Switching the disease off: Effects of spatial and temporal inactivation of mutant huntingtin in Huntington disease

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Name of Fellow
Institution
Funder

European Commission FP7-Seventh Framework Programme

Contact information of fellow
Country

EC

Title of project/programme

Switching the disease off: Effects of spatial and temporal inactivation of mutant huntingtin in Huntington disease

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€ 999,006

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The project/programme is most relevant to:

Huntington's disease

Keywords

Huntington Disease | striatum | hypothalamus | BACHD rat | Cre-recombinase

Research Abstract
Huntington Disease (HD) is an autosomal dominantly inherited neurodegenerative disorder,

which is caused by an expanded CAG repeat in the huntingtin gene. Mutant Huntingtin (mhtt) is expressed ubiquitously in the brain but the most affected region is the striatum. Marked atrophy also occurs in other brain regions such as the cortex and the hypothalamus. The latter has been linked to disturbances in sleep pattern, energy metabolism and emotion, all of them non-motor symptoms of HD that occur early in disease. In the project SWITCH-HD we will, for the first time, analyse, which mhtt containing brain area is causing which symptom in HD. We will investigate the role of mhtt in the striatum and in the hypothalamus by down-regulating mhtt in these brain regions at different disease stages using lentiviral delivery of Cre recombinase into a unique transgenic rat model of Huntington disease (BACHD rats). The effect of switching off mhtt in these brain regions on behaviour, metabolism, neuropathology, neuroimaging and potential biomarkers will be investigated. Translation of the results of this study to humans will allow to develop more specified treatments against HD. SWITCH-HD brings together the expertise of the SME JSW Life Sciences (JSW, Austria) and the University of Tübingen (EKUT, Germany). JSW will contribute and transfer its expertise regarding stereotactic applications, gene delivery by lentiviral vectors and behavioural studies. EKUT will contribute and transfer their expertise in regard to generation and comprehensive characterisation of transgenic rat models of HD. Co-operation and transfer of knowledge is dedicated to gaining new insights relevant to pathogenesis and treatment of HD as well as the establishment of new tools. Transfer of knowledge will be implemented through secondments, recruitments and meetings. There will be a structured exchange of researchers from both partners at project milestones facilitating the training of newly recruited team members

Types:
Fellowships

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