Long-term efficacy of GPi DBS for craniofacial dystonia: a retrospective report of 13 cases

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Received: 29 November 2020 / Revised: 1 May 2021 / Accepted: 10 June 2021 / Published online: 29 June 2021
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Abstract
This study evaluated the long-term efficacy of globus pallidus internus (GPi) deep brain stimulation (DBS) in the treatment of craniofacial dystonia (Meige syndrome) and investigated the correlation between the volume of tissue activated (VTA) in the GPi and each subregion and movement score improvement. We retrospectively analyzed the clinical data of 13 patients with drug-refractory Meige syndrome who were treated with GPi DBS. The pre- and postoperative Burke–Fahn–Marsden Dystonia Rating Scale (BFMDRS) scores were compared. The relationships between the preoperative baseline variables and improvement in the BFMDRS-Movement (BFMDRS-M) score were analyzed. LEAD-DBS software was used for the three-dimensional reconstruction of the GPi and implanted electrodes. The correlations between the GPi-VTA and score improvement were analyzed. The average follow-up period was 36.6 ± 11.0 months (18–55 months). At 3 months after the stimulation and the final follow-up visit, the improvements in the BFMDRS-M score were 58.2 and 54.6%, and the improvements in the BFMDRS-Disability (BFMDRS-D) score were 53.6 and 51.7%, respectively. At the final follow-up visit, the improvements in the BFMDRS-M scores of the eye, mouth, and speech/swallowing were significant (P < 0.001). Age was an independent predictor of improvement in the BFMDRS-M score after DBS (P = 0.005). A decrease in the BFMDRS-M score was significantly positively correlated with the GPi-VTA (r = 0.757, P = 0.003). GPi DBS is an effective method for treating drug-refractory Meige syndrome. LEAD-DBS software can be used as an effective aid for visualization programming after DBS.

Keywords Craniofacial dystonia · Meige syndrome · Deep brain stimulation · Globus pallidus internus · LEAD-DBS

Introduction
Meige syndrome is a segmental dystonia characterized by blepharospasm and involuntary contractions of muscles in the face and jaw. The first-line treatments include oral medications (anticholinergics, benzodiazepines, and baclofen) and injection of botulinum toxin. Surgical treatment becomes an option when conservative treatment is ineffective. Deep brain stimulation (DBS) has been widely used to treat primary dystonia [5, 22, 23], but only a few studies investigating DBS in Meige syndrome are available [10, 12, 15, 16, 24, 27]. To evaluate the long-term efficacy and safety of globus pallidus internus (GPi) DBS in the treatment of Meige syndrome, we summarized the long-term follow-up data of 13 patients with Meige syndrome who underwent GPi DBS.

During the follow-up, the spatial relationships between the contact of the implanted intracranial electrodes and the GPi and its subregions can be directly visualized using LEAD-DBS, which is a reconstruction software [11]. The volume of tissue activated (VTA) in the GPi and its subregions can be directly calculated through electrical field simulations [6]. An analysis of the correlation between the VTA in the GPi and each subregion and the Burke–Fahn–Marsden Dystonia Rating Scale (BFMDRS) scores can enhance post-DBS visualization programming.

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Materials and methods

Clinical data

We retrospectively analyzed the clinical data of 13 patients with drug-refractory Meige syndrome who were treated with GPI DBS at West China Hospital of Sichuan University from August 2014 to June 2018. There were five men and eight women, with an average age of 46.9 ± 7.2 years (range 38–65 years). The average course of the disease was 9.2 ± 2.1 years (range 6–13 years), and all symptoms manifested as blepharospasm and oromandibular dystonia. The initial symptom was present in the eye in 10 patients and the mouth in 3 patients. The muscle tension disorder symptoms progressed to the neck in 12 patients (Table 1).

A movement disorder specialist diagnosed this disease according to the diagnostic criteria for primary Meige syndrome [7]. All 13 patients received oral medication before surgery, and 11 patients received an intramuscular injection of botulinum toxin. Their symptoms were not alleviated indefinitely. All patients underwent brain MRI to rule out intracranial structural abnormalities. No history of brain trauma or cerebrovascular accidents was reported. All

