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Patient and Caregiver Quality of Life in Huntington's Disease

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Abstract

Little is known about subjective perceptions of quality of life (QOL) in Huntington's disease (HD). The current study determined correlates of patient and caregiver QOL and assessed change over time. Participants were 22 patient-caregiver dyads, who rated QOL at baseline and six months later. Overall, patient functional and cognitive impairment were significantly correlated with patient and caregiver QOL. Neuropsychiatric symptoms had differential impact on patient and caregiver QOL. Furthermore, when patients recalled their QOL about a previous time, their recall may have been negatively biased. Treatment implications of results are discussed. Future work is needed because subjective QOL is an important outcome measure in therapeutic trials.

Keywords

Huntington's disease; quality of life; caregiver; neuropsychiatric symptoms; self-report

Huntington's disease (HD) is a genetic neurodegenerative disorder that has pervasive effects on patient functioning. Impaired motor functions are cardinal features of the disease and include abnormal eye movements and involuntary choreiform movements early in the disease and rigidity and bradykinesia later in the disease [1]. Cognitive impairment is common and is characterized by psychomotor slowing, visuospatial impairment, and memory decline [2]. Neuropsychiatric symptoms also are pervasive in HD, with depression, agitation, irritability and apathy being the most common symptoms [3, 4].

Quality of Life in Huntington's Disease

Preliminary research also suggests that HD has detrimental effects on patient quality of life (QOL). That is, using generic health-related QOL (HR-QOL) measures, mild to moderately impaired HD patients report QOL lower than population norms [5, 6]. Before exploring issues related to QOL and HD, however, it is important to clarify the different measures and definitions of QOL that abound in the literature.

One of the most important distinctions is between HR-QOL and QOL. HR-QOL measures tend to assess the *functional impact* of symptoms and are useful for cross-disease or cross-age comparisons because the same items are used to assess QOL regardless of the disorders or age groups being studied [7]. For example, the generic Sickness Impact Profile (SIP), which has been used in two previous HD studies [5, 6], assesses psychosocial and physical health behaviors as they pertain to 12 categories of function [8]. HR-QOL measures also

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have been useful in determining the economic impact of disease because of their emphasis on functionality [9].

In contrast, QOL measures are less concerned with functional impact of symptoms and take a broader perspective on QOL, recognizing that a wide variety of life domains and contextual factors can impact QOL, in addition to the functional impact of disease symptoms. This subjective conceptualization of QOL is consistent with the World Health Organization (WHO) definition of QOL, which states that QOL is an "individuals' perception of their position in life in the context of the culture and value systems in which they live and in related to their goals, expectations, standards and concerns," [8, p. 1405]. Thus, for assessment of QOL, it is a person's subjective experience of their life that takes precedence over a more circumscribed focus on functional impact. QOL measures vary widely in breadth and depth and also in format [10]. For example, some QOL measures consist of single-items ratings [11], whereas others are full scales [12, 13].

To date, studies of QOL in HD have been limited to HR-QOL measures. As previously stated, HR-QOL measures have a number of benefits but they also offer a limited functional perspective on QOL. To obtain a more global and subjective assessment of QOL, one that is consistent with the WHO QOL definition, the current study use a single item measure to assess how persons affected by HD rated their QOL. This type of global measure has the benefit that is well-suited to determine which symptoms of HD correlate most strongly with QOL, whereas analyses of associations between symptoms and QOL is hampered by HR-QOL measures, which often confound the measurement of QOL with disease symptoms in a single instrument.

The single item measure used in this study has several other advantages for measuring QOL in HD. Single item measures are accessible to a wider variety of patients than are more lengthy scales and single item measures are quick and portable, allowing for greater clinical and research utility, and they have been used with patients and caregivers [11, 14]. Single item measures also are easily adapted to assess QOL from different vantage points. For example, they can be used to assess patient self-report QOL and also to ask patients to rate their QOL from their caregivers perspective, or vice versa [11, 14]. Finally, patient and caregiver single item measures of QOL demonstrate construct validity similar to full-scale measures of QOL [15] and they correlate strongly with a full-scale QOL measure in AD [11]. The current study will be the first to determine the feasibility and utility of a global item QOL measure for patients and caregivers affected by HD.

