

Prior Thalamotomy Diminishes the Response of Pallidal Deep Brain Stimulation for Primary Dystonia

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Objective: Deep brain stimulation (DBS) at the globus pallidus pars internus (GPI) improves the motor function of patients with generalized primary Dystonia. Patients with the DYT1 gene defect are reported to be particularly responsive. Presently, little is known about the impact of previous neuroablative procedures on the response of Dystonia patients to GPI DBS. Herein, we report our experience with three such cases.

Methods: Retrospective analysis of three patients with DYT1+ primary generalized Dystonia who underwent microelectrode-guided GPI DBS surgery at our institution. All had undergone bilateral thalamotomies during childhood with limited results and all were deemed to be medically refractory by our movement disorders neurologist. The response to surgery was assessed using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS), which was administered one week prior to surgery and 3, 6, and 12 months after the onset of stimulation.

Results: All three patients are females of Ashkenazi Jewish descent and each possesses the DYT1 gene defect. At the time of surgery they were aged 40, 28, and 40 years, respectively. Their ages at the onset of symptoms were 7, 8, and 3 years. Patients 1 and 3 underwent bilateral GPI DBS implantations; patient 2 received a left GPI implant. Baseline BFMDRS scores were: 99, 40, and 98 (mean: 79). Six months after surgery the scores were: 90, 37, and 73 (mean: 67), representing a marginal improvement (15%). Patient 1 has been followed for three years. At her 36-month follow-up her BFMDRS score was 72, a 27% improvement as compared to baseline. Patient 2 has received one year of therapy. Her 12-month BFMDRS score was 22.5, an improvement of 44%. Patient 3 has been followed for 6 months. Her BFMDRS score is improved 25.5%. In contrast, five patients with DYT1+ generalized primary Dystonia who had not undergone previous neuroablative surgery have experienced a mean improvement in their BFMDRS scores of 69.8% (range: 48-85%).

Conclusions: These results in a limited series of patients suggest that previous neuroablative surgery and thalamotomies in particular, may interfere with the response of DYT1+ Dytsonia patients to GPI DBS therapy.