**ABSTRACT:** Deep brain stimulation of the globus pallidus internus (GPI DBS) is effective in the treatment of primary segmental and generalized dystonia. Although limb, neck, or truncal dystonia are markedly improved, orofacial dystonia is ameliorated to a lesser extent. Nevertheless, several case reports and small cohort studies have described favorable short-term results of GPI DBS in patients with severe Meige syndrome. Here, we extend this preliminary experience by reporting long-term outcome in a multicenter case series, following 12 patients (6 women, 6 men) with Meige syndrome for up to 78 months after bilateral GPI DBS. We retrospectively assessed dystonia severity based on preoperative and postoperative video documentation. Mean age of patients at surgery was 64.5 years, and mean disease duration 8.3 ± 4.4 years. Dystonia severity as assessed by the Burke–Fahn–Marsden Dystonia Rating Scale showed a mean improvement of 45% at short-term follow-up (4.4 ± 1.5 months; P < 0.001) and of 53% at long-term follow-up (38.8 ± 21.7 months; P < 0.001). Subscores for eyes were improved by 38% (P = 0.004) and 47% (P < 0.001), for mouth by 50% (P < 0.001) and 56% (P < 0.001), and for speech/swallowing by 44% (P = 0.058) and 64% (P = 0.004). Mean improvements were 25% (P = 0.006) and 38% (P < 0.001) on the Blepharospasm Movement Scale and 44% (P < 0.001) and 49% (P < 0.001) on the Abnormal Involuntary Movement Scale. This series, which is the first to demonstrate a long-term follow-up in a large number of patients, shows that GPI DBS is a safe and highly effective therapy for Meige syndrome. The benefit is preserved for up to 6 years. ©2011 Movement Disorder Society

**Key Words:** deep brain stimulation; globus pallidus; dystonia; Meige syndrome

Meige syndrome is a manifestation of segmental dystonia comprising blepharospasm and dystonia of lower facial, jaw, and neck muscles. In idiopathic forms, symptoms usually start in the fifth or sixth decade of life with a twofold higher incidence in women. Blepharospasm is the most frequent initial complaint of patients as by time dystonic movement patterns may spread. Apart from primary forms, Meige syndrome has been described as a tardive movement disorder after neuroleptic treatment. Similar to other idiopathic dystonias, systemic treatment options in Meige syndrome include...
anticholinergics, benzodiazepines, and tetrabenazine, but are mostly of low clinical efficacy and limited by side effects. Selective chemical denervation of the affected muscles by botulinum toxin injections can be effective, but is often complicated by the large number and variability of muscles involved as well as the complexity of the dystonic movements.

Deep brain stimulation (DBS) of the globus pallidus internus (GPI) has become a first choice treatment for drug refractory, primary, segmental, or generalized dystonias. In these studies, the improvement of orofacial dystonia including speech and swallowing problems was typically inferior to that of limb, neck, or truncal dystonia. Nevertheless, several case reports and small cohort studies have described favorable short-term results of pallidal DBS in patients with severe Meige syndrome.

The aim of this study was to evaluate long-term outcome of a multicenter case series, following 12 patients with Meige syndrome, with bilateral GPI-DBS for up to 78 months.

**Patients and Methods**

**Patients**

Twelve patients (6 women and 6 men) were recruited from three German movement disorders centers (Berlin, 4 patients; Mannheim/Hannover, 4 patients; and Kiel, 4 patients). Mean age at surgery was 64.5 ± 4.4 years; mean disease duration was 8.6 ± 4.4 years. A detailed clinical overview of the patient data is given in Table 1. All patients provided written informed consent for the analysis of anonymized outcome data. Short-term follow-up of patient 7 was reported earlier. Idiopathic Meige syndrome was diagnosed by movement disorder neurologists when clinical criteria for blepharospasm combined with dystonia of the lower facial muscles were fulfilled. Involvement of cervical muscles was present in all but one patients, but did not dominate the clinical picture. All other topographical areas remained unaffected. Secondary causes were excluded by standard cranial magnetic resonance imaging (MRI) scans.

