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Dystonia and DBS: The Jury Arrives

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1. Introduction

Even before the deep brain stimulation (DBS) era, stereotactic functional neurosurgery such as thalamotomy and pallidotomy had been used for control of medically intractable dystonia (Cooper, 1976; Lozano et al., 1997). However, unreliability and variability in the results and furthermore, needs for bilateral surgery in most patients with generalized dystonia and the occurrence of unacceptable adverse effects including dysarthria and cognitive impairment have greatly limited their use. In this regard, DBS, which provides a more stable response with fewer side effects, has revolutionized the treatment of dystonia. The first report of DBS for dystonia was by Mundinger in 1977 (Mundinger, 1997). Since then, over the past few decades, bilateral globus pallidus internus (GPi) DBS has emerged as the best therapeutic option for medication-refractory dystonia (Lang, 2011).

Generally, bilateral GPi DBS is effective and safe for primary dystonias whether it is generalized or segmental (Bronte-Stewart et al., 2011). However, its effects on secondary dystonias are variable and generally less favorable (Eltahawy et al., 2004).

This Chapter will focus on factors that should be considered before and after DBS in patients with dystonia and the outcome of GPi DBS for the different forms of dystonia.

2. Mechanism of GPi DBS in dystonia

Since the pathophysiological mechanism of dystonia is not clearly understood, the mechanism by which GPi DBS improves dystonia remains elusive. The proposed mechanism of GPi DBS involves (1) silencing of stimulated neurons, which results in blocking of the pathological outflow from the target structure (i.e. GPi), and (2) introduction of new activity in the network (Hammond et al., 2008). Neuronal activity is altered in the GPi, thalamic ventral oral posterior nucleus (Vop), and subthalamic nucleus (STN) in dystonia (Zhuang et al., 2004); thus, it is suggested that GPi DBS modulates the activity of GABAergic GPi efferent exons, which inhibits Vop neurons through one or both of the above mentioned mechanisms (Hammond et al., 2008).

3. General consideration: Presurgical

3.1 Patient selection

When facing a surgical decision, several factors should be taken into account (Volkmann & Benecke, 2002). First and most important, the diagnosis of dystonia should be correct. It is

especially true for patients with phasic hyperkinetic movement or patients with dystonic tremor because sometimes very careful evaluation is needed to differentiate these conditions from chorea and tremor disorders, respectively. Second, DBS should be considered only when medical treatment has proven to be ineffective. Third, it should be determined whether the target symptom is the predominant source of the disability and severe enough to do surgery despite its cost and the risk of adverse events. Finally, the patient should have the realistic goals and expectations because not all the dystonic symptoms that the patient has had before surgery will disappear or improve after DBS.

3.2 Target selection

GPi is an established and the most commonly used target for DBS in the treatment of dystonia. Many studies have shown that GPi DBS improves motor symptoms and quality of life in patients with medically intractable dystonia.

Recently, several reports showed that STN DBS also improved dystonia and suggested that it may be an alternative target. Bilateral STN DBS improved primary cervical dystonia with an efficacy comparable to that of GPi DBS (Ostrem et al., 2011). Improvements in secondary dystonia such as neurodegeneration with brain iron accumulation (NBIA) also have been reported (Ge et al., 2011; Zhang et al., 2006). Moreover, it has been claimed that STN is a better target than GPi for segmental dystonia because stimulation-related adverse effects such as bradykinesia, which has been repeatedly reported in GPi DBS, does not occur with STN DBS (Ostrem et al., 2011). However, STN is still a novel target for dystonia and further studies are needed to see whether STN DBS is an effective and safe therapy for dystonia. Successful treatment of writer's cramp with thalamic DBS has been reported (Fukaya et al., 2007), but generally, it is not considered as a therapeutic option for dystonia (Andrews et al., 2010).

