

# Good outcome in refractory cervical dystopia with combination of Deep Brain Stimulation on globus pallidus internus nucleus and delayed selective periphery rhizotomy with five years follow up

## Abstract

Cervical dystonia is the most common form of focal dystonia and it has poor options for treatment like botulin toxin, Baclofen, kinesics, which most times are ineffective or have complications. As second line treatments destructive surgical procedures are currently tried first and Deep Brain Stimulation (DBS) by stereotactic implant, later. We present a case in which cervical neurology was done after DBS, with long term good results.

**Keywords:** cranio cervical dystonia, deep brain stimulation, dystonia, peripheral neurotomy

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## Introduction

Cervical dystonia is a common presentation of segmental dystonia, and is currently manifested by involuntary sustained contractions of cervical territory and abnormal head movements or postures, visible and often palpable, what become harmful and disabling. Medical treatment of cervical dystonia often renders limited benefit and side effects are common. Dystonia can be treated by means of medication, botulinum toxin injections, and physiatry.<sup>1</sup> In refractory cases or those who worsens with time in despite of the treatment, surgery could a valid option.<sup>2</sup> The surgical treatment for refractory torticollis has been continuously developed with the aim of both, improvement of outcome and reduce complications. Most of destructive procedures have been less used because their current complications while some other ones have been increasingly more employed because considered safer and more effective.<sup>3-6</sup>

## Case report

As far as we could know, through bibliographical search in Medline, Pubmed and other current international sources, this was the first publication that reports the effect of GPi-DBS in combination with delayed bilateral rhizotomy in cervical dystonia and we have published it in a previous paper.<sup>7</sup> The actual presentation is the continuation of the patient's follow up.

We present a women 58y, in the present days without hereditary history of dystonia; she developed at 52y progressive contracture of muscles of the neck and a feeling that his head was moving backwards. The symptoms were gradually exacerbating to take a stand permanent retrocollis. Treatment with levodopa and botulinum toxin injections were performed (up to 500 IU) without satisfactory results. Secondary causes of dystonia were discarded. As she rapidly showed disabling clinical manifestations none responding to medical therapy, a surgical

approach was suggested, targeting the globus pallidus internus (GPi) nucleus.

The preoperative neurology examination was performed with Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) (severity 20/disability 25/pain 17.75) and Burke-Fahn-Marsden Rating Scale (segmental 20/disability 11).

By micoregistration guided technique a quadripolar electrode (model 7428, Medtronic) was stereotactically implanted bilaterally on GPi and fixed with the Stimloc system. The Magnetic resonance images were processed with WinNeus program to identify coordinates. On a posterior step, the electrodes were connected to a pulse generator (Kinetra; Medtronic). We are currently employing this technique for the treatment of refractory cases in several movement disorders.<sup>8</sup>

In the post – operatory assessment we registered a clear improvement in the dystonic signs. Six months later, there was a 69% mean improvement in the BFMDRS total movement score what meant a  $P < 0.031$ , (Wilcoxin signed rank test). The mean BFMDRS disability score clearly improved ( $P < 0.06$ ). The total TWSTRS score improved 58% ( $P < 0.044$ ). There were no adverse events following the procedure for surgical implant.<sup>7</sup>

Five years after, the dystonic symptoms dramatically returned despite a the correction of the stimulation parameters, so it was indicated a selective peripheral bilateral rhizotomy with Bertrand technique<sup>9</sup> at the left side and Taira technique<sup>10</sup> at the right, in order to reduce risks of complications on neck muscles voluntary movements.

In the immediate post - operative control, one month after surgery, the results returned to values very close as after DBS implantation. In five years of monthly follow up her clinical improvement has not significantly changed.

## Discussion

Palidum stimulation by DBS implantation is a current treatment for Spasmodic Torticollis,<sup>11</sup> but it hasn't solved all problems related to the disease and seldom results are incomplete or worsen with time.<sup>12</sup>

Efforts are being made to improve results by means of technological advances<sup>13</sup> and target selection.<sup>14</sup> It is a common practice in refractory Cranio, cervical Dystonia, to perform DBS after peripheral surgery has failed,<sup>15</sup> but not the converse, as in our case.

## Conclusion

We conclude that we need more studies and long-term monitoring to determine more adequate indications for the beneficial combination of these two procedures in this sequential order, as shows the long-term successful outcome in this case.

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## Conflict of interest

Author declares that there is no conflict of interest.

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