

Deep Brain Stimulation Found to be Effective in Children with Treatment-Resistant Generalized Primary Dystonia

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Dystonia is a very complex, highly variable neurological movement disorder characterized by involuntary muscle contractions. As many as 250,000 people in the United States have dystonia, making it the third most common movement disorder behind essential tremor and Parkinson's disease. The prevalence of generalized primary dystonia is estimated at 34 in 100,000 people worldwide, but cited statistics vary. This figure is most likely an underestimate, since many cases go undiagnosed for years.

Dystonia results from abnormal functioning of the basal ganglia, a deep part of the brain which helps control coordination of movement. [Patients](#) with generalized primary dystonia suffer from repetitive twisting movements or abnormal postures due to involuntary muscle contractions. This can be a severely disabling disease which starts around age 10, with initial misdiagnosis fairly common. Further, it can be a socially debilitating disease, especially for adolescents, since peer acceptance plays an important role, particularly during this period of development.

Researchers at Ludwig-Maximilians University in Munich investigated the safety and efficacy of [deep brain stimulation](#) (DBS) in children with severe cases of treatment-resistant primary dystonia. The results of this study, Globus Pallidus Internus DBS in Pediatric Patients with Generalized Primary Dystonia: Long-Term Efficacy and Safety, will be presented by Jan H. Mehrkens, MD, 5:00 to 5:07 pm, Tuesday, May 5,

2009, during the 77th Annual Meeting of the American Association of Neurological Surgeons in San Diego. Co-authors are Kai Boetzel, MD, Ingo Borggäfe, MD, and Florian Heinen, MD.

“Unfortunately, conservative medical treatment options are extremely limited and have proven ineffective for patients with severe cases,” stated Dr. Mehrkens. In light of this dilemma, the quest for effective therapeutic approaches has been ongoing for many years. With the growing application of chronic high-frequency DBS in the treatment of movement disorders including dystonia, this neurosurgical procedure has become safer and control of side effects has widely improved.

Over the last decade, the globus pallidus internus (GPi) of the basal ganglia of the brain, has been identified as the most promising target for treating dystonia. However, very little is still known about long-term efficacy and side effects of utilizing this treatment at early disease onset in children suffering from primary generalized dystonia. “Since 2000, we have treated 48 patients at our institution utilizing this approach, with long-term data available on a smaller group of pediatric patients,” said Dr. Mehrkens.

In this study, immediate postoperative results and long-term follow up data were analyzed in a group of five patients, age 16 or younger, suffering from primary generalized dystonia. All patients had cases of dystonia that were resistant to conservative approaches, and had undergone early GPi-DBS treatment, with a focus on long-term efficacy and safety. Electrode-implantation was performed under general anaesthesia with MRI-guided stereotaxy followed by the implantation of the internal pulse generator.

- Correct electrode position was verified by MRI and clinical outcome was assessed using a special scoring system (Burke-Fahn-Marsden (BFM) motor and disability scores).

- Mean age at surgery: 13, age range: 8-16 years.
- Two female, 3 male.
- Mean follow-up: 58 months, follow-up range: 36-85 months.

The entire patient group experienced significant improvement in symptoms as early as the first week, which further improved in the months to follow:

- The mean improvement in the BFM movement score was 67.4 percent (range 47.0-87.5 percent), 75.4 percent (range 61.5-91.7 percent) and 83.5 percent (range 72.0-93.3 percent) at 3 months, 12 months and long-term follow-up, respectively.
- Problems with the implanted devices occurred in two patients (electrode dislocation and breakage of extension cable in one, imminent perforation of extension cable in another). This was resolved through an immediate operative revision.
- There was very mild dysarthric speech disturbance in two patients, with no other therapy-related morbidity observed.

“Looking at the results of this study, we conclude that GPi-DBS offers both a very effective and safe therapy in children suffering from primary dystonia. Stable relief of symptoms can be achieved on a long-term basis. The children in this study were able to return to school and interact with their peers with very few restrictions. It seems crucial to operate in the early course of the disease to prevent irreversible impairment of motor function,” remarked Dr. Mehrkens.

“As physicians, the most rewarding aspect for us is that the beneficial results of DBS provide these young patients with a significantly

improved quality of life, enabling renewed and positive social integration,” concluded Dr. Mehrkens.

Source: American Association of Neurological Surgeons

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