Patients included in this study did not achieve satisfactory benefit after treatment with various drugs and local botulinum toxin injections (see Table 1). Moreover, patients deteriorated in activities of daily living and showed social withdrawal during the course of their disease as they suffered from apparent hyperkinesia of facial muscles and functional blindness secondary to severe blepharospasm. Cognitive impairment, dementia, severe psychiatric diseases as well as higher surgical risks due to comorbidities were exclusion criteria.

Patient 3 had a history of neuroleptic treatment, which had been discontinued many years before the onset of dystonic movements. Nevertheless this raises the remote possibility of a tardive etiology. The same patient was bilaterally implanted in the GPI in another center in 2002 and was admitted to the Kiel department in 2005 as chronic DBS was ineffective. Cranial MRI scans revealed bilateral electrode malpositioning. Electrodes were reimplanted in 2006 and all data presented here refer to the second implantation.

**Surgery and Postoperative Determination of Active Stimulation Contacts**

Details of the surgical procedures have been described elsewhere. Stereotactic procedures were performed under general anesthesia with propofol and remifentanil (Kiel) or under local anesthesia (Mannheim/Hannover and Berlin).

The initial stereotactic coordinates to target the posteroverentral lateral GPI were 20 to 22 mm lateral to, 3 mm anterior to, and 2 to 4 mm below the midcommissural point as determined by MRI-stereotactic (Kiel and Berlin) or CT-stereotactic (Mannheim/Hannover) planning adjusted to the individual patient’s anatomy.

Intraoperatively, neuronal recordings were obtained using parallel multichannel (Kiel), or sequential single channel (Mannheim/Hannover) microelectrode recordings (Medtronic, Minneapolis, MN) or via a tetrode (Berlin) (Thomas Recording, Giessen, Germany). Starting at 10 to 15 mm above the intended target, the border between the external (GPe) and internal part of the pallidum (GPI) was delineated by reduced spontaneous neuronal activity and background noise and the occasional presence of border cells. The ventral border of the Gpi was again determined by reduction of background noise.

Sensorimotor-associated neurons were identified by passive limb movements (Mannheim/Hannover). Macrostimulation was used to verify adequate distance to the inner capsule (all centers), and to evoke phosphenes to identify the optic tract (Mannheim/Hannover; Berlin).

Quadripolar electrodes (model 3389 or 3387, Medtronic) were implanted in a way that at least two contacts were within the sensorimotor GPI. Implantable pulse generators (Kineta or bilateral Soletra, Medtronic) were implanted in a second surgical procedure following directly the first procedure (Kiel) or a few days later (Mannheim/Hannover; Berlin).

Postoperative stereotactic CT or MRI scans were used to verify electrode position within the GPI and to exclude asymptomatic cerebral hemorrhage. The location of the active electrode contacts were derived from postoperative imaging data following anatomical normalization into the standard Montreal Neurological Institute (MNI) stereotactic space. In patients from the Hannover center, postoperative CT scans were transformed to MNI-stereotactic space using point-matching coregistration based on anatomical landmarks. In bipolar stimulation settings, the center of the cathodal contact was assessed to account for similar field distributions with respect to monopolar
### TABLE 1. Patients’ individual demographic profiles, medical history, adverse events, and implanted devices

