Research Report

Marriage as Protector for Nursing Home Admission in Huntington's Disease

Jessica Y. Winder[a]*, Wilco P. Achterberg[b,c] and Raymund A.C. Roos[a]
[a]*Department of Neurology, Leiden University Medical Center, Leiden, The Netherlands*
[b]*Department of Public Health and Primary Care, Leiden University Medical Center, Leiden, The Netherlands*
[c]*Huntington Center Topaz Overduin, Katwijk, The Netherlands*

Abstract.
Background: Huntington's disease is a progressive, incurable neurodegenerative disorder and it is not possible to delay onset or progression of the disease. Consequently, the disease leads to functional decline and loss of independency and finally to institutionalization.
Objective: The aim of this study is to identify factors which are associated with nursing home admission in patients with Huntington's disease.
Methods: The Unified Huntington's Disease Rating Scale (UHDRS) and the UHDRS-For Advanced Patients (UHDRS-FAP) were administered in 28 nursing home residents and 12 patients receiving day-care. Comparisons between the two groups were performed using Mann-Whitney U tests and Chi-square tests. The significantly different findings were fitted in individual univariate logistic regression models to determine which components were most predictive of institutionalization.
Results: Day-care participants were more often married than nursing home residents ($p = 0.006$) and were functionally more independent: the Functional Assessment Scale ($p = 0.022$) of the UHDRS was significantly higher. Not being married was more predictive for nursing home admission than functional capacity in the regression models.
Conclusions: Our results suggest that being married is protective for nursing home placement. Possibly, a caregiver living with a patient can assist with activities of daily living which the patient could not have done independently, resulting in being able to live at home longer. Providing support to unmarried patients, who do not have a caregiver living with them, by home care services specialized in Huntington's disease might increase the chance of the best possible care before institutionalization and postpone nursing home admission.

Keywords: Huntington's disease, advanced stage, nursing home admission, functional assessment, marriage

INTRODUCTION

Huntington's disease (HD) is a hereditary, progressive neurodegenerative disorder caused by a cytosine-adenine-guanine (CAG) trinucleotide repeat expansion in the Huntingtin gene on chromosome 4 [1]. The disease is clinically characterized by a triad of motor, cognitive, and psychiatric symptoms. These symptoms ultimately lead to functional decline and loss of independency as HD progresses into a more advanced stage. The mean age at disease onset is 30–50 years, with a mean disease duration of 17–20 years [2]. HD is presently incurable and it is not possible to delay either onset or progression of the disease. Due to the progressive nature of the disease, it eventually becomes more difficult for care to be provided at home, which leads to admission in a nursing home. However, guidelines for management and care of advanced HD patients in long-term care

*Correspondence to: Jessica Y. Winder, MD, Department of Neurology, Leiden University Medical Center, Albinusdreef 2, 2333 ZA Leiden, The Netherlands. Tel.: +31 71 526 5300; E-mail: j.y.winder@lumc.nl.

facilities are limited and only discuss different views on nursing home placement and treatment [3].

Demographically HD patients in nursing home facilities are younger (45–59 years) [4–7] than the average nursing home resident (83–85 years) [8, 9], and therefore the need for care is different. Previous studies have shown that motor dysfunction, impaired activities of daily living (ADL), and reduced scores on the Mini-Mental State Examination (MMSE) were predictors of institutionalization in HD [5, 6]. Psychiatric symptoms did not predict nursing home placement. However, these studies were conducted retrospectively and only one study [5] used the Unified Huntington's Disease Rating Scale (UHDRS), which is most commonly used for the clinical assessment of symptoms and signs in HD [10].

Identification of predictors for institutionalization may lead to interventions and treatment strategies that can postpone the need for nursing home placement. Additionally, characterization of HD specific problems in nursing home residents could optimize their care. Therefore, the aim of this present study is to examine the differences between nursing home residents and day-care participants with HD, using the UHDRS and UHDRS-For Advanced Patients (UHDRS-FAP), which is developed for patients with late stage HD [11]. We hypothesize that both nursing home residents as well as day-care participants are in an advanced stage of HD, and therefore we aim to examine which factors are associated with institutionalization.

