JNS

CLINICAL ARTICLE

J Neurosurg 130:84–89, 2019

Long-term outcome of pallidal stimulation for Meige syndrome

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OBJECTIVE Meige syndrome is characterized by blepharospasm and varied subphenotypes of craniocervical dystonia. Current literature on pallidal surgery for Meige syndrome is limited to case reports and a few small-scale studies. The authors investigated the clinical outcomes of deep brain stimulation (DBS) of the globus pallidus internus (GPi) in patients with Meige syndrome.

METHODS Sixteen patients who underwent GPi DBS at the Tokyo Women's Medical University Hospital between 2002 and 2015 were included in this study. Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) movement subscale (BFMDRS-M) scores (range 0–120) obtained at the following 3 time points were included in this analysis: before surgery, 3 months after surgery, and at the most recent follow-up evaluation.

RESULTS The patients' mean age (± SD) at symptom onset was 46.7 ± 10.1 years, and the mean disease duration at the time of the authors' initial evaluation was 5.9 ± 4.1 years. In 12 patients, the initial symptom was blepharospasm, and the other 4 patients presented with cervical dystonia. The mean postoperative follow-up period was 66.6 ± 40.7 months (range 13–150 months). The mean total BFMDRS-M scores at the 3 time points were 16.3 ± 5.5, 5.5 ± 5.6 (66.3% improvement, $p < 0.001$), and 6.7 ± 7.3 (58.9% improvement, $p < 0.001$).

CONCLUSIONS The results indicate long-term efficacy for GPi DBS for the majority of patients with Meige syndrome.

https://thejns.org/doi/abs/10.3171/2017.7.JNS17323

KEY WORDS Meige syndrome; craniocervical dystonia; globus pallidus internus; deep brain stimulation; functional neurosurgery

Meige syndrome is characterized by blepharospasm and varied subphenotypes of craniocervical dystonia, of which oromandibular dystonia is the most common.[21] Spontaneous remission of Meige syndrome is relatively rare (< 10%).[2] Treatment options for focal dystonia in these patients include injection of botulinum toxin and treatment with oral medication (anticholinergics, benzodiazepenes, or zolpidem).[8] Pallidal deep brain stimulation (DBS) is used in the event of failure of conservative treatment.

DBS of the internal segment of the globus pallidus (globus pallidus internus [GPi]) has been shown to be an effective treatment for generalized and segmental dystonia in several high-evidence-class studies.[10,23] However, current literature on surgical interventions for Meige syndrome is limited to case reports and a few small-scale studies.

We herein report the long-term outcomes of GPi DBS in 16 patients treated for Meige syndrome.

Methods

Patient Population

Sixteen patients (9 men and 7 women) with Meige syndrome who underwent GPi DBS at the Tokyo Women's

ABBREVIATIONS BFMDRS = Burke-Fahn-Marsden Dystonia Rating Scale; BFMDRS-M = BFMDRS movement subscale; DBS = deep brain stimulation; GPi = globus pallidus internus; IPG = implantable pulse generator.

SUBMITTED February 7, 2017. **ACCEPTED** July 6, 2017.

INCLUDE WHEN CITING Published online January 19, 2018; DOI: 10.3171/2017.7.JNS17323.

