Chapter 4

The Huntington's Disease Dysphagia Scale

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Abstract

Background

Little is known about the swallowing disturbances of patients with Huntington's Disease; therefore, we developed the Huntington's Disease Dysphagia Scale.

Methods

The scale was developed in four stages: 1) item generation, 2) comprehension testing, 3) evaluation of reliability, 4) item reduction and validity testing. The questionnaire was presented twice to 50 Huntington's disease patients and their caregivers. The Kruskal-Wallis test was used to evaluate whether the severity of swallowing difficulties increased with advancing disease. Pearson's correlation coefficient was used to examine the construct validity with the Swallowing Disturbance Questionnaire.

Results

The final version contained 11 items with five response options and exhibit a Cronbach's alpha coefficient of 0.728. The severity of swallowing difficulties was significantly higher in more advanced Huntington's disease. The correlation with the Swallowing Disturbance Questionnaire was 0.734.

Conclusion

We developed a valid and reliable 11-item scale to measure the severity of dysphagia in Huntington's disease.

Introduction

Huntington's disease (HD) is a progressive neurodegenerative disease characterized by disturbed movements, behaviour, and cognition.[1] Patients with HD suffer from dysphagia, and patients have swallowing difficulties in all phases of ingestion.[2-7] Carefully designed longitudinal studies on dysphagia in HD are required to clarify when dysphagia emanates, what the dysphagia symptoms in the different stages of ingestion are, and whether the severity of dysphagia increases with advancing HD. This may lead to increased awareness and a treatment that is more tailored to the patient's individual needs. A first requirement to address these questions is the availability of a good measurement instrument tailored to the specific swallowing impairments of this population, but because such an instrument does not exist, we had a goal of developing a patient-reported outcome measure, the Huntington's Disease Dysphagia Scale (HDDS).

Patients and methods

Expert team

The task of the HD expert team was to generate items that evaluated swallowing difficulties in general and to add items that were specific to HD. The items were complemented with items drawn from the literature. The experts were then asked to rate the relevance of each item (0=not important, 5=very important). Items with a score of 3 or greater were included in the initial scale. The professions of the experts were neurologists (n=2), speech-and-language therapists (n=4), an ear-nose-throat specialist (n=1), radiologist (n=1), nursing home physicians (n=2), and a methodologist (n=1). Almost all experts were experienced with HD, except one speech-and-language therapist, the ear-nose-throat specialist, and the radiologist, but they were experienced in dysphagia.

Participants

Patients with a CAG repeat size of 36 or greater and without other diseases that could affect swallowing were included. Patients from all three clinical stages[8] - in our study defined as stage I: patients living at home (n=23); stage II: patients living at home who attended day care (n=18); and stage III: patients living in a nursing home (n=14)- were enrolled. Stage I patients were consecutively selected from the outpatient neurology department of the Leiden University Medical Centre (LUMC), whereas patients in stage II and III were consecutively

selected from a Huntington Care center, a center specializing in care and treatment of HD patients. The center provides specialized units for HD patients who are in the mid or end-stage, and also offers the possibility for early HD patients to receive day care.
Proxy information on the patient's swallowing problems was obtained from the partner or caregiver, or from a close friend.
The study was approved by the medical ethics committee of the LUMC. All patients gave informed consent.

Scale development

The scale was developed in Dutch in four stages: (1) item generation, based on a literature search and input by experts; (2) item comprehension testing in patients; (3) evaluating reliability in patients and relatives; (4) item reduction and construction of the final scale, followed by examining the scale for the presence of floor and ceiling effects (i.e., 15% or greater of patients scoring the minimum or maximum possible total score), overall reproducibility, as well as construct validity by comparison with the Swallowing Disturbance Questionnaire (SDQ).[9]

Item generation

A large pool of questions regarding dysphagia in relation to HD at the level of disability was collected. Items were drawn from other existing dysphagia scales,[9-21] the literature on dysphagia in HD,[2-7] and from our clinical experience with HD.

Item comprehension testing

Intelligibility of questions was investigated in 10 HD patients sampled from the various clinical stages using the "Thinking aloud" procedure. Patients were also encouraged to add items. During this phase, items were rephrased continuously until all items were easily understood and interpreted correctly. The response options addressed the frequency of the problem: (0) almost never, (1) seldom, (2) sometimes, (3) frequently, (4) almost always.

Evaluating reliability in HD patients

The initial questionnaire was sent to all of the included 55 patients with HD. To measure intra-rater reliability, the same questionnaire was sent again to these patients 2 weeks after the first questionnaire had been received. Test-retest reliability of the individual items and total score were analysed with the Intraclass Correlation Coefficient (ICC; one-way random effects model). Items with ICC's less than 0.30 were removed. Relatives of the included

patients also received the questionnaire twice and rated their impression of the patients' swallowing difficulties. The aim of this proxy information was to find out whether close relatives were able to identify the patient's swallowing difficulties. When patients did not return the first questionnaire, but their relatives did, both patients and relatives received the second questionnaire after 2 weeks. The ICC was used to calculate the agreement between both outcomes. A total score was calculated if three or fewer values were missing, in which the mean of the other values was used to replace the missing values; no total score was computed if four or more values were missing.

Item reduction, scale construction, and construct validity

After removal of items with low intra-rater reliability, additional item reduction was based on the strength of the inter-item correlation, where items with high (>0.80; indicating redundancy) or low (<0.20; indicating incoherence) item-total correlations were removed. Additionally, items whose presence resulted in a decrease of Cronbach's alpha were also removed.

To examine the construct validity, the final questionnaire was correlated with the SDQ[9] (Pearson's correlation coefficient), which was completed simultaneously with the HDDS by all patients and relatives. The SDQ was chosen because this scale emerged as a valid tool to detect early dysphagia in patients with Parkinson's disease.

