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Understanding the Need for Assistance with Survey Completion in People with Huntington Disease

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Abstract

Purpose—In Huntington disease (HD), motor, cognitive and psychiatric changes can have a detrimental impact on health-related quality of life (HRQOL). The purpose of this paper is to describe the extent and type of assistance needed to complete online HRQOL surveys, and the impact of assistance on HRQOL scores.

Methods—A patient-reported outcome measurement system was developed for HD-specific HRQOL. Individuals across the prodromal and diagnosed disease severity spectrum ($n=532$) completed surveys by computer, and reported the amount and type of assistance they received.

Results—Some participants ($n=56$; 10.5%) did not complete all surveys; this group had larger proportions with late stage disease, racial/ethnic minority status, low education and single marital status, and poorer motor, independence and cognitive function compared to those who completed

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Compliance with Ethical Standards

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Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent: Informed consent was obtained from all individual participants included in the study.

all surveys ($n=476$). Overall, 72% of individuals did not receive assistance, 11% received computer assistance only, and 17% received assistance answering the survey questions. The majority of late stage individuals (78%) received some assistance compared to early stage (29%) and prodromal individuals (<1%). Those who received assistance had higher proportions with late stage disease, were older, had less education, and had poorer functional and cognitive skills. Before and after adjustment for sociodemographic and clinical characteristics, those who received assistance had poorer scores on some HRQOL outcomes than those who did not receive assistance.

Conclusions—Computer-based assessments are feasible for many persons with HD, although other methods may also be needed. Clinicians and researchers should develop strategies to assist people with HD to complete HRQOL surveys.

Keywords

health-related quality of life; Huntington disease; patient-reported outcomes; surveys and questionnaires

Health-related quality of life (HRQOL) is critically important for people with Huntington disease (HD) because this devastating disease typically strikes individuals during the prime of their lives [1]. HD is a hereditary neurodegenerative disorder with a prevalence of about 5.70 per 100,000 people in North America, Europe and Australia, and 0.40 per 100,000 people in Asia [2–4]. Each child of a parent who carries the HD expansion mutation gene has a 50% chance of inheriting this mutation, which inevitably leads to HD. Motor, cognitive and psychiatric decline begins more than a decade before diagnosis (prodromal HD) and progressively worsens over the disease course [5]. Although the rate of progression may differ for each person, HD is generally fatal within 15–20 years of clinical diagnosis [6].

The Huntington Disease Health-Related Quality of Life (HDQLIFE) study was designed to address the need for a patient-reported outcome (PRO) measurement system for HD [7; 8]. HDQLIFE utilizes the methodology of the Patient-Reported Outcomes Measurement Information System (PROMIS) [9; 10] and the Quality of Life in Neurological Disorders (Neuro-QOL) measurement system [11; 12]. These state-of-the-science PRO systems use item response theory to develop item banks, including computerized adaptive testing and fixed-length short forms with low response burden [13; 14]. Low burden of assessment is particularly important in HD because motor, psychiatric and cognitive symptoms can impede the ability to respond to long surveys. Even with short forms, some persons with HD may require assistance responding to questions due to disease symptoms. The purpose of this paper is to describe the extent and type of assistance required by HD participants to complete the HDQLIFE PRO surveys, to compare sociodemographic and HD-symptom-related differences between participants according to their assistance needs, and to examine the impact of such assistance on PRO scores. It was hypothesized that participants with greater disease severity would require more assistance than those who were asymptomatic or had less disease severity. It was unknown whether there would be differences in PRO scores between participants who did and did not receive assistance. Results could provide information about the feasibility and validity of computer-based HRQOL testing in the HD population.

Methods

HDQLIFE is a mixed-methods study to develop and validate HD-specific measures of HRQOL, which is described in detail elsewhere [7; 8; 15–17]. Briefly, this involved qualitative methods to determine relevant aspects of HRQOL, several iterations of item development and review, field testing of items, and final development of HRQOL measures with 532 individuals across the HD prodromal and diagnosed HD disease severity spectrum.

Participants and Recruitment

Participants were 18 years of age or older, able to understand English, and had either a positive test for the HD gene mutation and/or a clinical diagnosis of HD. They were recruited in the U.S. using a variety of recruitment strategies, including advertisements in HD-specific newsletters and websites, in-person recruitment at eight HD clinics, and in conjunction with other ongoing research studies (e.g., Predict-HD), HD support groups and HD specialized nursing home units. Participants received a \$40 incentive (check or gift card) for participating in this study. Institutional Review Board approval was obtained at each institution prior to the start of this project, and all participants provided informed consent.

