Clinical Outcome of Pallidal Deep Brain Stimulation for Various Types of Dystonia

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Background: *Deep brain stimulation of the globus pallidus interna (GPi-DBS) has been approved as a surgical treatment for severe dystonia.*
Objective: *To study and compare the efficacy of GPi-DBS for various types of dystonia, and to identify predictive factors of surgical outcome.*
Material and Method: *Fifteen dystonic patients who received bilateral GPi-DBS were included in the study. Clinical outcomes were evaluated by Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS).*
Results: *Seven cases with primary dystonia had consistent dramatic improvement in pain, motor and bulbar functions though all were negative for DYT1 gene mutation. The mean improvement of BFMDRS was 71.02%. Eight with secondary dystonia had inconsistent improvement of BFMDRS with a mean improvement of 30.49%. Post-stroke dystonia and tardive dystonia had significant sustained improvement of 100% and 97%, respectively. Secondary dystonia from traumatic brain injury had modest sustained improvement of 47%. Dystonia secondary to cerebral palsy, Huntington's disease and CNS infection, showed no improvement. Patients with segmental dystonia improved greater than generalized dystonia regardless of primary or secondary type (82.45% and 20.49%, respectively). Patients whose main symptom was mobile dystonia had more improvement than mixed dystonia and fixed dystonia.*
Conclusion: *Primary dystonia was a strong, good predictive factor. Secondary dystonia from stroke and tardive disorder, segmental and mobile dystonia seemed to be good predictive factors. Dystonia, secondary to cerebral palsy, Huntington's disease and CNS infection was a poor predictive factor.*

Keywords: *Dystonia, Globus pallidus internus, Deep brain stimulation*

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Dystonia is a chronic neurological disorder that causes muscles contracting involuntarily, forcing certain parts of the body, including face, neck, vocal cords, trunk, legs, and arms into abnormal appearance and sometimes causes painful movements or postures. Some forms of dystonia, termed "primary dystonia", are inherited. Other secondary forms of dystonia are caused by traumatic brain injury, stroke, tumor, infection, and exposure to certain drugs or toxins. These conditions, in which a pathological process occurs in the basal ganglia, induce a decrease in the inhibition of the brain cortex[1,2].

Dystonia involving focal area of the body, such as task-specific focal hand dystonia or cervical dystonia can usually be treated effectively by botulinum toxin injection. However, in patients with widely pervasive dystonia, such as generalized dystonia, segmental dystonia or hemidystonia, botulinum toxin injection and other medical treatments are not effective for rare cases of dopamine responsive dystonia[3].

These patients typically suffer life-long from motor dysfunction, bulbar dysfunction, pain and finally get worse from secondary orthopedic complications. Recently, deep brain stimulation of the globus pallidus interna (GPi-DBS) has emerged as a new treatment for

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these seemingly hopeless patients[4-10]. It significantly relieves abnormal movements and postures associated with dystonia. Surgical outcomes are especially excellent for primary general dystonia with DYT1 mutation[4,9,11,12]. This surgical treatment was approved by the Food and Drug Administration in 2003. After that there were studies of surgical outcome of GPi-DBS[4,6-10,13-18]. However, the number of enrolled patients in the studies was small, and physicians could not predict which patients with dystonia respond to the surgical therapy. In contrast to DBS for Parkinson's disease for which the surgical outcome can be predicted by pre-operative levodopa challenge test[19,20], there is no challenge test for dystonia. It is always uncertain how much dystonia will improve after surgery. Most studies tended to show reproducible improvement in patients with primary general dystonia[4,5,9,12,17,21], but inconsistent and inconclusive outcomes were found in patients with secondary dystonia[7,12-15,18,22-24].

The primary objective of our study is to evaluate the efficacy of GPi-DBS from a relatively large single center study. The secondary objective is to investigate predictive factors of outcome, if there are any.

