Successful Treatment of Blepharospasm by Pallidal Neurostimulation

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Blepharospasm, a focal dystonia characterized by involuntary orbicularis oculi muscle spasms,1,2 may vary in severity from a slightly bothersome condition to a disabling disorder that renders patients functionally blind.3 Botulinum toxin (BTX) injection, the gold-standard treatment,1 has a beneficial effect in 95% of patients.4 Oral medication is of limited efficacy. For pharospasm.

globus pallidus interna (GPi)-DBS surgery for isolated blepharospasm,1,3,4 Herein, we present the first case report of possible therapeutic options, although not always with acceptable results.1,3,4

In individuals that do not respond, eyelid myectomy or upper-lid orbicularis muscle strip with subsequent eyebrow suspension are alternative options.3,5 In this case, the patient was considered a candidate for DBS surgery after repeated BTX injections and hospitalization in the movement disorder clinic. During a 4-year period, he received repeated BTX injections and was submitted to two ocularpastic surgeries (myectomy and frontal suspension). Nonetheless, his condition was aggravated and rendered him functionally blind with a Jankovic rating scale of 4 (severity and frequency) bilaterally (see Video 1, Segment A). In October 2013, he was considered refractory to medical therapy and bilateral GPi-DBS was performed. As described previously,5 stereotactic surgery was undertaken under general anesthesia with a Leksell G frame (Elekta Instruments AB, Stockholm, Sweden). Images of stereotactic CT scan and previous MRI were fused. Target coordinates were chosen by direct visualization in MRI. Standard burr holes and dura mater incisions were made, and central, anterior, and lateral recording electrodes were introduced. Intraoperative microrecording was used to define the neurophysiological borders of GPi. Macrostimulation was performed to find the threshold for internal capsular response. The lateral trajectory was the most favorable bilaterally. Definitive electrodes (model 3389; Medtronic, Inc., Fridley, MN) and an Activa RC IPG (Medtronic) were implanted. An implantable pulse generator was programmed on the first postoperative day (left: GPi 8 - 3V / 60μs / 130Hz; right: GPi 0 - 3V / 60μs / 130Hz) and blepharospasm improved gradually over the subsequent weeks achieving a Jankovic rating scale of 1 (severity) and 2 (frequency) on the left eye and of 1 (severity and frequency) on the right eye (see Video 1, Segment B). However, a progressive loss of benefit for the left eye was noted at the seventh month postsurgery. Fusion of postoperative stereotactic CT to preoperative MRI using FrameLink and OPTIVISE (Medtronic) demonstrated a correct positioning of the left electrode within the posteroverentral lateral GPi, whereas the right electrode took a more lateral trajectory ending within the GPc (Fig. 1). Based on image fusion, we selected the two contacts of the right electrode in closest proximity to the GPi and tried an interleaving stimulation protocol (right: GPi 0 - 3.5V / 60μs / 125Hz; 1 - 4V / 90μs / 125Hz; left: GPi 8 - 5.5V / 90μs / 125Hz). After changing the stimulation parameters, the patient’s condition improved, achieving a Jankovic rating scale of 1 (severity) and 2 (frequency) on the left eye and of 1 (severity and frequency) on the right eye (see Video 1, Segment B). However, a progressive loss of benefit for the left eye was noted at the seventh month postsurgery. 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and frequency) on the right eye. The progressive reduction of symptoms granted him total independence for activities of daily living. Hence, up to the time of writing (21 months after surgery), it was not necessary to proceed to electrode reposition given that the same score on the Jankovic scale was maintained.

There is a growing interest in using DBS on patients with craniocervical dystonia (Meige syndrome) who became refractory to other forms of therapy. In 2003, Capelle reported on the first case of pallidal DBS for isolated Meige syndrome with marked improvement of oromandibular dystonia and blepharospasm (2 years after surgery Burke-Fahn-Marsden Dystonia Rating Scale [BFMDRS] score improved by 92% for eyes). Reese demonstrated sustained benefit (BFMDRS subscore for eyes was improved by 47%) of pallidal neurostimulation in 12 patients with this condition for up to 6 years. In the case series of Ghang, (11 patients), Ostrem (6 patients), Sako (5 patients), and Limota (6 patients), positive results were also achieved in patients with Meige syndrome treated with DBS; blepharospasm improved in all them (improvement of 63% in BFMDRS subscores for the eyes at 12 months, 72% in BFMDRS total movement score at 6 months, 84% in BFMDRS total movement score at 49.1 months, and 61.8 ± 30.9% in BFMDRS total movement score at 12 months, respectively).

BTX is still the main treatment option for blepharospasm. When it fails, there are not many options. Given the experience of blepharospasm improving with bilateral GPI-DBS in patients with Meige syndrome, DBS surgery can be an acceptably effective therapy for patients with isolated blepharospasm, like it was in our case. The risk of procedure should be weighed cautiously against the potential benefit.

**Author Roles**


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A.V.: 1A, 1B, 1C, 3A, 3B
L.A.: 1B, 1C, 3B
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References


Supporting Information

A video accompanying this article is available in the supporting information here.

Video 1. Video captured and broadcasted with the patient’s written consent, illustrating the most relevant clinical aspects. Segment A: preoperative video showing a bilateral blepharospasm with Jankovic rating scale of 4 (severity and frequency) on both eyes, rendering the patient functionally blind. Segment B: postoperative video showing much improved bilateral blepharospasm with a Jankovic rating scale of 1 (severity) and 2 (frequency) on the left eye and of 1 (severity and frequency) on the right eye.