DBS Effective in Early-Onset Form of Dystonia

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CHICAGO — The efficacy of deep brain stimulation can be maintained for up to 10 years in DYT1 dystonia, according to data from a study in 26 consecutive patients.

This prospective study is the first to report on more than 3 years of follow-up of deep brain stimulation (DBS) in DYT1 dystonia—a form of primary dystonia that typically presents in early childhood and is caused by a mutation in the DYT1 gene.

Significant decreases in Burke-Fahn-Marsden dystonia rating scale motor and disability scores were observed 1 year after DBS surgery. No significant difference was found when the 1-year scores were compared with the scores at 3, 5, and 6 years for the whole population, Dr. Laura Cif and associates reported in a poster at the 12th International Congress of Parkinson's Disease and Movement Disorders.

Efficacy of DBS therapy was maintained in the two patients who were followed as long as 10 years.

Long-term disease progression is even more important in DYTI dystonia than in other disorders treated with DBS, such as Parkinson's disease, because patients are much younger at the time of surgery and therefore require significantly longer-term follow-up, according to Dr. Cif of the University of Montpellier (France). The age at onset ranged from 6 to 20 years in the study. Moreover, DYT1 dystonia can develop into a life-threatening condition.

Eighteen patients were implanted with a single pair of electrodes in the internal globus pallidus (GPi) and eight patients had a second pair of GPi electrodes implanted because of incomplete initial response or subsequent worsening.

In spite of there being no significant difference 1 year after surgery, a significant difference was observed at 5 years between patients with a single lead vs. those with double leads in motor (8.95 vs. 31.5) and disability (3.61 vs. 7.85) scores. After implantation of additional pairs of leads, only four of the eight patients showed subsequent improvement.

During the follow-up, no patient died, including several patients with status dystonicus, who usually are not expected to survive for more than a few weeks, Dr. Cif said in an interview. ■

IVIG May Slow Progression of Alzheimer's

CHICAGO — Continuous intravenous infusions of immunoglobulin for 9 months stabilized cognition and function for Alzheimer's patients enrolled in a small placebo-controlled trial.

The 18-month-long phase II study included 24 patients with mild to moderate Alzheimer's. For the first 6 months, they were randomized either to placebo or to one of four intravenous immunoglobulin (IVIG) doses.

For the remaining 12 months, all the participants were switched to IVIG, but raters were still blinded to the dosing, said Dr. Norman Relkin, who presented the study's 9-month results at the International Conference on Alzheimer's Disease.

Patients who had received IVIG continuously for 9 months showed significantly better scores on measures of cognition and activities of daily living than did those taking placebo.

When the IVIG arms were analyzed by dose, 0.4 g/kg of body weight every 2 weeks provided the best results in global functioning, cognition, and activities of daily living, Dr. Relkin said at the meeting, which was sponsored by the Alzheimer's Association.

Planning for a larger, phase III trial is underway; it is to be conducted at 35 academic centers in the United States.

Dr. Relkin of Cornell University, New York, coauthored another IVIG study published earlier this year that found that improvements in Alzheimer's patients treated with IVIG lasted only as long as the treatment continued.

The trial of continuous IVIG treatment was cosponsored by the National Institutes of Health and Baxter International Inc.

Dr. Relkin said he received grants and research support from Baxter.

—Michele G. Sullivan