<table>
<thead>
<tr>
<th>Patient no. (sex)</th>
<th>Age at surgery (yr)</th>
<th>Disease duration (yr)</th>
<th>Insufficient medical trials</th>
<th>Medication preoperative</th>
<th>Medication at FU2</th>
<th>Adverse events</th>
<th>Implanted device</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (F) 72 12</td>
<td>Trihexyphenidyl, clonazepam, tetrabenazine, botulinum toxin</td>
<td>Amtriptyline, clonazepam, tetrabenazine, botulinum toxin</td>
<td>Botulinum toxin at irregular intervals</td>
<td>Local infection</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 (M) 65 6</td>
<td>Levodopa, lorazepam, baclofen, trihexyphenidyl, zolpidem</td>
<td>Zolpidem</td>
<td>Zolpidem</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 (F) 68 12</td>
<td>Trihexyphenidyl, tetrabenazine, clozapin, tiapride, clonazepam, botulinum toxin</td>
<td>Amtriptyline, diazepam</td>
<td>Mirtazapine, diazepam</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 (M) 66 4</td>
<td>Trihexyphenidyl, tetrabenazine, botulinum toxin</td>
<td>None</td>
<td>None</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 (M) 62 5</td>
<td>Botulinum toxin, trihexyphenidyl</td>
<td>Trihexyphenidyl</td>
<td>Trihexyphenidyl, sulpirid</td>
<td>None</td>
<td>Medtronic 3387 bilateral IPG: Soletra bilateral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 (M) 71 4</td>
<td>Trihexyphenidyl</td>
<td>Diazepam</td>
<td>None</td>
<td>None</td>
<td>Medtronic 3387 bilateral IPG: Soletra bilateral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7 (F) 60 5</td>
<td>Botulinum toxin, tiapride, trihexyphenidyl, zolpidem</td>
<td>Amtriptyline, zolpidem</td>
<td>Amtriptyline, zolpidem</td>
<td>None</td>
<td>Medtronic 3387 bilateral IPG: Soletra bilateral</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8 (M) 64 18</td>
<td>Botulinum toxin, trihexyphenidyl</td>
<td>Botulinum toxin periorbital</td>
<td>Botulinum toxin periorbital</td>
<td>None</td>
<td>Medtronic 3387 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9 (F) 61 7</td>
<td>Tiapride, trihexyphenidyl, botulinum toxin, levodopa</td>
<td>Trihexyphenidyl</td>
<td>None</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10 (M) 62 13</td>
<td>Trihexyphenidyl, pimozid, tetrabenazine, botulinum toxin A + B, lorazepam</td>
<td>Escitalopram, lorazepam</td>
<td>Lorazepam</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11 (F) 66 10</td>
<td>Botulinum toxin A + B, clozapin, valproic acid, clonazepam, levodopa</td>
<td>Clonazepam, mirtazapin</td>
<td>Mirtazapin</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12 (F) 57 7</td>
<td>Trihexyphenidyl, botulinum toxin, biperiden</td>
<td>Biperiden, reboxetine</td>
<td>Doxepin</td>
<td>None</td>
<td>Medtronic 3389 bilateral IPG: Kinetra</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
stimulation. The mean and the standard deviation of the stereotactic coordinates of the active contacts at the final visit were calculated separately for each side. Localizations of contacts were superimposed on the probabilistic Harvard-Oxford structural subcortical atlas to assess their position with respect to nuclear boundaries of the target structure (Fig. 1).

**Postoperative Programming of DBS**

Postoperative programming of the DBS system was done 4–5 days after surgery (center Kiel) or 1 day after the staged implantation of the pulse generator (centers Hannover/Mannheim and Berlin). Each of the four contacts was separately tested in a unipolar manner for both hemispheres (case served as anode; constant voltage, 90–210 μs pulse width, 130 Hz frequency). Acute clinical improvement and voltage-limiting side effects were assessed. For chronic stimulation, the lowest contact within the GPi with acceptable thresholds for side effects (typically ~3–4 V) was chosen for unipolar stimulation and programmed with a voltage at 10–20% below the threshold for side effects. In the Hannover/Mannheim/Berlin group, the lowest contact was located ventrally to the mean extent of the globus pallidus. In the Kiel group, the contact was located more rostrally to the mean extent of the globus pallidus.