Significance of QOL Research

QOL research made significant strides in the past decade, particularly because of greater emphases on holistic, integrated, and comprehensive patient care [16]. Prior to QOL research, treatment outcome studies often focused narrowly on improvement of cardinal symptoms of a disease, such as cognition in Alzheimer's disease (AD) [17] or motor symptoms in Parkinson's disease (PD) [18]. However, over time, it became evident that symptom improvement, or even extended survival, does not necessarily lead to better life quality [10, 19]. Currently, there are increasing pressures from regulatory agencies to include patient-reported outcomes in therapeutic trials and especially to assess QOL [20]. QOL is used as an outcome measure in clinical trials for several neurodegenerative diseases, including PD [9, 21], AD [22], and Amyotropic lateral sclerosis (ALS) [23].

Predictors of Quality of Life in Huntington's Disease

Previous research found that depression was a strong predictor of HR-QOL in HD and that motor impairment, independent functioning, and cognitive impairment had lesser yet

significant associations with functional life quality [5, 6]. This is consistent with findings that neuropsychiatric symptoms tend to be the most robust predictors of QOL in many neurodegenerative diseases [15, 24–27].

There is reason to suspect that in comparison to other neurodegenerative disorders, neuropsychiatric symptoms might be particularly detrimental to QOL in HD. That is, neuropsychiatric symptoms are common in HD [3, 4, 28] and psychopathology includes higher rates of depression, suicide, and Obsessive Compulsive Disorder, as compared to population base rates [28, 29]. Personality change is common in HD [30] and psychopathology tends to be more severe in HD than some other neurodegenerative diseases [31]. Furthermore, patients with HD deal with unique psychological issues due to the autosomal dominant inheritance pattern of the disease. Patients often know they are at risk or may be at risk for HD long before symptom onset, which may lead to anticipatory anxiety, stressful life planning, hopelessness, and uncertainty about the future [32]. Thus, in addition to the symptoms of the disease, patients may be dealing with a host of other unique psychosocial stressors. These factors emphasize the utility of a broad measure of QOL for HD, in contrast to HR-QOL measures that emphasize functional impact of HD.

Quality of Life in Caregivers for Huntington's Disease

Complicated issues stemming from the inherited nature of HD also affect caregivers [33]. Thus, in addition to the well known burden, stress, and morbidity impacting caregivers for persons with neurodegenerative diseases [24, 34], caregivers for patients with HD face particularly challenging issues, such as hopelessness associated with impending disease [32] and stress that HD might develop in other family members [33]. Caregivers also have to contend with neuropsychiatric symptoms that can be of a particularly antisocial orientation (e.g., irritability, aggression) [30, 33]. Thus, a second goal of the current study is to gather data about caregiver QOL and to contribute the literature on caregiver QOL [33, 35, 36] by determining associations between caregiver QOL and patient symptoms.

The Current Study

The current study seeks to determine associations between caregiver QOL, patient QOL, and patient symptoms of HD. Patient QOL ratings came from self- and caregiver-report. Patient self-report is generally regarded as the "gold standard" QOL measure [10, 19] but caregiver perspectives on patient QOL are important because caregivers assist with healthcare decision-making and provide important information about patients for clinical care and in research studies. Caregivers also are relied upon as proxies when patients lose the ability to communicate about their subjective states.

Thus, analyses determined associations between patient and caregiver QOL and symptoms of HD. Based on previous research [3, 11, 24–27, 37], we predicted that neuropsychiatric symptoms would be the strongest correlate of QOL. In addition, we explored actual and perceived changes in QOL over time from several perspectives.

