

Deep brain stimulation in Huntington's disease

Roberta Bonomo^{1,2}, A.E. Elia¹, G. Bonomo³, L.M. Romito¹, C. Mariotti⁴, G. Devigili¹, R. Cilia¹, R. Gioi^{5,6}, R. Eleopra¹

¹Fondazione IRCCS Istituto Neurologico Carlo Besta, Department of Clinical Neurosciences, Parkinson and Movement Disorders Unit, Milan, Italy

²Experimental Neurology Unit, School of Medicine and Surgery, University of Milano-Bicocca, Monza, Italy

³Neurosurgery Department, Fondazione IRCCS Istituto Neurologico Carlo Besta, Milan, Italy

⁴Unit of Medical Genetics, Fondazione IRCCS Istituto Neurologico Carlo Besta, Milan, Italy

⁵Neuroimmunology and Neuromuscular Diseases Unit, Fondazione IRCCS Istituto Neurologico Carlo Besta, Milan, Italy

⁶Department of Oncology and Onco-hematology, Postgraduate School of Clinical Pharmacology and Toxicology, University of Milan, Milan, Italy

Introduction: Huntington's disease (HD) is an inherited neurodegenerative disorder clinically characterized by involuntary movements, cognitive decline, and behavioral changes. The complex constellation of clinical symptoms still makes the therapeutic management challenging. In the new era of functional neurosurgery, deep brain stimulation (DBS) may represent a promising therapeutic approach in selected HD patients.

Objective: We evaluated the effects of DBS on patients affected by HD, providing a critical outlook on the achieved results and the possible developments.

Methods: Articles describing the effect of DBS in patients affected by HD were selected from Medline and PubMed by the association of text words with MeSH terms as follows: "Deep brain stimulation", "DBS", and "HD", "Huntington's disease", "Huntington". Details on repeat expansion, age at operation, target of operation, duration of follow-up, stimulation parameters, adverse events, and outcome measures were collected.

Results: Twenty eligible studies, assessing 42 patients with HD, were identified. The effect of GPi-DBS on Unified Huntington's Disease Rating Scale (UHDRS) total score revealed in 10 studies an improvement of total score from 5.4% to 34.5%, and in 4 studies an increase of motor score from 3.8% to 97.8%. Bilateral GPi-DBS was reported to be effective in reduction of Chorea subscore in all studies, with a mean percentage reduction of 21.4% to 73.6%.

Conclusions: HD patients with predominant choreic symptoms may be the best candidates for surgery, but the role of other clinical features and of disease progression should be elucidated. For this reason, there is a need for more reliable criteria that may guide the selection of HD patients suitable for DBS. Accordingly, further studies including functional outcomes as primary endpoint are needed.