Isolated dystonia

The term "isolated dystonia" is applied when dystonia is the only movement disorder identified, with or without tremor. ... When isolated dystonia is encountered in children and adolescents, the etiology is often genetic; whereas isolated dystonia in adults is most likely to be sporadic.

Surgical procedures involving deep brain stimulation (DBS) of the globus pallidus internus (GPi) or subthalamic nucleus (STN) are well-established treatments for isolated dystonia. However, selection of the best stimulation target remains a matter of debate.

Lin et al., from the Ruijin Hospital published a study comparing the effectiveness of DBS of the GPi and the STN in patients with isolated dystonia.

In this matched retrospective cohort study, they searched an institutional database for data on all patients with isolated dystonia who had undergone bilateral implantation of DBS electrodes in either the GPi or STN in the period from January 30, 2014, to June 30, 2017. Standardized assessments of dystonia and health-related quality of life using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) and SF-36 were conducted before and at 1, 6, and 12 months after surgery. No patients were lost to the 6-month follow-up; 5 patients were lost to the 12-month follow-up.

Both GPi (14 patients) and STN (16 patients) stimulation produced significant improvement in dystonia and quality of life in all 30 patients found in the database search. At the 1-month follow-up, however, the percentage improvement in the BFMDRS total movement score was significantly (p = 0.01) larger after STN DBS (64%) than after GPi DBS (48%). At the 12-month follow-up, the percentage improvement in the axis subscore was significantly (p = 0.03) larger after GPi DBS (93%) than after STN DBS (83%). Also, the total amount of electrical energy delivered was significantly (p = 0.008) lower with STN DBS than with GPi DBS (124 ± 52 vs 192 ± 65 µJ, respectively).

The GPi and STN are both effective targets in alleviating dystonia and improving quality of life. However, GPi stimulation may be better for patients with axial symptoms. Moreover, STN stimulation may produce a larger clinical response within 1 month after surgery and may have a potential economic advantage in terms of lower battery consumption ¹⁾.

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Lin S, Wu Y, Li H, Zhang C, Wang T, Pan Y, He L, Shen R, Deng Z, Sun B, Ding J, Li D. Deep brain stimulation of the globus pallidus internus versus the subthalamic nucleus in isolated dystonia. J Neurosurg. 2019 Mar 8:1-12. doi: 10.3171/2018.12.JNS181927. [Epub ahead of print] PubMed PMID: 30849756.

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