Clinical and Self-Report Assessments

The Unified Huntington's Disease Rating Scale (UHDRS) [18] was completed by an HD specialist certified in the administration of the scale; three measures were used for this study: the Total Functional Capacity (TFC) scale, the Total Motor Score and the Independence Scale. The TFC provides an index of day-to-day functioning across the domains of occupation, finances, domestic chores, activities of daily living and care level [19]. TFC scores range from 0 to 13, with higher scores indicating better functioning; scores can be used to create categories corresponding to HD disease stage [20]. For this study, symptomatic participants with an HD diagnosis were classified as either early stage (TFC score: 7–13, stages I–II) or later-stage HD (TFC score: 0–6, stages III–IV). The Total Motor Score provides a composite measure of oculomotor function, dysarthria, chorea, dystonia, gait and postural stability; scores range from 0 (no motor signs) to 120 (extensive and severe motor signs). The Independence Scale ranges from 0 to 100, with higher scores indicating better functioning and greater levels of independence. Two measures of cognition were administered by certified clinical examiners: the Symbol Digit Modalities Test [21] and the Stroop Color Word Interference test [22]; higher scores indicate better performance.

Participants completed generic and HD-specific measures of HRQOL. The World Health Organization Disability Assessment Schedule 2.0 (WHODAS 2.0) is a 12-item generic measure of functioning and disability; scores range from 0 to 48 and higher scores indicate worse HRQOL [23]. Participants also completed the three newly developed HDQLIFE item pools: 48 chorea items, 64 speech and swallowing difficulties items, and 47 end of life items. Item responses were used in the development and calibration of five HD-specific item banks: Chorea, Speech Difficulties, Swallowing Difficulties, Concern with Death and Dying, and Meaning and Purpose [15–17]. All HDQLIFE measures use a T-score metric (mean=50; standard deviation=10). Higher scores for negative PROs indicate worse functioning

(Chorea, Speech Difficulties, Swallowing Difficulties, Concern with Death and Dying); higher scores for positive PROs indicate better functioning (Meaning and Purpose).

Study measures (clinician-administered and PROs) were usually completed within a two-week time frame. Participants were asked to complete PRO surveys through the PROMIS Assessment Center platform (<http://www.assessmentcenter.net>), either at a designated computer during the research visit, or on a personal or publically available computer with an internet connection. Participants who did not complete the PROs during the visit, or shortly after, were called by study staff with reminders. Staff assisted some participants in completing the measures during this phone call. Participants were instructed that they could ask a caregiver, friend, family member or study staff member to assist them in using the computer, e.g., logging into the online study, reading questions aloud and/or clicking response options. They were told, “We want to make sure that the answers reflect what you feel and believe” and “It is not okay for the caregiver, friend or family member to answer questions for you.” At the end of the online assessment, participants were presented with a final survey asking about the amount and type of assistance they received: (1) Did you have assistance in completing this survey? (No, I completed the survey independently; Yes, a caregiver (family member, friend) helped me complete the survey; Yes, study staff at my site helped me complete the survey (in-person or via phone interview); (2) What type of assistance did you receive? (I needed help using the computer/iPad; My caregiver (family member, friend) helped explain questions to me; My caregiver (family member, friend) helped me answer questions by reminding me of important information; My caregiver (family member, friend) helped by answering questions for me); (3) How often did you receive assistance? (Rarely (0–25% of the time), Sometimes (26–50% of the time), Often, (51–75% of the time), Always (76–100% of the time)). Since these three assistance questions were presented at the end of the online assessment, they were not presented to participants who stopped before completing all of the PRO surveys. The amount of time needed to complete each PRO item was automatically recorded by the assessment platform. Additional study procedures and assessments occurred but were not used for this paper. Specifically, clinical and sociodemographic information was collected before the online PRO assessment, and participants completed several other generic PRO instruments after the HDQLIFE items.

