* ICH = intracerebral hemorrhage; NB = newborn; T2W = T2-weighted.

**TABLE 2: Lead location and stimulation parameters**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Lt Lateral (mm)</th>
<th>Lt AP (mm)</th>
<th>Lt Depth (mm)</th>
<th>Rt Lateral (mm)</th>
<th>Rt AP (mm)</th>
<th>Rt Depth (mm)</th>
<th>Postop Imaging</th>
<th>Active Contact†</th>
<th>Voltage (V)†</th>
<th>Pulse Width (μsec)†</th>
<th>Frequency (Hz)†</th>
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<td>180/180</td>
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<td>4.7</td>
<td>6.9</td>
<td>21.6</td>
<td>4.7</td>
<td>6.9</td>
<td>MRI</td>
<td>1-, 2/-9-, 10-</td>
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<td>120/120</td>
<td>180/180</td>
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<td>19.4</td>
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<td>1-, 2/-9,-, 10-</td>
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<td>21.8</td>
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<td>NA</td>
<td>19.9</td>
<td>NA</td>
<td>NA</td>
<td>CT</td>
<td>1, 2/-NA</td>
<td>3.0/NA</td>
<td>120/NA</td>
<td>180/NA</td>
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<td>160/160</td>
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</table>

* The Activa RC neurostimulator system was used in all cases. Lead location was calculated in reference to the midpoint of the anterior commissure–posterior commissure (AC-PC) line. Stimulation parameters are based on the most current settings at the last follow-up, with the exception of the patient in Case 5 whose settings were those recorded just prior to DBS removal. AP = anteroposterior; NA = not applicable. † Data on the left side of the virgule (/) refer to the left electrode; data on the right, to the right electrode. ‡ Right electrode removed due to cerebritis. § Entire DBS system removed due to persistent wound infection.
truncal hypotonia and spastic hypertonia of the legs. Compared with the more common spastic CP, the dyskinetic forms have greater motor impairment and functional disability, resulting in significant long-term morbidity and caretaker burden. And although this dystonic group has responded only modestly to DBS, even subtle improvements can significantly impact quality of life and caretaking, thus underscoring the importance of strict adherence to established classification schemes to identify patients who will benefit most.

The effect of DBS on dystonic CP has been difficult to ascertain, especially within the pediatric population, because reports are scarce and existing studies have small sample sizes and a heterogeneous group of patients. A recent meta-analysis revealed that only 61 patients with dyskinetic CP had adequate data for inclusion in a response analysis after 6 months of stimulation. In the first high-quality, multicenter, prospective study of bilateral pallidal stimulation in 13 adults with childhood-onset dystonia-choroeeathetosis CP, Vidailhet et al. found at the 1-year follow-up significant improvement in motor symptoms, as well as in functional disability, pain, and mental health–related quality of life. Subsequent series have demonstrated similar moderate levels of improvement (Table 6), and a recent meta-analysis of adult and pediatric cases revealed a 23.6% (p < 0.001) improvement in BFMDRS-M scores and 9.2% (p < 0.001) improvement in BFMDRS-D scores. However, the more severe the dystonia before DBS, the smaller the postoperative percentage of improvement in BFMDRS-M scores (p < 0.05). In a series of 14 patients with pediatric-onset disease, Marks et al. observed modest but statistically significant improvements in both BFMDRS-M (25.5%, p < 0.004) and BFMDRS-D (8.9%, p < 0.027) scores as well as in BADS scores (11.7%, p < 0.029) at 6 months and found a greater effect for those treated before skeletal maturity, as patients younger than 16 years old scored better (19.5%, p < 0.024) than those 16 years and older (1.9%, p < 0.589). When Marks et al. compared dystonic CP to DYT1 primary dystonia in children aged 7–15 years old, the dystonic CP cohort showed a 24.3% reduction in BFMDRS-M scoring compared to 6.6% in the DYT1 group. This discrepancy was not observed on BADS scoring, as both groups improved similarly (CP 19.3% vs DYT1 27.6%), and neither group showed improvement on the BFMDRS-D scores. Marks and colleagues, as well as others, have advocated for early intervention in both groups before musculoskeletal contractures limit the therapeutic response. Olaya et al. recently reported their experience with DBS in children and young adults with secondary dystonia. The patients had a more modest improvement of 9% in the BADS scores and 9.5% in the BFMDRS scores at a mean follow-up of 3.8 months, but most patients in the study had undergone DBS after achieving skeletal maturity.

