

FEAR OF CHOKING AND FEAR OF FALLING IN HUNTINGTON'S DISEASE: STUDY PROTOCOL FOR A MULTI-CENTER OBSERVATIONAL CROSS- SECTIONAL STUDY

K. KALKERS^{1,2}, J.M.G.A SCHOLS^{3,4}, J.C.L. NEYENS², R.A.C. ROOS¹

1. Department of Neurology, LUMC, Leiden, The Netherlands; 2. De Riethorst Stromenland, Raamsdonksveer, The Netherlands; 3. Department of Health Services Research, Caphri – Care and Public Health Research Institute, Maastricht University, Maastricht, The Netherlands; 4. Department of Family Medicine, Caphri – Care and Public Health Research Institute, Maastricht University, Maastricht, The Netherlands. Corresponding author: K. Kalkers, Leiden University Medical Center, Department of Neurology, Albinusdreef 2 (J3-R-141), 2333 ZA Leiden, The Netherlands. Tel- 0031653198462 E-mail-K.Kalkers@lumc.nl

Abstract: *Background:* Dysphagia and accidental falls are common in patients with Huntington's disease; they can have serious consequences and may, therefore lead to fear of choking and fear of falling. *Objectives:* In this article, a protocol is described to explore the prevalence of fear of choking and fear of falling in patients with Huntington's disease, to identify the relationship between fear of choking and fear of falling and, respectively, anxiety, awareness and cognitive functioning and to define the care demands with regard to fear of choking and fear of falling. In addition, the related problems encountered by their (in)formal caregivers are investigated. *Design:* This study protocol describes a multi-center observational cross-sectional study. *Participants:* Our aim is to include 150 patients with Huntington's Disease living in or attending day care in Dutch nursing homes specialized in Huntington's Disease and their (in)formal caregivers. *Setting:* Dutch nursing homes, specialized in long-term care for patients with Huntington's Disease. *Measurements:* The patients will be assessed by means of questionnaires enquiring about mobility, swallowing and their feelings and experiences related to these topics, a mobility task, a cognitive screening and a neurological examination. Other patient information will be derived from regular patient files, registered by formal caregivers. Data about fear of choking and fear of falling from the formal and informal caregiver's perspective will be obtained by means of self-administered questionnaires. *Conclusions:* This study may contribute to improving insight into the cognitive, emotional and behavioral functioning of patients with Huntington's disease. The findings may lead to relevant interventions for patients or support advice for caregivers, with regard to adequate coping strategies for risk-taking behavior.

Key words: Protocol, Huntington's disease, accidental falls, choking, fear.

Introduction

Huntington's disease (HD) is a progressive neurodegenerative autosomal dominant disease, caused by a mutant protein, huntingtin, and resulting from an expanded CAG repeat on the short arm of chromosome 4. The longer the CAG repeat, the earlier the mean age at onset of disease. The prevalence is estimated at 5-10/100.000 in the Caucasian population. Mean age at onset is 30-50 years with a wide range (2-85) and the mean duration of the disease is 17-20 years (1). At the time of institutionalization most HD patients are middle aged (2).

Characteristics of the disease are motor symptoms, cognitive dysfunction and psychiatric and behavioral symptoms (3). Disease progression leads to increased dependency in daily life and progressive care needs (1). Falls and swallowing disorders are both common symptoms in Huntington's disease and can have major consequences for patients.

Dysphagia is common in patients with HD, but little is known about its frequency or its development during the disease. Problems with swallowing include lack of coordination, rapid lingual chorea, repetitive swallows, frequent eructations, coughing when eating, and choking on liquids (4). Dysphagia can have serious consequences such as pneumonia, acute respiratory distress and subsequent death (5).

Pneumonia is the most prominent primary cause of death in HD and the cause is most likely dysphagia (6). Dysphagia can, therefore, potentially lead to fear of choking (FoC) (5). To our knowledge, no studies have been conducted to assess this type of fear in HD.