Statistical Analysis

Responses to the assistance questions were used to create three groups: no assistance, computer assistance only, and assistance with survey questions with or without computer assistance. In order to estimate the amount of time needed to complete each HDQLIFE PRO, participant responses to the four or six items in the short forms were used to sum the completion times. Chi-square tests, Fisher’s exact tests and analysis of variance methods were used to compare sociodemographic, clinical and PRO data across HD diagnostic groups and assistance groups, to compare participants who were excluded from the assistance analyses to those who were included, and to compare the amount of missing PRO data across assistance groups. A nominal significance level of 0.05 was used to interpret results. Bonferroni methods were used to adjust for multiple testing. A multivariable linear regression model was created for each PRO, and least-squares means were estimated to

determine the effects of assistance on PROs after adjustment for other covariates (gender, age, race/ethnicity and diagnostic group) [24]. Because missing data were generally very low (1% to 7% for clinician-rated assessments; 5% to 11% for the various PROs) [8] no adjustments were made for missing data.

Results

Of the 532 individuals who were enrolled in the HDQLIFE study, 56 (10.5%) stopped the assessment early and thus were not presented with the assistance questions at the end of the assessment. There were no differences in age or gender between participants who did ($n=476$) and did not ($n=56$) complete the assistance questions. However, the group that stopped the assessment early had larger proportions of people with late stage HD ($p<0.001$), racial/ethnic minority status ($p=0.003$), low education ($p=0.001$), and single marital status ($p<0.001$), poorer function based on the UHDRS measures ($p<0.001$), and poorer cognitive function ($p=0.017$). There were no significant differences between the two groups on most of the HDQLIFE PRO mean scores; however, this comparison could only be done using subsets of individuals who completed some of the HDQLIFE PROs before stopping the assessment (26 to 30 individuals out of 56).

Participants who completed all PROs included 191 individuals with prodromal HD, 186 with early stage HD and 99 with late stage HD (Table 1). These three HD diagnostic groups differed on many sociodemographic, clinical and PRO variables. Relative to the HD groups, the prodromal group was younger, had more education, had better functional and cognitive skills, and had better scores on all PROs except HDQLIFE Concern with Death and Dying and HDQLIFE Meaning and Purpose.

The majority of the late stage HD individuals (78%) received some assistance completing the PRO surveys, compared to 29% of early stage HD individuals and <1% of prodromal HD individuals (Table 2). Late stage HD individuals received more assistance with the computer and more assistance with the survey questions. Over half of those with late stage HD received assistance often or always.

Overall, 344 individuals received no assistance (72%), 53 received assistance with the computer only (11%), and 79 received assistance with the survey questions (17%) (Table 3). The three assistance groups differed on most sociodemographic, clinical and PRO variables. In particular, those who received assistance had higher proportions with late stage HD, were older, had less education, had poorer functional and cognitive skills, and had poorer scores on all PROs except HDQLIFE Concern with Death and Dying and HDQLIFE Meaning and Purpose. The range of missing PRO data was <1% to 4.6% for the no assistance group, 0 to 11.3% for the computer assistance group, and 0 to 8.9% for the survey assistance group; the proportion missing differed only for the WHODAS Disability Score ($p=0.004$).

Results of the multivariable linear regression models for each PRO are shown in Table 4. Least-squares adjusted PRO means are reported for each assistance group. Comparing the adjusted means in Table 4 to the unadjusted means in Table 3 shows that there are smaller differences between the assistance groups after adjustment for gender, age, race/ethnicity

and diagnostic group, but does not alter overall conclusions that those who received assistance had poorer scores on some PROs.

Table 5 provides the average time it took participants to complete the short forms. Participants who did not receive any assistance completing the PRO surveys took about a half-minute to complete a four- or six-item HDQLIFE short form. Those who received some assistance took about twice as long.

Discussion

To our knowledge, this is the first study to measure the extent to which HD research participants use assistance to complete online PRO surveys, and whether PRO scores differ between those who did and did not use assistance. It is important to obtain health-related information directly from patients to evaluate the effectiveness of treatments and interventions [25; 26]. This is especially relevant with regard to HRQOL, which is by nature a subjective domain. Since HD affects motor, behavioral, cognitive and day-to-day function, which impacts HRQOL [27–29], it is critical to assess HRQOL in this population. The purpose of the HDQLIFE PRO measurement system is to provide a low-burden, reliable and valid measure of HRQOL in persons with HD. Unfortunately, the symptoms that are characteristic of HD may also impair the ability of affected individuals to complete PRO surveys. For this reason, the HDQLIFE study allowed participants to have assistance in completing them. This paper examined whether there were important differences in personal characteristics and PROs between those who did and did not complete all of the PRO surveys, and differences between those who did and did not receive assistance.

