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Impact of Huntington's disease on working and driving

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Citation

Essink-Jacobs, M. (2019, May 8). *Impact of Huntington's disease on working and driving*. Retrieved from <https://hdl.handle.net/1887/72198>

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Author: Essink-Jacobs, M.

Title: Impact of Huntington's disease on working and driving

Issue Date: 2019-05-08

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Reasons why Huntington's disease gene carriers decide to stop working or driving

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Submitted

ABSTRACT

Background

In patients with Huntington's disease (HD), daily activities such as working and driving are affected at a relatively young age. Discussing the possibility of stopping work and driving is sensitive.

Objective

To explore why HD gene carriers decide to stop working or driving and to gain insight into their personal motives. Secondly, to investigate whether these motives differed between males and females.

Methods

Questionnaire responses of 191 HD gene carriers, who visited the outpatient clinic of the Leiden University Medical Center between 2016 and 2018, were used in this study. Self-reported reasons for work and driving cessation were explored and categorized according to gender.

Results

A total of 92 HD gene carriers (48% of the responders; 38 males and 54 females) had stopped working, due to reduced concentration (25%), problems with multi-tasking (22%), and slower reactions (21%). A total of 62 HD gene carriers (37%) had stopped driving a car (19 males and 43 females). They were significantly older ($p = 0.018$) and more often female ($p < 0.001$) compared to the active drivers. Males mainly reported difficulties with concentration (26.1%), whereas females stated anxiousness and insecurity as their primary reason for stopping driving (16.3%).

Conclusions

According to HD gene carriers, reduced concentration interferes with the decisions to stop working and stop driving. Gender differences and personal motives should be taken into consideration when discussing changes in daily life in the clinic.

INTRODUCTION

In patients with Huntington's disease (HD), activities of daily life, such as working and driving are affected at a relatively young age.^{1,2} Inability to work and decreased driving skills are among the first functional changes to be reported by HD gene carriers.^{3,4} Decreased occupational performance has been reported by more than two-thirds of the patients with HD and more than half of the HD patients are unable to work in their usual profession.³ In addition, HD gene carriers are concerned about the impact of their gene status on their employment.⁵ Not being able to work or a reduction in working time are associated with negative emotions, such as anger and a depressed mood. On the other hand, a reduction in the number of hours or the demands of work can also lower levels of stress.⁶ The clinical signs of HD, specifically apathy and executive dysfunction, have been shown to predict unemployment.⁷ Reasons why HD gene carriers decide to stop working have not been studied previously.

Patients with HD might be reluctant to stop driving, because this influences their independence and quality of life. Discussing in the clinic the possibility of stopping driving is, therefore, sensitive. In older adults, not being able to drive has been related to signs of depression.⁸ Furthermore, driving cessation does not only impact the independence of patients, but also has consequences for family members.⁹ Caregivers often have to compensate when the patient can no longer drive, influencing their lives and familial roles. In the Netherlands, patients with HD have the moral obligation to report a potential decline in driving performance to the national driver's licensing authority.¹⁰ By the means of a clinical and neurological examination, a neurologist or independent physician has to evaluate the patient's ability to drive. Based on this examination and the physicians' opinion, a formal driving test may become mandatory; patients have to undergo this at their own expense. If a patient passes the driving test, then the driver's license may be renewed for a maximum of five years. This process is comparable to that in other European countries.¹¹ Studies in older adults with dementia have shown that driving cessation is a gradual process with distinct stages, often starting with small alterations in driving behavior, such as avoiding certain situations (e.g., unknown roads, motorways).^{12,13} The number of driving restrictions usually increases, eventually resulting in complete cessation. Driving behavior is also influenced by self-monitoring and personal insight into driving capacity.¹⁴

Reasons for driving cessation have been classified into external and internal reasons. External reasons are those outside the individual, such as the influence of

family members, doctors, or lifestyle changes. Reasons within the individual, such as declining health or psychological factors (e.g., anxiety), are defined as internal reasons. In older adults, internal factors, especially a decline in health, seem to have the highest impact on changes in driving and the decision to stop.^{12,15} In general, women tend to stop driving at a younger age than men.^{15,16} Although previous studies have revealed driving impairments in HD, it remains, to our knowledge, unclear why HD gene carriers decide to stop driving.

The aim of this study is to explore why HD gene carriers stop working or driving, and to obtain a better understanding of their personal motives. Secondly, we aim to provide guidance for clinicians when discussing work and driving cessation with patients in the clinic and to increase awareness of these issues in the HD population.