TABLE 1. Clinical characteristics of 16 patients with Meige syndrome

Case No.	Age at Onset (yrs),* Sex	Disease Duration (yrs)†	Onset Site	Failed Preop Medication	Surgery	Implanted Device	Remarks
1	20, M	16	Neck	Trihexyphenidyl, clonazepam	Bilat GPi DBS	Soletra	
2	33, M	1	Neck	Botulinum toxin, MAB	Bilat GPi DBS	Soletra	Respiratory Sx (improved)
3	51, M	2	Neck	Tiapride, baclofen, clonazepam, etizolam, alprazolam, trihexyphenidyl	Bilat GPi DBS	Soletra	Erosion at lt pst auricle 3 yrs postop
4	50, M	7	Eye	Botulinum toxin, etizolam, haloperidol, clonazepam, trihexyphenidyl, quetiapine, baclofen, levodopa,	Bilat GPi DBS	Soletra	
5	58, F	14	Eye	Botulinum toxin, clonazepam	Bilat GPi DBS	Soletra	
6	60, M	9	Eye	Botulinum toxin, trihexyphenidyl	Bilat GPi DBS	Soletra	New Sx after surgery: rt foot
7	59, M	7	Eye	Botulinum toxin, clonazepam, alprazolam, chlorpromazine, haloperidol, trihexyphenidyl	Bilat GPi DBS	Soletra	
8	45, M	3	Eye	Botulinum toxin, amantadine, tiapride, clonazepam	Bilat GPi DBS	Soletra	
9	43, M	6	Eye	Botulinum toxin	Bilat GPi DBS	Soletra	
10	52, F	3	Eye	Botulinum toxin	Bilat GPi DBS	Soletra	
11	50, F	5	Eye	Botulinum toxin	Bilat GPi DBS	Activa SC	Respiratory Sx (improved)
12	49, M	5	Neck	Botulinum toxin, trihexyphenidyl, clonazepam	Bilat GPi DBS	Activa SC	
13	40, F	1	Eye	Botulinum toxin, trihexyphenidyl	Bilat GPi DBS	Activa SC	Respiratory Sx (improved)
14	39, M	1	Neck	Botulinum toxin	Bilat GPi DBS	Brio	
15	31, F	7	Eye	Trihexyphenidyl, haloperidol, clonazepam, diazepam, botulinum toxin, MAB	Bilat GPi DBS	Brio	Lead breakage, infection
16	52, F	4	Eye	Botulinum toxin, tiapride, baclofen, levodopa, biperiden	Bilat GPi DBS	Brio	New Sx after surgery: lt arm

MAB = monoclonal antibody; pst = posterior; Sx = symptom(s).

* Mean age at onset 46.7 ± 10.1 years.

† Mean duration of disease (from symptom onset to initial evaluation) 5.9 ± 4.1 years.

Medical University Hospital between 2002 and 2015 were included in this study. Detailed clinical characteristics are shown in Table 1. Their mean age at onset was 46.7 ± 10.1 years; the mean duration of disease at the time of our initial evaluation was 5.9 ± 4.1 years. In 12 patients, the initial symptom was blepharospasm, whereas 4 patients presented with cervical dystonia.

Surgical Procedures

Implantation of DBS leads (cases 1–13, model 3387, Medtronic; cases 14–16, model 6145, St. Jude Medical) was carried out under local anesthesia. The target was defined on MRI and CT with stereotactic frame by direct visualization of the GPi borders and optic tract. The stereotactic coordinates of the GPi were 2 mm anterior to the midcommissural point, 4 mm below the anterior commissure–posterior commissure (AC-PC) line, and 20–22 mm lateral to the midline. We did not use microelectrode recording. Macrostimulation was carried out to check for capsular responses and visual phosphenes by using DBS leads with an external neurostimulator (Medtronic model 3625). Postoperative MRI was performed for confirmation of electrode placement in cases 1–13 (Fig. 1). Implantable pulse generators (IPGs; cases 1–10, SOLETRA, Medtronic; cases 11–13, ACTIVA SC, Medtronic; cases 14–16, Brio, St. Jude Medical) were placed under general anesthesia 1–2 weeks after the first operation. Stimulation was initiated the day of IPG implantation.

Evaluation Procedures

Scores were obtained for the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)[1] movement subscale (BFMDRS-M) based on evaluation of video recordings obtained at 3 time points: before surgery, 3 months after surgery, and at the most recent follow-up visit. All patients

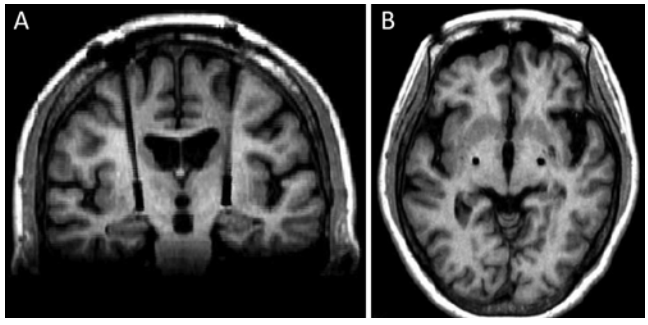

FIG. 1. Coronal (**A**) and axial (**B**) T1-weighted MR images obtained in a representative case (case 13) showing electrodes placed in the GPi bilaterally.