MATERIALS AND METHODS

Setting and participants

Our study was carried out at the Huntington Center Topaz Overduin (Katwijk, the Netherlands), which is a nursing home with 70 residents, 20 day-care patients, and over 100 outpatients, specialized in the care for HD patients, both in late and early stages. Specialized medical doctors, therapists, psychologists, and nurses provide long-term care and day-care, organize activities, and offer support for patients (and their family) who live at home. The long-term care facility comprises three departments: one department is specialized in psychiatric problems, one department provides care for patients highly dependent on care, and one department provides care for patients less dependent on care. Institutionalized patients and patients receiving day-care, with a clinically and/or genetically confirmed diagnosis of HD, were

asked to participate in this study. The local medical ethics committee approved the study and written informed consent was obtained from all participants or their caregivers. Assessments performed in this study include the UHDRS, followed by the UHDRS-FAP on the same day. The scales were administered by two medical doctors experienced with HD. They interviewed and examined the patients, preferably in the presence of their caregiver.

Assessments

The UHDRS consists of four domains, which are motor function, cognitive function, behavioral abnormalities, and functional capacity [10]. The motor section comprises 31 items assessing chorea, dystonia, eye movements, bradykinesia/rigidity, and gait/balance [10]. All items are rated on a 0 to 4 point scale with 4 indicating the most severe impairment. The range of the Total Motor Score (TMS) is 0–124, with higher scores indicating more motor disturbances. The cognitive domain consists of the Verbal Fluency test [12], the Symbol Digit Modalities test [13], and the Stroop test (color naming, word reading, and interference) [14]. Higher scores indicate better cognitive performance. The behavioral component assesses the frequency and severity of 11 items, such as depression, anxiety, irritability, apathy, and other behavioral symptoms [10]. The items are rated from 0 to 4 and the total score ranges from 0 to 88, with higher scores indicating more severe psychiatric abnormalities. The functional domain is composed of three subunits [10]: the Functional Assessment Scale (FAS) including 25 yes/no questions about common daily tasks (range 0–25), the Independence Scale (IS) measuring the level of independence with one single score (range 10–100), and the Total Functional Capacity (TFC) assessing occupation, finances, domestic chores, ADL, and care level (range 0–13) [15]. Overall, higher functional scores indicate better function.

The UHDRS-FAP is divided into four sections: motor, cognitive, somatic, and behavior [11]. Motor performance is measured by 14 items, such as walking around, capacity to transfer, eat, and wash independently, dysphagia, imitation synkinesias, and other motor components (range 0–52). The cognitive score includes functional and categorical matching of the Protocole Toulouse Montreal d'Evaluation des Gnosies Visuelles (PEGV) [16], pointing, simple commands, the Stroop test, orientation, participation in activities, imitation (apraxia), and automatic series.

The somatic domain comprises 10 items assessing hyperhidrosis, hypersalivation, incontinence, digestion, hypersomnia, and pressure ulcers (range 0–28). The behavioral subscale includes 8 yes/no questions about the presence of psychiatric abnormalities (range 0–8). Higher scores on the motor, somatic, and behavioral sections indicate more impairment, while higher scores on the cognitive domain indicate better performance.

Statistical analysis

Demographic differences between nursing home residents and day-care participants with HD were analyzed using Mann-Whitney U tests or Chi-square tests. Median scores of each section of the UHDRS and UHDRS-FAP were calculated for the institutionalized patients and the patients receiving day-care. We also calculated a modified TFC without the care level-item, because this item automatically distinguishes between patients who are admitted to a long-term care facility and patients who receive day-care. Comparisons of the scores between the two groups were performed with Mann-Whitney U tests. For the scores of the UHDRS and UHDRS-FAP sections that were significantly different between nursing home residents and day-care participants, we calculated the frequencies of the items within the domain. The significantly different findings between the two groups were fitted in individual univariate logistic regression models to determine which components were most predictive of institutionalization. In order to compare the outcomes of the univariate regression models the scores were dichotomized at the median in high and low scores. A p-value of <0.05 was considered statistically significant. Data analysis was performed using IBM Statistical Package for the Social Sciences (SPSS) version 23.

RESULTS

Forty HD patients participated in our study, including 28 nursing home residents and 12 day-care participants. Of the 28 nursing home residents, 12 came from the 21-bed low-care unit (57%), 9 from the 29-bed high-care unit (31%), and 7 from the 20-bed psychiatry unit (35%). Twelve of the 20 day-care patients chose to participate (60%). Demographic data of the nursing home residents and day-care participants are shown in Table 1. Day-care participants were more often married or had a domestic partnership than nursing home residents ($p = 0.006$). Marital

status changed for three residents during admission, but was also significantly different between day-care participants and residents at the time of admission ($p = 0.038$). Other demographics, such as age and disease duration, were not significantly different between the two groups. Age and gender of study participants were similar to the age and gender of the patients who did not consent to participate in the study. Likewise, only 15 of all 70 nursing home patients were married (21.4%) compared to 14 of 20 day-care patients (70.0%) ($p < 0.001$).