Material and Method

Patient population

From 2004 to 2010 the senior author (SN) has performed bilateral GPi-DBS in 19 patients with generalized or segmental dystonia. Of them, 15 cases operated in this period, had complete data and VDO records for retrospective analysis and were enrolled in the study. All patients met the criteria of dystonia by Fahn[25,26]. There were 14 adults and 1 child; 9 male and 6 female. Seven were primary (idiopathic) dystonia and 8 were secondary dystonia due to various etiologies. The distribution of dystonia was segmental in 7 and generalized in 8. The character of symptom was mobile dystonia in 4, fixed dystonia in 8 and mixed (or undetermined) pattern in 3.

Clinical evaluation

All data were collected, including age of onset, age at surgery, duration of symptoms, familial history of dystonia, distribution of dystonia (segmental or generalized), characters of dystonia (mobile, fixed or mixed), type of dystonia (primary or secondary), causes of secondary dystonia, such as head trauma, CNS infection, stroke, psychiatric medication, cerebral palsy. The presence of the mutation 946del GAG was used to screen for DYT1 mutation in all cases of primary dystonia[27]. Dystonia was assessed before and after GPi-DBS by an investigator who did not involve in the surgery and was unaware of the treatment status (PN) using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS)[28]. The BFMDRS is a 120-point scale used to rate the severity of dystonia in nine body regions, taking into account both severity of the dystonic movements and frequency which they are provoked. The higher the score, the greater the severity of dystonia.

Benefits of the procedure were calculated as a percentage of improvement by the following equation: (pre-operative BFMDRS score-postoperative BFMDRS score) x 100/pre-operative BFMDRS score

Surgical procedure

Axial acquisition inversion recovery MRI brain was done several days before the operation. This MRI pulse sequence allows visualization of all boundaries of the globus pallidus interna and optic tract. On operative day, a stereotactic frame was applied rigidly to patient's head along the inferior orbitomeatal line. Stereotactic CT brain was done and transported to a stereotactic planning workstation. The stereotactic CT images were fused volumetrically with the pre-operative inversion recovery MRI. The images then were reconstructed orthogonal to the commissural plane. Stereotactic targeting was done by a combination of indirect targeting and direct targeting. The Y Cartesian coordinate was set at 2.0 mm anterior to the midcommissural point in all patients. The X and Z Cartesian coordinate were defined individually at the dorsolateral border of the optic tract. The surgical procedures were performed under local anesthesia in most patients. However, general anesthesia was used in very young patients (case No. 12) and patients who had severe abnormal cervical posture or severe mobile dystonia (case No. 1, 2, 4 and 15). We used a true parasagittal trajectory inclining 60 to 70 degree to the commissural plane, with avoidance of vessels, sulci and ventricle. Microelectrode recording (MER) was done for intraoperative nuclear localization. Dystonia is a clinical syndrome of heterogeneous etiologies, there is no specific MER pattern of the target. Thus, the globus pallidus interna was defined as a nucleus structure between the silent zone of internal medullary lamina and ansa lenticularis. Typically, two tracks were required on each side. The first track attempted to find the typical pallidotomy target. Namely, a track in which the globus pallidus interna was at least 5-6 mm in length and an optic tract presented at its base. After

successful discovery of the typical pallidotomy track, the final track for implantation of stimulating lead was put 2 mm more laterally. This final position was quite close to the internal medullary lamina and was 3.5 to 4 mm away from the pallidocapsular border. Macrostimulation was done at the bottom of the globus pallidus interna by the recording electrode. The electrical parameters were constant current stimulation, negative monophasic square wave, pulse width 0.06 ms and frequency 130 Hz. The final target should not have electrical threshold for any adverse effects lower than 4.5 mA. For patients who were operated under general anesthesia, assessment of macrostimulation-induced adverse effects was unreliable and abandoned. These patients needed multiple MER tracks of at least three tracks on each side to comprehensively map the posterovental portion of globus pallidus interna and accurately place the lead 3.5 to 4 mm away from the pallidocapsular border. After bilateral lead insertion, an internal pulse generator was implanted under general anesthesia.