Psychometric performance SDQ

Test-retest reliability of the SDQ was calculated with an ICC (one-way random effects model). Cronbach's alpha of the SDQ was also calculated.

Dysphagia progression during the disease

To evaluate whether patients in more advanced stages of HD perceived more swallowing abnormalities than those in the earlier stages, a Kruskal-Wallis test was used.

Results

Item generation

In total, 14 items were considered relevant by the experts. These items related to the four different phases of ingestion: preparatory oral phase (3 items), oral phase (2 items), pharyngeal phase (7 items), and esophageal phase (2 items). The questions were all found intelligible by the patients.

Patients

Fifty (out of 55) patients (91%; 22 men; mean ± SD age: 52 ± 11 years; mean ± SD disease duration: 10 ± 5 years) and 50 relatives (29 partners, 15 carers, 6 close friends) returned the questionnaires (Table 1). The five nonresponding patients were in stage II (n=1), and stage III (n=4). Two (out of 55) patients partially completed the questionnaire (≥4 missings). Forty-eight (out of 55) patients (87%; 21 men; mean ± SD age: 54 ± 11 years; mean ± SD disease duration: 10 ± 5 years) and 45 relatives (26 partners, 15 carers, 4 close friends) completed the second set.

Table 1 Baseline sociodemographic, clinical and functional characteristics of the Huntington's disease patients

Patients	
N (Males/Females) (n=50)	50 (22/28)
Age, y	52.2 (11.3)
Age at onset, y	43.1 (9.8)
Disease duration, y	9.6 (5.2)
CAG repeat length (n=46)	43.9 (3.1)
N Stages of HD (n=50)	
Clinical stage I	23
Clinical stage II	17
Clinical stage III	10
N Proxies (n=50):	
Partners	29
Carergivers	15
Close friends	6

Unless otherwise stated, values are means (SD).
Abbreviations: SD, standard deviation; HD, Huntington's disease; CAG, repeat length triplets number (≥36, pathological range)

Item reduction, scale construction, and construct validity

The ICC of one question ("wet voice after swallowing") was very low (ICC=0.189) and therefore removed. Cronbach's alpha for the 13 remaining questions was 0.670, which increased to 0.728 after removal of the items "residue in the mouth, cheek or teeth" and "producing sounds during eating or drinking" (Table 2). Finally, 11 items were included in the HDDS (Table 2). These 11 items related to the various swallowing phases as follows: preparatory oral phase (3 items), oral phase (1 item), pharyngeal phase (5 items), and esophageal phase (2 items), indicating that all phases of ingestion were still covered.

Of all patients, 9.1% had a total score of 11, whereas none of the patients had the maximum possible score, indicating absence of floor and ceiling effects. The test-retest reliability of the

final HDDS as measured with the ICC was 0.754. The correlation with the SDQ was 0.734. The test-retest reliability (ICC) of the SDQ was 0.597.

Table 2 Final Huntington's Disease Dysphagia Scale

	Item	Cronbach's alpha if item deleted
1.	Do you drool during the day?	0.697
2.	Do you feel you have too much food in your mouth, so that you cannot swallow anymore?	0.705
3.	Does food come out of your mouth?	0.709
4.	Does food or beverage come out of your nose?	0.710
5.	Are you always successful in swallowing food or beverage?	0.714
6,.	Do you choke food or beverage?	0.693
7.	Do you cough while swallowing food or beverage?	0.701
8.	Do you sneeze while swallowing food or beverage?	0.718
9.	Do you sometimes unexpectedly breathe while you swallow food or beverage? In other words: Suddenly breathe when you swallow?	0.715
10.	Does food or beverage sometimes come back in your mouth, while you had actually swallowed it?	0.702
11.	Do you sometimes feel if there is a lump in your throat?	0.724

Cronbach's alpha of the final scale = 0.728 (n=48). 'Cronbach's alpha if item deleted' indicates Cronbach's alpha value of the total scale if the particular items were removed. This shows that further removal of any item would result in a decrease of Cronbach's alpha.
Response options;
Question 1-4 and 6-11: 1 No, almost never; 2 Yes, seldom; 3 Yes, sometimes; 4 Yes, frequently; 5 Yes, almost always
Question 5: 1 Yes, almost always; 2 Yes, frequently; 3 Yes, sometimes; 4 Yes, seldom; 5 No, almost never

Dysphagia severity across the disease stages

The Kruskal-Wallis test, used to evaluate whether the severity of the swallowing difficulties measured with the HDDS increased with increasing disease severity according to the three clinical stages, was significant (P = 0.037). Kruskal-Wallis was also significant for the proxy scores (P = .0005).

Discussion

We developed an 11-item scale to measure dysphagia in HD. The instrument can be used to monitor swallowing difficulties from early manifest to end-stage disease. The HDDS demonstrates good construct validity, as shown by the high correlation with the SDQ.

Nevertheless, for evaluating dysphagia in HD, the HDDS may be preferred, not only because it more fully covers the specific domain (i.e., better content validity), but also because of its superior reproducibility in this population. Agreement between the patients' and partners' HDDS scores was low, which indicates that relatives have a quite different perception of the patient's swallowing difficulties than the patients themselves. Because reproducibility and construct validity of the HDDS was assessed in patients and exhibited high values-indicating that one can be confident of the patients' ability to complete this scale- completion by proxies is not recommended. However, when HD patients in the very last stage are confronted with cognitive decline, it is recommended that carers fill out the HDDS. At present the HDDS scale is developed and validated in Dutch, but the scale has been translated into English.

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