An accidental fall is a common problem faced by HD patients in everyday life. Rates of fall incidents are reported to vary between 20.8% and 75.6% (7-11). A fall can result in hospitalization, early admission to a nursing home or subsequently death (12). In addition to physical injuries, a fall can lead to fear of falling (FoF) and hence avoiding activities (13, 14). Individuals who fall often develop a protective fear for falling in the future. Paradoxically, FoF and consequently avoiding activities are predictive of future falls in both fallers and non-fallers (13, 15). There is some evidence that patients with HD, suffering from falls, experience less FoF compared to other patient groups (8, 11).

In general, anxiety can be a useful emotion. It arises when experiencing or thinking about danger and may lead to preventive actions (16). This ability to act safely may be influenced by experiencing less fear. Reduced fear responses have been reported in HD (17, 18). A possible cause of experiencing less fear might be unawareness. The terms insight, self-awareness, unawareness, anosognosia, and denial are often used interchangeably in literature for HD (19). According to

McCusker and Loy (2014), the following definition can be used: '1) the manifestations of disease and the consequences are obvious to and can be documented by an observer, but 2) the individual underestimates or does not report experiencing symptoms or notice these changes or the impact'. In the literature, reduced self-awareness is described as a possible cause of falling in the elderly. As a result of this reduced self-awareness, the person will take more risks (20). Unawareness can appear at all stages of the disease and is differentiated across different domains, e.g. physical and mental changes (21). Presumably the loss of awareness of deficits in HD is caused by a combination of cognitive, physiological and psychological factors (21). Also, cognitive factors, such as the ability to evaluate possible consequences of behavior (8), the ability to comprehend information and the awareness of task demands, (22) can affect the extent to which fear is experienced.

In addition to the fear experienced by HD patients, their caregivers' fear of the patient falling or choking is also a factor. This is true for both formal and family or other informal caregivers. In dementia (23, 24), the fearful attitudes of caregivers can result in restriction of the care recipient's activities and even the use of restraints by the caregiver to prevent falls. The caregiver's fear may, in fact, increase the patient's anxiety.

There is little information in the literature about falls, choking, FoC or FoF in HD patients, or about the related problems in formal and informal caregivers. We, therefore, aim to explore the prevalence of FoC and FoF in HD patients and the related problems in their caregivers and to identify the relationship between FoC and FoF and anxiety, unawareness and cognitive functioning. Finally, we will define the care demands with regard to FoC and FoF. The objectives of the study are, therefore:

In patients with HD and their formal caregivers:

- To assess the prevalence of dysphagia symptoms and accidental falls in HD patients
- To assess the prevalence of FoC and FoF in HD patients
- To identify the relationship between FoC and FoF and emotional and cognitive functioning in HD patients
- To define the care needs of patients with HD with regard to FoC and FoF

In formal and informal caregivers of patients with HD:

- To assess the prevalence of FoC and FoF from the formal and informal caregiver's perspective
- To define the care needs with regard to FoC and FoF from formal and informal caregiver's perspective

The assessments may contribute to a better insight into the cognitive, emotional and behavioral functioning of the patient during the disease. The findings may lead to interventions for patients or guidelines for caregivers with regard to coping strategies for risk taking behavior.

Methods

Study design

This is designed as a multi-center observational cross-sectional study.

Participants and recruitment

Our aim is to include at least 150 patients; because the total number of patients with HD living in or attending day care in Dutch nursing homes specialized in HD is approximately 300, we consider this to be the maximum achievable number in the Netherlands. The inclusion criteria for the patients are: 18 years of age and older, diagnosis of HD confirmed clinically and/or genetically, with an CAG expansion of ≥ 36 , resident in or attending day care in a nursing home which specializes exclusively in long-term care of patients with HD. The exclusion criteria for the patients are: concurrent neurological or serious psychiatric disorders which might interfere with the assessment, other causes of balance disturbance, terminal or serious illness, participation in intervention trials. By using these inclusion criteria, not all HD patients will be able to answer questions themselves. In that case information will be provided by proxy observations, meaning their nurses, speech language therapists and informal caregivers, as described in the next paragraphs. This also concerns information regarding fear of falling and fear of choking.

