

Globus pallidus internus-deep brain stimulation in Huntington's disease: a case report

Alessandro Bertini^{1,2}, M. Vizziello^{1,2}, F. Pirone^{1,2}, E. Zacarias^{1,2}, G.D. Oggioni^{1,2}, C. Gambini^{1,2}, T. Bocci^{1,2}, R. Ferrucci^{1,2}, F. Motta³, G. Iess³, M. Egidi³, A. Priori^{1,2}

¹Clinical Neurology Unit I, Department of Health Sciences, San Paolo University Hospital, Aldo Ravelli Research Center, University of Milan, Milan, Italy

²ASST Santi Paolo e Carlo Hospital, Milan, Italy

³Unit of Neurosurgery, Azienda Socio Sanitaria Territoriale Santi Carlo e Paolo, San Carlo Hospital, Milan, Italy

Introduction. Deep brain stimulation (DBS) of the internal globus pallidus (GPi) has been proposed for the treatment of drug-resistant chorea in selected patients affected by Huntington's disease (HD), but long-term efficacy and optimal stimulation protocols are not yet well established.

Case presentation. We report the case of a 61 y.o. man with 20-year history of genetically confirmed HD. He presented severe and disabling axial and limbs chorea associated to mild bradykinesia. Pharmacological treatments for chorea (tetrabenazine, haloperidol, valproate, atypical antipsychotic) were ineffective or not tolerated. Pre-surgery neuropsychological assessment found out a quite well-preserved cognitive status except for moderate executive, memory and language deficits along with mild anxiety and depression. He underwent to bilateral GPi-DBS in November 2021, no post-operative motor or cognitive side effects occurred. With 130 Hz stimulation hyperkinetic movements slowly decreased, but parkinsonism, freezing of gait and unbalance, correlating with stimulation intensity and increasing frequencies, appeared. However, lower frequency and low voltage stimulation (75Hz, 1.5V) improved chorea by 42% compared to pre-surgery (from 12 to 7 according to Unified Huntington's Disease Rating Scale-chorea subscore) without affecting gait and global movement quality significantly. Therapeutic effect on chorea was objectively assessed by multichannel electromyography at various stimulation voltages. Videolaringscopy showed an improvement of swallowing in the On-DBS state, with better bolus propulsion compared to Off-DBS. Neuropsychological assessment, repeated at 1&3 months, was not affected by surgery and DBS status, but anxiety and depression improved, allowing antidepressant reduction. Subjective motor improvement is maintained 6 months after surgery.

Discussion. GPi-DBS surgery was well tolerated by our patient, with improvement on motor function, swallowing and mood. The main stimulation challenge was finding a balance between therapeutic effect on chorea and parkinsonism, as an unavoidable side effect. This case supports the possible benefit from DBS in HD patient with predominant motor phenotype.