It is not surprising that participants who received assistance were older, more advanced in disease, and had greater clinician-rated cognitive and functional impairment than those who did not receive assistance. This emphasizes the potential impact of HD on the ability to complete PROs independently. These findings suggest that when using PROs, clinicians and researchers should be aware of the possibility that some participants will need assistance, particularly older and more severely impaired persons. A recent meta-analysis of diverse adult samples reported overall equivalence in PRO scores between self-completion and assisted completion [30]. The meta-analysis did detect some evidence of bias attributable to the setting (home vs. clinic). The HDQLIFE study did not record the setting, so this issue will require further investigation. Participants who received assistance in the HDQLIFE study were also less likely to have post-high school education than persons who did not require assistance. Computerized PRO surveys may be more challenging for people with lower levels of education. Multimedia methods of survey administration have been developed to accommodate people with a range of literacy, language and computer skills in primary care and chronic disease populations [31–36]. These methods might be useful for people with HD.

It is important to acknowledge that persons providing assistance may have influenced participants' responses. Over half of participants with late stage HD indicated they received assistance with the questions, and over half needed assistance often or always. In retrospect, one response choice was somewhat ambiguous: "My caregiver helped by answering

questions for me.” This wording leaves open the possibility that some caregivers answered the questions without input from the participants. Evidence from other studies indicates that companion and participant ratings of HD function can differ, especially as the disease progresses [37]. Anosognosia (lack of awareness of one’s health) is one explanation for this phenomenon in people with HD [38]. Therefore, PRO surveys that are completed with caregiver assistance might be a less valid representation of self-reported HRQOL in later stage HD. Nevertheless, only 17 participants indicated that their caregiver helped by answering questions for them. It would be useful for future studies to explore the type and amount of assistance in more detail.

While the functional PRO scores (WHODAS Disability; HDQLIFE Chorea, Speech Difficulties and Swallowing Difficulties) were worse in the assistance groups than the no assistance group, Concern with Death and Dying and Meaning and Purpose scores did not differ between the groups. There were also no differences in these PRO scores between the three HD disease stages. More data are needed regarding end-of-life concerns and meaning and purpose in persons with HD and how these beliefs change over the disease course. The findings reported here suggest these beliefs remain relatively stable despite independence function and disease stage; however, the current analysis is cross-sectional. Future analyses could include a longitudinal examination of these PROs.

An important component of PRO measurement is to reduce response burden by limiting the time needed to complete the surveys [25; 39; 40]. Receiving assistance completing the HDQLIFE study surveys approximately doubled the amount of time it took to complete the short forms. This could be due to the extra time it requires for those assisting participants to read each item aloud. Also, since those who received assistance were more likely to have diagnosed HD and worse cognitive function, the longer time might be partly due to decreased cognitive processing speed.

Participants in this study might not be representative of most persons with HD. Participants were recruited through other research studies and through large, established HD clinics; thus, these participants may not represent the HD population at large who may often be seen by non-specialists. Prodromal participants might not be representative of most persons who have the HD gene mutation, but are not yet diagnosed, because they knew their HD gene status prior to study participation, while the majority of people at risk for HD do not undergo presymptomatic genetic testing [41].

The HDQLIFE PRO measurement system offers a brief, HD-specific alternative to currently available lengthier and/or generic assessments of HRQOL. Among its advantages, HDQLIFE can be used in the clinical setting, fostering the ability of patients to effectively communicate symptoms of concern to treatment providers. HDQLIFE can also enhance communication when HD specialists are geographically distant from patients because patients can use their home computers to complete the HDQLIFE measures; thus, this approach may be useful for patient care via telemedicine approaches [42]. The results of this study provide useful information about the feasibility of computer-based testing in three HD diagnostic groups. More data are needed to determine the best assessment strategies for individuals with more advanced HD who are likely to require some assistance completing

PRO surveys. Studies evaluating PROs administered by multimedia platforms, paper or interviews could help researchers and clinicians plan the best strategies for assessing HRQOL in people with HD.