**TABLE 2. Patients’ individual follow-up, clinical scores and stimulation parameters (C = case)**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Follow up (mo)</th>
<th>BFMDRS base/FU1/FU2</th>
<th>AMS base/FU1/FU2</th>
<th>BMS base/FU1/FU2</th>
<th>Stimulation parameters at final FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3/51</td>
<td>22/14/10</td>
<td>12/9/6</td>
<td>10/8/6</td>
<td>GPi right: 3 + 1 – 4.8 V, 60 μs/180 Hz; GPi left: 7 + 5 – 4.0 V, 90 μs/180 Hz</td>
</tr>
<tr>
<td>2</td>
<td>5/41</td>
<td>22/5/2.5</td>
<td>14/5/3</td>
<td>10/4/2</td>
<td>GPi right: C + 1 – 4.1 V, 120 μs/210 Hz; GPi left: C + 4 – 2.5 V, 120 μs/210 Hz</td>
</tr>
<tr>
<td>3</td>
<td>6/33</td>
<td>20/9/8</td>
<td>14/4/4</td>
<td>8/4/4</td>
<td>GPi right: C + 0 – 4.0 V, 90 μs/160 Hz; GPi left: C + 4 – 4.5 V, 90 μs/160 Hz</td>
</tr>
<tr>
<td>4</td>
<td>~/16</td>
<td>15~/7</td>
<td>14~/9</td>
<td>10~/6</td>
<td>GPi right: C + 0 – 2.7 V, 90 μs/210 Hz; GPi left: C + 4 – 2.2 V, 90 μs/210 Hz</td>
</tr>
<tr>
<td>5</td>
<td>6/72</td>
<td>26/18/16</td>
<td>13/10/8</td>
<td>10/9/8</td>
<td>GPi right: 3 + 0 – 2.4 V, 210 μs/130 Hz; GPi left: 3 + 0 – 2.3 V, 210 μs/130 Hz</td>
</tr>
<tr>
<td>6</td>
<td>6~/~</td>
<td>26/14~/~</td>
<td>16/10/~</td>
<td>10/8/~</td>
<td>GPi right: 2 + 1 – 3.3 V, 210 μs/130 Hz; GPi left: 6 + 5 – 3.3 V, 210 μs/130 Hz</td>
</tr>
<tr>
<td>7</td>
<td>~/24</td>
<td>20~/~</td>
<td>16~/11</td>
<td>12~/9</td>
<td>GPi right: 2 + 1 – 3.9 V, 210 μs/130 Hz; GPi left: 2 + 1 – 4.7 V, 210 μs/130 Hz</td>
</tr>
<tr>
<td>8</td>
<td>~/28</td>
<td>18~/~</td>
<td>8~/3</td>
<td>8~/5</td>
<td>GPi right: 2 + 1 – 4.9 V, 210 μs/130 Hz; GPi left: 6 + 5 – 4.9 V, 210 μs/130 Hz</td>
</tr>
<tr>
<td>9</td>
<td>3/12</td>
<td>22/9/14</td>
<td>13/4/8</td>
<td>8/7/8</td>
<td>GPi right: C + 0 – 1 – 2.6 V, 90 μs/180 Hz; GPi left: C + 4 – 5 – 2.6 V, 90 μs/180 Hz</td>
</tr>
<tr>
<td>10</td>
<td>3/24</td>
<td>22/16/11</td>
<td>14/10/5</td>
<td>12/10/6</td>
<td>GPi right: 0 + 1 + 2 – 3 – 4.0 V, 150 μs/210 Hz; GPi left: 4 + 5 + 6 – 7 – 4.0 V, 150 μs/210 Hz</td>
</tr>
<tr>
<td>11</td>
<td>3/48</td>
<td>20/14/14</td>
<td>10/7/7</td>
<td>8/7/8</td>
<td>GPi right: C + 0 – 4.0 V, 180 μs/130 Hz; GPi left: C + 4 – 4.0 V, 180 μs/130 Hz</td>
</tr>
<tr>
<td>12</td>
<td>~/78</td>
<td>24~/~</td>
<td>12~/~</td>
<td>8~/~2</td>
<td>GPi right: C + 1 – 2 – 5.0 V, 90 μs/235 Hz; GPi left: C + 5 – 6 – 5.0 V, 90 μs/235 Hz</td>
</tr>
</tbody>
</table>
Mannheim center, the initial setting used a bipolar stimulation mode of two adjacent contacts. Follow-up visits were typically performed around 4–6 weeks, 3 and 6 months after initial programming, and thereafter according to individual patient’s requirements. Further adjustments of stimulation parameters according to the individual patient’s response to stimulation were done at follow-up visits (Table 2).

**Data Acquisition**

A movement disorders neurologist (RR) not involved in the regular treatment of the patients evaluated dystonia severity in all patients based on the video documentation of the preoperative and postoperative neurological assessments. Symptoms were scored on the global Burke–Fahn–Marsden Dystonia Rating Scale (BFMDRS) and separately on selected subscores (eyes, mouth, speech/swallowing and neck), the Blepharospasm Movement Scale (BMS), and the Abnormal Involuntary Movement Scale (AIMS) excluding item 10 which cannot be assessed by video scoring (Table 2).