Statistical analysis

The mean ± SD and median (range) pre-operative and postoperative absolute scores of the BFMDRS and the percentage of improvement were calculated and tested with paired t-test and Mann-Whitney U test. The statistical software, SPSS 10.0 for window (SPSS. Inc., Chicago IL, USA), was used for statistical analysis.

Results

Summary of patients' data is presented in Table 1. The mean (±SD) age at onset of symptoms was 35.8±13.7 years, and the mean age at surgery was 35.3±14 years. The postoperative follow-up period ranged from 2 months to 5 years. The mean baseline BFMDRS score was 53.1±33 (Table 2). All patients (case No. 1 to 7) with primary dystonia were negative for the DYT1 gene mutation. There were 2 patients whose MRI revealed abnormal findings (case No. 9 and No. 11). One with secondary dystonia (case No. 9) had undergone prior intracranial surgery for hemorrhagic arterovenous malformation.

At the time of surgery, all patients were severely disabled in their performance of daily activities. After GPi-DBS in primary dystonia group, abnormal postures and dystonic movements decreased considerably. The motor functions greatly improved with a mean improvement of 71±15.4% (Table 2). However, in the secondary dystonia group, there was variability in the context of improvement with a lower mean improvement of 30.5±45% (Table 2). Dystonia, secondary to stroke and tardive disorder, had significant sustained improvement of 100% and 97%, respectively. Dystonia secondary, to traumatic brain injury, had modest sustained improvement of 47%. Dystonia, secondary to cerebral palsy (3 cases), Huntington's disease and CNS infection, showed no improvement. We compared the percentage of improvement between the primary and secondary group, but found no significant differences (p = 0.126, Table 2 and Fig. 1).

In addition to etiology, other factors, such as age of onset, duration of symptom, distribution of dystonia and character of dystonia were analyzed. Age of onset and duration of symptom showed no relationship with the surgical outcome, but distribution and character of dystonia showed some predictive value. Segmental dystonia and mobile dystonia tended to be good predictive factors regardless of primary or secondary type (Fig. 2 and 3). Patients with segmental dystonia had an average improvement of 82.5±15.4%, which was significantly higher than 20.5±29.2% improvement in patients with generalized dystonia. Patients whose main symptom was mobile dystonia had an average improvement of 75.1±8.2% while mixed dystonia and fixed dystonia had an average improvement of only 51.9±48.8% and 35.6±42.4%, respectively.

Discussion

Our data confirmed efficacy of GPi-DBS in the treatment of patients with severe dystonia. There were improvement in pain, motor and bulbar functions after the surgery. Bulbar symptoms, such as spasmodic dysphonia and laryngopharyngeal dystonia, which did not improve or even got worse after deep brain stimulation (DBS) for Parkinson's disease, were noticed improving very well in dystonic patients.

The improvement of BFMDRS score was higher in primary dystonia than in secondary dystonia. There was no statistical difference (p = 0.126) because some patients in secondary dystonia group showed excellent improvement after implantation as well. The superior efficacy in primary dystonia over secondary dystonia has previously been observed[11,14,29]. Reported series showed DBS being effective in most cases of primary dystonia, though these series was composed of a limited number of patients and had short follow-up periods[9,10,29].

When any patients with Parkinson's disease

Table 1. Clinical characteristics

Case No.	Age at surgery (years)	Dystonia type	Distribution of symptom	Characteristic of symptom	Duration of symptom (years)	BFMDRS score		
						Preop	Postop	Improvement (%)
1	27	Primary, DYT1-	Generalized	Fixed	13	92	50	46.67
2	31	Primary, DYT1-	Segmental	Mobile	1	33	4.5	86.36
3	39	Primary, DYT1-	Segmental	Mobile	2	28	9	67.85
4	20	Primary, DYT1-	Generalized	Mobile	2	69	20.5	70.28
5	55	Primary, DYT1-	Segmental	Mixed	23	17	7	58.82
6	42	Primary, DYT1-	Segmental	Mobile	7	66	16	75.75
7	51	Primary, DYT1-	Segmental	Fixed	11	41	3.5	91.46
8	52	Secondary, tardive	Segmental	Mixed	7	32.5	1	96.92
9	35	Secondary, stroke	Segmental	Fixed	3	8	0	100
10	47	Secondary, TBI	Generalized	Fixed	10	68	36	47.05
11	31	Secondary, CNSI	Generalized	Fixed	7	15.5	15.5	0
12	14	Secondary, CP	Generalized	Fixed	12	116.6	116.6	0
13	22	Secondary, CP	Generalized	Mixed	9	52	52	0
14	52	Secondary, HD	Generalized	Fixed	6	53	53	0
15	19	Secondary, CP, CNSI	Generalized	Fixed	17	106	106	0