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Table 1

Sociodemographic and Clinical Characteristics, and Health-Related Quality of Life Outcomes of Study Participants, by Huntington Disease (HD) Diagnosis^a

	Prodromal (n=191)	Early Stage HD (n=186)	Late Stage HD (n=99)	p-value ^b
Sociodemographics				
Female Gender	122 (63.9%)	101 (54.3%)	55 (55.6%)	0.137
Age in Years	42.6 (12.2)	51.8 (12.4)	55.3 (11.5)	<0.001 ^c
Ethnicity, Race				
Hispanic, any race	3 (1.7%)	7 (3.9%)	1 (1.0%)	0.066
Non-Hispanic, White	173 (96.6%)	170 (94.4%)	90 (92.8%)	
Non-Hispanic, Other	3 (1.7%)	3 (1.7%)	6 (6.2%)	
Highest Education				
Less than HS	0 (0.0%)	4 (2.2%)	2 (2.0%)	0.001 ^c
HS/GED	20 (10.5%)	47 (25.4%)	21 (21.2%)	
More than HS	170 (89.5%)	134 (72.4%)	76 (76.8%)	
Married/Partner	137 (71.1%)	109 (58.6%)	66 (66.7%)	0.032
Clinician-Rated Disease Severity				
Unified Huntington's Disease Rating Scale (UHDRS)				
Independence Scale	97.9 (5.8)	85.2 (9.7)	61.2 (12.2)	<0.001 ^d
Total Motor Dysfunction Score	5.5 (6.1)	30.8 (14.7)	54.4 (21.3)	<0.001 ^d
Cognitive Performance Tests				
Symbol Digit Modalities Test, raw score	52.6 (10.5)	32.5 (11.5)	18.4 (10.7)	<0.001 ^e
Stroop Interference Test, # Correct	46.6 (15.6)	29.1 (10.1)	18.0 (9.2)	<0.001 ^e
Patient-Reported Outcomes				
WHODAS Disability Score	3.8 (5.3)	9.5 (8.7)	19.7 (11.2)	<0.001 ^f
HDQLIFE Chorea	41.6 (5.6)	52.4 (7.6)	57.2 (7.3)	<0.001 ^f
HDQLIFE Speech Difficulties	42.4 (6.8)	50.4 (7.6)	55.0 (7.5)	<0.001 ^f
HDQLIFE Swallowing Difficulties	44.2 (6.5)	51.2 (8.2)	55.8 (7.6)	<0.001 ^f
HDQLIFE Concern with Death and Dying	49.6 (8.8)	50.5 (9.3)	49.7 (10.8)	0.573

	Prodromal (n=191)	Early Stage HD (n=186)	Late Stage HD (n=99)	p-value^b
HDQLIFE Meaning and Purpose	50.5 (9.1)	50.4 (9.7)	49.2 (8.6)	0.477

Entries in the table represent the number of participants (percentage), or the mean (SD).

^aProdromal: positive gene test for HD, not symptomatic; Early Stage HD: Total Functional Capacity (TFC) scale [19] score 7–13; Late Stage HD: TFC scale score 0–6

^bp-value: analysis of variance or chi-square test, excluding missing data

^cmeets criterion for Bonferroni adjustment for 5 comparisons, $p < 0.01$

^dmeets criterion for Bonferroni adjustment for 2 comparisons, $p < 0.025$

^emeets criterion for Bonferroni adjustment for 2 comparisons, $p < 0.025$

^fmeets criterion for Bonferroni adjustment for 6 comparisons, $p < 0.008$

GED: General Educational Development Test (High School Equivalency Diploma); HDQLIFE: Huntington Disease Quality of Life; HS: High School; SD: standard deviation; WHODAS: World Health Organization Disability Assessment Schedule 2.0

Table 2Assistance in Completing PRO Surveys, by Huntington Disease (HD) Diagnosis^a

	Prodromal (<i>n</i> =191)	Early Stage HD (<i>n</i> =186)	Late Stage HD (<i>n</i> =99)	Total (<i>n</i> =476)	<i>p</i> -value ^b
Assistance					
None	190 (99.5%)	132 (71.0%)	22 (22.2%)	344 (72.3%)	<i>p</i> <0.001
Some assistance provided	1 (0.5%)	54 (29.0%)	77 (77.8%)	132 (27.7%)	
Type of Assistance					
None	190 (99.5%)	132 (71.0%)	22 (22.2%)	344 (72.3%)	<i>p</i> <0.001
Assistance with computer only	1 (0.5%)	28 (15.1%)	24 (24.2%)	53 (11.1%)	
Assistance with survey questions ^c	0	26 (14.0%)	53 (53.5%)	79 (16.6%)	
Detailed type of assistance ^d					
Assistance with computer	1	37	51	89	---
Explain questions to me	0	21	41	62	
Remind me of important information	0	9	29	38	
Answer questions for me	0	4	13	17	
How Often Assistance Needed					
Never	190 (99.5%)	132 (71.0%)	22 (22.2%)	344 (72.3%)	<i>p</i> =0.010
Rarely (0–25% of the time)	0	20 (10.8%)	12 (12.1%)	32 (6.7%)	
Sometimes (26–50% of the time)	1 (0.5%)	9 (4.8%)	10 (10.1%)	20 (4.2%)	
Often (51–75% of the time)	0	9 (4.8%)	12 (12.1%)	21 (4.4%)	
Always (76–100% of the time)	0	16 (8.6%)	43 (43.4%)	59 (12.4%)	