4. Primary dystonia

4.1 Primary generalized dystonia

Primary generalized dystonia responds well to GPi DBS. Actually, it is the only form of dystonia, in which, the effect of GPi DBS was confirmed by randomized controlled trials. The mean improvement in the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) movement score was 46% at 6 months in one study (Kupsch et al., 2006) and 51% at 1 year and 58% at 3 year in another study without permanent adverse effects (Vidailhet et al., 2005, 2007). A recent long-term follow-up study showed that improvement by GPi DBS was sustained for up to 8 years (Isaias et al., 2009). Although results from early studies suggested that patients positive for DYT1 mutation have a greater benefit (Coubes et al., 2000; Krauss et al., 2003), it is now widely accepted that there is no difference in the outcome between DYT1-posivie and DYT1-negative patients (Isaias et al., 2008, 2011; Kupsch et al., 2006; Vidailhet et al., 2005). Results from a small group of DYT6-positive patients were less favorable, with 16-55% of motor improvement (Groen et al., 2010).

The magnitude of response to GPi DBS varies considerably among patients, and factors possibly associated with poor or good outcomes have been suggested. Patients with diffuse phasic hyperkinetic movements tend to improve more rapidly and better than patients with severe tonic posturing (Kupsch et al., 2006; Vidailhet et al., 2005; Wang et al., 2006). Fixed skeletal deformity, longer disease duration at surgery, older age at surgery, and more severe

motor symptoms at surgery have been associated with a poor outcome (Andrews et al., 2010; Isaias et al., 2008, 2011). Speech and swallowing symptoms are less responsive than axial or limb dystonia (Isaias et al., 2009; Vidailhet et al., 2007), even within an individual patient.

4.2 Cervical dystonia (spasmodic torticollis)

Many case reports and several studies indicate that bilateral GPi DBS is an effective treatment for cervical dystonia (Jeong et al., 2009; Kiss et al., 2007; Kupsch et al., 2006; Pretto et al., 2008). Two long-term follow-up studies showed 67% and 55% improvement in Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) severity scores at 38 and 32 months after surgery, respectively (Cacciola et al., 2010; Hung et al., 2007). TWSTRS pain scores were also reduced by more than 50% and TWSTRS disability score improved by 81% and 59%.

Usually the age of the patient at surgery is greater in cervical dystonia than in generalized dystonia and DBS in older subjects in their 60s and 70s appears to be safe. Until now, there has been not enough data to prove that the age or duration of disease at surgery affects the outcome in cervical dystonia. However, since a longer duration of disease may run a risk of fixed skeletal deformities, DBS should be considered before these problems occur (Bronte-Stewart et al., 2011).

As described above, a recently study reported that STN DBS improved cervical dystonia with an efficacy comparable to that of GPi DBS (Ostrem et al., 2011).

4.3 Craniofacial and craniocervical dystonia (Meige syndrome)

Data from the literature suggest that GPi DBS is an effective and safe treatment for Meige syndrome. Recent case reports with long-term follow-up (1 to 4 years) show sustained improvement in cranio-facio-cervical dystonia by GPi DBS. Improvement in terms of the BFMDRS movement score was 53% in one report and 82 – 86% in the other reports (Ghang et al., 2010; Lyons et al., 2010; Reese et al., 2011; Sako et al., 2011). However, in a recent case series, the effect of GPi DBS on Meige syndrome was variable with some patients having less than 20% improvement (Limotai et al., 2011). Speech and swallowing did not improve. The authors pointed out that a careful re-examination of the selection criteria for surgery for Meige syndrome is needed.

5. Secondary dystonia

5.1 Myoclonus-dystonia (ε-Sarcoglycan mutation, DYT11)

Several case reports showed that bilateral GPi DBS improves motor symptoms in patients with myoclonus-dystonia with an overall improvement of 60% to 90% (Cif et al., 2004; Foncke et al., 2007; Jog & Kumar, 2009; Kurtis et al., 2010). A recent case series of 5 patients with myoclonus-dystonia reported that both myoclonus and dystonia improved with GPi DBS more than 80% and this improvement was sustained after 15-18 months of follow-up (Azoulay-Zyss et al., 2011). Improvement in myoclonus but not in dystonia by thalamic DBS was reported in 2 cases of myoclonus-dystonia (Kuncel et al., 2009; Trottenberg et al., 2001).