Method

Participants

Participants ($N = 22$) were patients diagnosed with HD (Table 1). The majority of the sample was male (72.7%) and Caucasian (96%). Caregivers also participated ($N = 22$; Table 1) and the majority were female (86.4%) and Caucasian (90%). Caregivers were spouses (59.1%) or had parent-child (22.7%) or other relationships (18.2%). Caregivers had known patients for an average of 29.9 years ($SD = 13.8$) and the majority lived with the patient

(72.7%). Follow-up assessments were conducted 6.5 months (range 5 - 10) after the first assessment. Four caregivers and four patients declined participation or were lost (i.e., patient moved without forwarding address) to follow-up (drop-out rates of 18%). Caregivers and patients who dropped did not significantly differ from participants who stayed in the study for age, length of relationship, patient QOL self- or caregiver-rating, or any of the UHDRS subscales. However, in a *t*-test with equal variances not assumed, caregivers who dropped had higher self-report QOL at time 1 ($M = 4.0$, $SD = 0.0$) than caregivers who stayed in the study ($M = 3.2$, 1.0; $t = -3.3$, $df = 17$, $p < .05$). Patients who dropped from the study had significantly lower education ($M = 12.0$, $SD = 1.0$) than persons who did not drop ($M = 14.4$, $SD = 1.8$; $t = 2.3$ $df = 18$, $p < .05$).

Measures

QOL rating scales—Both patients and caregivers were asked to rate their overall current QOL (i.e., "Overall, how would you rate your quality of life?") on a 5-point response scale: 1 = bad, 2 = fair, 3 = good, 4 = very good, 5 = excellent. QOL ratings were taken twice, once during the initial assessment (T1 QOL) and again at a six month follow-up (T2 QOL). At times 1 and 2, the item was administered verbally by a trained research assistant; time 1 assessment was in-person and time 2 assessment was over the phone. The question and response options were repeated as often as necessary. Caregivers also were asked to provide a rating for the patient's QOL at both times. Finally, during the follow-up assessment, all participants were asked to make a retrospective rating of QOL (Retro QOL). Specifically, they were asked to think back to the initial assessment and retrospectively re-rate QOL at that time using the same scale.

Unified Huntington's Disease Rating Scale (UHDRS)—The UHDRS is a clinical research tool that assesses four symptom domains in HD: motor, cognitive, neuropsychiatric, and functional [38]. The UHDRS Motor score is the sum of 31 motor items, with higher scores indicating more severe impairment [39]. The UHDRS Functional score [39] assesses a patient's ability to perform basic and instrumental activities of daily living, which is derived from reports of the patient and his/her companion, with higher scores indicating better functioning. The UHDRS Neuropsychiatric score [40] is the sum of the product of frequency and severity for 11 neuropsychiatric symptoms (e.g., anxiety, depression), with higher scores indicating increased psychiatric symptoms. The UHDRS Cognitive score is the sum of the age and education corrected T-scores ($M = 50$, $SD = 10$) for three cognitive tests (verbal fluency, symbol digit modalities, and Stroop interference) that assesses executive functioning; higher scores indicate better cognitive abilities.

Procedure

Participants were recruited from the Huntington's Disease Clinic at the University of Iowa Hospitals and Clinics. During regularly scheduled clinic appointments, patients and caregivers were invited to participate in a study about QOL and HD. If interested, all patients and caregivers provided written informed consent. The initial assessment consisted of the UHDRS and the QOL rating scales. The follow-up assessment was conducted 6 months later via phone and included only the QOL ratings. On both occasions, patients and caregivers provided QOL ratings independent from one another.

Results

Preliminary Analyses

Descriptive statistics—Descriptive statistics for the UHDRS indicate that patients are in the mild to moderate stages of HD (Table 1). QOL descriptive statistics (Table 2) are consistent with a large body of research [e.g., [15, 41]] and t-tests indicated that caregivers

rated patient QOL significantly ($t(21) = -3.22$, $p < .05$) lower than patients at T1; the difference at T2 indicated a trend ($t(15) = -2.00$, $p < .10$).