CNSI = central nervous system infection; CP = cerebral palsy; DYT1- = DYT1-negative; HD = Huntington's disease; TBI = traumatic brain injury; Postop = postoperative; Preop = preoperative

Table 2. Comparison of preoperative, postoperative BFMDRS score and percentage of improvement between primary, secondary and overall dystonia groups

	Primary dystonia			Secondary dystonia			Overall cases			*p*-value
	Mean±SD	Median	Range	Mean ± SD	Median	Range	Mean ± SD	Median	Range	
Preop BFMDRS	49.4±26.8	41	17-92	56.4±39.2	52.5	8-116	53.1±33.0	52	8-116	0.908
Postop BFMDRS	15.8±16.0	9	4-50	47.4 ±44.3	44	0-116	32.7±36.9	16	0-116	0.247
Improvement (%)	71.0±15.4	70.3	46.6-91.5	30.5±45.0	0	0-100	49.4±39.4	58.8	0-100	0.126

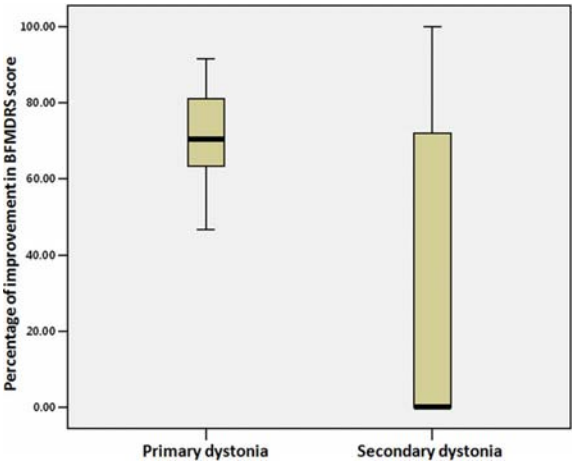

Fig. 1 Box-and-Whisker plots show a comparison of percentage of improvement in BFMDRS score between primary and secondary dystonia, the dark transverse line within the box indicates median percentage of improvement.

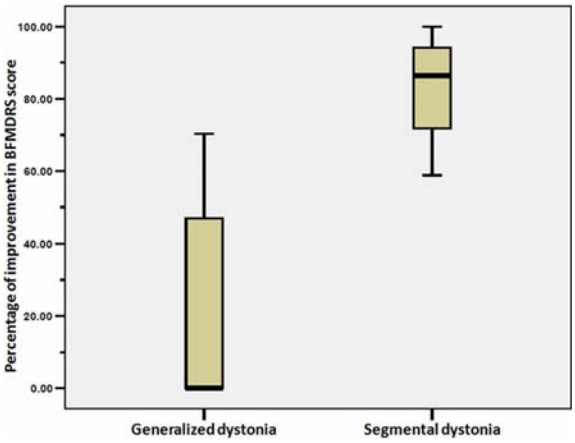

Fig. 2 Box-and-Whisker plots show a comparison of percentage of improvement in BFMDRS score between generalized and segmental dystonia, the dark transverse line within the box indicates median percentage of improvement.

undergo DBS, surgical outcome can be predicted by their character of symptoms and response to levodopa challenge test. Unfortunately, there is no such an analogous test for dystonia. As a result, many patients with dystonia undergo just trials of surgery without any improvement. Considering high cost of the device and risks of brain surgery, it is necessary to investigate for criteria of proper patient selection. Base on the results of our study and extensive review of literature[8,9,13-15,22,29,30-35], we propose a concept of