MATERIALS AND METHODS

Between 2016 and 2018, HD gene carriers who visited the outpatient clinic of the Neurology Department at the Leiden University Medical Center in the Netherlands were asked to fill in a short survey containing items on symptoms and potential functional changes due to HD. The questionnaire was designed to provide the treating physician with insight into the patients' self-perception and general functioning.

The items on occupation (i.e., current working status, reason for work cessation, year of work cessation) and driving a car (i.e., type of driver's license, current driving status, reasons for driving cessation, year of driving cessation, and, for those still driving, preferred driving conditions (e.g., urban driving, not driving by themselves)) were used in the current study. Ethical approval for the use of the anonymous survey responses was obtained from the local medical ethics committee of the LUMC. Only those who worked in a paid job were selected for inclusion in the workers group. Gene carriers who stopped working due to HD-related symptoms were defined as not working. Gene carriers who stopped working because of other reasons (e.g., age) and those who had never obtained a driver's license were excluded from the analyses.

Reported reasons for work and driving cessation were described and percentages of these reasons were explored, separately for males and females. Age and gender differences between working and not working and drivers and not drivers were analyzed using separate logistic regression analysis with working or driving status as dependent variable. All analyses were performed using SPSS (version 23.0) for Windows.

RESULTS

Data of 191 HD gene carriers (77 males, 114 females) who filled in the questionnaire were used. The mean age was 50.2 years.

Work cessation

Of the 191 responses, 92 HD gene carriers (48%; 38 males, 54 females) retired from work due to signs of HD and 55 (25 males, 30 females) were still working in a paid job (Table 1). The median number of years of not working, defined as the difference between year of retirement and year the questionnaire was filled in, was 4.0 years. Women had been retired longer compared to men (5.5 years versus 3.0 years, respectively). HD gene carriers who stopped working were significantly older (OR = 1.08, $p < 0.001$) compared to gene carriers who were still working. There was no significant difference in gender (OR = 0.84, $p = 0.623$) between the working and not working group. Reduced concentration (25%), problems with multi-tasking (22%), and slower reactions (physical and mental; 21%) were the primary reasons noted for stopping working (Table 2). Here, no differences were observed between males and females; both mentioned reduced concentration most frequently.

TABLE 1 Demographics

	Working (N=55)	Not working (N=92)	Drivers (N=105)	Not driving (N=62)
Age (years)	44.4 ± 9.2	51.0 ± 9.3	48.5 ± 9.8	56.3 ± 10.2
Gender male/female	25/30	38/54	52/53	19/43
Years of not working (male/female)	NA	4.0 (3.0 / 5.5)	NA	NA
Years of not driving (male/female)	NA	NA	NA	3.5 (2.0 / 5.0)

Data are mean ± SD for age, number for gender, and median for number of years not driving and number of years not working for all participants and separately for males and females. NA: not applicable

TABLE 2 Self-reported reasons for stopping working by HD gene carriers

Reason for stopping working	Total number of times reported
Reduced concentration and attention/vigilance	N = 58 (25.4%)
Problems multi-tasking	N = 50 (21.9%)
Slower reactions (physical and mental)	N = 47 (20.6%)
Forgetfulness	N = 29 (12.7%)
Physical burden of the job became too heavy	N = 28 (12.3%)
Unspecified reason	N = 16 (7.0%)

Note: total number is not equal to the total number of gene carriers who stopped working N=92) because most gene carriers reported multiple reasons which were counted in all categories.

Driving cessation

Of the 191 HD gene carriers who filled in the questionnaire, 168 (97 females, 71 males) had obtained a driver's license. One hundred and five gene carriers were still driving, whereas 62 (37%) reported they had stopped of whom 43 (69.3%) were female and 19 (30.6%) were men (Table 1). One female temporarily stopped driving while waiting for a formal driving test. These data were not included in the analyses. HD gene carriers who stopped driving were significantly older (OR = 2.22, $p = 0.018$) and more often female (OR = 1.08, $p < 0.001$) compared to gene carriers who were still active drivers. The median period since driving cessation, defined as the difference between year of driving cessation and year the questionnaire was filled in, was 3.5 years for all HD gene carriers, 2.0 and 5.0 years for males and females, respectively.