| Case number, sex | DU (years) | AS (years) | Follow-up (Ms) | BFMDRS-M Pre-op; 3 Ms; last FU | Parameters at last follow-up |
|-----------------|-----------|------------|----------------|-------------------------------|-----------------------------|
| 1, male         | 8         | 42         | 24             | 16; 6; 8                      | Contact 1(−) Case(+) 120 μs, 180 Hz, 2.9 V |
| 2, female       | 10        | 47         | 32             | 13; 6; 4                      | Contact 0(−)1(−) Case(+) 90 μs, 160 Hz, 3.0 V |
| 3, female       | 11        | 38         | 26             | 18; 2.5; 10                   | Contact 1(−) Case(+) 150 μs, 135 Hz, 2.5 V |
| 4, male         | 9         | 50         | 45             | 15; 13; 3.5                   | Contact 1(−) Case(+) 120 μs, 170 Hz, 3.2 V |
| 5, male         | 8         | 41         | 48             | 14; 5; 4.5                    | Contact 0(−) Case(+) 70 μs, 125 Hz, 3.8 V |
| 6, female       | 12        | 44         | 49             | 17.5; 6; 7.5                  | Contact 1(−)2(−) Case(+) 160 μs, 180 Hz, 3.4 V |
| 7, female       | 13        | 54         | 28             | 15; 3; 12                     | Contact 1(−) Case(+) 130 μs, 150 Hz, 3.1 V |
| 8, male         | 13        | 65         | 38             | 20.5; 8; 10                   | Contact 0(−)1(−) Case(+) 110 μs, 160 Hz, 2.8 V |
| 9, female       | 7         | 42         | 35             | 14; 1.5; 1.5                  | Contact 0(−) Case(+) 160 μs, 110 Hz, 3.6 V |
| 10, female      | 10        | 52         | 36             | 16; 8; 10.5                   | Contact 0(−)1(−) Case(+) 100 μs, 180 Hz, 3.2 V |
| 11, male        | 6         | 42         | 42             | 15; 7; 4                      | Contact 1(−) Case(+) 100 μs, 180 Hz, 3.2 V |
| 12, female      | 8         | 49         | 55             | 17; 15.5; 15                  | Contact 0(−)1(−) Case(+) 150 μs, 115 Hz, 2.8 V |
| 13, female      | 11        | 44         | 18             | 21; 6.5; 7.5                  | Contact 0(−) Case(+) 60 μs, 160 Hz, 3.6 V |

DU disease duration, AS age at surgery, FU follow-up time, Pre-op pre-operation, Ms months
patients denied the use of antipsychotic drugs. No secondary dystonia with other etiologies was indicated.

**DBS surgical procedure**

One day before the operation, a 3.0-T MRI system (DISCOVERY, MR750 W; GE) was used to scan the brain. On the day of the operation, after installing the Leksell stereotactic head frame (Elekta Instrument AB, Sweden), the brain MRI scan was repeated using the same setting as previously described. The detail scan parameters were as follows: (1) T2 scan parameters—slice thickness $= 2.0$ mm, spacing $= 0$ mm, TR $= 3500$ ms, TE $= 102.0$ ms, NEX $= 2.00$, FOV $= 24.0$ cm, $24.0$ cm; (2) T1 scan parameters—slice thickness $= 1.0$ mm, TR $= 7.5$ ms, TE $= 2.8$ ms, NEX $= 1.00$, FOV $= 24.0$ cm $0$, 24.0 cm. The images obtained from the two scans were fused to determine the target site in the GPi, which was usually at the site 1–3 mm anterior, 3–4 mm inferior, and 20–22 mm lateral to the midpoint of the anterior/posterior commissure. Under local anesthesia, the tip of the electrode (Model 3387; Medtronic) was placed at the bottom of the globus pallidus and the upper edge of the optic tract. Intraoperative test stimulation was performed to determine the efficacy and detect any adverse effects (internal capsule effect or visual hallucinations) to confirm the accuracy of the target site. Under general anesthesia, an implantable pulse generator (IPG, ACTIVA RC; Medtronic) was placed subcutaneously under the clavicle. Postoperative MRI or CT was performed to verify the location of the electrode within the GPi and exclude intracranial hemorrhage.

**3D reconstruction of the GPi and electrodes and VTA calculations**

Based on the preoperative MRI and CT images, LEAD-DBS software was used for the 3D reconstruction of the GPi and implanted electrodes. The VTA was calculated by stimulation contact and stimulation parameters, which were input into the software during the follow-up to analyze the correlations between the VTA in the GPi and its subregions and BFMDRS score changes.

**Imaging data**

Preoperative head MRI and postoperative head thin-slice CT images in DICOM format were collected after the operation.

