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BILATERAL GPI DBS FOR OFF DYSTONIAS AND ON DYSKINESIAS IN JUVENILE PD



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Abstract

Data supporting preference of DBS target choice in juvenile PD are lacking. In particular, correlation between disabling symptoms and choice of target (between STN and GPi) represents an underinvestigated area.

Our patient was originally diagnosed with DRD at 10 years of age. He had dramatic response to levodopa, which helped him transition from being wheelchair-bound to near normal functioning. The diagnosis was subsequently revised to juvenile PD as he developed progressively worsening signs of parkinsonism, requiring increasing quantities of levodopa. The patient is from the Middle East and no genetic etiology for his juvenile PD was identified.

When he was first seen in our Center at 25 years of age, he was greatly disabled from grade 4 ON dyskinesias and severe OFF dystonias. Presurgically, the patient had virtually no ON time without dyskinesias, despite taking levodopa no less frequently than hourly during waking hours, up to 3.5 grams a day.

Bilateral GPi DBS electrodes were implanted, with moderate microlesioning benefit. There was no levodopa dose reduction. Multiple programming sessions allowed stable and sustained 70-90% reduction of OFF dystonias and complete resolution of dyskinesias. The patient now spends most of the day ON without dyskinesias.

Many experts consider GPi a better suited target when the most disabling symptoms include dyskinesias and dystonias. While prospective studies are needed to address site selection as a function of most disabling symptoms, our case supports the notion that GPi stimulation can work exceptionally well in reducing ON dyskinesias and OFF dystonias in juvenile PD.

Background

Deep brain stimulation has been an established, standard-of-care treatment option for Parkinson's disease¹. Target selection practices have not been fully backed by data collected in prospective controlled studies. As the clinical and etiological heterogeneity of PD becomes more obvious and intensely researched, practically no data are available to back the practice to guide DBS target selection as a function of different PD subtypes.

VIDEO

Case Report

- 27 years old man (as of 12/2009)
- First symptoms include foot dystonia around age 10
- Diagnosis of DRD a couple of years later (already wheelchair-bound at the time)
- Dramatic initial levodopa response
- Gradual escalation of levodopa dose over the years with acceptable response and good functioning
- At the time of seeing us for the first time the patient had almost no ON time w/o dyskinesias and essentially transitioned between severe levodopa-induced dyskinesias and OFF dystonias
- Had a tremendous anxiety component fearing OFF dystonias and took levodopa on a half-hour interval (up to grams a day)
- Genetic testing revealed homogenous deletions in PARK2 (exon 4)
- There also was a deletion in PINK1 of unknown significance
- Because of the severe dystonias and dyskinesias, bilateral GPi electrodes were implanted
- Dramatic response to DBS with 90% reduction of dyskinesias and 70% reduction of dystonias
- Levodopa overuse remains an issue

Conclusions

- Our patient with juvenile PD due to homogenous PARKIN mutation and prominent levodopa response (and apomorphine response) did very well with bilateral GPi stimulation
- Prospective studies correlating the choice of STN versus GPi targets in various forms of PD, including PARK2 mutations, as well as a function of leading/most disabling symptomatology are needed.

References

1. Kluger BM, Klepitskaya O, Okun MS. Surgical treatment of movement disorders. *Neurol Clin* 2009;27(3):633-677,