



Deep Brain Stimulation or Pallidotomy for Dystonia in Neurodegeneration with Brain Iron Accumulation? A Call for Justice to the Balance

David Armando Guach-Hevia, MD*

Medical Sciences University, Holguin, Cuba



Letter

Neurodegeneration with brain iron accumulation (NBIA) comprises a group of rare but devastating inherited neurological diseases with unifying features of gradual cognitive and motor decline, and progressive degeneration of basal ganglia, globus pallidus, and reticular part of substantia nigra, produced by brain iron accumulation [1]. The most common disorders presented in infancy and childhood are Betapropeller Protein-Associated Neurodegeneration, Pantothenate Kinase-Associated Neurodegeneration, Phospholipase A2-Associated Neurodegeneration, and Mitochondrial membrane Protein-Associated Neurodegeneration [2].

Clinical presentations include dystonia, dysarthria, dysphagia, dementia, severe mental retardation, and severe movement disability at later stages [1]. Among the movement disorders described in the majority of these patients dystonia is a practically constant clinical feature that progresses along with the neurodegeneration. Treatment in most patients is often challenging, and remains mainly symptomatic for the majority of cases [2]. Since the definitive therapies are still lacking, symptomatic management by means of surgical interventions have played an immense role in improving the quality of living of these patients.

With the refinement of surgical techniques, the past few decades have witnessed a significant upsurge in the role of surgical interventions as a part of treatment protocol in medically refractory movement disorders of various aetiologies [3]. Surgical treatments have proven to be efficacious for a number of the NBIA's. Although these are by nature palliative, they can bring marked symptomatic improvement and can be targeted and individualized. Cerebral ablative surgery, also called functional or lesional surgery, mostly applied since the late part of the twentieth century for movement disorders and historically used for the treatment of advanced Parkinson's Disease, have been also considered on the therapy of refractory dystonia in several rare diseases with motor compromise, including NBIA's, having the Globus Pallidus pars interna (Gpi) as the main target [4].

In addition, as medical technology progresses deep brain stimulation (DBS) has showed up as the leading neurosurgical treatment for many movement disorders including

Parkinsonism, of course, and a wide range of dystonic related conditions, including those due to brain iron accumulation. There are a number of case series reporting outcomes after DBS in NBIA [3,5-9].

Since then, DBS has become the gold standard surgical procedure for practically all types of dystonia, leading to a decrease in the number of publications related to current ablative surgeries, partly due to DBS's advantages well promoted by the industry such as reversibility, programmability and bilateral targeting precision. Subsequently, the popularity of DBS increased and the charm of pallidotomy decreased [4]. Concerned about this intriguing topic, Hariz M. in his article Pallidotomy for Dystonia: A Neglected Procedure?, claims that the introduction of DBS for parkinsonian and essential tremor in 1987, and the subsequent global spread of DBS have overshadowed all previous ablative surgical procedures for movement disorders including posteroventral pallidotomy for dystonia [10]. Therefore, a question emerges: why has this happened?

About this issue very few studies comparing DBS and ablative surgery outcomes while treating dystonia have been carried out, all showing a quite similar clinical improvement in post-surgery evaluation while using the Burke-Fahn-Marsden-Dystonia-Rating Scale (BFMDRS). Dhar D et al. in their cohort exposed the results of ten patients that underwent surgeries primarily for dystonia (DBS-7, pallidotomy-3), including NBIA's cases. Irrespective of the type of surgical intervention, statistically significant improvement was noted in the BFMDRS motor scores at 6-months ($p < 0.001$) and 12-months interval ($p = 0.02$) compared to the baseline, and

***Corresponding author:** David Armando Guach-Hevia MD, Medical Sciences University, Holguin, Cuba

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the comparison between GPi DBS and bilateral pallidotomy showed no significant difference in mean BFMDRS motor scores at baseline ($p = 0.66$), 6 months ($p = 0.93$) or 12 months follow-up period ($p = 0.48$) [3]. Furthermore, authors like Centen et al. on their review about bilateral pallidotomy for dystonia highlights the potential of this ablative surgery, not only as first choice when DBS is not available, but also in cases when DBS fails or has to be withdrawn because of infection and other hardware complications such as an arrest of chronic DBS [11].

However, despite these remarkable and well oriented results, its controversial the lack of studies about treating dystonia by applying pallidotomy or other targets of ablative surgery, instead of using DBS, even when considering how expensive this last procedure is, and that in fact, it is not available in a wide list of countries with low income economies.

