

Deep brain stimulation for Meige's syndrome

Thirty-two patients with primary Meige syndrome underwent [GPi-DBS](#). Burke-Fahn-Marsden Dystonia Rating Scale scores (BFMDRS-M) were obtained for the evaluation of clinical symptoms 3 days before DBS (baseline), 24 h after DBS surgery, once weekly for 1 month until electrical stimulation, 6 months postoperatively and 12 months after surgery. Twenty-seven patients had MLE after GPi-DBS. The mean time of BFMDRS-M scores maximal improvement from MLE was 35.9 h postoperatively (range, 24-48 h), and the mean scores improved by $49.35 \pm 18.16\%$. At 12 months after surgery, the mean BFMDRS-M scores improved by $50.28 \pm 29.70\%$. There was a positive correlation between the magnitude of MLE and the motor score at 12 months after GPi-DBS ($R^2 = 0.335$, $p < 0.05$). However, there was no correlation between the duration of MLE and DBS improvement. Most Meige syndrome patients who underwent GPi-DBS and had MLE benefited from MLE. For Meige syndrome, MLE might be a predictive factor for patient clinical symptom improvement from DBS ¹⁾.

10 patients experienced improved Meige's syndrome-related symptoms after extracranial radiofrequency ablation of the cranial and/or mandibular branches of the extracranial [trigeminal nerve](#). Adverse events included class II-III facial paralysis and/or mandibular skin numbness. Two patients had recurrences at the 18th and 22nd months postoperatively, respectively; the other patients were being followed up.

These results show that CT-guided radiofrequency ablation of bilateral stylomastoid foramen facial nerve and/or oval foramen trigeminal mandibular branch can effectively treat the corresponding types of Meige's syndrome. According to preliminary observations, the therapeutic effect may last more than 18 months ²⁾.

A well-established therapeutic option is [deep brain stimulation](#) (DBS), and the target in bilateral [globus pallidus internus](#) (GPi DBS) demonstrated satisfactory short- and long-term [efficacy](#). However, some patients present minor or suboptimal responses after GPi DBS, and in those cases, rescue DBS may be appropriate ³⁾.

A retrospective study to assess the efficacy and safety of bilateral GPi stimulation in 40 patients with primary Meige syndrome who responded poorly to medical treatments or botulinum toxin injections. All participants were postoperatively followed up at the outpatient clinic, and their motor functions were assessed using the [Burke-Fahn-Marsden Dystonia Rating Scale](#) (BFMDRS). The severity of patients' dystonia was evaluated before surgery and at follow-up neurostimulation.

Results: The implanted stimulator was turned on 1 month after surgery. All 40 patients received monopolar stimulation using the following parameters: voltage 2.5-3.5 V (average: 2.6 ± 0.8 V), frequency 60-160 Hz (average: 88.0 ± 21.3 Hz), and pulse width 60-185 μ S (average: 90.0 ± 21.1 μ S). In 28 of 40 patients, the symptoms had significantly improved within 1 week of stimulation. Most of the patients had been followed up for 6-24 months (average: 15.0 ± 7.8 months). The clinical symptoms of all patients had significantly improved. At 6, 12, and 24 months after surgery, the BFMDRS subscores of eyes, mouth, speech, and swallowing were significantly lower, and subscores of

mouth movement showed progressively decreased with prolonged stimulation time. The overall improvement rate was 83%. Five adverse events occurred in the 40 patients; all of these events resolved without permanent sequelae.

Bilateral GPi-DBS demonstrated satisfactory long-term efficacy in the treatment of primary Meige syndrome and could serve as an effective and safe option ⁴⁾.

A study explored the predictors of [clinical outcome](#) in patients with [Meige syndrome](#) who underwent DBS.

Twenty patients who underwent DBS targeting the bilateral subthalamic nucleus (STN) or globus pallidus internus (GPi) at the Chinese People's Liberation Army General Hospital from August 2013 to February 2018 were enrolled in the study. Their clinical outcomes were evaluated using the Burke-Fahn-Marsden Dystonia Rating Scale at baseline and at the follow-up visits; patients were accordingly divided into a good-outcome group and a poor-outcome group. Putative influential factors, such as age and course of disease, were examined separately, and the factors that reached statistical significance were subjected to logistic regression analysis to identify predictors of clinical outcomes.

Four factors showed significant differences between the good- and poor-outcome groups: 1) the DBS target (STN vs GPi); 2) whether symptoms first appeared at multiple sites or at a single site; 3) the sub-item scores of the mouth at baseline; and 4) the follow-up period ($p < 0.05$). Binary logistic regression analysis revealed that initial involvement of multiple sites and the mouth score were the only significant predictors of clinical outcome.

The severity of the disease in the initial stage and presurgical period was the only independent predictive factor of the clinical outcomes of DBS for the treatment of patients with Meige syndrome ⁵⁾.