**Transform format**

The program dcm2niix (Rorden, Li 2016) in LEAD-DBS software or SPM 12 (The Wellcome Center for Human Neuroimaging, UCL Queen Square Institute of Neurology, London, UK) was used to convert the DICOM-format image data into NiTi-format image data.

**Image fusion**

The NiTi-formatted images were imported into LEAD-DBS for image fusion.

**Image normalization and correction**

The fused images were normalized using Advanced Normalization Tools (Avants 2008), and then, the Coarse mask (Schonecker 2008) method was used to correct the cortex and subcortex structures on the normalized images.

**Reconstruction of the lead trajectory**

A Medtronic 3387 electrode was used. Using previously fused and corrected images, the PaCER (Husch 2017) program was able to automatically extract the locations and directions of the implanted electrodes. Moreover, the program could determine the spatial relationships between the contact of the implanted electrodes and the GPi.

**3D reconstruction**

The DISTAL Minimal (Ewert 2017) atlas was selected for the 3D visualization. Finally, the reconstructed results were used to show the corresponding electrode contact and GPi positions (Fig. 1).

**Calculation of the VTA**

LEAD-DBS displays a 3D spatial image of the electrical field of corresponding electrodes and a simulated active stimulation area, which are created by inputting stimulation contact and stimulation parameters into LEAD-DBS software (SimBio/FieldTrip (Horn 2017) method). By opening LEAD GROUP, the software can calculate the electric field stimulation zone and the VTA in the GPi and its subregions (Fig. 1). Based on different efferent nerves, the GPi is divided into the GPi sensory, GPi primary motor, GPi premotor, GPi postparietal, GPi occipital, GPi temporal, and GPi prefrontal regions (Fig. 2). Of these regions, the GPi sensorimotor (GPi sensory + GPi primary motor + GPi premotor) is considered a relay station for motor fibers and the best implantation area for GPi DBS for the treatment of Meige syndrome [7].

**Correlation analysis**

The appropriate stimulation contact and stimulation parameters were selected for each side of the brain after the
procedure. Each patient’s GPi-VTA is the sum of the left and right VTAs. The differences in the BFMDRS scores before and after surgery were calculated based on the BFMDRS score at each follow-up visit. The correlation between the VTA in the GPi and each subregion and the changes in the BFMDRS scores was further analyzed.

**Efficacy evaluation**

Stimulation started 4 weeks after the operation, and the symptom changes and adverse effects corresponding to the stimuli of each contact point were documented to select the best contact. The stimulation parameters were gradually adjusted according to the patient’s response to the stimulation. All scoring was performed by the same rater. The degree of dyskinesia was evaluated based on the BFMDRS, which includes the BFMDRS-Movement scale (BFMDRS-M) (0–120 points) and BFMDRS-Disability Scale (BFMDRS-D) (0–30 points) [1]. The evaluation was performed before surgery, 3 months after stimulation, and at the final follow-up visit. During the follow-up period, time to symptom alleviation, oral medication, stimulation parameters, and complications were documented.

**Statistics**

A Mann–Whitney U test was used to compare the BFMDRS score after surgery with the baseline BFMDRS score before surgery. A Wilcoxon test was used to analyze the relationship between BFMDRS score improvement and the preoperative baseline variables. A Spearman correlation test was used to analyze the correlations between the movement score changes and the VTA in the GPi and its subregions. The correlation coefficient was determined based on the R value. An R value greater than 0.5 was considered indicative of a significant correlation between the two variables, an R value between 0.3 and 0.5 was considered indicative of a weak correlation between the two variables, and an R value < 0.3 was considered indicative of no correlation between the two variables. P < 0.05 was considered statistically significant.

SPSS 23.0 statistical software was used for the statistical analysis.

**Results**

**BFMDRS score improvement**

The average follow-up period was 36.6 ± 11.0 months (range 18–55 months). Three months after the stimulation, the BFMDRS-M score decreased by an average of 58.2% compared to the baseline before surgery (16.3 ± 2.4 vs. 6.8 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000). By the final follow-up visit, the BFMDRS-M score decreased by an average of 54.6% compared with baseline (16.3 ± 2.4 vs. 7.5 ± 3.9, P = 0.000), and the BFMDRS-D score decreased by an average of 53.6% compared to baseline (6.2 ± 1.8 and 2.8 ± 1.1, P = 0.000).