Entries in the table represent the number of participants (percentage).

^aProdromal: positive gene test for HD; Early Stage: Total Functional Capacity (TFC) scale [19] score 7–13; Late Stage: TFC scale score 0–6^b*p*-value: chi-square or Fisher's exact test for comparison of the diagnostic groups^cSome participants who needed assistance with the survey questions also needed computer assistance (*n*=36)^dParticipant could indicate more than one type of assistance

Table 3

Sociodemographic and Clinical Characteristics, and Health-Related Quality of Life Outcomes, by Type of Assistance Needed

	No Assistance (n=344)	Assistance with the Computer Only (n=53)	Assistance with the Survey Questions ^a (n=79)	p-value ^b
Huntington Disease (HD) Diagnosis^c				
Prodromal	190 (55.2%)	1 (1.9%)	0 (0.0%)	<i>p</i> <0.001 ^d
Early Stage HD	132 (38.4%)	28 (52.8%)	26 (32.9%)	
Late Stage HD	22 (6.4%)	24 (45.3%)	53 (67.1%)	
Sociodemographics				
Female Gender	210 (61.0%)	26 (49.1%)	42 (53.2%)	0.150
Age in Years	46.7 (12.5)	57.1 (11.3)	52.7 (14.3)	<0.001 ^d
Ethnicity, Race				
Hispanic, any race	8 (2.4%)	2 (3.8%)	1 (1.3%)	0.079
Non-Hispanic, White	315 (99.0%)	48 (92.3%)	70 (92.1%)	
Non-Hispanic, Other	5 (1.6%)	2 (3.8%)	5 (6.6%)	
Highest Education				
Less than HS	1 (0.3%)	2 (3.8%)	3 (3.8%)	0.002 ^d
HS/GED	54 (15.8%)	14 (26.4%)	20 (25.3%)	
More than HS	287 (83.9%)	37 (69.8%)	56 (70.9%)	
Married/Partner	238 (69.4%)	27 (50.9%)	47 (59.5%)	0.014
Clinician-Rated Disease Severity				
Unified Huntington's Disease Rating Scale (UHDRS)				
Independence Scale	91.8 (11.1)	70.8 (16.2)	66.6 (14.6)	<0.001 ^e
Total Motor Dysfunction Score	16.1 (15.9)	50.0 (22.6)	50.2 (19.9)	<0.001 ^e
Cognitive Performance Tests				
Symbol Digit Modalities Test, raw score	44.0 (14.6)	22.0 (11.3)	20.8 (11.5)	<0.001 ^f
Stroop Interference Test, # Correct	39.1 (15.8)	21.2 (8.0)	19.2 (10.2)	<0.001 ^f
Patient-Reported Outcomes				
WHODAS Disability Score	6.3 (7.5)	14.2 (11.3)	19.0 (11.3)	<0.001 ^g

	No Assistance (<i>n</i> =344)	Assistance with the Computer Only (<i>n</i> =53)	Assistance with the Survey Questions ^a (<i>n</i> =79)	<i>p</i> -value ^b
HDQLIFE Chorea	46.2 (8.4)	54.9 (7.2)	57.2 (7.1)	<0.001 ^g
HDQLIFE Speech Difficulties	47.5 (8.2)	53.4 (8.2)	54.8 (8.0)	<0.001 ^g
HDQLIFE Swallowing Difficulties	46.0 (8.1)	51.7 (8.0)	54.8 (8.3)	<0.001 ^g
HDQLIFE Concern with Death and Dying	50.3 (9.0)	49.0 (9.9)	49.2 (10.9)	0.482
HDQLIFE Meaning and Purpose	50.3 (9.3)	50.4 (8.7)	49.8 (9.5)	0.905

Entries in the table represent the number of participants (percentage), or the mean (SD).