TABLE 2. BFMDRS-M total and subitem scores at the 3 study time points

BFMDRS-M	Preop	3 Mos Postop	p Value*	% Impr†	Last FU	p Value‡	% Impr†	p Value§
Total	15.5 (11.75–22.0)	2.5 (1.4–7.5)	<0.001	66.30%	3.8 (1.0–10.3)	<0.001	58.90%	<0.001
Subitem								
Eye	4.0 (2.0–8.0)	0.5 (0.0–2.0)	<0.001	67.35%	0.3 (0.0–2.3)	0.006	59.0%	<0.001
Mouth	4.0 (2.0–4.9)	0.3 (0.0–1.0)	<0.001	81.0%	0.0 (0.0–1.0)	0.001	75.8%	<0.001
Speech/swallow	4.0 (1.5–6.0)	0.0 (0.0–1.3)	0.002	72.0%	0.5 (0.0–2.5)	0.002	64.70%	<0.001
Neck	2.5 (0.9–6.0)	0.5 (0.0–1.0)	0.004	62.50%	0.5 (0.0–1.3)	0.008	56.20%	<0.001

FU = follow-up; impr = improvement.

Data presented as median (interquartile range). Significance was accepted at $p < 0.01$. Bonferroni correction was used for multiple comparisons.

* For comparison of mean preoperative and 3-month postoperative scores; Wilcoxon signed-rank test.

† Calculated based on mean scores.

‡ For comparison of mean preoperative and last follow-up scores; Wilcoxon signed-rank test.

§ For comparison of mean preoperative, 3-month postoperative, and last follow-up scores; Friedman test.

were asked about the time of best symptomatic improvement during clinical follow-up. Percent improvement values were calculated based on the mean scores.

Statistical Analysis

The data were considered nonparametric, and the Wilcoxon signed-rank test and Friedman test were used to compare the BFMDRS-M total score and subitem scores with their respective baseline scores. Bonferroni correction was used for multiple comparisons.

Significance was accepted at $p < 0.01$.

Results

The mean (± SD) duration of follow-up was 66.6 ± 40.7 months (range 13–150 months). The mean preoperative, postoperative, and last follow-up BFMDRS-M scores were, respectively, 16.3 ± 5.5, 5.5 ± 5.6 (66.3% improvement, $p < 0.001$), and 6.7 ± 7.3 (58.9% improvement, $p < 0.001$). On analysis of subitem scores, there was a significant reduction in subitem scores for eyes (67.35%, $p < 0.001$ at 3 months after surgery, and 59.0%, $p = 0.006$ at the last available follow-up examination), mouth (81.0%, $p < 0.001$ and 75.8%, $p = 0.001$), speech/swallow (72.0%, $p = 0.002$ and 64.7%, $p = 0.002$), and neck (62.5%, $p = 0.004$ and 56.2%, $p = 0.008$) (Table 2).

Detailed subscale scores of BFMDRS-M from the preoperative, postoperative, and latest follow-up assessments are shown in Table 3. Nine patients showed deterioration in BFMDRS-M scores at the most recent follow-up in comparison with scores obtained 3 months after surgery. Nine patients reported that the timing of best symptomatic improvement was within the first 3 months of surgery. During the clinical follow-up period, 2 patients developed new dystonic symptoms in regions that were previously unaffected. Three patients who had respiratory dystonic symptoms before DBS showed remarkable improvement with GPi DBS. One patient developed a DBS device infection and lead breakage that required removal of the DBS system; another patient developed erosion but did not require removal of the DBS system.