At the final follow-up visit, regarding the individual BFMDRS-M scores, the score of the eye decreased by an average of 56.8% (5.9 ± 1.4 to 2.5 ± 1.7, P = 0.000), the score of the mouth decreased by an average of 61.5% (4.9 ± 1.9 to 2.0 ± 1.8, P = 0.002), the score of speech and
swallowing decreased by an average of $51.9\%$ ($3.5 \pm 1.0$ to $1.6 \pm 1.0$, $P = 0.000$), and the score of the neck decreased by an average of $35.9\%$ ($2.0 \pm 1.3$ to $1.2 \pm 0.8$, $P = 0.081$) (Table 2).
Predictors of DBS efficacy

The Wilcoxon test was used to analyze the relationships between the BFMDRS-M score improvement and baseline variables before surgery. Only age was a predictor of improvement in the BFMDRS score after DBS surgery (age, \( P = 0.005 \); sex, \( P = 0.414 \); course of disease, \( P = 0.705 \); follow-up period, \( P = 0.655 \); BFMDRS-M score before the operation, \( P = 1.000 \)).

Follow-up for programming

The follow-up for programming showed that stimulating the first or second contact at the bottom of the electrode produced the best clinical effect. The time to symptom alleviation was 5.6 days (range 1–14 days) after the stimulation, and the effect gradually stabilized. The stimulation parameters at the final follow-up visit were as follows: average pulse width 113 μs (60–160 μs), average frequency 153 Hz (110–180 Hz), and average voltage 3.1 V (2.5–3.8 V) (Table 1). By the final follow-up visit, three patients were taking a small amount of benzodiazepines, and no patient required treatment with botulinum toxin injections.

GPi-VTA and the difference in the BFMDRS scores

The VTA is the sum of the VTAs in the left and right sides of the brain. The GPi-VTA is mainly concentrated in the GPi sensory region, the GPi primary motor region, and the GPi premotor region. The average GPi-VTA, GPi sensorimotor-VTA, GPi primary motor-VTA, and GPi premotor-VTA were 493 ± 117.1 mm³, 90.6 ± 35.5 mm³, 168.3 ± 48.7 mm³, and 182.3 ± 48.1 mm³, respectively. By the latest follow-up, the BFMDRS-M score and the BFMDRS-D score decreased by an average of 11.77 ± 3.47 and 2.69 ± 1.32, respectively, compared to the preoperative baseline scores. Among the BFMDRS-M scores of different sites, the score of the eyes decreased by an average of 3.38 ± 1.79, the score of the mouth decreased by an average of 2.84 ± 1.60, the score of speech and swallowing decreased by an average of 1.85 ± 1.46, and the score of the neck decreased by an average of 0.85 ± 0.69 (Table 3).

Correlations between the GPi-VTA and BFMDRS scores (Table 4)

The decrease in the BFMDRS-M score was significantly correlated with the GPi-VTA and the subregional VTA. The decrease in the BFMDRS-M score was significantly positively correlated with the GPi-VTA (r = 0.757, \( P = 0.003 \)), GPi sensory-VTA (r = 0.812, \( P = 0.001 \)), GPi primary motor-VTA (r = 0.726, \( P = 0.005 \)), GPi sensorimotor-VTA (r = 0.713, \( P = 0.006 \)), GPi occipital-VTA (r = 0.843, \( P = 0.000 \)), and GPi temporal-VTA (r = 0.655, \( P = 0.015 \)).

The decrease in the eye score was significantly correlated only with the GPi occipital VTA (r = 0.581, \( P = 0.037 \)).

The decrease in the speech and swallowing scores was significantly positively correlated with the GPi-VTA (r = 0.800, \( P = 0.001 \)), GPi sensory-VTA (r = 0.775, \( P = 0.002 \)), GPi primary motor-VTA (r = 0.757, \( P = 0.003 \)), GPi premotor-VTA (r = 0.724, \( P = 0.005 \)), GPi sensorimotor-VTA (r = 0.703, \( P = 0.007 \)), GPi occipital-VTA (r = 0.651, \( P = 0.016 \)), and GPi prefrontal-VTA (r = 0.754, \( P = 0.003 \)).