Though small, our series is similar in size to several other studies evaluating DBS in the pediatric population with dystonic CP and revealed commensurate levels of improvement on both BFMDRS-M and BADS scores. Although not statistically significant, improvement in scores occurred in the entire cohort (5 patients; 16.0% on BADS and 28.5% on BFMDRS-M scoring), and when selecting for patients stimulated for at least 23 months (4 patients), scores improved 18.3% and 30.5%, respectively. The results were durable over a mean long-term follow-up of 26.6 months.

**Improvement per Muscle Group**

The mean percentage of overall improvement by muscle group yielded conflicting results. The BADS revealed that the axial muscle group (neck and trunk) showed greater improvement than the extremities and the least improvement in the face group (eyes and mouth), whereas the BFMDRS-M revealed the greatest improve-

<table>
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<th>Case No.</th>
<th>Age at Surgery (yrs)</th>
<th>Preop</th>
<th>Postop</th>
<th>% Improvement</th>
<th>Preop</th>
<th>Postop</th>
<th>% Improvement</th>
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* FU = follow-up.
ments in the face group, followed in descending order by the extremities, speech, and axial group. The literature is sparse and unclear on which muscle groups show the greatest improvements. Vidailhet et al. found that axial and limb groups fared better than face and speech groups, whereas Marks et al. found that extremities improved more than the axial group. These discrepancies may simply reflect small sample sizes, but they may also underscore the fact that the tools used for measuring improvement in the dystonic CP population may not capture subtle changes within each muscle group. There were no significant differences in the effect on laterality, except in the patient in Case 4 who demonstrated less improvement on the BFMDRS-M in the upper extremity contralateral to the removed electrode. This patient also exhibited speech improvements, which suggests that unilateral stimulation can still be efficacious. These differences were not detected on BADS scoring.

Unexpectedly, the BFMDRS-M scores for speech demonstrated improvements comparable to those for other muscle groups (Table 5). This manifested as increased nonverbal vocalization (Cases 2 and 4), diversity of sounds (Case 3), and ability to count and verbalize (Cases 1–4), albeit subtle with severe dysarthria. Significant speech improvements have not been reported in the DBS-dystonia literature, though it has been an area of focus in the Parkinson’s literature. Studies have been equivocal regarding subthalamic nucleus stimulation in Parkinson’s disease (PD), as some have demonstrated improvements in certain aspects of speech and others have shown deterioration, especially regarding dysarthria. The trend is toward worsened speech in subthalamic nucleus DBS for PD; however, GPI DBS in a small series of 7 patients with PD showed improved speech overall. While studies are inconclusive, it appears that the basal ganglia is linked to speech and linguistic processing. It has been postulated that stimulation of neighboring corticobulbar pathways for laryngeal motor control may account for dysarthria, and pallidofugal or cerebellothalamic fibers may account for speech alterations. Basal ganglionic correlates to speech and language should continue to be studied and reported.

Dystonia Rating Scales

Although the degree of improvement in our cohort was considerably less on BFMDRS-M scoring than that reported in the primary dystonia literature, both BADS and BFMDRS-M scores compared well with the scores in other published pediatric dystonic CP studies (Table 6). Because primary dystonia is rarely evaluated using the BADS, it is unclear if the same discrepancy would exist between the dystonic CP and primary dystonia groups. Furthermore, the modest degree of improvement in scores in the present study suggests that neither scale is appropriately tailored for dystonic CP patients, who may have coexisting mixed-motor symptoms, and does not reflect the subtle improvements in quality of life and caretaking. For example, the 22%–48% range of improvement in the patients stimulated for at least 23 months translates into an improved ability to sit upright without assistance, better gross and fine motor control that allows the use of assistive devices, improved speech and/or verbalizations, and an ability to ambulate with assistance.