Details of the stimulation parameter settings are described in Table 3.

Poor-Results Group

In 4 cases (cases 5, 14, 15, and 16), the results qualified as "poor" (< 30% improvement in BFMDMS) despite repetitive stimulation settings including low-frequency stimulation (450–500 μsec, 60–90 Hz, 2.0–4.0 mA/2.0–3.5 V). Stimulation-induced dysarthria was observed in 3 patients (cases 14, 15, and 16), but it did not influence the postoperative programming. We investigated the possibility of misplacement of electrodes in all 4 patients with poor outcomes using postoperative frame-based CT (1-mm slice thickness), but there was no significant misplacement with respect to the preoperatively selected targets. Development of dystonic symptoms in hitherto unaffected body regions was observed in case 5 (speech symptoms) and case 16 (arm dystonia). Deterioration of existing symptoms after surgery occurred in case 15 (eyes) and case 16 (eyes). The patient in case 14 experienced a return to his baseline condition after transient initial improvement. All of these patients experienced their best symptomatic improvement within 2–3 weeks of the surgery, followed by a gradual reversal of symptomatic improvement. We tried to identify variables (age at onset, age at surgery, disease duration, preoperative BFMDRS-M scores, onset site, etc.) that might serve as predictive factors, but we were unable to identify any statistically significant association, possibly because of the small sample size.

Discussion

In this study, GPi DBS resulted in a 58.9% improvement in the overall mean BFMDRS scores for our patient group during the follow-up period, a result that is consistent with previous reports. The patient with the longest follow-up period of 150 months maintained a significant improvement in his total BFMDRS-M score (94.4%). Our results suggest that GPi DBS may be a promising procedure with long-term benefits for many patients with Meige syndrome.

Current literature on surgical interventions for Meige syndrome is limited to case reports and a few small-scale studies. A total of 22 published reports have documented a total of 75 cases in which Meige syndrome patients underwent DBS and pallidotomy. According to a review