One formal and one informal caregiver, related to each participating patient, will also be asked to take part in the study if the patient approves. Consequently, we intend to include 150 formal and 150 informal caregivers. The formal caregiver is the nurse with primary responsibility for the patient, or, if unavailable, a nurse on the patient's ward who works at least 24 hours a week. The informal caregiver is the formal representative or legal guardian, or, when unavailable, a closely involved informal caregiver of the patient.

Dutch nursing homes, specialized in long-term care for patients with HD and associated with the Huntington Network Netherlands (HNN) which endorses the importance of scientific research, will be asked to participate in the study. Nursing homes offer HD patients multidisciplinary treatment, guidance and support and nursing care (25). Besides nursing staff, these nursing homes employ medical, paramedical and psychosocial staff (26); participating in the study will place demands on the employees' time and requires commitment. All patients and their (in)formal caregivers in participating nursing homes will be approached to take part.

Written informed consent will be obtained from all participants. If a patient is incompetent, the legal guardian will be able to sign the informed consent form. Inclusion is planned for 2018-2019. The results are expected in 2020.

Ethical approval

The study protocol has been reviewed by the Medical Ethics Review Committee of VU University Medical Center

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(2017.445). They declared that the Medical Research Involving Human Subjects Act (WMO) does not apply and hence official approval by the committee is not required (27).

Procedures

Patients

The patients will be examined in the nursing homes. This involves administration of a questionnaire by standardized (semi-) structured interview and a cognitive screening by the researcher or an assistant, both psychologists. To standardize the interviews, all question and answer options have been processed in a flowchart. Furthermore, patients will undergo a brief neurological examination by a physician who, prior to the examination, will attend an online training session and a mobility task by a physiotherapist. Additional patient information will be derived from regular patient files and observations and registered by a nurse and speech-language therapist.

The expectation is that it will take approximately one hour and fifteen minutes to complete the assessments. The examination can be spread over multiple moments of contact during a four-week period. The data collection starts with the registration of patient information by the nurses, followed by the patient interview by the researcher. Then the physician, speech-language therapist and physiotherapist will collect and record their data in random order. For the nurse and speech-language therapist, both paper and web-based forms are available. The researcher, physician and physiotherapist will use paper forms

(In)formal caregivers

Data on FoC and FoF and related care needs from the formal and informal caregivers' perspective, will be obtained by means of self-administered questionnaires; It is expected that the caregiver assessments will take approximately twenty minutes to complete. They have the option of using paper or web-based forms, which they will receive by (e-)mail.

Study measurements

The quantitative questionnaires and measurements regarding HD patients and (in)formal caregivers are respectively presented in tables 1 and 2. If present, questionnaires have been selected that have been validated for patients with Huntington's disease (28-33). In other cases, questionnaires were chosen that have been validated for institutionalized patients (34-36), patients with other neurological disorders (37, 38) and patients with dysphagia (39).

Patients

Patient Demographics and clinical characteristics

Demographic and clinical data, such as date of birth, gender, current medication and comorbidity, are registered

for each patient. Functional capacity is examined with the Total Functional Capacity Scale (TFC), which is a subscale of the Unified Huntington Disease Rating Scale (UHDRS) (32), a 5-item clinician rating scale for function in HD. It is used to make a global assessment of capacity to work, handle finances, perform domestic chores and self-care tasks. Care dependency is examined using the Care-Dependency Scale (CDS) consisting of 16 items based on fifteen basic needs and one subjective judgment of the extent to which the patient is care-dependent (40). The Total Motor Score (TMS) (32), also part of the UHDRS, is administered to the patient by a physician and is used for the motor type classification (41). Based on these scores, the patients are classified as hypo-, hyperkinetic- or mixed motor subtype.

