**INTRODUCTION**

A dystonic storm, or status dystonicus, is defined as a progressively increasing frequent and severe episode of generalized dystonia. It is a life-threatening condition that may arise in the course of primary and secondary dystonic syndromes (7). Uncontrolled, this disorder can lead to serious complications such as, respiratory and renal failures. The majority of research demonstrates that DBS implantation shows favorable results in drug-resistant cases. However, only a few articles reported DBS use in young children, with the youngest reported age being 8 years old (2,8). In performing DBS implantation in youngsters, there are possible limitations such as the potential growth of the brain, head and trunk, the small amount subcutaneous fat and the loose covering skin. Therefore, DBS is FDA approved for only patients of 7 years old and more.

In this report, the authors are presenting a 5-year-old boy with dystonic storm treated with DBS, making him the youngest patient treated with DBS to be reported. DBS implantation was considered as a last line life-saving treatment in this patient.

**CASE REPORT**

A 5-year-old boy with medical history of epilepsy and mental retardation who presented with dystonic storm is discussed in this case report. The patient was under treatment with Valproic acid 300mg/day with effective control of his epilepsy. Neurologic examination revealed generalized persistent dystonia with hyperactive deep tendon reflexes. Due to tense masseter muscles and nasopharyngeal dystonia oral intake was absent and therefore, nasogastric (NG) tube placement failed, and intravenous (IV) fluids and total parenteral...
nutrition (TPN) was started. Ophthalmologic examination was normal, with no iris and pupil pathologies. Brain computed tomography (CT) scan and magnetic resonance imaging (MRI) showed bilateral calcification located in the pallidum (Figure 1A-B). Complete blood count (CBC) revealed low hemoglobin levels (9.7gm/dL) suggesting the necessity of iron replacement. Blood and urine biochemistries revealed no pathological findings with the normal levels of ammoniac, amino acids, ceruloplasmin, copper and hormones. Serologic tests and TORCH screening showed negative results. Lumbar puncture was performed for cerebrospinal fluid (CSF) analysis. Opening pressure, direct microscopic analysis, biochemistry, culture and serologic tests of the CSF were all normal. Electroencephalography (EEG) revealed central continuous sharp waves activity. Neuropsychological tests could not be administered to the patient due to his dystonicus state. However, before the presentation of the dystonic storm the patient was administered Denver II Developmental Test at the outpatient clinic where he was monitored. He showed performances of 3 months for personal-social interaction, 4 months for fine motor skills and 2 years for language. Fahn-Marsden Score was 106.

Treatment with Valproic acid 40 mg/kg/day was continued. Trials of oral Akineton 0.4 mg/kg/day, bolus IV Akeniton 10mg/day, IV midazolam infusion 40mg/kg/day, L-Dopa 8.3mg/kg/day, Carbamazepine 40mg/kg/day, oral Baclofen 0.6 mg/kg/day and intrathecal (IT) baclofen infusion test with 50mg/day were carried out for the treatment of dystonia symptoms; however, all were found ineffective. Repeated injections of botulinum toxin-A into the left extremities, cervical and masster muscles were performed; but they also were ineffective in reduction of the dystonic symptoms. The only effective intervention in controlling the dystonia was sedating the patient with prolonged continuous Midazolam infusion, which was considered a temporarily management and not appropriate as long-term treatment. For this reason, surgical treatment with DBS was decided to be utilized. The surgical procedure was performed under general anesthesia using the Leksell (Elekta, Stockholm, Sweeden) stereotactic frame with a ring system. The surgical target was GPI and its position was localized anatomically by direct visualizing on T2-weighted and IR MRI. FrameLink 2.0 software (Medtronic, Inc., Minneapolis, MN) was used for applying CT-MR fusion technique and reformatting the MR image to the AC-PC plane to yield the calculated coordinates for the GPI, with the aid of Schaltenbrand and Wahren stereotactic atlas. Quadripolar electrodes (Model 3389, Medtronic, Minneapolis, MN, USA) were implanted bilaterally in GPI in a single session. The electrodes were directed ventromedially making their trajectory passing lateral and ending ventral to the calcifications. Electrode targeting was confirmed using microelectrode recordings (MER) demonstrating bursting neuronal patterns that are archetypal in dystonic patients. Keeping in mind the young age of the patient and the potential growth of the head, the distal ends of the electrodes were implanted 1cm inferior to the targeted GPI, with the trajectory of the electrode poles including the GPI. After the localization of GPI and the recording of the bursting patterns, the electrode was advanced more ventrally. Bursting pattern was followed during the advancement of the electrode until the appearance of a pattern of low noise and decreased background activity, which indicated the inferior end of the GPI. The electrode was the advanced more inferior to GPI with the final position of contact of 0 was inferior the GPI and the electrode tip ventroanterior to the calcifications. A dual-channel pulse generator (Kineta Model 7428, Medtronic, Inc.) that is subclavicularly implanted to the right side of the patient’s chest during the same operation was connected to the leads. No complication occurred during or after the surgery. Postoperative cranial CT scan was obtained in order to confirm the correct positions of the electrodes (Figure 2). The activation of the electrodes took place 3 days after the surgery. The stimulation parameters were 1.5V, 130 Hz, 90µs, with monopolar stimulation on contact 1. Four weeks after the initiation of stimulation dystonia showed improvement.
A dystonic storm, or status dystonicus, is a life-threatening condition which requires aggressive control; otherwise it may result in bulbar and ventilatory dysfunction resulting with the risk of aspiration pneumonia and respiratory failure, severe pain, and metabolic derangements such as acute renal failure and rhabdomyolysis (4). In our case, the dystonic storm was secondary to the pathological calcifications. As these calcifications are located in the pallidum, the dysfunction will result in the reduction of the activity along the putamen-GPi connection and increase of the inhibition of the STN and GPi by GPe efferents (9). In spite of this possible mechanism, the certain pathophysiology of the disease is unknown. Cases of drug-resistant dystonic storm are reported to be well responding to surgical treatment, such as thalamotomy (7), bilateral pallidotomy (6), bilateral GPi DBS (11) and pallidotomy with GPi DBS (4). Radiofrequency ablation of the globus pallidus internus, or pallidotomy, result in removal of the inhibitory output from pallidum to the cortex (1). DBS of GPi has also been safely used for symptoms of dystonia in many different conditions (4,14).

