A European Study to identify the needs of individuals and families affected by Juvenile Huntington's Disease

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identification of the needs of families with juvenile Huntington's disease

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

Summary

ID

NL-OMON30947

Source
ToetsingOnline

Brief title
Juvenile Huntington's Disease

Condition

- Neurological disorders congenital
- Movement disorders (incl parkinsonism)

Synonym
Huntington's disease in childhood

Research involving
Human

Sponsors and support

Primary sponsor: European Huntington's Disease Network
Source(s) of monetary or material Support: European Huntington's Disease Network

Intervention

Keyword: care, Huntington's disease, Juvenile, psychology

Outcome measures

Primary outcome

none

Secondary outcome

none

Study description

Background summary

Huntington*s Disease (HD) is an inherited neurodegenerative condition, for which there is currently no cure and management is by way of symptomatic treatment and support only. Individuals with HD develop a number of symptoms, including a movement disorder, cognitive dysfunction and affective disturbance. HD is typically a late-onset disorder, with many individuals developing the condition between the ages of 30-50 years (Kremer, 2002), although the age at onset varies greatly and it can affect children and teenagers. Juvenile-onset HD (JHD) has been defined arbitrarily as having an onset occurring under 20 years (Bruyn, 1968). The youngest symptomatic child reported in the literature had an age of onset of 2 years (Nance et al., 1999), while some individuals with JHD may now be over 20 years old although having developed symptoms while they were still in the teenage years. HD is a life-limiting condition, usually lasting 15-20 years from onset (Kremer, 2002).

Huntington*s Disease affects between 7 and 10 in every 100,000 (Harper, 2002). The proportion of individuals with JHD in previous surveys has varied between 1-10% of those affected by HD (Hayden, 1981) and it is therefore a rare condition. In young people with JHD the clinical presentation varies from that seen in adults, and patients with JHD are more likely to present with a failure at school or behavioural disturbance. Clumsiness and unsteadiness in gait as well as rigidity are often the early predominant motor abnormalities seen in children with JHD, with the choreic movements (involuntary, dance-like movements) often seen in adult-onset HD being less common (Nance, 1997). They are also more likely to develop epilepsy (Brackenridge, 1980). In about 75% of cases of JHD the affected parent was the father, although males and females are affected equally by JHD (Telenius et al., 1993), and this means that in many

cases the sole carer is the mother.

As a condition JHD is generally less well-recognised than HD, and as such even less is known about it and the impact it has on the individual and their family. There have been very few studies that have focused on JHD and, until recently, there have none that have considered psychosocial issues arising from the condition. An awareness of the psychosocial impact is particularly important where there is currently little possibility of a cure. The Huntington*s Disease Association in the UK (HDA) recently carried out research looking at the impact of JHD on the family. Semi-structured interviews were carried out with twelve parent/guardian carers of young people with JHD and a number of issues were identified, meaning that more appropriate services could be developed for this group (Smith et al., 2006; Brewer et al., in press). In particular, families highlighted how their experience and their needs were different from that of those affected by adult-onset HD in a number of ways (Brewer et al., under submission). This suggests that studies which focus on the experiences of this group may be particularly relevant. However, given the small number of participants included in the research in the UK, questions still remain as to whether the experiences of these families can be generalised to other countries and other cultural groups.

A European Huntington*s Disease Network (EHDN) has recently been established (www.euro-hd.net). This network has been developed to establish links between clinicians/researchers with an interest in HD and families affected by the condition across Europe, in an attempt to provide the infrastructure for HD research across Europe. As part of this network a number of working groups have been formed, including one focused on JHD. EHDN and the working group provide the opportunity to extend the UK project further within Europe. This research will give us further information about the psychosocial impact of this rare condition. In particular, it will enable the experiences of families from a variety of countries across Europe and from different cultural backgrounds to be elicited. In addition, the experiences and views of a larger number of families will be collected, which will enable more generalised statements about the issues affecting this group to be made.

Study objective

identification of the needs of families with juvenile Huntington's disease

Study design

explorative interview study

Study burden and risks

none

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

families with a patient with diagnosed juvenile Hungton's disease (onset under 18 years)

Exclusion criteria

absence of informed consent

Study design

Design

Study type: Observational non invasive

Masking:	Open (masking not used)
Control:	Uncontrolled
Primary purpose:	Other

Recruitment

NL

Recruitment status:	Pending
Start date (anticipated):	01-03-2007
Enrollment:	2
Type:	Anticipated

Ethics review

Approved WMO

Application type:	First submission
Review commission:	METC Leids Universitair Medisch Centrum (Leiden)

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	NL16501.058.07