5.2 X-linked dystonia parkinsonism (DYT3, 'Lubag')

In the literature, 5 case reports of GPi DBS on X-linked dystonia parkinsonism are available (Aguilar et al., 2011; Evidente et al., 2007; Martinez-Torres et al., 2009; Oyama et al., 2010; Wadia et al., 2010). All cases showed improvement in dystonia. Of note, dysarthria, oromandibular dystonia, and stridor, which usually show poor response to GPi DBS in primary generalized dystonia, also improved. Interestingly, improvements in dystonia were immediate in all cases. However, the effect on parkinsonism was variable: parkinsonism improved in 3 patients but not in the other 2 patients.

5.3 Rapid-onset dystonia parkinsonism (ATA1A3 mutation, DYT12)

Only 2 case reports of bilateral GPi DBS in this rare disease are available. One patient did not receive any benefit from the surgery (Deutschländer et al., 2005) and the other patient had only mild (30%) improvement in dystonia, mainly in the craniocervical and truncal area. Limb dystonia and parkinsonism did not improve (Kamm et al., 2008).

5.4 Tardive dystonia

DBS is a very effective treatment for tardive dyskinesia. Recent studies showed that GPi DBS improves tardive dystonia motor symptoms for more than 80% and this benefit was sustained during long-term follow-up up to 80 months (Capelle et al., 2010; Gruber et al., 2009; Trottenberg et al., 2005). In contrast to primary generalized dystonia, patients experienced distinct improvement within days or even hours after stimulation. Improvements of tardive dystonia with STN DBS also have been reported (Sun et al., 2007; Zhang et al., 2006).

5.5 NBIA

Responses to GPi DBS are variable in NBIA. There are reports of favorable (65-91%) responses (Castelnau et al., 2005; Clement et al., 2007; Krause et al., 2006; Mikati et al., 2009; Umemura et al., 2004), but others reported only 20-30% improvement (Isaac et al., 2007; Shields et al., 2007). A single case report of STN DBS on a NBIA patient showed 84% improvement at 3 years after surgery (Ge et al., 2011). It is surprising that motor symptoms in NBIA can improve with DBS, given that structural abnormalities in the brain MRI usually meet the exclusion criteria for DBS in primary dystonia (Kupsch et al., 2006; Vidailhet et al., 2005).

5.6 Cerebral palsy

A wide range of responses has been reported on the effect of GPi DBS in cerebral palsy. Some patients had favorable outcomes but others experienced no or only minimal improvement (Alterman and Tagliati, 2007; Pretto et al., 2008; Zorzi et al., 2005). This variability in response is most likely due to the heterogeneity of this condition. Recently, a multicenter prospective study investigating the effect of bilateral GPi DBS on dystonia-choreoathetosis cerebral palsy showed 24% improvement in the BFMDRS movement score at 1 year after surgery (Vidailhet et al., 2009). However, as the authors mentioned, cerebral palsy patients who meet the criteria of this study (i.e. prominent dystonia-choreoathetosis, little or no spasticity, unimpaired intellectual function, and only slight abnormalities of the basal ganglia on MRI) was only about 10% of the cerebral palsy population.

5.7 Other secondary dystonias

There are many causes of secondary dystonias and the number of patients with each secondary dystonia who underwent DBS is small. There are reports of DBS in postanoxic dystonia, postencephalitic dystonia, and posttraumatic dystonia (Eltahawy et al., 2004; Pretto et al., 2008; Katsakiori et al., 2009; Zhang et al., 2006; Krause et al., 2004; Ghika et al., 2002). Improvements in dystonia in Lesch-Nyhan syndrome (Cif et al., 2007; Pralong et al., 2005) and GM1 gangliosidosis (Roze et al., 2006) also have been reported. Generally, the effects of DBS on secondary dystonias are variable and less favorable.

