Interventional MR Guided DBS in Pediatric Dystonia: Technical and Clinical Outcomes

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Introduction:

The placement of deep brain stimulator (DBS) electrodes in adults is traditionally performed using stereotactic methods. Stereotactic methods alone, however, lack the requisite accuracy and therefore physiological confirmation of lead location via micro-electrode recordings is commonly performed. These physiologically based measures require the patient to be awake, cooperative and off medications that could mask their clinical condition. These requirements largely preclude pediatric populations due to the challenge of confirming electrode locations intra-operatively. We have developed an intra-operative MR methodology that permits highly accurate delivery of DBS electrodes based entirely on anatomical characterization [1]. The approach does not require patients to be awake, cooperative, or off their medications. The method has been widely used in Parkinsonian patients, where clinical outcomes have been similar to patients that received their electrodes with stereotactic methods and physiological confirmation [2]. Shorter procedure durations and fewer brain penetrations have also been demonstrated with MR guidance. In this study we apply the approach to children with dystonia, a rare syndrome of sustained muscle contractions that produces writhing movements and abnormal postures. We report on technical accuracy and assess clinical benefit based on changes in a standardized disease rating scale.

Methods:

Six pediatric patients with primary dystonia were prospectively enrolled. Patients were candidates for surgery if they had marked disability and medical therapy was ineffective. Five patients had a mutation to the DYT1 gene, and mean age at surgery was 11.0 ± 2.8 years. All patients were anesthetized throughout surgery, which was performed entirely within the magnet bore, and no physiological testing was performed. Patients received bilateral DBS electrodes in the globus pallidus interna (GPi, n=5) or subthalamic nucleus (STN, n=1). Inversion recovery turbo spin echo (GPi) or T_2-weighted turbo spin echo (GPi and STN) imaging was used to identify anatomical targets (**Figure 1**). The electrodes were implanted in a 1.5T interventional MR suite (Philips Acheiva, Cleveland, OH) using a novel skull mounted trajectory guide in conjunction with dedicated software (Clearpoint, MRI interventions, Irvine, CA) [1]. The trajectory guide contains a linear fluid filled alignment indicator with an open central lumen through which devices can be inserted. Imaging methods are used to manipulate the trajectory guide until it is precisely aimed at the intended target. The device is then inserted and the target definition scan repeated to assess targeting accuracy.

Accuracy was assessed by comparing the intended versus achieved electrode position in the axial plane used for anatomic targeting. Since the DBS electrode is flexible, a rigid ceramic mandrel within a peel-away sheath was initially inserted. If acceptable positioning was achieved the mandrel is swapped for the DBS electrode and the peel-away sheath removed. Clinical outcomes were measured with the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS), which was performed at baseline, 6 months and 12 months post-operatively. The difference between the BFMDRS scores at baseline compared to 12 months (on stimulation) was assessed using the Wilcoxon matched-pairs sign rank test. Further measures, included procedure durations, quality of life, adverse events, and stimulation settings, were monitored.

Results:

All 12 electrodes were successfully implanted and only a single brain penetration was required for each electrode. The mean ($\pm$ SD) difference between the intended target location and the actual lead location, in the axial plane passing through the intended target, was 0.6 ± 0.5 mm. The mean surgical time, measured from skin incision to skin closure was 190 ± 26 minutes. Mean percent improvement in the BFMDRS movement score was $86.1\% \pm 12.5\%$ (n=6, p=0.028) at 6 months, and $87.6\% \pm 19.2\%$ (p=0.028) at 12 months. For comparison, prior studies that assessed bilateral DBS in dystonia reported mean BFMDRS improvements that ranged from 62-85%. The mean stimulation settings at 12 months were 3.0V, 83µs, 135Hz (GPi DBS), and 2.1V, 60µs, 145Hz (STN DBS). There were no serious adverse events such as hemorrhage, prolonged hospitalization or additional surgery.

Conclusions:

MR guided DBS electrode implantation is extremely accurate and provides real-time anatomical confirmation of DBS placement. The procedure does not require an awake, cooperative patient and requires relatively short operative durations. Clinical outcomes for pediatric dystonia are comparable with the best reported results using traditional frame-based stereotaxy.

Figure 1: The selection of the target within the GPi is demonstrated. Inversion-recovery turbo spin echo imaging is used to provide contrast in deep brain structures. The insert demonstrates how the target is selected: a point 1/3 of the way along, and 3-4 mm in from the pallidocapsular border. Actual electrode positions (red arrows) are demonstrated on post-operative imaging.

References

[1] Larson PS, Starr PA, Bates G, Tansey L, Richardson RM, Martin AJ. An optimized system for interventional magnetic resonance imaging-guided stereotactic surgery: preliminary evaluation of targeting accuracy. Neurosurgery. Mar 2012;70(1 Suppl Operative):95-103. **[2]** Starr PA, Martin AJ, Ostrem JL, Talke P, Levesque N, Larson PS. Subthalamic nucleus deep brain stimulator placement using high-field interventional magnetic resonance imaging and a skull-mounted aiming device: technique and application ac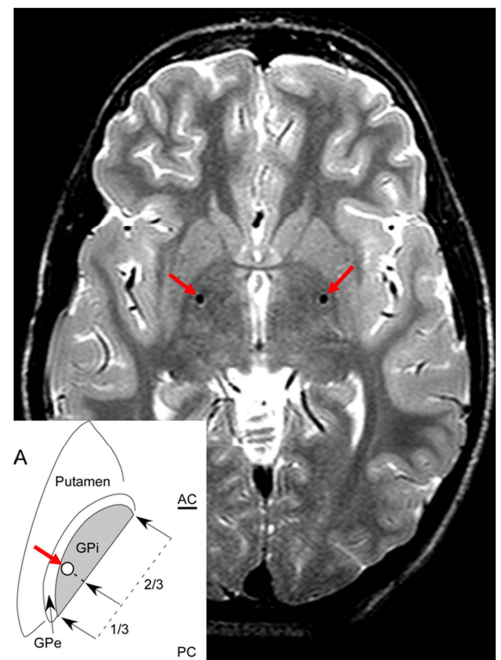curacy. J Neurosurg. Mar 2010;112(3):479-490.