Patient characteristics regarding falls and mobility

Information regarding use of spectacles and use of hearing aids is registered for each patient. Fall incidence is obtained retrospectively for a period of 30 days on the basis of the daily reports, and additionally for day care patients by interviewing the patient and informal caregiver. The incidence represents the number of residents who have fallen at least once during the 30 days prior to the measurement day and the number of falls by each patient. A fall is defined as an event when the resident accidentally ends up on a lower level or on the ground (42). One item of the Braden scale for predicting pressure sore risk (43) is used to specify the degree of physical activity. One item of the motor section of the Unified Huntington Disease Rating Scale For Advanced Patients (UHDRS-FAP) (29) is used to specify the need for assistance with walking or use of walking aids. The Performance Oriented Mobility Assessment (POMA) (44) is used to assess balance and gait status of the patients. FoF is conceptualized as low-perceived self-efficacy in the Short Falls Efficacy Scale-International (Short FES-I) (45). This scale is used to assess concerns about falling during seven physical and social activities. Additionally, a single question about FoF, with a yes-no answer ('Are you afraid of falling?'), is asked (46). This section ends with questions about care needs with regard to falling, providing qualitative data. The first question is: what measures have been taken to prevent falling? The answer may be selected from a variety of possible measures, but there is also space for alternatives. Then the patient is asked whether the measures taken have had an effect, with five possible answers ('Yes, I fall less often', 'yes, walking is improved', 'yes, I am less afraid of falling', 'yes, I feel more comfortable while walking' 'no, there are no changes in this area'). This part is concluded with the question: 'What measures do you miss?' The answer may be selected from a variety of possible measures, but there is also space for alternative ideas.

Patient characteristics regarding dysphagia

Information on height, weight and presence of dentures is registered for each patient. One item of the motor section of the

Table 1
Quantitative measurements and questionnaires concerning HD patients

| | Variable | Questionnaire | Number of items | Number of response categories (scoring) | Total score range (interpretation) | Source | Administered by |
|----------------------------------|-------------------------|-------------------------------------|--|--|--|-----------------------------------|-------------------------------------|
| Clinical characteristics | Functional Capacity | TFC (part of UHDRS) | 5 | 4 (0-3): item 1-2, 4 3 (0-2): item 3, 5 | 0-13 (higher=better) | Nurse | Self-completion |
| | Care Dependency | CDS | 16 | 5 (1-5) | 16-80 (higher=better) | Nurse | Self-completion |
| | Motor features of HD | TMS (part of UHDRS) | 15/31* | 5 (0-4) | 0-124 (higher=worse) | Patient | Physician |
| Falls and mobility | Activity level | Braden scale | 1 | 4 (1-4) | 1-4 (higher=better) | Nurse | Self-completion |
| | Assistance with walking | UHDRS-FAP (1 Item of motor section) | 1 | 5 (0-4) | 0-4 (higher=worse) | Nurse | Self-completion |
| | Balance and gait | POMA | Balance: 9 Gait: 7/10 † | 2 (0-1) items 1,7,10-13, 16 3 (0-2) items 2-6, 8-9, 14-15 | 0-28 (higher=better) | Patient | Physiotherapist |
| Dysphagia | Concerns about falling | Short FES-I | 7 | 4 (1-4) | 7-28 (higher=worse) | Patient | Researcher |
| | Fear of Falling | Single question | 1 | 2 (yes/no) | 0-1 (higher=worse) | Patient | Researcher |
| | Assistance with eating | UHDRS-FAP (1 Item of motor section) | 1 | 7 (0-6) | 0-6 (higher=worse) | Nurse | Self-completion |
| | Oral intake | FOIS | 1 | 7 (1-7) | 1-7 (higher=better) | Speech-language therapist | Self-completion |
| | Dysphagia symptoms | HDDS | 11 | 5 (1-5) | 11-55 (higher=worse) | Patient | Researcher |
| | Concerns about choking | Subscale fear of SWAL-QoL | 4 | 5 (1-5) | 4-20 (higher=better) | Patient | Researcher |
| | Fear of Choking | Two questions | 2 | 2 (yes/no) | 0-2 (higher=worse) | Patient | Researcher |
| Awareness, Cognition and Anxiety | Awareness of fall risk | SAFR part E and A Patient version | Part E: 6 Part A: 10 | Part E: 5 (1-5) Part A: 7 (1-7) | Part E: 6-30 (higher=worse) ‡ Part A: 10-70 (higher=better) § | Patient | Physiotherapist (E), Researcher (A) |
| | Awareness of fall risk | SAFR part E and A Clinician version | Part E: 6 Part A: 10 | Part E: 5 (1-5) Part A: 7 (1-7) | Part E: 6-30 (higher=worse) ‡ Part A: 10-70 (higher=better) § | Nurse (A) and Physiotherapist (E) | Self-completion |
| | Dysphagia symptoms | HDDS Clinician version | 11 | 5 (1-5) | 11-55 (higher=worse) | Nurse | Self-completion |
| | Cognition | MOCA | | | 0-30 (higher=better) | Patient | Researcher |
| | Anxiety | HADS | 14 (7 items for both Anxiety and Depression) | 4 (0-3) | 0-21 for both Anxiety and Depression (higher=worse) | Patient | Researcher |

