An observational neuroimaging and neurophysiological study investigating the visual pathway in Huntington*s disease.

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Primary objectives:* To improve the understanding of alterations in the visual pathway in patients with HD in different disease stages.* To investigate possible changes in brain function and structure in rest and in response to visual stimulation in...

Ethical review	Approved WMO
Status	Recruiting
Health condition type	Neurological disorders congenital
Study type	Observational non invasive

Summary

ID

NL-OMON45518

Source
ToetsingOnline

Brief title
Visual pathway in HD.

Condition

- Neurological disorders congenital
- Movement disorders (incl parkinsonism)

Synonym
hereditary neurodegenerative movement disorder, Huntington's disease

Research involving
Human

Sponsors and support

Primary sponsor: Leids Universitair Medisch Centrum
Source(s) of monetary or material Support: andere projecten uit 3e geldstroom en nog aan te vragen fondsen (seed fund EHDN) en donaties

Intervention

Keyword: Huntington's disease, MRI, Visual cortex, Visual-Evoked Potentials

Outcome measures

Primary outcome

* Group differences and correlations between pre- and manifest HD and healthy controls on the functional and structural MRI outcome parameters.

* Correlation between scores on neuropsychological assessments and MRI outcome parameters in all groups.

* Differences in VEP outcome measures (amplitudes and latencies) between pre-, manifest HD and healthy controls.

* Correlation between VEP outcome measures and outcome parameters of different imaging modalities (e.g. fMRI, DTI and structural MRI).

Secondary outcome

Not applicable

Study description

Background summary

Huntington*s disease (HD) is a rare autosomal dominant inherited progressive neurodegenerative disorder. Besides the well-known triad of clinical signs such as motor disturbances, cognitive dysfunction and psychiatric symptoms, are impairments of the visual system also frequently described in HD patients. These impairments include visuomotor and visuospatial dysfunctions, impaired visual attention, and deficits in naming visual objects.

Neuropathological studies demonstrated evidence of specific neuronal loss in the visual system, which is consistent with structural neuroimaging studies that show extensive thinning and atrophy of the visual cortex in different disease stages. Also, neurophysiological investigation of the visual pathway using visual-evoked potentials (VEP) showed reduced amplitudes and normal latencies in manifest HD compared to controls suggesting involvement of the visual cortex when the disease progresses.
Still, the visual pathway in the human brain has not yet been investigated systematically in patients with HD. This explorative observational study will therefore combine different neuroimaging modalities with neurophysiological visual evoked potentials to improve our understanding about the pathophysiological degenerative processes in HD.In addition, we want to investigate if there is a correlation with clinical cognitive assessments involving a visual component.

Study objective

Primary objectives:
* To improve the understanding of alterations in the visual pathway in patients with HD in different disease stages.
* To investigate possible changes in brain function and structure in rest and in response to visual stimulation in patients with HD and healthy controls.
* To explore if visual cognitive impairments are related to changes in brain structure and brain function in premanifest and manifest HD.

Secondary objectives:
* To examine occipital cortical function in premanifest and manifest HD using visual-evoked potentials.
* To evaluate if there is a correlation between outcome measures of neurophysiological and neuroimaging techniques.

Study design

This is a prospective, cross-sectional, observational explorative study in Huntington's disease and healthy individuals that are not at risk for HD.

Study burden and risks

Participants will undergo different clinical and neuropsychological assessments during a single visits of approximately 3 hours. These assessments will examine the general physical and cognitive functioning.
Furthermore, each participant will undergo a MRI-scan of the brain and a neurophysiological test (VEP). Both assessments are non-invasive and are also commonly used in the daily clinical practice for general clinical care.
Therefore, there is no immediate risk associated with participation in this study. Participants receive no immediate benefit from participation in this

study. The only potential benefit is a better understanding of the pathophysiology of HD. The information obtained in this study may potentially benefit the development of new treatments and help to plan future studies.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adults (18-64 years)
Elderly (65 years and older)

Inclusion criteria

* Between 18 and 65 years of age at time of visit
* Ability to undergo MRI scanning
* Written informed consent must be obtained from the participant;If the participant is a manifest HD gene carrier:
* Gene positive tested with CAG repeat length of * 36 in the HTT gene
* UHDRS-TMS > 5;If the participant is a premanifest HD gene carrier:
* Gene positive tested with CAG repeat length of * 36 in the HTT gene

* UHDRS-TMS * 5;If the participant is a control subject:
* Gene negative tested for HD or without family history of HD
* No other known cognitive, neurological or psychiatric disorders

Exclusion criteria

* Impaired primary visual ability (<0.5) or ophthalmic disorders
(including amblyopia or color vision deficiency)
* Additional major co-morbidities not related to HD
(e.g. cardiovascular diseases, hypertension, diabetes mellitus, and/or other neurological disorders)
* History of severe head injury
* Contra-indication for MRI (metallic implants/devices, claustrophobia)
* Participation in intervention trials
* Pregnancy
* Inability to understand the information about the protocol

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)

Primary purpose: Basic science

Recruitment

NL

Recruitment status:	Recruiting
Start date (anticipated):	16-01-2017
Enrollment:	60
Type:	Actual

Ethics review

Approved WMO

Date:	15-12-2016
Application type:	First submission
Review commission:	METC Leiden-Den Haag-Delft (Leiden) metc-ldd@lumc.nl

Approved WMO Date:	12-04-2017
Application type:	Amendment
Review commission:	METC Leiden-Den Haag-Delft (Leiden) metc-ldd@lumc.nl

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL58661.058.16