Improvement of ≥25% score change was defined as the minimal clinically useful change and necessary to justify surgical risks.

**Statistics**

For each rating scale, baseline scores and scores of the short- and long-term follow-up visit were compared using a one-way repeated measures analysis of variance (RM-ANOVA) followed by a post hoc Tukey test if appropriate. For data not normally distributed, a RM-ANOVA on ranks was performed as data were not normally distributed. Percentage improvement is shown as well as mean score ± standard deviation.

For the BFMDRS subitem neck, a RM-ANOVA on ranks was performed as data were not normally distributed. Percentage improvement is shown as well as mean score ± standard deviation.

**Results**

**Effects of GPI DBS on Motor Function**

The three centers followed their patients at slightly different time intervals, but in 11 cases long-term follow-up [FU2: 12–78 (38.8 ± 21.7) months after surgery] was available for review and in 8 cases short-term postoperative evaluation [FU1: 3–6 (4.4 ± 1.5)]. GPI DBS significantly reduced dystonia severity as measured by the BFMDRS, AIMS, and BMS. Mean improvement in BFMDRS was 45% at FU1 (12.4 ± 4.3 compared to baseline 21.4 ± 3.2; \(P < 0.001\) post hoc Tukey) and 53% at FU2 (9.8 ± 4.1; \(P < 0.001\)). Subitem analysis showed 38% and 47% for eyes (3.3 ± 1.7, \(P = 0.004\) and 2.6 ± 1.6, \(P < 0.001\), respectively, compared to baseline 5.8 ± 2.2), 50% and 56% mouth (3.7 ± 2.3, \(P < 0.001\) and 3.2 ± 2.3, \(P < 0.001\) to baseline 6.8 ± 1.8), 44% and 64% speech/swallowing (2.1 ± 1.6, \(P = 0.038\) and 1.4 ± 1.4, \(P = 0.004\) to baseline 4.3 ± 3.1). There was 25% and 38% improvement in BMS (7.1 ± 0.8, \(P = 0.006\) and 5.8 ± 0.7, \(P < 0.001\) to baseline 9.5 ± 0.4) and 44% and 49% in AIMS (7.4 ± 1.0, \(P < 0.001\) and 6.5 ± 0.8, \(P < 0.001\) to baseline 13.0 ± 0.7; Table 3 and Fig. 2).

As data of the BFMDRS subitem “neck” were not normally distributed, we performed a RM-ANOVA on ranks which failed to show statistical significance in the overall comparison (relative improvement of 35% at FU1 (3.3 ± 1.5 compared to baseline 4.3 ± 1.9) and 38% at FU2 (2.5 ± 1.8; Table 3).

Clinically relevant improvement (≥25%) was observed in all patients according to the BFMDRS motor score. In the BMS, a score change of ≥25% was apparent in 2 of 8 patients at FU1 and in 8 of 11 patients at FU2. Using the AIMS score, 7 of 8 patients at FU1 and all patients at FU2 showed such improvement.

Scores of short- versus long-term follow-up failed to show statistically significant differences in all rating scales. Baseline data and relative improvement were not different among surgical centers (data not shown).
Concomitant Medication

Patients were encouraged to discontinue medical treatment after surgery and in most cases appreciated lower medication doses or discontinuation (Table 1). Before and after surgery, Patient 1 received 8 units of Dysport (Ipsen Pharma, Germany) in each thyroarytenoideus muscle every 3 months to treat spasmodic dysphonia but lacking efficacy. She was also treated with periorbital injections of Dysport 45 months after implantation showing no clinical effect. This should not have interfered with the last follow-up 6 months after botulinum toxin treatment. Patient 8 received small amounts of botulinum toxin for residual blepharospasm on long-term follow-up.

Adverse Events

The surgical procedure was overall well tolerated. No intracranial hemorrhage was determined on postoperative MRI or CT scans. No delayed hardware complications occurred except an infection of the right DBS electrodes in patient 1 two years postoperatively. The electrode was temporarily removed and reimplanted 6 months later. Dysarthria could be elicited in some instances with stimulation intensities above the therapeutic threshold.

