Fear of choking and fear of falling in middle and end stage patients with Huntington's disease

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SUMMARY

The aim of this thesis is to explore fear of choking (FoC) and fear of falling (FoF) in patients with Huntington's Disease (HD) and their caregivers. These patients were either living in or attending day care in Dutch nursing homes, specialized exclusively in long-term care of HD patients. HD is a neurodegenerative autosomal dominantly inherited disease of the central nervous system. Characteristics of the disease are motor symptoms, psychiatric and behavioral disturbances, and dementia. The prevalence is estimated at 5-10/100,000 in the Caucasian population. The mean age at onset is 30-50 years with a wide range (2-85) and a mean duration of 17-20 years. Disease progression leads to increased dependency in daily life and to progressive care needs. Treatment is multidisciplinary and based on symptomatic therapy with the aim of improving quality of life.

The introduction of this thesis **(chapter 1)** describes how dysphagia and falls are common in HD and can lead to FoC and FoF. However, knowledge of FoC and FoF in both HD patients and their caregivers is mostly lacking, as well as knowledge about the relationship between cognitive and emotional factors and these types of fear.

We explored the incidence of falling, and the association with fear of falling and avoiding activities, in nursing home residents with Huntington's disease compared to other nursing home residents (**chapter 2**). We used data, recorded in 2012, 2013 and 2014, from the cross-sectional multi-centre point prevalence study of relevant care problems, including falls, known as the Dutch National Prevalence Measurement of Care Problems. Fifty-seven HD residents and 404 non-HD residents were included over a three-year period. The participants lived in eight Dutch nursing homes belonging to one organization. One of these nursing homes cares exclusively for Huntington residents. Our study illustrated that the fall incidence in a 30-day period was significantly higher in residents with HD (30%) than in non-HD residents (10%). In contrast, the prevalence of FoF in HD residents was significantly lower (14%), compared to non-HD residents (30%). Although there was a lower rate of avoiding activities in the HD group (11% versus 18%), this difference was without statistical significance. A significant relationship between fall incidence and FoF or activity avoidance was not found in this study. A predictor of FoF was an interaction between the group and age. Fewer HD residents experienced FoF and this difference increased with age. Activity avoidance was predicted by group and care dependency. Based on our results, we recommend that in future research, cognitive functioning and insight into cognitive deficits should be included as factors which possibly contribute to fear of falling.

Chapter 3 presents a literature review exploring the relationship between cognition, awareness and anxiety and experiencing FoC or FoF in HD, Parkinson's disease (PD) and dementia. As the literature on HD is limited, the study population was expanded

with studies on PD and dementia, both of which are neurodegenerative diseases sharing symptoms and signs with HD. Studies on PD and dementia might, therefore, offer some reference points for HD. Although dysphagia and accidental falls are common symptoms in patients with HD, PD and dementia, data on emotional and cognitive factors and their relationship with FoC and FoF are limited. For HD, no reports were found related to the objective of the study. In PD and dementia, only studies addressing the relation between FoF and cognition, respectively, anxiety were found. However, the results of these studies were contradictory. Therefore, we recommend that future research focuses specifically on the relationship between cognition, awareness and anxiety and experiencing FoC or FoF. Better insight might result in therapeutic options for patients and/or guidelines for caregivers, regarding prevention and adequate coping strategies, and thus improve quality of care.

Based on the findings reported in chapters 2 and 3, we have written a protocol for a multi-center, observational, cross-sectional study in Dutch nursing homes, specialized in long-term care of patients with Huntington's Disease **(chapter 4)**. Our aims were to explore the prevalence of fear of choking and fear of falling in HD patients, 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, we wanted to explore related problems encountered by their (in)formal caregivers. This study might 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.

The results of this comprehensive study are reported in the chapters on FoF **(chapter 5)** and FoC **(chapter 6).**

The overall study confirms that the fall prevalence in individuals with HD is high, with a mean prevalence of almost 30% for all 158 HD patients included **(chapter 5)**. FoF is also a common problem, not only in individuals with HD where the prevalence is approximately 50%, but also in their (in)formal caregivers. In the current study, most FoF was found among informal caregivers (63%); less among formal caregivers (26%). Anticipatory awareness of fall risks and gender appeared to be predictors of FoF in care-independent HD patients, though not in the group of care-dependent HD patients. Physiotherapy and assistive devices were the most frequently used fall preventive measures, but in most individuals with HD, a combination of measures was used. In addition to the use of individual multifactorial fall prevention strategies, we recommend supporting both formal and informal caregivers to cope with falls.

Dysphagia symptoms and signs were present in 91% of all 158 HD patients **(chapter 6)**. The prevalence of FoC in HD patients was 52%. Most FoC was found in the informal

caregivers and less in formal caregivers within the groups of care-independent (60% and 19%, respectively) and care-dependent (78% and 50%, respectively) HD patients; between the groups, most FoC was reported by caregivers of the more severe and care-dependent HD patients, compared to caregivers of the care-independent HD patients. We found no association between FoC and lack of awareness, cognition and anxiety. However, the score on the Huntington's Disease Dysphagia Scale was a predictor for FoC. Speech-language therapy, supervision while eating or drinking, and adapted consistencies of food and drinks were the measures most frequently used to manage dysphagia in this study; in most patients, a combination was applied.

Finally, in **chapter 7** the most important findings of this thesis and implications for clinical practice and future research are discussed. As a result of the studies, we have learned that, in addition to falls and dysphagia, FoC and FoF are common problems in HD. Because there is no cure for HD, the focus is on improving quality of life and providing tailored support to patients and families. Because FoC and FoF are a real burden for both patients and caregivers, and because they may also have an impact on physical functioning, it is important to address these issues in daily practice. Therefore, it is recommended that FoC and FoF are continually discussed during the course of the disease and that possible avoidance behaviors of patients be identified, allowing timely (multi-disciplinary) interventions, which may prevent incipient signs and symptoms from worsening. In addition, even after admission to the nursing home, it is still important to continually involve informal caregivers and support them to cope with dysphagia and falls.

Future studies can improve both the management of dysphagia and FoC and falls and FoF. Improved knowledge of the predictors of FoC and FoF can lead to a better management of the identified risk factors. Another topic for future research is the development of diagnostic tools for FoC and FoF for this patient group, because questioning HD patients themselves or the use of questionnaires may be difficult, due to speech- and cognitive problems. Finally, tailored interventions should be assessed in order to reduce dysphagia and FoC, as well as falls and FoF.