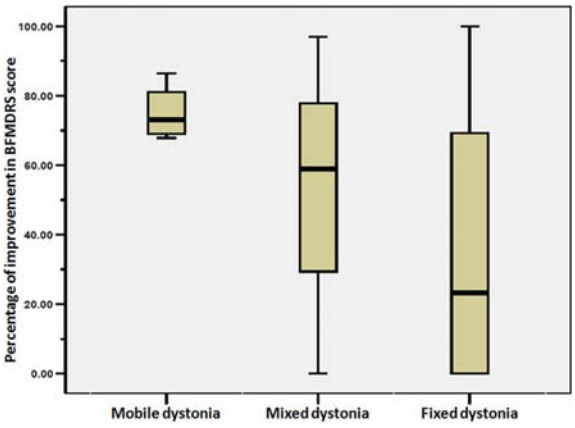

Fig. 3 Box-and-Whisker plots show a comparison of percentage of improvement in BFMDRS score between mobile, mixed and fixed dystonia, the dark transverse line within the box indicates median percentage of improvement.

Table 3. Factors predicting surgical outcome following GPi-DBS

Good predictive factors
Primary dystonia
Segmental dystonia
Mobile dystonia
DYT1 dystonia[4,9,11,12]
Tardive dystonia[2,3,14,22,35]
Fair predictive factors
Pantothenate kinase-associated neurodegeneration (PKAN) or Hallervorden-Spatz syndrome[5,9,22,30]
Hemidystonia[16,31,33]
Post-stroke dystonia[31]
Post-traumatic dystonia[19,22]
Poor predictive factors
Huntington's disease[34]
Post-infectious dystonia[22]
Dystonic cerebral palsy[22]

patient selection as shown in Table 3. It is not possible to verify the accuracy of the concept by this study and larger surgical data to validate this concept are required.

Even though pallidal DBS yields excellent results in DYT1 primary dystonia, several studies revealed favorable outcomes of DBS in non-DYT1 primary dystonia[7,36-38]. Our study also showed marked clinical improvement in the patients with DYT1-negative primary dystonia after GPi-DBS. These results indicate that DBS surgery is useful in patients suffering from primary dystonia regardless of a known genetic cause.

Limitation of the present study is the number of patients. Because the disease is uncommon in Thai and the cost of surgical device is high, the number of generalized or segmental dystonic patients operated by DBS have been relatively scant. A large number of surgical data will never be possible in single center in Thailand and we call for a co-operative multicenter study.

Conclusion

GPi-DBS effectively alleviates disabling dystonic symptoms in patients with severe dystonia. This study shows that primary dystonia, regardless of DYT1-mutation, segmental dystonia and mobile dystonia, are good surgical candidates for GPi-DBS.

What is already known on this topic?

Dystonia is a rare entity of movement disorders. GPi-DBS has become an alternative treatment for severe and refractory cases. DYT1 dystonia often responds well to GPi-DBS, whereas secondary dystonia renders less improvement after the surgery.

What this study adds?

DYT1 dystonia was not found in the present study. Factors predicting favorable surgical outcome following GPi-DBS include primary, segmental and mobile dystonia, whereas poor surgical predictor is secondary dystonia caused by Huntington's disease, CNS infection and dystonic cerebral palsy.

Potential conflicts of interest

None.

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ผลของการรักษาโรคดิสโทเนียชนิดต่าง ๆ โดยการผ่าตัดกระตุ้นสมองส่วนโกลบัสพาลิดัส

ศรัณย์ นันทอารี, ปรีดิ์ นิมมานนิตย์, บรรพต สิทธินามสุวรรณ, อภิชาติ พิศาลพงศ์, กนกวรรณ บุญญพิสิฏฐ์

ภูมิหลัง: *การผ่าตัดกระตุ้นสมองส่วนโกลบัสพาลิดัสอินเทอร์นาได้รับการยอมรับอย่างแพร่หลายสำหรับรักษาโรคดิสโทเนียที่เป็นรุนแรง*