The median scores of the UHDRS and UHDRS-FAP sections for the institutionalized patients and patients receiving day-care are given in Table 2. The FAS ($p = 0.022$) and TFC ($p = 0.011$) of the UHDRS functional domain were significantly lower for nursing home residents compared to day-care participants, indicating a worse function. However, the modified TFC did not show a significant difference ($p = 0.163$). The scores of the other UHDRS sections and all UHDRS-FAP sections did not show any differences between the two groups.

Figure 1 shows how many patients were able to perform the common daily tasks of the FAS independently, demonstrating that particularly number 13 (take own medications without help), 18 (walk to places in neighborhood without help), and 24 (use toilet/commode without help) were less frequently performed independently by nursing home residents. The logistic regression model with married/domestic partnership as independent variable showed that not being married (i.e. not having a partner) was predictive of institutionalization in patients with HD ($p = 0.010$, OR 7.50, 95%CI 1.60–35.08), meaning that non-married patients were 7.50 times more likely to being institutionalized than married patients. The univariate regression model with the dichotomized FAS score as independent variable showed that lower scores were also associated with institutionalization ($p = 0.047$, OR 4.64, 95%CI 1.02–21.00), but this association is not as strong as the association between not being married and nursing home admission.

DISCUSSION

This study identified associations between clinical characteristics of patients with advanced HD in day-care and in a nursing home. Our results showed that lower scores on the FAS of the UHDRS functional domain and not being married were related to institutionalization. Probably, lower functional capacity

Table 1
Demographics for nursing home residents and day-care participants with Huntington's disease

	Nursing home residents ($n = 28$)	Day-care participants ($n = 12$)	*p*-value
Age, years	57.5 (46.0–66.8)	51.5 (45.3–63.8)	$p = 0.512$
Gender, male/female (%male)	8/20 (28.6%)	6/6 (50.0%)	$p = 0.193$
CAG repeat length	44.0 (42.0–47.3)[a]	44.0 (42.0–47.5)	$p = 0.938$
Educational level, years	12.0 (12.0–14.5)	14.5 (12.0–16.8)	$p = 0.074$
Age of disease onset, years	40.0 (31.3–49.8)	40.5 (33.3–49.3)	$p = 0.998$
Disease duration, years	13.0 (10.0–18.8)	11.5 (8.3–16.3)	$p = 0.202$
Residence duration, years	3.0 (1.3–5.0)	NA	NA
Medication for HD, yes/no (%yes)	26/2 (92.9%)	12/0 (100.0%)	$p = 0.342$
Married, yes/no (%yes)	8/20 (28.6%)	9/3 (75.0%)	**$p = 0.006$**

Data are median (IQ-range); except for gender, medication for HD, and married, which are number (%). *p*-values were calculated using Mann-Whitney U tests; except for gender, medication for HD, and married, which were calculated using Chi-square tests. [a]CAG repeat length was missing for two nursing home residents; they tested positive for HD through linkage analysis. CAG, cytosine-adenine-guanine; HD, Huntington's disease; NA, not applicable; IQ, interquartile.

Table 2
Clinical characteristics of nursing home residents and day-care participants with Huntington's disease

	Nursing home residents ($n = 28$)	Day-care participants ($n = 12$)	*p*-value
UHDRS			
Motor score	75.0 (40.5–88.0)	49.0 (29.8–82.3)	$p = 0.115$
Cognitive score	51.5 (10.0–96.8)	102.0 (51.5–148.8)	$p = 0.069$
Behavioral score	12.5 (8.0–24.8)	17.0 (9.0–25.0)	$p = 0.610$
Functional Assessment Scale	7.5 (1.3–13.8)	13.5 (8.3–17.0)	**$p = 0.022$**
Independence Scale	60.0 (36.3–65.0)	62.5 (52.5–70.0)	$p = 0.163$
Total Functional Capacity	1.0 (0.0–3.8)	3.5 (2.3–6.8)	**$p = 0.011$**
Modified TFC	1.0 (0.0–3.8)	2.5 (1.3–5.8)	$p = 0.163$
UHDRS-FAP			
Motor score	13.0 (4.3–25.3)	8.0 (6.0–17.8)	$p = 0.512$
Cognitive score	89.8 (51.8–139.9)	141.0 (90.3–167.5)	$p = 0.096$
Somatic score	5.0 (1.3–13.0)	5.5 (2.3–9.0)	$p = 0.805$
Behavioral score	2.0 (1.0–3.0)	1.0 (0.3–3.0)	$p = 0.531$