* Item 1-3, 6-7, 9 consist of 2 sub-questions; item 11 consists of 5 sub-questions and item 12, 7 sub-questions; † Item 11 consists of 4 sub-questions; ‡ A higher score means the performance of the patient is rated as being better than that of an average healthy person (evaluated by patient and caregiver); § A higher score means less assistance is required to complete activities (evaluated by patient and caregiver); Abbreviations: CDS Care Dependency Scale; FOIS Functional Oral Intake Scale; HADS Hospital Anxiety and Depression Scale; HD Huntington's Disease; HDDS Huntington's Disease Dysphagia Scale; MOCA Montreal Cognitive Assessment; POMA Performance Oriented Mobility Assessment; SAFR Self-awareness of fall risk Questionnaire; Short FES-I Short Falls Efficacy Scale-International; SWAL-QoL Swallowing Quality of Life questionnaire; TFC Total Functional Capacity; TMS Total Motor Score; UHDRS Unified Huntington Disease Rating Scale; UHDRS-FAP Unified Huntington Disease Rating Scale For Advanced Patients

Unified Huntington Disease Rating Scale for advanced patients (UHDRS-FAP) (29) is used to specify the need for assistance with eating. Information about consistency of the food or the use of a PEG tube is registered using the Functional Oral Intake Scale (FOIS) (37). The prevalence of dysphagia symptoms is measured with the Huntington's Disease Dysphagia Scale (HDDS) (31) which contains eleven questions on dysphagia

in relation to HD (E.g. 'Do you drool during the day?'). For FoC, the subscale fear of the Swallowing Quality of Life questionnaire (SWAL-QoL-NL) is used (39). This measures quality of life of patients with oropharyngeal dysphagia, but in the present study, only the subscale fear is used. Patients are presented with concerns that people with swallowing problems sometimes mention (E.g. 'I fear I may start choking when I

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eat food'). They indicate how often they experienced each feeling during the last month. A further two questions about FoC, with yes-no answers ('Are you afraid of swallowing the wrong way?' and 'Are you afraid of choking?') are asked. This section concludes with questions about care needs with regard to dysphagia, resulting in qualitative data. The first question is: what measures have been taken to prevent choking? The answer can be selected from a variety of possible measures, but there is also space for alternatives. This is followed by the question about whether the measures taken have had an effect, with five possible answers ('Yes, I choke less often', 'yes, swallowing has been improved', 'yes, I am less afraid of choking', 'yes, I feel more confident while eating or drinking', 'no, there are no changes in this area'). This part is concluded with: 'What measures do you miss?' The answer can be selected from a variety of possible measures, but there is also space for other ideas.