Patient-caregiver agreement—Agreement between patient self-rated QOL and caregiver ratings about patients were low to moderate but not significant. At T1, there was a trend for the association between self- and caregiver-rating (Pearson $r = .39$, $p < .10$) but agreement for the T2 and Retro ratings were lower (rs = .12 and .14, respectively, ps > .60). These low to moderate agreement ratings are consistent with previous studies [15, 37] and suggest that patients and caregivers have unique, yet each potentially valid, perspectives on patient QOL.

Associations between QOL and Symptoms of HD

Pearson correlations between QOL ratings and UHDRS symptom categories were calculated (Table 3). Overall, functional capacity and cognitive scores had the strongest associations with patient and caregiver self-report QOL. Contrary to expectations, no significant associations were found between neuropsychiatric symptoms and QOL. In part, this may have been due to the small sample size because some correlations between neuropsychiatric symptoms and QOL were moderately strong (e.g., rs = −.40). However, to follow-up on our null results, post hoc analyses were run to determine if specific neuropsychiatric symptoms were associated with QOL. Results indicated that patient self-report QOL at T1 was negatively associated with frequency of irritability ($r = -.43$, $p < .05$). Patient self-report QOL at T2 was negatively associated with apathy frequency and severity (rs = −.65 and −.55, respectively, $p < .05$).

Caregiver Retro ratings about the patients' QOL were positively associated with the severity of obsessional thinking ($r = .53$, $p < .05$). Furthermore, T2 caregiver-reported patient QOL was significantly ($p < .05$) and negatively correlated with severity of depressed mood ($r = -.48$), severity of delusions ($r = -.53$), and frequency and severity of hallucinations ($rs = -.51$).

Caregiver self-reported Retro rating was negatively associated with the frequency of patient suicidal thoughts ($r = -.58$, $p < .05$) and frequency of distruptive/aggressive behavior ($r = -.58$, $p < .05$). T2 caregiver self-report QOL also was negatively associated with the frequency of distruptive/aggressive behavior ($r = -.50$, $p < .05$).

Changes and Response Shift in QOL over Time

To determine actual QOL change over time, we conducted t-tests to determine differences in T1 and T2 QOL ratings. For patients self-ratings, there was not a significant difference between their T1 and T2 QOL self-ratings ($t(17) = 1.38$, $p > .15$). There also were no significant differences for caregiver QOL self-ratings ($t(17) = 0.29$, $p > .70$) or patient-ratings ($t(17) = 1.23$, $p > .20$).

To determine if respondents perceived change over time, we tested for differences between the T2 current QOL ratings and the Retro QOL ratings. That is, we were testing to see if current QOL was rated differently than retrospective QOL. Both of these ratings were made at T2, so we presume that the same general standard of evaluation was used. For patients, there was not a significant difference between their T2 QOL rating and their Retro QOL rating ($t(16) = 1.17$, $p > .25$). There also were no significant differences for caregiver QOL self-ratings ($t(17) = -1.14$, $p > .25$) or patient-ratings ($t(17) = -0.90$, $p > .35$).

A third aim was to determine if there was evidence of a 'response shift' over time in how QOL was rated. That is, we wanted to determine if there was evidence that Retro T1 ratings had somehow shifted from the original T1 QOL current ratings.

For patients, there was evidence for a response shift. That is, patient T1 QOL ratings were significantly greater than their Retro QOL ratings of T1 ($t(16) = 2.68$, $p < .05$), suggesting that patients recalled their QOL as being worse than was actually the case. In contrast, the difference between caregiver QOL self- and patient-ratings for T1 and Retro T1 were not significant ($t(17) = -0.52$, $p > .60$; ($t(17) = 0.49$, $p > .60$), respectively).