Fourteen percent of the HD gene carriers reported multiple reasons for their decision to stop driving, and some (6.9%) reported a generic reason, such as "I prefer walking" or "I just stopped driving" (Table 3). Internal reasons for driving cessation were endorsed by 58.5% of the HD gene carriers (Figure 1A). For males, difficulties concentrating (26.1%) was the primary reason for stopping driving, whereas the most frequently answered reason for cessation reported by females was feeling of anxiety and insecurity (16.2%) (Figure 1B). Of the active drivers, 21.9% made minor alterations in their driving behavior; only driving in their own neighborhood and no motorway

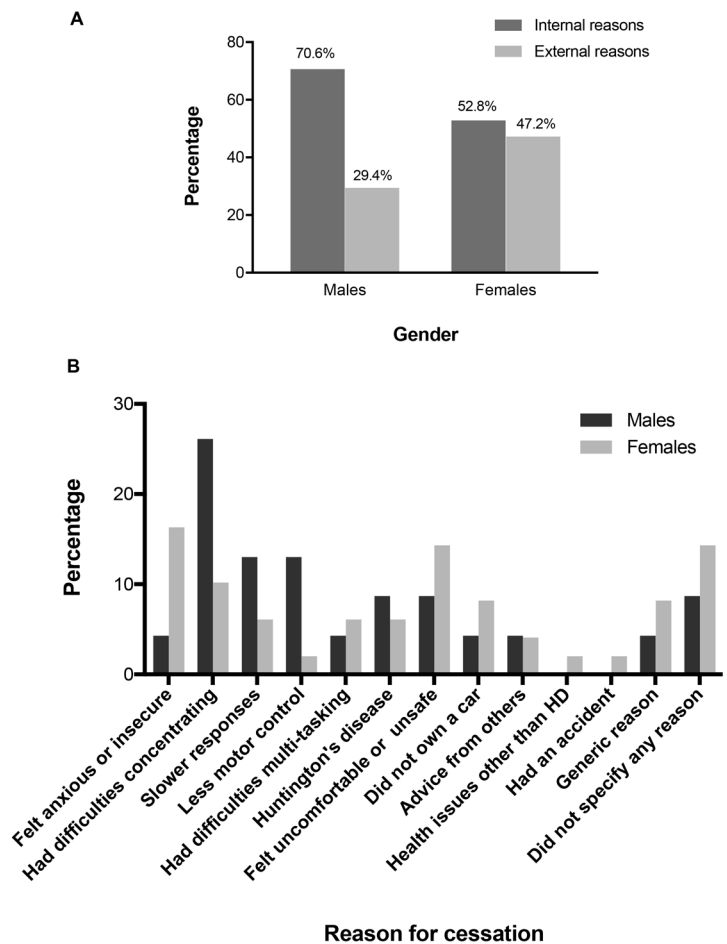
driving being most frequently noted (10.5%). Forty-six HD gene carriers (13 males, 33 females) stopped both working and driving. The mean age of this group was 53.9 years, with median period of unemployment of 7 years and median period of driving cessation of 3 years. In this group, the primary reported reason for stopping work was reduced concentration (26%), and the main reasons for stopping driving were reduced concentration (15%), anxiety (15%), and feeling uncomfortable/unsafe (15%).

TABLE 3 Reported reasons for driving cessation by HD gene carriers

Reason for stopping driving	Total (N=62)		Males (N=19)		Females (N=43)	
Had difficulties concentrating	11	15.3%	6	26.1%	5	10.2%
Felt anxious or insecure	9	12.5%	1	4.3%	8	16.3%
Felt uncomfortable or unsafe	9	12.5%	2	8.7%	7	14.3%
Slower responses	6	8.3%	3	13.0%	3	6.1%
Huntington's disease	5	6.9%	2	8.7%	3	6.1%
Did not own a car	5	6.9%	1	4.3%	4	8.2%
Less motor control	4	5.6%	3	13.0%	1	2.0%
Had difficulties multi-tasking	4	5.6%	1	4.3%	3	6.1%
Advice from others (e.g., family or physician)	3	4.2%	1	4.3%	2	4.1%
Health issues other than HD	1	1.4%	0	0%	1	2.0%
Had an accident	1	1.4%	0	0%	1	2.0%
Generic reason (e.g., 'prefer walking', 'just stopped driving')	5	6.9%	1	4.3%	4	8.2%
Did not specify any reason	9	12.5%	2	8.7%	7	14.3%

Note: Some HD gene carriers reported multiple reasons, therefore, the total number in the first column does not equal the number of gene carriers. The most frequently reported reasons per gender are presented in bold.

FIGURE 1 Reported reasons for driving cessation for males and females



Note: Figure 1A shows the percentage of internal and external reasons for driving cessation separately for males and females. Figure 1B shows percentages of all reported reasons for males and females separately.