A total of 6 patients seen between 2002 and 2010 with craniofacial and craniocervical dystonia symptoms were identified from the University of Florida Institutional Review Board approved database. Patients were videotaped using a standardized protocol, and tapes were randomized and blindly reviewed by a movement disorders neurologist. The Unified Dystonia Rating Scale improved $31.6 \pm 23.2\%$ (range: 3.4-63.2%) at 6 months and $63.7 \pm 35.3\%$ (range: 6.3-100%) at 12 months. The Burke-Fahn-Marsden Dystonia Rating Scale improved $45.3 \pm 29.5\%$ (range: 4.7-75.0%) at 6 months and $61.8 \pm 30.9\%$ (range: 16.6-100%) at 12 months. One patient significantly had a very large improvement with little evidence of residual dystonia. Blepharospasm improved in all patients, whereas speech and swallowing did not improve in this cohort. Two patients improved with unilateral GPi-DBS, although one required a contralateral DBS later in the disease course. Two patients were managed with low-frequency stimulation (<100 Hz). Two patients had less than 20% benefit. GPi-DBS for cranio-facial and craniocervical symptoms is an effective strategy to manage a subset of patients who remain unresponsive to optimized medical management. Unilateral stimulation may be an option for some patients, but it remains unclear whether response to single-sided stimulation will be sustainable. The mixed results of this GPi-DBS case series highlight the need for a careful re-examination of selection criteria, alternative brain targets, and possibly rescue leads for patients who are non-responders to the GPi target ⁶⁾.

Lyons report the long-term results of bilateral globus pallidus internus (GPi) or subthalamic nucleus (STN) stimulation in 3 patients with Meige syndrome and 1 patient with Parkinson's disease and associated craniofacial dystonia treated at their center.

Initial 12-month and long-term follow-up Burke-Fahn-Marsden scores were substantially improved in all 4 patients compared with preoperative scores.

Bilateral GPi DBS may be an effective and safe treatment for medically refractory Meige syndrome. The results are comparable with those reported in the literature. Sustained and long-term improvement in symptoms does appear to be reproducible across reports. The authors' patient with Parkinson's disease and associated craniofacial dystonia syndrome undergoing bilateral STN DBS noted immediate and sustained improvement in his symptoms. Further study is required, but these results, along with the other reports, suggest that bilateral GPi DBS is an effective treatment for medically refractory Meige syndrome ⁷⁾.

Sobstyl M, Ząbek M, Mossakowski Z, Zaczyński A. Pallidal deep brain stimulation in the treatment of [Meige syndrome](#). Neurol Neurochir Pol. 2014;48(3):196-9. doi: 10.1016/j.pjnns.2014.05.008. Epub 2014 Jun 6. PubMed PMID: 24981184.

¹⁾
Liu J, Ding H, Xu K, Wang D, Ouyang J, Liu Z, Liu R. Micro lesion effect of pallidal deep-brain stimulation for Meige syndrome. Sci Rep. 2022 Nov 21;12(1):19980. doi: 10.1038/s41598-022-23156-2. PMID: 36411289; PMCID: PMC9678874.

²⁾
Huang B, Du XD, Yao M, Lin HD, Yu WH, Zhou QH. CT-guided radiofrequency ablation of the extracranial cranial nerve for the treatment of Meige's syndrome. Front Neurosci. 2022 Oct 6;16:1013555. doi: 10.3389/fnins.2022.1013555. PMID: 36278012; PMCID: PMC9582605.

³⁾
Aragão VT, Barbosa Casagrande SC, Listik C, Teixeira MJ, Barbosa ER, Cury RG. Rescue [Subthalamic Deep Brain Stimulation](#) for Refractory [Meige's syndrome](#). Stereotact Funct Neurosurg. 2021 Apr 23:1-3. doi: 10.1159/000515722. Epub ahead of print. PMID: 33895729.

⁴⁾
Tian H, Yu Y, Zhen X, Zhang L, Yuan Y, Zhang B, Wang L. Long-Term Efficacy of Deep Brain Stimulation of Bilateral Globus Pallidus Internus in Primary Meige Syndrome. Stereotact Funct Neurosurg. 2019;97(5-6):356-361. doi: 10.1159/000504861. Epub 2020 Jan 10. PMID: 31927550.

⁵⁾
Wang X, Mao Z, Cui Z, Xu X, Pan L, Liang S, Ling Z, Yu X. Predictive factors for long-term clinical outcomes of [deep brain stimulation](#) in the treatment of primary [Meige syndrome](#). J Neurosurg. 2019 Apr 5:1-9. doi: 10.3171/2019.1.JNS182555. [Epub ahead of print] PubMed PMID: 30952124.

⁶⁾
Limotai N, Go C, Oyama G, Hwynn N, Zesiewicz T, Foote K, Bhidayasiri R, Malaty I, Zeilman P, Rodriguez R, Okun MS. Mixed results for GPi-DBS in the treatment of cranio-facial and cranio-cervical dystonia symptoms. J Neurol. 2011 Nov;258(11):2069-74. doi: 10.1007/s00415-011-6075-0. Epub 2011 May 7. PMID: 21553081.

⁷⁾
Lyons MK, Birch BD, Hillman RA, Boucher OK, Evidente VG. Long-term follow-up of [deep brain stimulation](#) for Meige syndrome. Neurosurg Focus. 2010 Aug;29(2):E5. doi: 10.3171/2010.4.FOCUS1067. PMID: 20672922.

Last
update:
2024/06/07 02:49 deep_brain_stimulation_for_meige_s_syndrome https://neurosurgerywiki.com/wiki/doku.php?id=deep_brain_stimulation_for_meige_s_syndrome

From:
<https://neurosurgerywiki.com/wiki/> - **Neurosurgery Wiki**

Permanent link:
https://neurosurgerywiki.com/wiki/doku.php?id=deep_brain_stimulation_for_meige_s_syndrome

Last update: **2024/06/07 02:49**