Patient characteristics regarding awareness, cognition and anxiety

To examine awareness of personal fall risk, parts E and A of the Self-awareness of fall risk Questionnaires (SAFR) (34) are used. In these subscales, the Emergent awareness (part E) and Anticipatory awareness (part A) of fall risks are examined, using a calculation of discrepancy scores between patient-rated ability and informant-rated ability on parallel questionnaires (34). Emergent awareness is defined as the ability to identify a problem as it is occurring. Items evaluate aspects such as steadiness while walking. The patient's performance will be rated 'in comparison to an average healthy person of the same age' (34). Anticipatory awareness is defined as the individual's ability to anticipate a problem occurring due to some deficit (34). Items were generated to explore the patient's ability to understand the level of assistance required to complete activities, indicating the amount of assistance needed when trying to perform a certain activity. (e.g. 'Standing up from a chair') (34). In order to obtain a Dutch translation of this questionnaire, forward and back translations were used. Translation guidelines, derived from the translation manual of the Prevention of Falls Network Europe (47) were used in this process. To assess awareness of dysphagia, the same method of questionnaire-based discrepancy is used. In addition to the HADS (31) administered to a patient, a separate caregiver's version is administered to a nurse and the discrepancy calculated. Cognition is examined using the Montreal Cognitive Assessment (48), a brief cognitive screening tool. Finally, the Hospital Anxiety and Depression Scale HADS (49) is used to measure the level of anxiety a patient is experiencing.

Informal and formal caregivers

Caregiver Demographics

First of all, the relationship the informal caregiver has with the patient (e.g. partner, parent, sibling) is registered. The

formal caregiver is asked which job role they have and how many years they have provided care to HD patients in this role.

Caregiver characteristics regarding fear of falling and care needs

The prevalence of FoF from the formal and informal caregiver's perspective is assessed with a parallel, caregivers' version of the Short FES-I and a single question about FoF ('Are you afraid the patient will fall?': Yes/No). A supplementary question enquires whether the caregiver suspects that the patient is afraid of falling. 'Do you think the patient is afraid to fall?' This topic ends with questions about care needs with regard to falling, in parallel with the patients' version, providing qualitative data.

Caregiver characteristics regarding fear of choking and care needs

The prevalence of FoC from the formal and informal caregiver's perspective is assessed with a parallel, caregivers' version of the SWAL-QoL and two single questions, with Yes-No answers, about FoC ('Are you afraid the patient will swallow the wrong way?' and 'Are you afraid the patient will choke?'). A supplementary question enquires whether the caregiver suspects that the patient is afraid of swallowing the wrong way or choking. 'Do you think the patient is afraid to swallow the wrong way?' and 'Do you think the patient is afraid to choke?' Finally, questions about care needs with regard to dysphagia are asked in parallel with the patients' version, providing qualitative data.

Statistical analysis

Analyses will be performed using the Statistical Package for the Social Sciences (SPSS).

Patients

Patients' characteristics will be presented as mean \pm standard deviation (SD). If outcomes are categorical, percentages will be given. Prevalences of dysphagia symptoms, accidental falls, FoC and FoF in HD patients will be described as percentages with a 95% confidence interval. For the association between FoC, respectively FoF and unawareness, cognition and anxiety, Pearson's and Spearman's correlation coefficient will be used for the univariate analysis and multiple linear regression for the multivariate analysis. Potential confounders will be included in the regression model of the primary study parameters.

(In)Formal caregivers

Caregivers' characteristics will be presented as mean \pm standard deviation (SD); for categorical outcomes percentages will be given. Prevalences of FoC and FoF from the formal and informal caregiver's perspective will be described as percentages with a 95% confidence interval.

Table 2
Quantitative questionnaires concerning (in)formal caregivers of HD patients

| | Variable | Questionnaire | Number of items | Number of response categories (scoring) | Total score range (interpretation) | Source | Administered by |
|-----------------|---|---|-----------------|---|------------------------------------|--|-----------------|
| Fear of Falling | Concerns about falling from caregiver's perspective | Short FES-I (caregiver version) | 7 | 4 (1-4) | 7-28 (higher=worse) | Formal caregiver Informal caregiver | Self-completion |
| | Fear of Falling from caregiver's perspective | Two questions | 2 | 2 (yes/no) | 0-2 (higher=worse) | Formal caregiver Informal caregiver | Self-completion |
| Fear of Choking | Concerns about choking from caregiver's perspective | Subscale fear of SWAL-QoL (caregiver version) | 4 | 5 (1-5) | 4-20 (higher=better) | Formal caregiver Informal caregiver | Self-completion |
| | Fear of Choking from caregiver's perspective | Four questions | 4 | 2 (yes/no) | 0-4 (higher=worse) | Formal caregiver Informal caregiver | Self-completion |

