Globus Pallidus Deep Brain Stimulation in Dystonia

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BACKGROUND: Since pharmacological and surgical treatment is often ineffective in generalized dystonia, globus pallidus deep brain stimulation (GPI-DBS) has become a useful alternative. METHODS: All patients selected for GPI-DBS were prospectively rated pre- and post-DBS with the Unified Dystonia Rating Scale (UDRS). In addition, “blinded” videotape assessments using the Abnormal Involuntary Movement Scale (AIMS) were performed. RESULTS: Eleven patients, 7 male, with mean age at onset of dystonia 18.4 years and mean duration of symptoms 7.5 years, had adequate follow-up of up to one year. Compared with pre-DBS scores there were significant improvements in mean total UDRS score: 15.3±11.64% (p=0.002) and in the following subscores: neck 18.2±22.6% (p=0.03), trunk 19.9±16.9% (p=0.003), arm 17.9±12.4% (p=0.04), leg 19.9±16.9% (p=0.003). Six subjects underwent staged implantation. Compared with pre-DBS scores there were significant improvements in mean total UDRS score: 15.31±11.64% (p=0.002) and in the following subscores: neck 18.2±22.6% (p=0.03), trunk 19.9±16.9% (p=0.004), arm 17.9±12.4% (p=0.04), and leg 19.9±16.9% (p=0.003) (Figure 1). In addition, there was a 15.8% (p=0.003) improvement in AIMS motor score on follow-up. The mean improvement in AIMS motor score measured by rater 1 was 15.69% (p=0.003) and by rater 2 15.72% (p=0.003). Inter-rater reliability was assessed using weighted kappa scores. The Mann-Whitney U test was used to compare the pre-DBS and post-DBS scores. A p<0.05 was considered statistically significant. RESULTS: Eleven subjects had prospective UDRS and blinded video assessments with the Unified Dystonia Rating Scale (UDRS) and the Abnormal Involuntary Movement Scale (AIMS). The mean improvement for those patients was 15.7±12.3% (p=0.003) on the total UDRS score. The mean improvement in AIMS motor score measured by rater 1 was 15.69% (p=0.003) and by rater 2 15.72% (p=0.003). Inter-rater reliability was assessed using weighted kappa scores. The Mann-Whitney U test was used to compare the pre-DBS and post-DBS scores. A p<0.05 was considered statistically significant.

METHODS

All subjects with severe generalized or hemidystonia who were refractory to oral medication and botulinum toxin injections and were considered for GPI-DBS at Baylor College of Medicine underwent prospective pre-DBS and post-DBS assessments with the Unified Dystonia Rating Scale (UDRS). Functional disability was determined for speaking, writing, drinking, and walking using a 5 point scale (0=none, 1=minimal interference with task, 2=mild interference with task, 3=moderate interference with task, 4=marked interference with task, 5=unable to perform task). When available, videotape assessments using the Abnormal Involuntary Movement Scale (AIMS) were performed by 2 blinded raters. The two-tailed Wilcoxon Signed Ranks Test was used to compare the pre-DBS and post-DBS scores. All subjects with severe generalized or hemidystonia who were refractory to oral medication and botulinum toxin injections and were considered for GPI-DBS at Baylor College of Medicine underwent prospective pre-DBS and post-DBS assessments with the Unified Dystonia Rating Scale (UDRS). Functional disability was determined for speaking, writing, drinking, and walking using a 5 point scale (0=none, 1=minimal interference with task, 2=mild interference with task, 3=moderate interference with task, 4=marked interference with task, 5=unable to perform task). When available, videotape assessments using the Abnormal Involuntary Movement Scale (AIMS) were performed by 2 blinded raters. The two-tailed Wilcoxon Signed Ranks Test was used to compare the pre-DBS and post-DBS scores. All subjects with severe generalized or hemidystonia who were refractory to oral medication and botulinum toxin injections and were considered for GPI-DBS at Baylor College of Medicine underwent prospective pre-DBS and post-DBS assessments with the Unified Dystonia Rating Scale (UDRS). Functional disability was determined for speaking, writing, drinking, and walking using a 5 point scale (0=none, 1=minimal interference with task, 2=mild interference with task, 3=moderate interference with task, 4=marked interference with task, 5=unable to perform task). When available, videotape assessments using the Abnormal Involuntary Movement Scale (AIMS) were performed by 2 blinded raters. The two-tailed Wilcoxon Signed Ranks Test was used to compare the pre-DBS and post-DBS scores.

RESULTS

Eleven subjects (7 male) with mean age at onset of dystonia 18.4 years and mean duration of symptoms 7.5 years, had adequate follow-up of up to one year. Compared with pre-DBS scores there were significant improvements in mean total UDRS score: 15.3±11.64% (p=0.002) and in the following subscores: neck 18.2±22.6% (p=0.03), trunk 19.9±16.9% (p=0.004), arm 17.9±12.4% (p=0.04), and leg 19.9±16.9% (p=0.003) (Figure 1). In addition, there was a 15.8% (p=0.003) improvement in AIMS motor score on follow-up. The mean improvement in AIMS motor score measured by rater 1 was 15.69% (p=0.003) and by rater 2 15.72% (p=0.003). Inter-rater reliability was assessed using weighted kappa scores. The Mann-Whitney U test was used to compare the pre-DBS and post-DBS scores. A p<0.05 was considered statistically significant.

DISCUSSION

In our population of patients with heterogeneous etiology of dystonia, GPI-DBS was found to be a safe and effective treatment, consistent with previous reports.1,2,4 The variable response of cranial dystonia in our patients could be related to genetic heterogeneity, globus pallidus somatotopic organization, or electrode positioning.2,4 One patient had a serious adverse event consisting of skin erosion requiring surgical debridement. This low complication rate is consistent with the experience in other centers.2,4,15 The lack of improvement in functional disability in our patient was possibly because of the short follow-up. A delay in clinical response after GPI-DBS has been observed by others.20

The lack of statistical significance with the blinded video evaluation was likely due to the relatively low number of patients. Another possibility is that the AIMS is not sensitive enough to detect post-DBS changes. GPI-DBS is a safe and effective treatment in medically resistant patients with dystonia. Long-term follow up is needed to determine whether the benefits of the procedure are sustained.

REFERENCES