Decoding Presymptomatic white matter changes in Huntington Disease (WIN-HD)

https://neurodegenerationresearch.eu/survey/decoding-presymptomatic-white-matter-changes-in-huntington-disease-win-hd-3/

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Multiple

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Country

France|UK

Title of project or programme

Decoding Presymptomatic white matter changes in Huntington Disease (WIN-HD)

Source of funding information

CoEN

Total sum awarded (Euro)

611,601

Start date of award

01/06/2015

Total duration of award in years

2

The project/programme is most relevant to:

Huntington's disease

Keywords
Research Abstract

Huntington disease (HD) is an inherited neurodegenerative disease caused by a single mutation in the huntingtin gene. Onset occurs in adults, with slow progression and a combination of motor signs and cognitive changes, including dementia, as a result of underlying neurodegeneration of brain tissue. The mutation is an expansion of a CAG sequence within the gene and the length of the expansion has a strong effect on disease onset and course.

Early postmortem and imaging studies in HD have focused on grey matter changes in the subcortical brain structures such as the caudate and putamen. However, there is increasing recognition that white matter changes also play a role in the disease from the very early stages, many years before any clinical signs are evident. The exact nature of these white matter changes are not fully understood. Most imaging techniques used to date do not have the power to determine the specific mechanisms underlying white matter degeneration. For example, loss of white matter integrity may be due to degeneration of the axonal section of the neurone, or a reduction in the amount of myelin surrounding it. Myelin is produced by oligodendrocytes and it is thought that dysfunction within these cells may play an important role in the underlying neuropathology in HD.

We aim to improve our understanding of the role of oligodendrocytes in the white matter degeneration which typifies HD by:
a) examining white matter changes in individuals who carry the HD mutation but are not yet affected by the symptoms using advanced neuroimaging techniques
b) using mouse models of HD to examine disease-related changes in living animals (using imaging) and in postmortem brain tissue
c) examining postmortem human brain tissue to understand the nature of white matter changes in terms of characteristics such as axonal loss, demyelination and numbers of oligodendrocytes
d) determining whether other genes which cause dysfunction in oligodendrocytes have an impact on the disease course in HD mouse models

Lay Summary
Further information available at:

Types:
Investments > €500k

Member States:
France, United Kingdom

Diseases:
Huntington's disease

Years:
2016

Database Categories:
N/A

Database Tags:
N/A