DISCUSSION

Our study showed that reduced concentration, problems with multi-tasking, and slower reactions influenced the decision to stop working. No differences were observed between males and females. HD gene carriers who stopped driving were older and more often female compared to gene carriers who were still active drivers. Difficulty concentrating was the primary reason why males stopped driving, whereas females reported feelings of anxiety and insecurity as their main reason. Occupational changes have previously been reported as one of the earliest functional declines observed in HD, emphasizing the need for early discussion.³ Our results support the idea that cognitive functioning interferes more with activities of daily life than the characteristic motor symptoms of HD. Previous studies have also shown that cognitive decline, specifically in executive functions, is related to unemployment in HD.^{7,17} Most gene carriers reported a combination of multiple reasons for stopping working, implying that there is not a single factor that influences the decision to stop. In our study, women retired earlier than men, which is comparable to findings of studies in Parkinson's disease.¹⁸ It is important to educate patients, caregivers and employers to ensure that patients can maintain gainful employment for as long as possible. Making adjustments to the demands of work may offer a solution, but this might not be possible for all professions.

The fact that HD gene carriers worry about the effect of their gene status on their employment suggests that they might be reluctant to address work-related difficulties during a consultation with their physician.⁵ However, an early discussion about the future necessity to stop working and driving will allow the patient to anticipate functional changes due to HD, especially as the onset of clinical symptoms of HD occurs during mid-adulthood, a period of life when patients rely on the financial benefit of employment and the independence afforded by driving a car. The mean age of the HD gene carriers who stopped work in our study was 51 years, well below the general retirement age of 65-67 years. In addition, the mean age of the former drivers in our study was relatively young compared to other studies focusing on older adults and dementia.^{16,19}

Although males generally have the tendency to externalize their behavior, our study showed that males mostly reported internal reasons for driving cessation.²⁰ An explanation might be that males are more reluctant to stop driving because they rely more heavily on their car for work. In addition, males tend to be less risk averse compared to females.¹⁵ Decreased cognitive functioning has been related to driving impairments and might influence the decision to stop.^{21,22} This could also explain

the fact that, in our study, most males reported reduced concentration as their primary reason for driving cessation. Previous studies in older adults showed that their own opinion was the deciding factor to stop driving.^{19,23} Our findings suggest that gender influences this decision and that females might be more sensitive than males to external factors, such as the opinions of family or their physician. In addition, females also have the tendency to stop driving at a younger age and while in better health compared to males, which could explain our result that the duration of driving cessation was longer in females than in males.²⁴

We agree with previous studies stating that driving cessation is a gradual and dynamic process that not only involves patients and their relatives, but also the treating physician and the driver's licensing authority.^{12,23,25} In our cohort, gene carriers who still drove reported adaptations in their driving behavior, confirming this suggestion of gradual changes. The treating physician should include the topic of driving safety during routine visits and provide the patient with the right information to ease decision-making. Changes in the ability to drive safely should be discussed in an early phase of HD, without suggesting the patient simply stops driving following genetic confirmation. A study in patients with Alzheimer's disease (AD) showed that transportation and law-enforcement professionals were of the opinion that a diagnosis of AD, regardless of disease severity, was an indicator for driving cessation.²⁶ This suggests that driver's licensing authorities need to be well educated about the different stages of HD. We recommend an individual approach to discussing driving safety, acknowledging the differences between male and female perspectives observed in our study. The moral obligation to report changes in health has to be emphasized, because if the patient does not inform the national driver's licensing authority, this can affect their insurance if they are involved in a collision or traffic violation. Here, it is also important to consider the possible limited self-insight of patients with HD.^{27,28} Decreased awareness of impairments (i.e., anosognosia) could lead to an overestimation of the patient's own capacity or an under-reporting of functional impairments such as driving.²⁹ This could also have affected the results of the current study.

A limitation of our study is that the questionnaire was designed for use in the outpatient clinic as a screening questionnaire with different response options (multiple choice and open questions), and not specifically for research purposes. It contained global items on driving and not much information about driving restrictions by gene carriers who are still active drivers. Including more specific items or focus groups could be useful to further explore whether the decision to stop driving is indeed a process with distinct stages, as suggested in previous

studies.^{23,25,26} In addition, it would be interesting to compare our results with those of other European countries in view of the differences in regulations. Using a validated questionnaire is recommended. A more detailed approach, taking different types of professions into consideration and dividing patients into more working categories than the dichotomous employed versus unemployed categories, could provide more in-depth knowledge regarding the influence of HD on the ability to work. Due to the study design, we were unable to distinguish between different HD disease stages and no information was available regarding symptom severity. Nevertheless, our study provides greater insight into the daily life issues that patients with HD are confronted with and has implications for clinical care.

In conclusion, according to HD gene carriers, reduced concentration interferes with both the ability to work and drive. We identified that both internal and external reasons influence the decision to stop driving and that these reasons differ between males and females. Gender differences and personal situations should be taken into account in the clinic when discussing changes in daily life due to HD.

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