Previous reports show that surgical intervention has good results in cases of refractory dystonic storm. Our results in this specific case were also consistent with the previous literature. In addition, the patient in our case was a 5 year old, and to our best knowledge this is youngest patient to be treated with DBS mentioned in the literature. Other articles also reported treatment with DBS for young children, however, the age of the youngest patients was 8 years old (2,8). The brain grows in size and volume considerably during the early years of life (3). Extensive brain development is believed to occur in the first 10 years, with increased rates between 3 to 6 years (12). Also, the functional development of the brain occurs early in life and continues to mature and develop into the teen years and perhaps beyond. Also, a child is physically smaller and will undergo several periods of physical growth (5,13). For this reason, one must keep in mind if the device will provide the same physiologic fit, when implanted in a child as compared to an adult. If it does not, alternate techniques of implantation must be considered (10). In addition, the growth and development of adolescents, particularly in girls where procedures involving the chest may impact the development of breast buds. Also, less subcutaneous fat in a child may result in increased device erosion. Another important issue is the possibility that the child will outgrow the device. Thus, plans for revisions or device design changes must be considered (10). For the above reasons, FDA had approved the use of DBS for children of 7 years old and over (10).

In the present case, the patient presented with dystonic storm which was resistant to all modalities of medical treatment. As mentioned before, untreated cases carry high risk of mortality and morbidity, due to bulbar and ventilatory dysfunction with the risk of aspiration pneumonia and respiratory failure, severe pain, and metabolic derangements, such as acute renal failure and rhabdomyolysis (4). For this reason, using DBS for this patient was our last chance as a life-saving procedure. The young age of the patient and the potential growth of the head, the distal ends of the electrodes we implanted 1cm inferior to the targeted GPi, with the trajectory of the electrode poles including the GPi was taken into consideration. There were no side effects related to the stimulation. The patient was relieved from the dystonic storm state and the dystonia showed improvement with 81% reduction of the total Fahn-Marsden score after 15 months of surgery, which was even better than the score of the patient before the dystonic storm.

**DISCUSSION**

A dystonic storm, or status dystonicus, is a life-threatening condition which requires aggressive control; otherwise it may result in bulbar and ventilatory dysfunction resulting with the risk of aspiration pneumonia and respiratory failure, severe pain, and metabolic derangements such as acute renal failure and rhabdomyolysis (4). In our case, the dystonic storm was secondary to the pathological calcifications. As these calcifications are located in the pallidum, the dysfunction will result in the reduction of the activity along the putamen-GPi connection and increase of the inhibition of the STN and GPi by GPe efferents (9). In spite of this possible mechanism, the certain pathophysiology of the disease is unknown. Cases of drug-resistant dystonic storm are reported to be well responding to surgical treatment, such as thalamotomy (7), bilateral pallidotomy (6), bilateral GPi DBS (11) and pallidotomy with GPi DBS (4). Radiofrequency ablation of the globus pallidus internus, or pallidotomy, result in removal of the inhibitory output from pallidum to the cortex (1). DBS of GPi has also been safely used for symptoms of dystonia in many different conditions (4,14).

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**REFERENCES**

Aydin S. et al: Pallidal Deep Brain Stimulation in a 5-Year-Old Child with Dystonic Storm