วัตถุประสงค์: *เพื่อศึกษาและเปรียบเทียบประสิทธิภาพของการผ่าตัดกระตุ้นสมองส่วนโกลบัสพาลิดัสอินเทอร์นา สำหรับโรคดิสโทเนียชนิดต่าง ๆ และหาปัจจัยที่บ่งชี้การตอบสนองต่อการรักษา*

วัสดุและวิธีการ: *การศึกษานี้รวบรวมผู้ป่วยดิสโทเนีย 15 ราย ซึ่งได้รับการผ่าตัดกระตุ้นสมองส่วนโกลบัสพาลิดัสอินเทอร์นาทั้งสองข้างโดยใช้ Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) สำหรับประเมินผลการรักษาโดยการผ่าตัด*

ผลการศึกษา: *ผู้ป่วย 7 รายเป็นดิสโทเนียซึ่งไม่ทราบสาเหตุ ทุกรายตรวจไม่พบการกลายพันธุ์ของจีน DYT1 ผู้ป่วยกลุ่มดังกล่าวมีการดีขึ้นของอาการปวดระบบประสาทสั่งการและการทำงานของก้านสมองพบว่าคะแนน BFMDRS ดีขึ้นโดยเฉลี่ยร้อยละ 71 หลังผ่าตัดผู้ป่วย 8 รายเป็นดิสโทเนียซึ่งทราบสาเหตุมีคะแนน BFMDRS ดีขึ้นโดยเฉลี่ยร้อยละ 30.5 หลังผ่าตัด โดยผู้ป่วยดิสโทเนียซึ่งมีสาเหตุจากโรคหลอดเลือดสมองมีคะแนน BFMDRS ดีขึ้นร้อยละ 100 และผู้ป่วยดิสโทเนียซึ่งมีสาเหตุจากการใช้ยารักษาโรคจิตเวชมีคะแนน BFMDRS ดีขึ้นร้อยละ 97 ผู้ป่วยดิสโทเนียซึ่งมีสาเหตุจากการบาดเจ็บของสมองมีคะแนน BFMDRS ดีขึ้นเพียงร้อยละ 47 ในขณะที่ผู้ป่วยดิสโทเนียซึ่งมีสาเหตุจากโรคสมองพิการโรคฮันติงตันและการติดเชื้อของระบบประสาทส่วนกลางไม่มีรายใดมีคะแนน BFMDRS ดีขึ้นเลย ผู้ป่วยดิสโทเนียบางส่วนของร่างกายมีคะแนน BFMDRS ดีขึ้นร้อยละ 82.5 เมื่อเปรียบเทียบกับผู้ป่วย ดิสโทเนียทั้งร่างกายพบว่าผู้ป่วยกลุ่มนี้มีคะแนน BFMDRS ดีขึ้นเพียงร้อยละ 20.5 โดยไม่เกี่ยวข้องกับการทราบหรือไม่ทราบสาเหตุของดิสโทเนีย นอกจากนี้พบว่าผู้ป่วยดิสโทเนียแบบเคลื่อนไหวมีอาการดีขึ้นมากกว่าผู้ป่วยดิสโทเนียแบบผสมและแบบอยู่นิ่ง*

สรุป: *โรคดิสโทเนียที่ไม่ทราบสาเหตุเป็นตัวพยากรณ์ที่สำคัญสำหรับผลการรักษาที่ดีหลังผ่าตัดโรคดิสโทเนียซึ่งมีสาเหตุจากโรคหลอดเลือดสมอง และการใช้ยารักษาโรคจิตเวช โรคดิสโทเนียบางส่วนของร่างกายและแบบเคลื่อนไหวเป็นตัวพยากรณ์ผลการรักษาที่ดีหลังผ่าตัดเช่นเดียวกัน ในขณะที่โรคดิสโทเนียซึ่งมีสาเหตุจากโรคสมองพิการ โรคฮันติงตัน และการติดเชื้อในระบบประสาทส่วนกลางเป็นตัวพยากรณ์ผลการรักษาที่ไม่ดีหลังผ่าตัด*