Adverse events

At the final follow-up, poor efficacy was observed in two patients (BFMDRS-M score improvement < 30% [10]). The postoperative CT imaging fusion showed no obvious deviation of the electrode from the intended target site. One patient only had 5-month symptom alleviation after the stimulation. The symptoms in this patient relapsed to the preoperative baseline level and were not alleviated after multiple programming adjustments. One patient had dystonia in the right upper limb that was not present before the operation. In addition, during the programming adjustment, two

### Table 3 The basic information of VTA and the difference between the BFMDRS score of each part

| VTA (mm³)               | Mean ± SD | Median (interquartile spacing) |
|-------------------------|-----------|--------------------------------|
| GPi VTA                 | 493 ± 117.1 | 518 (219)                    |
| GPi sensory VTA         | 90.6 ± 35.5 | 88 (70)                      |
| GPi primary motor VTA   | 168.3 ± 48.7 | 164 (92)                    |
| GPi premotor VTA        | 182.3 ± 48.1 | 168 (95)                    |
| GPi sensorimotor VTA    | 265.2 ± 85.6 | 226 (165)                  |
| GPi postparietal VTA    | 111.6 ± 60.9 | 127 (127)                  |
| GPi occipital VTA       | 42.3 ± 26.3  | 42 (36)                      |
| GPi temporal VTA        | 4.2 ± 8.2   | 0.75 (7.5)                   |
| GPi prefrontal VTA      | 22.1 ± 19.7 | 18 (40)                      |

Score difference difference in scores before surgery and at the last follow-up, VTA volume of tissue activated
patients developed a facial twitch, and one patient developed dysarthria. These symptoms subsided after adjusting the stimulation parameters. One patient had an infection in the pulse generator pouch 1.5 years after the surgery. The pulse generator was removed, and the pouch position was changed after anti-infective treatment. No intracranial hemorrhage was reported. No patient experienced slow movements or frozen gait.

### Discussion

Meige syndrome is a segmental dystonia. The first-line treatments include oral medication and intramuscular injection of botulinum toxin. If patients have severe symptoms and are not responsive to conservative treatment, DBS surgery may be considered. A few studies have found that GPi DBS can significantly relieve patients’ movement symptoms and improve quality of life [10, 12, 15, 16]. In this study, the average decrease in the BFMDRS-M score is consistent with the results of previous studies [10, 12, 16] (Table 5).

After GPi DBS, the BFMDRS-M scores were improved by different degrees in different regions in the patients with Meige syndrome. At the final follow-up visit in this study, the BFMDRS-M scores of the eye, mouth, and speech/swallowing were significantly improved \((P < 0.001)\), whereas the BFMDRS-M score of the neck was not significantly improved \((P = 0.081)\). These results are consistent with the results of previous studies [10, 12, 17]. In contrast, Chang et al. reported that the best improvement in the BFMDRS-M score was observed in the neck assessment [9]. The inconsistent improvement in the BFMDRS-M scores in different parts may be related to the position of the electrodes on the posterior ventral aspect of the GPi. Muscle movement of the head and neck may correspond to different regions in the posterior ventral aspect of the GPi. A small difference in the electrode location may lead to different degrees of improvement. A study with a larger sample size is needed to confirm that the stimulation of different subregions in the posterior ventral aspect of the GPi can alleviate symptoms in different segments of the head and neck.

Various factors may contribute to BFMDRS-M score improvements, including the electrode location, stimulation contact, and stimulation parameters. In some studies,
imaging techniques have been used to explore the relationships between the locations of the stimulation contact in the GPi and its subregions and the effectiveness of treatment. A study by Yao et al. included 15 patients with Meige syndrome who were treated with subthalamic nucleus (STN)-DBS and showed that the effective stimulation contacts were primarily located on the dorsolateral side of the STN. The improvement in motion symptoms was significantly positively correlated with the VTA in the STN motion zone [26]. Reich et al. analyzed 87 dystonia patients treated with GPi DBS and found that the GPi-VTA had some predictive value for improvement in movement symptoms [13]. The results of this study demonstrate that improvements in the BFMDRS-M score are closely correlated with the VTAs in movement and sensory areas. Positive correlations with the VTAs in other subregions were also identified, and the reasons may be related to anatomical and fibrotic connections in GPi subregions. Thus, LEAD-DBS can be an effective aid in postoperative DBS visualization programming. Larger stimulation parameters should be set for the GPi-VTA while ensuring no apparent side effects.

