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TÍTOL

Huntington's Disease: from therapeutics to Cognition

PONENT

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DIA

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ORGANITZA

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PER A MÉS
INFORMACIÓ

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Resum:

Huntington's disease from therapeutics to cognition

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Huntington's disease is a rare neurodegenerative genetic disease whose natural evolution, at a stage where the patients are still available for treatment, is poorly understood. Patients are difficult to follow and to evaluate because of the entanglement of motor, psychiatric and cognitive disorders. The disease leads towards dementia and death in approximately 20 years.

The progressive neuronal degeneration in HD primitively localized in the striatum made it possible to develop a "substitute" cell therapy with the aim of rebuilding the brain circuitry by the functional and anatomical replacement of neuronal loss and a "neuroprotective" therapy, intended to maintain the viability of the endogenous neurons still present. This research addresses the problem of the functional restoration and its relation with brain reconstruction in patients. We undergone the first pilot study that showed, in 3 out of 5 HD patient that brain transplant could improve the motor abilities of the patients from 4-5 years and more than 6 years for their cognitive functions. We are now coordinating the MIG-HD trial (2001-2010) for a validation of transplant at large scale outside from pilot centers. Because of the transient effect of the transplant procedure, we develop trials on neuroprotection in order to stop the evolution of the disease. Thus, we ran a phase I pilot study using CNTF delivered by encapsulated cells of baby hamster kidney in the brain (coll. P. Aebischer). This shows the feasibility and the safety of the procedure but calls for new methods of CNTF delivery. Nevertheless, because only transplants allows the recovery of lost functions in symptomatic patients via neuroreconstruction, the future probably lies on a synergy between neuroprotection and neuroreconstruction in order to restore but also maintain recovery.

In addition to the need for a treatment for this devastating disease, the primary degeneration of striatum at the early stage of HD makes it a very useful model of striatal lesion, which can be used as such for research purposes. The role of the striatum is known in articulation, memory, attention and executive functions - all of them constituting the abilities required for successful performance on complex tasks in daily life. However, despite the obvious language difficulties and the social disturbance in HD patients, its role in language and social cognition, domains in which the animal models are not suitable, remains to be elucidated. This is why to progress in this knowledge we have so far followed a double approach: basic research on the development of structured models of these functions and explorations aimed at identifying the components to integrate in the models, starting from the disorders observed in patients. We associated a methodology of classical neuropsychology based on the observation of the decline of performance in single cases or in groups of brain-damaged patients (lesional neuropsychology) and experimental psychology through the study of the performance of healthy subjects, patients with HD and with focal lesions. We show that HD patients suffer from language disorders at early stages and that the striatum should be included in the models of language processing. Finally, we will combine our expertise in transplant and in cognition, to determine which components of language and social cognition are managed by the striatum and to which extent we are able to rebuild them, using human foetal cells.