There were no significant correlations between the UHDRS subscales and different indices of change in QOL over time. That is, regression analyses were used to create standardized difference scores (i.e., standardized residuals) for the three change scores (i.e., actual change, perceived change, and response shift). However, there were no significant associations between these change scores and the UHDRS subscales.

Discussion

HD patient functional and cognitive capacities appear to have the most detrimental effect on subjective QOL for patients and caregivers. Results are consistent with previous research in PD, indicating that functional disability and cognitive impairment were associated with lower patient QOL [24, 42, 43]. An important and novel contribution of this initial study of subjective QOL in HD, however, is demonstrating that similar factors influence *both* patient and caregiver QOL. These results have clear implications for treatment. Interventions that target functional and cognitive capacities of patients have the most promise to simultaneously improve QOL in patients and caregivers.

A surprising finding was that an overall index of neuropsychiatric symptoms was not significantly associated with patient or caregiver QOL. Previous research has shown that neuropsychiatric symptoms are the most robust predictor of QOL in many neurodegenerative diseases [15, 24–27], including HD [5, 6]. Furthermore, there were not particularly low rates of neuropsychiatric symptoms in our sample. For example, depressive symptoms were present in 50% of patients and only 5 persons (23% of sample) reported no neuropsychiatric symptoms, which is fairly typical for HD [3, 29, 30].

Since no significant results were found for the neuropsychiatric index, in post hoc correlational analyses, we determined if specific neuropsychiatric symptoms were associated with HD. Our preliminary results suggest that neuropsychiatric symptoms may differentially affect QOL for patients and caregivers. Patients may experience the loss of positive affect and engagement as detrimental to their QOL, whereas caregivers may suffer the most when patient behaviors are difficult to manage. These findings are preliminary but if they are replicated in future research, there are several clinical implications. First, patients and caregivers may differentially suffer the impact of neuropsychiatric symptoms. On the one hand, patients who are apathetic may be easy to manage for caregivers but patients themselves may experience lack of engagement as more detrimental to their life quality. Thus, behavioral or pharmacologic intervention to improve patient apathy may be particularly effective to improve their QOL. On the other hand, patient behaviors that are difficult to manage (e.g., psychosis, obsessions, aggressive and disruptive behavior), not surprisingly, may be particularly detrimental to caregiver QOL because caregivers may personally suffer as a result of these behaviors, which are difficult to manage and can be time-intensive. Thus, our results indicate that overall indices of neuropsychiatric symptoms will be less clinically informative for QOL in persons HD and their caregivers; clinicians

may need to differentially focus on and treat specific symptoms to improve QOL for both parties.

This was the first study to gather patient and caregiver ratings of global QOL. Agreement was low to moderate, which is consistent with research in AD [10] and suggests that patients and caregivers have unique perspectives on patient QOL. A logical question to follow is, "Who is more accurate, patients or caregivers?" Whereas future validity studies will be useful in answering this question more definitely, our data suggest that both perspectives likely have some validity, due to convergent associations with HD symptoms for patient and caregiver reported QOL. Differences likely are due to the fact that patients and caregivers have different experiences and priorities when it comes to HD [36]. For example, as discussed above, we found differential associations between patient and caregiver-reported QOL and neuropsychiatric symptoms. Thus, in clinical practice, it would be useful to gather perspectives of QOL from each party and not to assume that caregivers are able to provide substituted judgments for patients, or vice versa. Each person is uniquely affected by HD and it is likely that their perceptions of QOL can not be readily divorced from these different experiences.

Change in QOL over Time

A second goal of this study was to determine change over time in QOL. Little change in QOL was found over six months, probably because this time frame is too short to detect change in a slowly progressive disease like HD. For example, it takes several years for significant changes in cognitive abilities to become evident in HD [44–46]. Although not the goal of the current study, the general stability of QOL over time might speak the general reliability of our brief QOL measure. Thus, test-retest studies of our measure, as well as longer studies that are better equipped to detect change over time, are warranted.