Indeed, any indirect methods used to visualize electrodes have some limitations [20]. The method used in our study has the following limitations. (1) Preoperative MRI needs to be registered in the standard atlas, which may change the patient's nuclear anatomical structure to some extent, and the reconstructed anatomical structure subregion does not completely conform to the patient's nuclear anatomical structure. (2) It is difficult to achieve 100% fusion of the preoperative MRI and the postoperative CT, which may lead to a certain deviation in the relative position of the electrode and the nucleus. (3) Although the process of the reconstructed electrode was registered by software and confirmed by the researchers, it was difficult to match the real electrode perfectly. (4) Based on the above three issues, there might be some deviation between the reconstructed stimulus contacts and the nucleus structure. However, our team's previous study suggested that screening contacts using this method was consistent with blind screening contacts during testing symptom improvement. Therefore, to some extent, this reconstruction method can reflect the relationship between contacts and the nucleus and may explain the efficacy of DBS using the VTA. Our present study preliminarily explored the relationship between the VTA of the nucleus subregion and the improvement in clinical symptoms, hoping to provide some theoretical guidance for postoperative visualization programming. Notably, however, any indirect method used to explore the relationship between stimulus contacts and nucleus structure has some limitations. Therefore, more studies are needed in the future to explore more powerful methods for us to reconstruct the nucleus and contacts.

Whether baseline factors (sex, age, and course and severity of disease) are independent predictors of the BFMDRS-M score after DBS surgery remains controversial [2, 9]. Some studies have reported that baseline factors are not related to motor function improvement [9, 22, 27]. A study by Xin Wang et al. involving 20 patients with Meige syndrome who underwent DBS showed that the severity of the disease at onset and before surgery is an independent predictor of motor function improvement after DBS surgery [25]. In the current study, only age was an independent predictor of motor function improvement after surgery ($P=0.005$).

In the present study, after the electrode implantation, most patients' symptoms were alleviated after surgery due to the effect of microdamage. The time required for symptom alleviation was 5.6 days (1–14 days) after starting the stimulation, and the effect gradually stabilized. This study demonstrated that the ventral contact point of the electrodes (the lowest first or second contact point) with high-frequency stimulation greater than 100 Hz is associated with the best alleviation of motor symptoms. Two studies [15, 16] reported that low-frequency stimulation (60 Hz) alleviated motor symptoms in Meige syndrome. In this study, low-frequency stimulation (less than 100 Hz) was applied to two patients with poor efficacy, and the movement disorder symptoms were not alleviated.

Good tolerance with few complications was observed in patients with Meige syndrome who underwent GPi DBS surgery. The common complications related to stimulation include limb twitching, dysarthria, and bradykinesia. Some studies have reported that patients undergoing GPi DBS treatment for dystonia develop bradykinesia, difficulty moving, and other symptoms similar to those of Parkinson’s disease [3, 4, 14, 18]. In addition, GPi DBS may cause cognitive and emotional changes and even serious adverse effects in the limbic system, such as mania, depression, and suicidal impulses [19, 21]. In 2006, Foncke et al. reported that two patients committed suicide 3 weeks and 14 months after undergoing GPi DBS for dystonia [8]. In the present study, one patient had an infected pulse generator pouch, leading us to change its position. Two patients developed facial twitches, and one patient developed dysarthria. These symptoms subsided after adjusting the stimulation parameters.

**Study limitations**

The shortcomings of this study include the lack of an evaluation of the effect of GPi DBS on cognition and emotion. This study was a retrospective study with a small sample size. Our team is conducting a prospective, multicenter, randomized controlled study to compare the alleviation in motor disorder symptoms, the improvement in the quality of life, and the effect on emotional cognition between GPI DBS and STN DBS for the treatment of Meige syndrome.
and fully assess the efficacy and safety of DBS (two target points) (registration number ChiCTR2000032852).

**Conclusion**

GPI DBS treatment for Meige syndrome can significantly alleviate the severity of dystonia and patient dysfunction. LEAD-DBS software can be used as an effective aid for visualization programming after DBS.

**Author contribution** All authors contributed to the study conception and design. The material preparation, data collection, and analysis were performed by H.R., R.W., and D.L. The first draft of the manuscript was written by H.R. and R.W. W.W., D.L., M.W., Y.G., Y.X., and Y.W. read and approved the final manuscript.

**Funding** This study was funded by the 135 Project of Outstanding Development of West China Hospital, Sichuan University (No. ZY2017307).

**Data availability** The data used and/or analyzed during the current study are available from the corresponding author on reasonable request.

**Code availability** Not applicable.

**Declarations**

**Ethical approval** The research protocol was approved by the Ethics Committee on Biomedical Research, West China Hospital of Sichuan University (No. 202002).

**Consent to participate** Informed consent was obtained from all individual participants included in the study.

**Consent for publication** Informed consent for publication was obtained.

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**Conflicts of interest** The authors declare no competing interests.

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