^aSome participants who needed assistance with the survey questions also needed computer assistance (*n*=36)

^b*p*-value: analysis of variance, chi-square test or Fisher's exact test for comparison of the assistance groups, excluding missing data

^cProdromal: positive gene test for HD; Early Stage: Total Functional Capacity (TFC) scale [19] score 7–13; Late Stage: TFC scale score 0–6

^dmeets criterion for Bonferroni adjustment for 6 comparisons, *p*<0.008

^emeets criterion for Bonferroni adjustment for 2 comparisons, *p*<0.025

^fmeets criterion for Bonferroni adjustment for 2 comparisons, *p*<0.025

^gmeets criterion for Bonferroni adjustment for 6 comparisons, *p*<0.008

GED: General Educational Development Test (High School Equivalency Diploma); HDQLIFE: Huntington Disease Quality of Life; HS: High School; SD: standard deviation; WHODAS: World Health Organization Disability Assessment Schedule 2.0

Table 4

Adjusted Health-Related Quality of Life Outcomes, by Type of Assistance Needed

	No Assistance (<i>n</i> =344)	Assistance with the Computer Only (<i>n</i> =53)	Assistance with the Survey Questions ^{<i>a</i>} (<i>n</i> =79)	<i>p</i> -value ^{<i>b</i>}
WHODAS Disability Score	10.0 (1.2) ^{<i>d,e</i>}	13.5 (1.5) ^{<i>d</i>}	16.0 (1.4) ^{<i>e</i>}	<0.001 ^{<i>c</i>}
HDQLIFE Chorea	51.5 (1.1)	54.3 (1.4)	55.4 (1.3)	0.001 ^{<i>c</i>}
HDQLIFE Speech Difficulties	50.9 (1.1)	51.4 (1.4)	53.9 (1.3)	0.039
HDQLIFE Swallowing Difficulties	51.7 (1.1)	52.7 (1.4)	53.4 (1.3)	0.363
HDQLIFE Concern with Death and Dying	51.1 (1.5)	49.5 (1.9)	49.1 (1.8)	0.370
HDQLIFE Meaning and Purpose	49.5 (1.4)	50.0 (1.8)	50.5 (1.7)	0.805

Entries in the table represent the least-squares means (standard error), adjusted for gender, age, race/ethnicity and diagnostic group.

^{*a*} Some participants who needed assistance with the survey questions also needed computer assistance (*n*=36)

^{*b*} *p*-value: multivariable linear regression model, Type III tests

^{*c*} meets criterion for Bonferroni adjustment for 6 comparisons, *p*<0.008

^{*d*} pairwise comparison, *p*<0.017

^{*e*} pairwise comparison, *p*<0.017

Table 5

Estimated Time to Complete HDQLIFE PRO Short Forms, by Type of Assistance Needed

	No Assistance (<i>n</i> =344)	Assistance with the Computer Only (<i>n</i> =53)	Assistance with the Survey Questions ^{<i>a</i>} (<i>n</i> =79)	<i>p</i> -value ^{<i>b</i>}
6-item short form				
HDQLIFE Chorea	32.8 (25.6)	61.9 (26.6)	64.0 (32.2)	<0.001 ^{<i>c</i>}
HDQLIFE Speech Difficulties	32.1 (23.7)	62.2 (28.4)	63.3 (32.2)	<0.001 ^{<i>c</i>}
HDQLIFE Swallowing Difficulties	32.5 (25.2)	72.2 (38.8)	65.0 (31.5)	<0.001 ^{<i>c</i>}
HDQLIFE Concern with Death and Dying	38.4 (22.3)	56.1 (22.2)	60.1 (33.2)	<0.001 ^{<i>c</i>}
4-item short form				
HDQLIFE Meaning and Purpose	30.0 (18.0)	49.2 (23.0)	46.4 (23.8)	<0.001 ^{<i>c</i>}

Entries in the table represent the mean (SD) seconds to complete the short form.

^{*a*}Some participants who needed assistance with the survey questions also needed computer assistance (*n*=36)

^{*b*}*p*-value: analysis of variance, excluding missing data

^{*c*}meets criterion for Bonferroni adjustment for 5 comparisons, *p*<0.01