An intriguing finding in the current study was that patients may look back on their QOL at a previous time point as being worse than was actually the case. We found a significant response shift of this nature in our patient sample. Perhaps viewing the past as worse helps one to cope with a chronic illness for example, by allowing one to view their current situation as better or as improving, compared to prior circumstances.

Response shift in patient QOL ratings has implications for the design of future clinical trials in HD. That is, classic pre-test/post-test designs may mask treatment gains if patients' standards for rating QOL are shifting over time [47]. Thus, study designs that can assess and accommodate for changing standards of evaluation hold the most promise for detecting true treatment gains or losses in QOL (see [48] for a review of relevant methodologies).

A potential response shift in patient QOL reports also has clinical implications. For example, in clinical evaluations patients often are asked if they are doing better, worse, or the same. These types of questions involve implicit or explicit comparisons to past states, similar to the type of retrospective QOL judgment that was part of this study. It is important to stress that patients are rarely asked to veridically *recall* past states and they were not required to do so in the current study. Rather they are asked to look back on the past from their current vantage point and provide a new judgment about their situation or life quality at that time. The fact that this judgment may be biased so that the past situation is judged worse than it was, as stated above, may simply be a positive coping response and clinicians should be aware that this could be occurring. Alternately, it could reflect a distortion that is secondary to cognitive impairment in HD. Future research will be very useful to further address these issues.

Limitations

This study was an initial project to investigate subjective QOL in HD. However, the sample was small and 18% were lost to follow-up. Furthermore, caregivers who dropped the study tended to have higher QOL than persons who stayed and it is unclear what effect this issue may have had on results. In addition, patients who dropped from the study had lower education than those who remained.

Additionally, although our single item measure has many benefits, including ease, economy, repeatability, adaptability, and accessibility to a wide range of patients and disorders [11], it also has some limitations and a broader disease-specific measure of QOL for HD might illuminate more specifically how different symptoms of HD differentially impact QOL. Thus, results of this project are regarded not as the final word about subjective QOL in HD but as the first step. We hope our results provide inspiration and hypotheses for future studies of QOL in HD and we regard our initial data as the foundation upon which to base more comprehensive studies.

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Table 1

Descriptive Statistics for Patient and Caregiver Characteristics

	M	*SD*	Range
Patient			
Age	47.3	15.2	19 – 79
Education	14.1	1.9	11 – 18
UHDRS Motor	38.3	14.3	10 – 56
UHDRS Functional	8.4	2.9	3 – 13
UHDRS Psychiatric	20.9	21.3	0 – 61
UHDRS Cognitive	−6.3	2.9	<14 – 42
Caregiver			
Age	50.2	12.6	31 – 77
Education	14.3	2.4	12 – 20

Table 2

Descriptive Statistics for the QOL Ratings

QOL Ratings	*M*	*SD*
Patient QOL Self-ratings		
T1	3.41	0.96
T2	3.11	1.13
Retro	2.88	1.05
Caregiver QOL Self-ratings		
T1	3.36	0.95
T2	3.17	0.86
Retro	3.33	0.97
Caregiver QOL Patient-ratings		
T1	2.73	0.83
T2	2.44	0.70
Retro	2.61	0.70

Table 3

Correlations between QOL Ratings and UHDRS Scores

	UHDRS Subscales			
QOL Ratings	**Functional**	**Cognitive**	**Motor**	**Neuropsychiatric**
Patient QOL Self-ratings				
T1	.54*	.57*	−.20	−.33
T2	.15	.20	.18	−.40
Retro	.46	.36	.13	−.40
Caregiver QOL Self-ratings				
T1	.26	.49*	−.14	−.09
T2	.47*	.41	.03	−.17
Retro	.55*	.60*	−.13	−.20
Caregiver QOL Patient-ratings				
T1	.36	.40	−.40	−.05
T2	.08	−.18	.10	−.25
Retro	.15	.03	.28	−.01

* $p < .05$

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