TABLE 3. Clinical outcomes in 16 patients with Meige syndrome

Case No.	BFMDRS-M						% Impr§	Duration of FU (mos)¶	Stimulation Parameters at Last FU		Timing of Max Impr
	Preop		3 Mos Postop		Last FU						
	Subitems	Total*	Subitems	Total†	Subitems	Total‡			Right	Left	
1	E:8/M:6/S:4/N:6	24	M:0.5/N:0.5	1	M:0.5/N:0.5	1	95.8	140	C(+)1(−), 3.2 V, 160 μsec, 210 Hz	C(+)1(−), 3.1 V, 160 μsec, 210 Hz	Contin
2	E:4/M:6/S:4/N:4	18	E:1/N:1	2	N:1	1	94.4	150	C(+)0(−)1(−), 2.0 V, 150 μsec, 150 Hz	C(+)0(−)1(−), 2.5 V, 150 μsec, 150 Hz	Contin
3	E:8/M:6/S:6/N:3	23	E:2/M:2/S:4/N:4	12	E:2/M:2/S:4/N:6	14	40.9	113	3(+)1(−)2(−), 3.3 V, 150 μsec, 130 Hz	3(+)1(−)2(−), 3.2 V, 150 μsec, 130 Hz	w/in 3 mos
4	E:2/M:4/S:4/N:1/A:15	26	M:2/S:2/N:1/A:8	13	M:0.5/S:1/N:0.5/A:8	10	61.5	99	3(+)1(−)2(−), 3.5 V, 210 μsec, 185 Hz	C(+)1(−)2(−), 3.2 V, 210 μsec, 185 Hz	w/in 3 mos
5	E:3/M:3/N:6	12	M:2/N:4	6	M:3/S:4/N:4	11	8.3	81	2(+)1(−), 3.2 V, 210 μsec, 185 Hz	2(+)1(−), 3.2 V, 210 μsec, 185 Hz	w/in 3 mos
6	E:1/M:2/S:6/A:2	11	E:0.5/M:1/S:4	5.5	S:4/A:1	5	54.5	68	1(+)0(−), 3.6 V, 210 μsec, 185 Hz	1(+)0(−), 3.6 V, 210 μsec, 185 Hz	NR
7	E:8/M:4/S:4/N:2/ A:2/T:2	22	E:1	1	E:1	1	95.5	68	2(+)0(−)1(−), 3.3 V, 200 μsec, 185 Hz	2(+)0(−)1(−), 3.3 V, 200 μsec, 185 Hz	w/in 3 mos
8	E:1/M:2/S:12/N:0.5	15.5	E:0.5/M:0.5/S:1	2	E:0.5/M:1/S:2	3.5	77.4	66	2(+)0(−)1(−), 3.6 V, 210 μsec, 145 Hz	2(+)0(−)1(−), 3.6 V, 210 μsec, 145 Hz	NR
9	E:4/M:2/N:1	7	E:2/M:1	3	E:3/M:1	4	42.9	62	2(+)0(−)1(−), 3.5 V, 210 μsec, 185 Hz	3(+)1(−)2(−), 3.5 V, 210 μsec, 185 Hz	w/in 3 mos
10	E:2/M:4.5/S:6/N:3	15.5	N:1	1	N:2	2	87.1	60	C(+)0(−)1(−), 3.2 V, 210 μsec, 185 Hz	2(+)0(−)1(−), 3.2 V, 210 μsec, 185 Hz	w/in 3 mos
11	E:8/S:2	10	S:1	1	S:1	1	90	47	2(+)0(−)1(−), 3.0 V, 210 μsec, 185 Hz	2(+)0(−)1(−), 3.2 V, 210 μsec, 185 Hz	Contin
12	E:1.5/M:2/S:8/N:6	17.5	E:0.5/M:0/S:1	1.5	E:0.5/S:2	2.5	85.7	39	3(+)1(−)2(−), 2.0 V, 210 μsec, 185 Hz	0(+)1(−), 2.5 V, 210 μsec, 185 Hz	NR
13	E:4/M:4/N:6	14	E:0.5/M:0.5/N:0.5	1.5	N:0.5	0.5	96.4	31	2(+)0(−)1(−), 3.1 V, 210 μsec, 180 Hz	2(+)0(−)1(−), 3.1 V, 210 μsec, 180 Hz	NR
14	E:8/N:6	14	E:8/N:6	14	E:8/N:6	14	0	15	3(+)1(−), 1.2 V, 450 μsec, 160 Hz	4(+)2(−)3(−), 6.7 V, 212 μsec, 140 Hz	w/in 3 mos
15	E:6/M:4	10	E:4	4	E:8	8	20	14	3(+)1(−)2(−), 2.5 V, 137 μsec, 140 Hz	2(+)1(−), 2.3 V, 137 μsec, 140 Hz	w/in 3 mos
16	E:2/M:6/S:6/N:2/A:6	22	E:3/M:3/S:6/N:1/A:6	19	E:6/M:6/S:6/N:1/A:10	29	–24.1	13	C(+)1(−)2(−), 2.5V, 212 μsec, 140 Hz	C(+)1(−)2(−), 2.5 V, 212 μsec, 140 Hz	w/in 3 mos

A = arm; contin = continuous; E = eyes; L = leg; M = mouth; N = neck; NR = not reported; S = speech and swallowing; T = trunk.

* Mean score 16.3 ± 5.5.

† Mean score 5.5 ± 5.6.

‡ Mean score 6.7 ± 7.3.

§ Mean 58.9% ± 38.1%.

¶ Mean 66.6 ± 40.7 months.

by Wang et al., GPi DBS was associated with 66.9% improvement in BFMDRS-M scores,[24] which is consistent with our results. In our study, 2 patients with over 10 years of follow-up after GPi DBS (140 and 150 months) maintained their best BFMDRS-M scores, representing, respectively, a 95.8% and a 94.4% improvement relative to their baseline scores. GPi DBS is already an established treatment for primary generalized dystonia. Although robust evidence of the efficacy of GPi DBS for Meige syndrome is lacking, it can be expected to provide a similar effect for Meige syndrome. GPi DBS can be an alternative treatment modality for cases that are refractory to botulinum toxin and oral medications.