Data are median (IQ-range). *p*-values were calculated using Mann-Whitney U tests. UHDRS, Unified Huntington's Disease Rating Scale; UHDRS-FAP, Unified Huntington's Disease Rating Scale-For Advanced Patients; TFC, Total Functional Capacity; IQ, interquartile.

causes more dependency on care for common daily tasks, which ultimately leads to institutionalization. The association between lower functional capacity and nursing home placement is consistent with previous findings that the ADL scale is related to institutionalization [6]. We did not find correlations between institutionalization and motor disturbances, cognitive performance, and behavioral abnormalities on the UHDRS and UHDRS-FAP. This finding indicates that nursing home residents are not more affected by the triad of symptoms characteristic for HD then patients in day-care, suggesting that these symptoms do not cause institutionalization. Our findings contrast with the results of previous studies, which indicated bradykinesia, impaired gait, and impaired tandem walking of the UHDRS-TMS [5], the Motor Impairment Score (MIS), and the MMSE [6] as predictors for nursing home placement. Like our study, psychiatric symptoms were not shown to predict institutionalization in HD in these

studies [5, 6]. However, in our study less patients from the nursing home department specialized in psychiatric problems chose to participate compared to the other departments, which may have underestimated the amount of psychiatric symptoms in the nursing home residents.

The results of the individual univariate logistic regression analysis suggest that not being married is more predictive of institutionalization in HD patients than lower scores on the FAS. This finding implies that having a partner protects better against nursing home admission than higher functional ability. Possibly, a caregiver living with a patient assists with ADL tasks which the patient could not have done independently, resulting in being able to live at home longer than a patient without a caregiver living with the patient. Assistance with ADL tasks and care can also be provided by home care services or caregivers not living with a patient (usually their children), but this care is not available 24 hours a day.
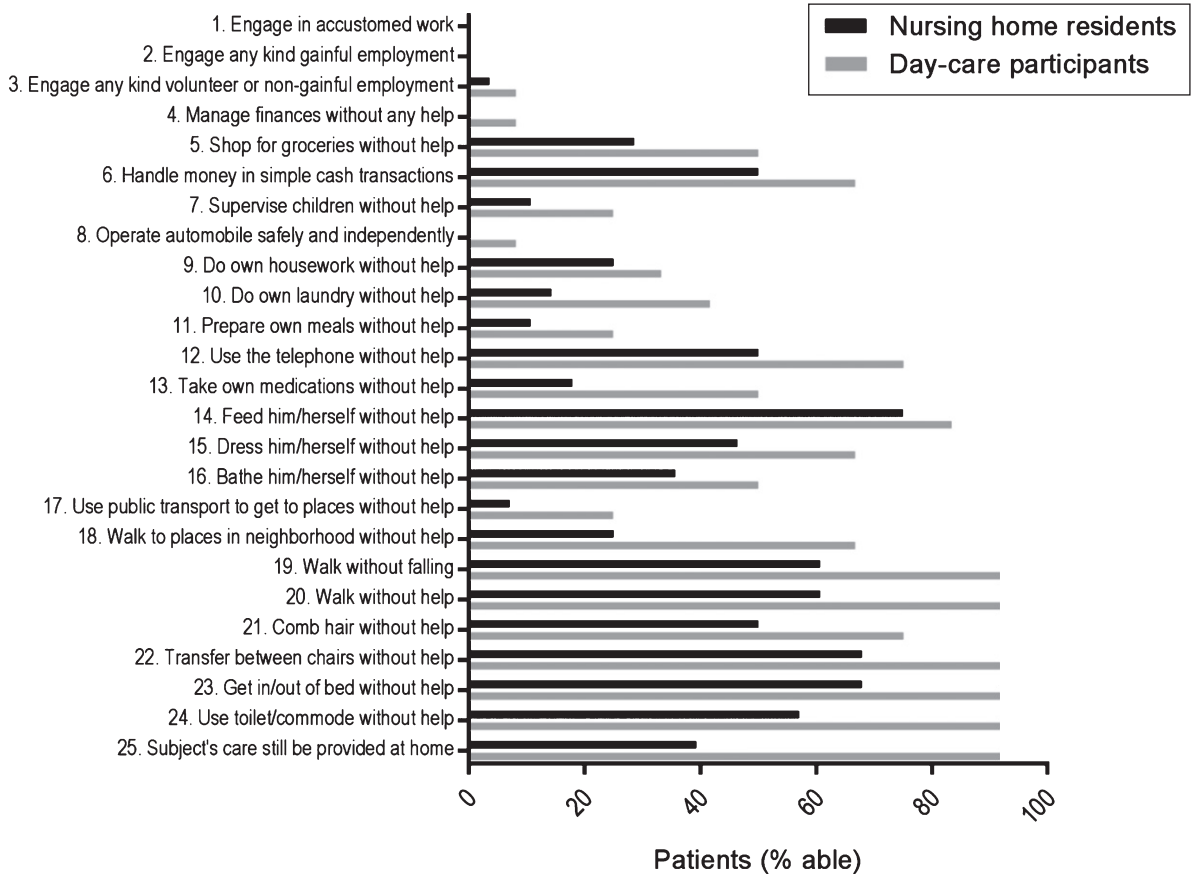
Fig. 1. Percentage of nursing home residents and day-care participants being able to perform the common daily tasks of the Functional Assessment Scale independently.