A comprehensive scoring system that not only takes into account the mixed motor components, but also cap-

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**TABLE 4: BADS subscores per body region**

<table>
<thead>
<tr>
<th>Muscle Group</th>
<th>Mean Preop Score (range)</th>
<th>Mean Postop Score (range)</th>
<th>% Improvement</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>axial (neck/trunk)</td>
<td>5.4 ± 2.4 (3–8)</td>
<td>4.45 ± 2.5 (2.5–8)</td>
<td>17.6</td>
<td>0.28</td>
</tr>
<tr>
<td>face (eyes/mouth)</td>
<td>4.0 ± 2.5 (3–5)</td>
<td>3.6 ± 0.7 (3–4.5)</td>
<td>10.0</td>
<td>0.24</td>
</tr>
<tr>
<td>bilat extremities</td>
<td>14.3 ± 1.9 (12–16)</td>
<td>11.95 ± 2.7 (8.5–15.5)</td>
<td>16.4</td>
<td>0.35</td>
</tr>
<tr>
<td>lt extremities</td>
<td>7.4 ± 0.8 (6.5–8)</td>
<td>6.2 ± 1.2 (4.5–7.75)</td>
<td>16.2</td>
<td>0.05</td>
</tr>
<tr>
<td>rt extremities</td>
<td>6.9 ± 1.2 (5.5–8)</td>
<td>5.75 ± 1.5 (4–7.75)</td>
<td>16.7</td>
<td>0.11</td>
</tr>
</tbody>
</table>

* Values expressed as the mean ± standard deviation, unless indicated otherwise.

**TABLE 5: BFMDRS-M subscores per body region**

<table>
<thead>
<tr>
<th>Muscle Group</th>
<th>Mean Preop Score (range)</th>
<th>Mean Postop Score (range)</th>
<th>% Improvement</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>axial (neck/trunk)</td>
<td>17.2 ± 6.4 (7.5–24)</td>
<td>11.9 ± 6.8 (6.5–20.5)</td>
<td>13.4</td>
<td>0.12</td>
</tr>
<tr>
<td>face (eyes/mouth)</td>
<td>4.7 ± 5.12 (0–10.5)</td>
<td>1.9 ± 2.7 (0–6.5)</td>
<td>59.6</td>
<td>0.16</td>
</tr>
<tr>
<td>speech/swallow</td>
<td>9.4 ± 4.3 (2–12)</td>
<td>7.6 ± 3.8 (2–12)</td>
<td>19.2</td>
<td>0.25</td>
</tr>
<tr>
<td>bilat extremities</td>
<td>42.0 ± 13.7 (25–56)</td>
<td>33.2 ± 13.1 (25–51)</td>
<td>21.0</td>
<td>0.16</td>
</tr>
<tr>
<td>lt extremities</td>
<td>22.2 ± 5.7 (15–28)</td>
<td>18.6 ± 13.1 (14–24)</td>
<td>16.2</td>
<td>0.18</td>
</tr>
<tr>
<td>rt extremities</td>
<td>19.8 ± 8.0 (10–28)</td>
<td>14.6 ± 7.3 (8–24)</td>
<td>26.3</td>
<td>0.16</td>
</tr>
</tbody>
</table>
Deep brain stimulation for childhood dystonic cerebral palsy

**TABLE 6: Literature review of studies on dystonic CP**

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Cases</th>
<th>Mean Age at Surgery (yrs)</th>
<th>Rating Scale(s) Used*</th>
<th>% Improvement</th>
<th>Average FU (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Katsakiori et al., 2009†</td>
<td>4</td>
<td>36.3</td>
<td>BFMDRS-M</td>
<td>24</td>
<td>19.8</td>
</tr>
<tr>
<td>Vidailhet et al., 2009†</td>
<td>13</td>
<td>33.0</td>
<td>BFMDRS-M</td>
<td>24</td>
<td>12</td>
</tr>
<tr>
<td>Air et al., 2011</td>
<td>6</td>
<td>13.8</td>
<td>BFMDRS-M, BADS</td>
<td>10, 22</td>
<td>12.8</td>
</tr>
<tr>
<td>Marks et al., 2011</td>
<td>8‡</td>
<td>10.7‡</td>
<td>BFMDRS-M, BADS‡</td>
<td>37.8, 19.5‡</td>
<td>6‡</td>
</tr>
<tr>
<td>Marks et al., 2013</td>
<td>9</td>
<td>11.0</td>
<td>BFMDRS-M, BADS§</td>
<td>9.0, 1.4§</td>
<td>6§</td>
</tr>
<tr>
<td>Gimeno et al., 2012</td>
<td>6</td>
<td>10.2</td>
<td>BFMDRS-M</td>
<td>5.9</td>
<td>12</td>
</tr>
<tr>
<td>Olaya et al., 2013</td>
<td>9</td>
<td>15.1</td>
<td>BFMDRS-M, BADS</td>
<td>9.3, 9</td>
<td>3.8</td>
</tr>
<tr>
<td>current study</td>
<td>5</td>
<td>11.0</td>
<td>BFMDRS-M, BADS</td>
<td>28.5, 18.3</td>
<td>26.6</td>
</tr>
</tbody>
</table>

