Deep Brain Stimulation for Children with Intractable Secondary Dystonia: 3 Cases

Douglas Anderson, Jacquelyn Hill, Kurt Grahnke, Xabier Beristain, Mary Keen
Loyola University Medical Center

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Introduction

Deep brain stimulation (DBS) of the globus pallidus interna (GPi) has been used in the treatment of intractable dystonias in the pediatric population and presently carries a humanitarian device exemption (HDE). While effective in the treatment of primary generalized dystonia, it has been more challenging to assess the initial and long term effects of DBS for secondary dystonia, especially in children. We have performed GPi DBS on three pediatric patients with intractable secondary dystonia refractory to medicines.

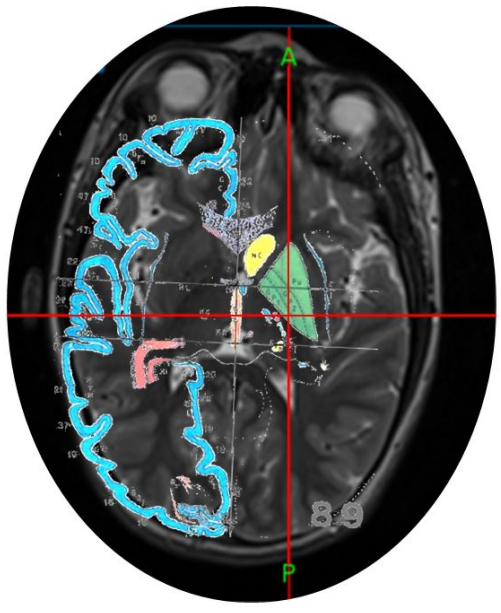
Methods

Our patients were evaluated by developmental pediatric specialists and referred for evaluation of surgery because of painful secondary dystonia associated with decreases in motor function. In conjunction with a movement disorders neurologist, we obtained IRB approval for this study. For those receiving surgery, DBS leads are placed under general anesthesia into the posterior medial GPi with neuronavigation and CRW frame (x= 20-23 mm lateral to the AC-PC line, -y= 2-3 mm anterior to the midcommissural point, -z= 2-5 mm below the AC-PC line). We used micro-electrode recording to supplement anatomical localization of the targets. Electrodes were implanted in one or two stages, followed by separate surgeries for each pulse generator/battery.

Methods cont.

Patients are evaluated post-operatively using the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) and follow-up clinic visits. Also, we sought observations via interviews from parents and teachers of the children, of functional changes observed.

Figure 1

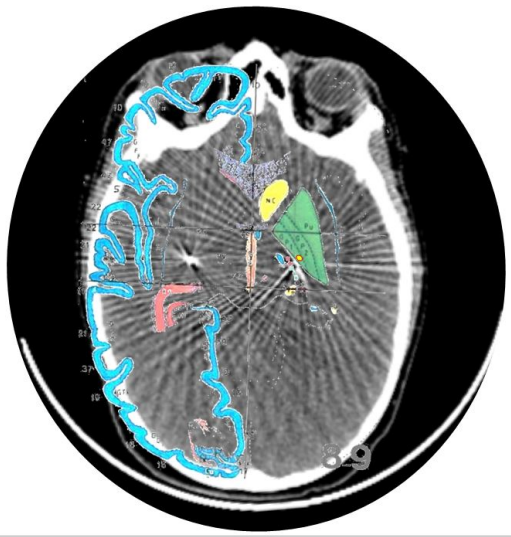


Stereotactic plan for GPi DBS with atlas overlay

Results

We have documented sustained but fluctuating improvement in motor function and spasticity. Changes in electrode settings were managed by our movement disorder neurologist. We have seen no complications. Parent and physician observations have been sources of information in addition to the BFMDRS.

Figure 2



Axial CT post-DBS with atlas overlay

Table 1

Patient	Pre-op Movement Score	Current Movement Score	Length of Follow-up	Percent Difference
#1	89.5	58.5	1 year	34%
#2	46	30	6 months	34%
#3	97	95	1 month	2%

BFMDRS movement scores, max 120

Table 2

Patient	Pre-op Disability Score	Current Disability Score	Length of Follow-up	Percent Difference
#1	30	27	1 year	10%
#2	18	13	6 months	28%
#3	30	30	1 month	0%

BFMDRS disability scores, max 30

Table 3

Patient #, age in years, sex	Pre-operative Neurological Status	Post-operative Outcomes (1 year, 6 months, 1 month)
#1 10,F	• Paraplegic, has never been able to walk • Has never been able to talk more than a few monosyllables • Constant spasms, which become so bad at times that she needs a fan to cool down (nurse has fan on-hand at school, parents at home) • Restraints on arms • Tracheostomy and GT tube for all feeding	• Able to form most words • Able to operate on a stander bicycle • Able to open her hands and make some purposeful movements • Dressing, grooming and feeding are significantly easier • Can roll over in bed – never was able to • Episodic spasms, fan not currently needed for cooling • No arm restraints • Eating some food – gaining weight
#2 10,F	• Wheelchair-bound with aid because she lost all purposeful function in her right arm • Posture erect in wheelchair but unable to stand	• Able to navigate wheelchair • Able to walk with walker
#3 9,M	• Never able to sit down unsupported • Never able to stand • Unable to form words • Tendency to keep tongue protruded • Almost constant thrust of tongue and teeth grinding • Initially able to swallow but overtime he has lost this ability and he had to receive a PEG tube	• Decreased grinding of his teeth • Improved sleep • Of note, these are early results without reprogramming

Patient outcomes as noted by parents and clinician

Conclusions

We believe DBS-GPi represents a useful treatment modality for carefully selected cases of severe intractable, progressive, spastic dystonia with neurological regression. Because of weighted number metrics, the BFMDRS may not fully or adequately characterize the full clinical benefit noted by patient, parents, and teachers.

References

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2. Burke RE, Fahn S, Marsden CD, Bressman SB, Moskowitz C, Friedman J. Validity and reliability of a rating scale for the primary torsion dystonias. Neurology. 1985;35:73-77.