The distribution of dystonia in Meige syndrome has varied combinations. Symptoms involving the neck or eyes were less likely to respond to GPi DBS than symptoms involving the mouth and speech/swallowing. The neck symptoms received the lowest scores among the BFMDRS-M subitems in this study. This trend is consistent with the results from other studies[16,19] and is potentially linked to the location of the DBS electrodes in the GPi. Usually, electrode placement for dystonia favors the posteroventrolateral GPi (Laitinen target).[11] In the somatotopic representation within the GPi, the face area is located more toward the posterior and ventral portion,[5] while the cervicoaxial representation is more anterior.[22] In light of this somatotopic representation, additional electrodes more anterior in the GPi might provide further improvement in intractable cases.

In this study, 4 patients responded poorly to GPi DBS, with < 30% improvement in BFMDRS-M scores. One of these patients had refractory blepharospasm (case 15), one showed deterioration of dystonic symptoms and development of dystonia in a previously unaffected region (case 16), one showed deterioration in speech and swallowing symptoms (case 5), and one experienced transient improvement but had returned to baseline status within a few weeks of surgery (case 14). Of these, 3 patients (cases 14, 15, and 16) experienced significant improvement of symptoms within 2 to 3 weeks of the implantation of DBS leads. Thereafter, the symptoms gradually worsened by varying degrees despite repeated adjustments of stimulation settings. The initial improvements were considered to be due to a microlesion effect (micropallidotomy), which is a commonly observed phenomenon of transient improvement of symptoms after electrode insertion. Basically, dystonia patients with microlesion effect are more likely to respond well to GPi DBS.[3] We do not have convincing evidence, however, for the cause of the initial improvement and subsequent worsening in these 3 patients, and it is possible that placebo effect or disease progression might have been involved.

Limotai et al.[12] suggested placement of rescue leads in the subthalamic nucleus (STN) or thalamus in patients who do not respond to GPi DBS. STN and thalamic DBS have been reported as effective treatments for dystonia, including that associated with Meige syndrome.[13,14,20] In *DYT-1* dystonia, disease progression after GPi DBS and loss of efficacy of DBS with time can occur. Patients with Meige syndrome may also show progression of dystonic symptoms even after GPi DBS, as occurred in 3 of our cases. Cif et al.[4] reported that placement of additional leads in the GPi provided further improvement in patients with *DYT-1* dystonia who experienced disease progression after GPi DBS. As mentioned above, placement of additional leads in the GPi may provide further improvement in refractory Meige syndrome. Sako et al.[17] reported on the effects of low-frequency stimulation (60 Hz) for Meige syndrome, but our trial of low-frequency stimulation in the intractable cases in this study did not lead to improvement.

A limitation of this study is the lack of evaluation of the effects of GPi DBS on cognition and mood. Some studies have reported severe limbic adverse effects of GPi DBS, such as mania, depression, and suicidal urges.[6,18] Meige syndrome is more likely to be complicated by depression.[21] In this study, depression was observed in 3 patients. Jahanshahi et al.[7] reported that GPi DBS for dystonia is only associated with deterioration of sustained attention, without any major adverse impact on cognition. Several other studies have also supported the safety of GPi DBS with respect to cognition.[9,15] Careful attention to mood should be paid in dystonia patients after GPi DBS.

Conclusions

Overall, the long-term outcomes of GPi DBS in this study are comparable to those reported previously. Further study is required to investigate additional treatment for Meige syndrome refractory to surgical intervention.

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Disclosures

Takaomi Taira reports receiving consulting fees from St. Jude Japan and lecture fees from Daiichi-Sankyo.

Author Contributions

Conception and design: Taira, Horisawa, Kawamata. Acquisition of data: Taira, Horisawa, Ochiai, Goto, Nakajima, Takeda. Analysis and interpretation of data: Horisawa. Drafting the article: Horisawa. Critically revising the article: Horisawa. Statistical analysis: Horisawa.

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