Abbreviations: Short FES-I Short Falls Efficacy Scale-International; SWAL-QoL Swallowing Quality of Life questionnaire

Discussion

Dysphagia and accidental falls are common in HD. They can have serious consequences (5, 12-14), which may cause FoC and FoF. Fear of choking may cause a reduction in the intake of food and beverages. But because HD often results in weight loss, it is necessary to pay extra attention to nutritional status (50). FoF can lead to avoidance of activities, which may result in social isolation and /or an increase in falling incidents. Although FoC and FoF would seem to be relevant areas of HD research, they have so far been neglected (51). Therefore, we want to provide greater insight into these subjects by means of the study as described in this protocol. We want to assess falls and dysphagia and the prevalence of FoC and FoF in HD from both the patient and caregivers' perspective. In addition, we want to distinguish which factors contribute to causing FoC and FoF, and finally to define the patients' and caregivers' care demands regarding FoC and FoF.

The authors chose to focus on patients with HD living in nursing homes or attending day care and therefore for patients with moderate and advanced stages of HD. The main reason is that floor and ceiling effects hamper the evaluation, when using the same measurements in early, moderate and advanced stages of HD (29, 52). In addition, studies show that motor dysfunction is the main contributor to institutionalization (2, 53).

Because HD is a rare disorder (1), a challenge during this research is the inclusion of an adequate number of participants. By involving all specialized nursing homes on HD-care in The Netherlands, we aim to reach as many patients as possible and we will include the maximum achievable number. In addition, the complex multi-dimensional nature of the study is also a challenge. Due to the multicenter aspect, the management of eight different care organizations must be approached to get permission for participation. Subsequently, per organization cooperation must be sought with a large part of the multidisciplinary team: nurses, physiotherapist, speech-

language therapist and nursing home physician. Contact must also be made with informal care providers (partner and family).

Finally, data will be gathered on multiple domains, requiring everyone's contribution to be achieved at the right time and in the right order. In order to achieve this, next to sufficient time, proper planning and tight monitoring are therefore required.

Questioning the residents themselves may be difficult (54). A common administration approach is self-completion of questionnaires. However, when using self-completion, cognitive decline may be a factor influencing the validity, response rate, and completion time (55). Evidence of advantages in terms of validity, reliability and completion percentages, when using standardized interview-based questionnaires in frail, older adults with and without cognitive impairments (55), made us decide to use this method. Also, the involvement of proxies in obtaining adequate information, as we did with the parallel versions of the FoC and FoF questionnaires for the (in)formal caregivers, helps to complete data on FoC and FoF in HD (56). In order to minimize between-interviewer variability, the number of interviewers administering the questionnaires is limited to two and calibrated standards for conducting the interviews are used. Experienced professionals, familiar with HD patients, conducted the other tasks including registering patient information in order to increase the inter-rater reliability.

In this multi-center study, differences in the care and treatment offered by the care organizations can lead to differences in outcomes. The association between the prevalence of FoC, respectively, FoF in the different organizations may, therefore, also be an area of interest. With regard to the prevalence of FoC, respectively, FoF from the informal caregiver's perspective, the different relationships that may exist between the informal caregiver and the patient may affect the level of concern the caregiver experiences. Similarly, the relationship between the formal caregiver's level of education, respectively, experience may again influence the degree of concern about the patient.

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The knowledge generated by the study described in this article will be of value in developing interventions aimed at improving the quality of life of and quality of care for HD patients. Therapeutic options to reduce fear can be considered, but also interventions that increase safety or coping strategies for risk-taking behavior. The interventions can be aimed at the patient, but also at the environment. Guidelines on adequate coping strategies and improving quality of care, may result in caregiving becoming less stressful.

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