Thus, a combination of, on the one hand, publications of a couple of randomized trials of DBS for dystonia providing “evidence-based” results and, on the other hand, relentless promotion of DBS by industry, leading to an almost complete loss of training in ablative surgery of younger neurosurgeons, has sealed the coffin over pallidotomy [10].

Subsequently some justice is needed on the balance. Although DBS is preferred, the disadvantages over pallidotomy include the risk of infection of the implants, especially in immunocompromised patients [12]. In addition, requirement for change of implantable pulse generator is higher in dystonia [13], and the patients need to be under close follow due to the frequent need of programming [4]. In fact, there are cases in which patients have developed fatal dystonic storm because the battery was depleted and a replacement battery was not readily available for financial issues or other reasons [14] or because a severe status dystonicus failed to respond to DBS [15]. Bilateral pallidotomy could then have saved the patients’ lives [10].

That is why further and larger studies about ablative surgery for the treatment of dystonia or other clinical features of neurodegenerative diseases, such as NBIA, are paramount to be performed in order to vindicate its proper training on current neurosurgery, and to reinforce its potential as a primary option when DBS is not available, or when the patient is at high risk of infection or have financial constraints, or in patients who cannot be adequately followed up.

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References

1. Labrada-Aguilera PE, Guach-Hevia DA, Almira-Gómez CR, et al. (2022) Type 1 neurodegeneration with brain iron accumulation: a case report. *J Med Case Rep* 16: 217.
2. Spaul RVV, Soo AKS, Hogarth P, et al. (2021) Towards Precision Therapies for Inherited Disorders of Neurodegeneration with Brain Iron Accumulation. *Tremor Other Hyperkinet Mov* 11: 51.
3. Dhar D, Holla VV, Kamble N, et al. (2022) Surgical Outcomes in Rare Movement Disorders: A Report of Seventeen Patients from India and Review of Literature. *Tremor Other Hyperkinet Mov* 12: 22.
4. Garg K, Samala R, Agrawal M, et al. (2020) Pallidotomy for Dystonia. *Neurol India* 68: S322-S324.
5. Castelnau P, Cif L, Valente EM, et al. (2005) Pallidal stimulation improves pantothenate kinase-associated neurodegeneration. *Ann Neurol* 57: 738-741.
6. Mikati MA, Yehya A, Darwish H, et al. (2009) Deep brain stimulation as a mode of treatment of early onset pantothenate kinase-associated neurodegeneration. *Eur J Paediatr Neurol* 13: 61-64.
7. Timmermann L, Pauls KAM, Wieland K, et al. (2010) Dystonia in neurodegeneration with brain iron accumulation: outcome of bilateral pallidal stimulation. *Brain* 133: 701-712.
8. Lumsden DE, Kaminska M, Gimeno H, et al. (2013) Proportion of life lived with dystonia inversely correlates with response to pallidal deep brain stimulation in both primary and secondary childhood dystonia. *Dev Med Child Neurol* 55: 567-574.
9. Artusi CA, Dwivedi A, Romagnolo A, et al. (2020) Differential response to pallidal deep brain stimulation among monogenic dystonias: systematic review and meta-analysis. *J Neurol Neurosurg Psychiatry* 91: 426-433.
10. Hariz M (2021) Pallidotomy for Dystonia: A Neglected Procedure? *Mov Disord* 36: 533-534.
11. Centen LM, Oterdoom DLM, Tijssen MAJ, et al. (2021) Bilateral Pallidotomy for Dystonia: A Systematic Review. *Mov Disord* 36: 547-557.
12. Singh M, Shabari Girishan KV, Bajaj J, et al. (2018) Deep Brain Stimulation for movement disorders: Surgical nuances. *Neurol India* 66: S122-S130.
13. Okun MS, Vitek JL (2004) Lesion therapy for Parkinson’s disease and other movement disorders: Update and controversies. *Mov Disord* 19: 375-389.
14. Rohani M, Munhoz RP, Shahidi G, et al. (2017) Fatal status dystonicus in tardive dystonia due to depletion of deep brain stimulation’s pulse generator. *Brain Stimul* 10: 160-161.
15. Nerrant E, Gonzalez V, Milesi C, et al. (2018) Deep brain stimulation treated dystonia-trajectory via status dystonicus. *Mov Disord* 33: 1168-1173.

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