For example, when a patient is not able to use the toilet independently or walk to places in the neighborhood independently (with the chance of wandering), (s)he usually needs supervision 24 hours a day by a partner or nursing home facility. HD patients are usually admitted to a nursing home when middle-aged [4–7]. Accordingly, partners of HD patients are generally younger than partners of average nursing home residents. Younger partners may have a better health status than older partners and may, therefore, be better capable to provide care for the patient at home. However, younger partners usually have a job and, consequently, are often not home to take care of the patient.

To our knowledge, no study has been performed investigating the influence of a partner/caregiver on institutionalization in HD. A systematic review in patients with dementia showed that married patients and patients living with their caregiver had a lower risk of nursing home placement [17]. Greater

dementia severity, older age, neuropsychiatric symptoms, impaired cognition, and more functional impairment were also predictors of long-term care admission [17, 18]. Additionally, caregiver burden and inability of the caregiver to care for the patient were stated as reasons for institutionalization by caregivers of patients with dementia. The reasons for admission varied between countries. For example, none of the French caregivers expressed inability to care as a reason for institutionalization in patients with dementia, while this category was mentioned by 30% of the Spanish caregivers [18]. Reasons for nursing home placement in HD could also differ between countries, due to the organization of nursing home care, financial costs, and cultural aspects. Therefore, our results may be hard to apply to other countries, where HD care is less well organized or more expensive than in the Netherlands.

Ideally, identification of predictors for institutionalization lead to interventions and treatment

strategies that postpone the need for nursing home placement. However, not having a partner cannot be changed. Providing support to unmarried HD patients, who do not have a caregiver living with them, by home care services specialized in HD might increase the chance of the best possible care in the own environment before institutionalization and postpone nursing home admission. The last question of the FAS is 'could subject's care still be provided at home?', which gives an overview of the examiner's perception of the patient's functional ability. The examiners answered this question with 'yes' in 39.3% of the institutionalized HD patients, suggesting that for almost 40% of the institutionalized patients care could be provided at home if the patient had a suitable caregiver/partner. After nursing home placement, the FAS can provide information about a patient's dependency on care easily and quickly, which could possibly optimize their care. For example, less than 50% of the nursing home residents were able to bath or dress themselves without help, but more than 50% of the patients were able to walk, use the toilet, or feed themselves without help.

The strength of our study is the use of the UHDRS and UHDRS-FAP for the clinical assessment of the patients. These scales were developed especially for HD patients. A limitation of our study is the small sample size and, therefore, this study might be underpowered. Due to the small sample size, we could not perform a multivariate regression analysis and in the univariate regression models the ranges of the confidence intervals of the odds ratios were very wide. The use of two examiners is another limitation and could have influenced our results due to interrater reliability. For some patients the caregiver could not attend the visit, which may have caused underreporting of symptoms due to reduced awareness of symptoms by the patient. Additionally, we did not collect information about the caregivers or questioned the caregivers independently from the patients. Further research is needed to investigate the role of caregivers. Ideally, cultural aspects, financial costs, and organization of HD care across different countries will also be explored.

In conclusion, our study in patients with advanced HD residing in a nursing home or receiving day-care showed that not being married/not having a partner and low functional abilities were associated with institutionalization. Motor impairment, cognitive function, and psychiatric symptoms were not related to nursing home placement. Marital status seemed more predictive for nursing home admission than functional capacity, suggesting that a partner can compensate for high dependency on care for ADL tasks.

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CONFLICT OF INTEREST

The authors have no conflict of interest to report relevant to this work. The institute of R.A.C. Roos received payment from Teva Pharmaceutical Industries Ltd outside the submitted work. He is also advisor for uniQure N.V. outside the submitted work.

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