* The BFMDRS-M and BADS were chosen for best comparison with our study. The BFMDRS-D (disability) scores were not used for comparison, as they were often not reported or did not demonstrate meaningful change.
† Study includes adults with childhood-onset dystonic CP. Pediatric dystonic CP cases were extracted from the reported data.
‡ Refers to patients younger than 16 years old.
§ Refers to patients 16 years of age and older.

Infections

Implantation of DBS electrodes in children appears to be safe, as there were no major complications. However, our infection rate was 40% (2 of 5 patients). Infection rates have been reported to range from 5%–33%, with higher rates reported in children.23 Air et al.,1 who has published the largest series of DBS implantations in children, reported a 13% rate of hardware infection on a per patient basis and a 57% rate for those younger than 10 years old. Marks et al. reported a 17% infection rate for children with dystonic CP,23 and Haridas et al. reported a 14% infection rate for children with primary dystonia.12

Children with dystonic CP, who are severely disabled and often wheelchair bound, may have a high risk of perioperative infectious complications. The high infection rates may simply be the result of small sample sizes, but it is imperative to identify factors that make this pediatric population more susceptible and establish better infection prophylaxis protocols. Olaya et al. reported no infections in their cohort of 9 patients with secondary dystonia who had been treated with 72 hours of intravenous vancomycin and ceftazidime followed by 2 weeks of oral dicloxacillin.27

Study Limitations

Although this series consists of a uniform cohort with long-term follow-up and is comparable in size to other series, the small sample size limits the power of the study and makes statistical analysis less meaningful. Additionally, those scoring the dystonia rating scales were not blinded, which has been shown to artificially overstate the treatment effect of DBS.34

Because DBS is expensive, a cost analysis should be conducted to determine if the modest benefits warrant its use in this population, as there are no published studies to our knowledge. For adult dystonia, a cost analysis revealed an overall gain of 0.94 quality-adjusted life years (QALYs) with a cost of €33,980 per QALY. While the cost was at the higher end of expenditure per patient per QALY in Great Britain, the study revealed that the “willingness-to-pay” value far outweighed its cost.38 We speculate that DBS would compare favorably to other therapies for dystonia, as even subtle improvements translate into highly valuable improvements in function and ease of caretaking.

Conclusions

Deep brain stimulation for dystonic CP in the pediatric population has rarely been reported, and its efficacy, though modest as compared with that for primary dystonia, remains unclear. We present a uniform population of pediatric patients with dystonic CP who underwent bilateral pallidal stimulation before both the age of 18 years and the onset of musculoskeletal deformities. Outcomes according to the BADS and BFMDRS were modest but...
comparable to other published secondary dystonia data and were durable over the long-term follow-up. Although muscle subgroup scoring yielded conflicting results, speech showed some of the greatest improvements and may be particularly affected by pallidal stimulation in this population. Deep brain stimulation should remain an early treatment option for children with dystonic CP; however, a more comprehensive rating system is needed to evaluate motor and nonmotor functional domains.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following: Conception and design: Keen, Przekop, Olaya, Hsu. Acquisition of data: Keen, Przekop. Analysis and interpretation of data: all authors. Drafting the article: Keen. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Keen. Administrative/technical/material support: Zouros. Study supervision: Keen, Przekop.

References

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Current affiliation for Dr. Oluya: Children’s Hospital of Orange County and Department of Neurological Surgery, University of California, Irvine, Orange, California.

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Supplemental online information:


Address correspondence to: Joseph R. Keen, D.O., Department of Neurosurgery, Loma Linda University Medical Center, 11234 Anderson St., Rm. 2562-B, Loma Linda, CA 92354. email: jkeen@llu.edu.