

Deep Brain Stimulation for Status Dystonicus: Multitargeting Method

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Introduction

Generalized dystonia may progress to a very severe persistence devastating dystonia that can be life threatening. This form of dystonia can induce rhabdomyolysis and respiratory dysfunction. Here, we report on two pediatric patients with generalized status dystonicus.

Methods

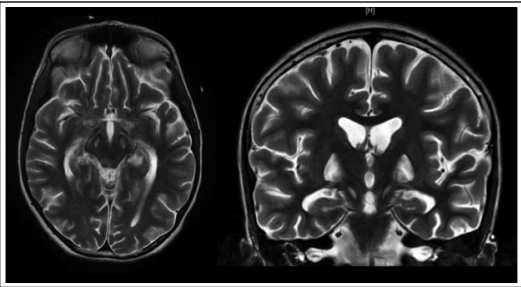
Two pediatric patients with status dystonicus (dystonic storm) who underwent urgent deep brain stimulation were analyzed. Dystonia severity (motor) and disability scales were applied using BFMDRS. Demographic data, type of dystonia, associated clinical features, Magnetic Resonance imaging (MRI) features, electrophysiology data, and stereotactic targets were analyzed.



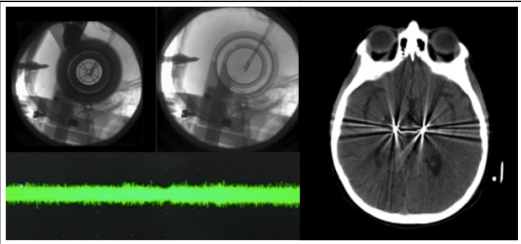
Patient 1: Severe dystonia and contractures that was not reduced by general anesthesia

Results

The first patient was a 10 year-old boy who developed transient febrile illness followed by neurological deterioration. Subsequently; he developed severe generalized dystonia and spasm, inability to verbalize. His BEMDRS score was 116 and creatinine kinase was around 2000. MRI brain depicts complete neuronal loss of globus pallidus internus (GPI) and substantia nigra (SNr). He underwent deep brain stimulation targeting bilateral thalamus (VOA) and zona incerta (ZI). Neurophysiology recording from GPI area revealed silent neuronal activities. He had a 70% improvement in his symptoms. After 3 months of DBS, he is able to walk, eat, and he regained speech function. The second patient is a 9 year-old boy who developed progressive generalized dystonia over 11 months till he developed a fixed twisted body position that lead to severe weight loss due to poor feeding. BFMDRS score was 108. MRI brain did not reveal any structural abnormalities. He underwent bilateral GPI DBS after which he improved over 4 months (88%). He was able to attend public school 5 months after DBS.



MRI brain depicting neuronal loss at globus pallidus area and SNr, STN



Microelectrode recording from GPI:
Silent neuronal activities
Four DBS electrodes implantaiton



Patient 1: DBS clinical benefit at 10 weeks and 4 months intervals



Patient 2: Preoperative clinical status and post DBS clinical benefit after 4 months

Conclusions

The pathophysiology of status dystonicus remains poorly understood. Urgent deep brain stimulation can be a life saving procedure that restore functional state. Multitargeting method may increase the chance of benefit from DBS.

Learning Objectives

Multitargeting method in cases with neuronal loss of certain known DBS targets can be an alternative method for treating dystonia.

Disclosure

The author has nothing to disclose

Consent for publication was obtained from patients' guardians