6. Task-specific dystonias

For writer's cramp, contralateral unilateral thalamic DBS has been tried with favorable results (Cho et al., 2009; Fukaya et al., 2007). Improvements were immediate in all cases. It appears that thalamic DBS is more effective than GPi DBS for writer's cramp (Fukaya et al., 2007).

7. Complications

There is no compelling evidence that DBS surgery- or device-related adverse effects are more common in dystonia than in Parkinson disease (PD). However, it has been suggested that lead migration and lead fracture is more common in dystonia than in parkinsonian patients (Yianni et al., 2003). Stimulation-related adverse effects specific for GPi DBS in dystonia include the development of reversible bradykinesia and parkinsonian gait problems in previously nondystonic body regions (Berman et al., 2009; Ostrem et al., 2007; Zauber et al., 2009).

8. Postsurgical management

Several points should be kept in mind when managing dystonic patients after DBS surgery (Kupsch et al., 2011).

In contrast to PD where maximal clinical effect of DBS occurs within hours of switching on of the device, the beneficial effects of DBS in dystonia are not immediate and slowly progress over weeks to months, possibly beyond 1 year after surgery. This protracted improvement is more prominent in older patients (Isaias et al., 2011). There is no evidence that tolerance develops with long-term stimulation (Tagliati et al., 2011).

Battery lifetime in GPi DBS for dystonia is usually shorter compared to that in DBS for PD because of higher voltages and greater pulse widths. Thus, a more frequent battery change is required. In a recent study, the mean battery life in patients with GPi DBS for dystonia was 25 months (Blahak et al., 2011). Regarding inadvertent depletion of the battery or discontinuation of stimulation during procedures for battery replacement, it should be noted that sudden bilateral cessation of stimulation can lead to acute and possibly life threatening rebound dystonia or respiratory difficulty (Grabli et al., 2009; Tagliati et al., 2011).

9. Conclusion

So, has the jury arrived at a verdict as to the usefulness of DBS in treatment of dystonia? The answer appears to be yes for primary dystonias. However, for secondary dystonias, more evidences are needed.

Literatures show that bilateral GPi DBS is an effective and safe therapy for medically intractable primary dystonia and it provides a sustained benefit. Not only good surgical technique, but also appropriate selection of patients and individualized postsurgical management are crucial for optimized patient care.

In secondary dystonias, its effects are heterogeneous, and at this stage, data are not enough to determine whether it can be considered as an effective therapy for each form of the disease. Further studies are needed for re-examination of the inclusion criteria and selection of targets other than GPi.

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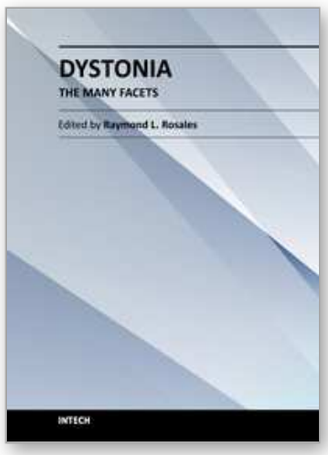
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Dystonia - The Many Facets

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Dystonia has many facets, and among those, this book commences with the increasingly associated genes identified, including a construct on how biology interacts with the dystonia genesis. The clinical phenomenology of dystonia as approached in the book is interesting because, not only were the cervical, oromandibular/lingual/laryngeal, task-specific and secondary dystonias dealt with individually, but that the associated features such as parkinsonism, tremors and spasticity were also separately presented. Advances in dystonia management followed, and they ranged from dopaminergic therapy, chemodenervation, surgical approaches and rehabilitation, effectively complementing the approach in dystonia at the clinics. A timely critical pathophysiologic review, including the muscle spindle involvement in dystonia, is highlighted at the book's end.

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