Electrode Localization and Battery Replacement

Assessment of active contacts of the electrodes showed correct localization in all patients within the posteroverentral GPi or ventrally adjacent to Gpi at the horizontal level of the emerging fibre tracts. The mean coordinates of the active contacts were [(mean ± SD) in MNI coordinates] x = 20.44 ± 1.52 mm, y = −8.08 ± 1.55 mm, and z = −6.98 ± 2.17 mm on the right side and x = −19.70 ± 1.64 mm, y = −7.88 ± 1.66 mm, and z =
−7.44 ± 1.94 mm on the left side (Fig. 1). Based on indirect reference with respect to the medio-commissural point of the MNI brain, these coordinates correspond to [(mean distance in x: medio-lateral, y: antero-posterior, z: dorso-ventral direction)] x = 19.86 mm, y = 3.67 mm, and z = 3.58 mm on the right side and x = 20.28 mm, y = 3.88 mm, and z = −4.04 mm on the left side in GPi.

Impulse generators were changed between 2 and 4 years after surgery due to battery depletion.

Discussion

Using a multicenter, retrospective analysis including video assessment by an independent rater we were able to study the long-term treatment response to GPi DBS in a relatively large cohort of patients suffering from Meige syndrome. All clinical scores indicated significant improvement in global dystonia severity and in the severity of blepharospasm and orofacial dystonia as constituting the major symptoms of Meige syndrome. Cervical dystonia failed to show significant improvement most likely due to a floor effect with minor severity and underrepresentation in BFMDRS. Improvements were already present at FU1 and remained stable or became even more pronounced in the long-term.

The extent of symptomatic improvement was comparable to previously published outcomes of GPi DBS in Meige syndrome11–18 as well as in segmental and generalized primary dystonia7–9 and tardive forms.20,30

Individual patients’ response was heterogeneous despite uniform clinical phenotype. Furthermore, probabilistic determination of electrode localization confirmed active contacts within the posteroverentral GPi or at its ventral border in all patients. The somatotopy of arm, leg, and trunk sensorimotor areas are presumed to overlap within the GPi31 but nevertheless even slight differences in electrode positioning may explain different clinical outcome.32 The optimal target point within the GPi for treating dystonia by DBS is still a matter of debate. In particular, it remains unclear whether different target areas would be more useful for treating dystonia in specific body parts. No differences in clinical outcome were noticed between patients who had bipolar and those who had unipolar DBS although the electrical power delivered from the impulse generator was lower in patients with unipolar stimulation.33

The surgical procedure was well tolerated in this group of patients. Transient morbidity was caused by an infection of the implanted material in a single patient. Stimulation-induced side effects resulted predominantly from unintended current spread to the internal capsule causing dysarthria and/or contractions of facial or extremity muscles. These symptoms typically showed a sudden onset when increasing stimulation intensity and also a sudden relief when reducing or switching stimulation off and they were mostly easily alleviated by modifying the stimulation parameters. Two recent reports suggested bradykinesia as a therapy-related adverse effect of palilidal neurostimulation in Meige syndrome at high voltage.4,8,34 No clinically relevant bradykinesia was observed in our cohort, although a patient with Meige syndrome who was operated on after this study was concluded, developed a mild parkinsonian gait disorder with freezing of gait upon increase of stimulation intensity.35

Limitations of our study result from the retrospective analysis of video-documented clinical evaluations, the lack of blinding, and the lack of patient-centered outcome parameters such as activity of daily living or quality of life scales, which were not consistently used by the different centers. Moreover, rating a video documentation means to assess an instantaneous clinical presentation in an artificial environment, rather than symptomatology in daily life. This could lead to either underestimation or overestimation of clinical efficacy.

As all three centers serve as tertiary nation-wide referral centers, we assume that our patient samples are not representative with regard to consider what percentage of patients with Meige syndrome would be appropriate candidates for DBS surgery. Overall, we think that only a minority of patients with Meige syndrome might be selected for DBS surgery, but more detailed analysis on long-term satisfaction of patients with Meige syndrome with botulinum toxin injections would be needed.

Nevertheless, our data show that GPi DBS in Meige syndrome is a highly effective and relatively safe therapy, if conservative treatment options have failed. The benefit is preserved in the long-term for up to 6 years. Randomized controlled studies are required to provide further evidence for the clinical efficacy of GPi DBS in Meige syndrome.

References


