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Health-Related Quality of Life in Huntington's Disease: A Comparison of Two Generic Instruments, SF-36 and SIP

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Abstract: **Whereas several clinical endpoints in monitoring the response to treatment in patients with Huntington's disease (HD) have been explored, there has been a paucity of research in the quality of life in such patients. The aim of this study was to validate the use of two generic health-related quality of life instruments (the Short Form 36 health survey questionnaire [SF-36] and the Sickness Impact Profile [SIP]) and to evaluate their psychometric properties. We found that both instruments demonstrated acceptable convergent validity and reliability for patients and carers. However, there was an advantage in using the SF-36 because of its more robust construct validity and test–retest reliability; furthermore, motor symptoms appeared to influence some strictly nonmotor dimensions of the SIP. On a pragmatic level, the SF-36 is shorter and quicker to administer and, therefore, easier for patients at various stages of the disease to complete. Thus, the SF-36 would appear to be the recommended instrument of choice for patients with HD and their carers, although further work needs to be done to investigate the sensitivity of this instrument longitudinally.**

Key words: Huntington's disease; quality of life; SF-36; SIP; carers

There is increasing emphasis on incorporating subjective patient-determined endpoints in the monitoring and treat-

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ment of chronic disease. In fact, more studies are now adopting health-related quality of life (HRQoL) as a main outcome measure because traditional clinical measures are often highly specific and may not always translate into perceptible changes on patients' view of their overall quality of life. For example, the Parkinson's Disease Questionnaire (PDQ-39), which is designed to examine HRQoL specifically in Parkinsonian patients, has been used in the evaluation of therapies for this population. However, no such disease-specific HRQoL assessment exists in Huntington's disease (HD), and in the light of emerging experimental intervention options for this disease, it will be of great importance to monitor patients' HRQoL over time. To accomplish this, it is essential to use instruments that are suited for this population. Surprisingly, the impact of HD on quality of life has largely been characterized by anecdotal evidence and clinical observation, and even less is known about which generic instrument is best suited for these patients and their carers. To our knowledge, there have only been two published studies examining quality of life in this patient group, one using the Sickness Impact Profile (SIP)[1] and the other using the Short Form 36 health survey questionnaire (SF-36),[2] but no comparison and assessment of the psychometric properties of these questionnaires has been performed to date.

In this study, we investigated HRQoL in a population of patients with HD using these two well-known and established generic HRQoL instruments, i.e., the SF-36 and SIP. The purpose of the study was to compare and evaluate the psychometric properties of these questionnaires to determine their suitability for HD patients at various stages of their disease and their carers.

PATIENTS AND METHODS

Participants and Procedure

A total of 214 patients with a genetic diagnosis of HD and their respective carers who had attended the regional HD clinic in the past 10 years were contacted by letter to participate in this study. A total of 79 patients responded yielding a response rate of 37%, which was within the expected range given previous questionnaire studies in HD patients.[3] A one-way analysis of variance on the data that was available for nonresponders showed that this group was not significantly different from responders on Unified Huntington's Disease Rating Scale (UHDRS) motor scores ($F(1,193) = 2.80$; $P = 0.096$) or clinician-rated UHDRS Independence scores ($F(1,167) = 0.147$; $P = 0.702$). However, it appeared that, although the nonresponders were younger ($F(1,208) = 7.36$; $P = 0.007$) and had had HD for a longer period of time ($F(1,111) = 6.22$; $P = 0.014$). Therefore, this younger but more affected group may have been more disinclined to volunteer given the time commitment required by this study, and/or a significant worsening of their condition since last attending the clinic, which would have made their data unreliable. Fourteen patients who were not showing clinical symptoms at their most recent neurological examination were excluded from the study ($n = 14$). Where patients were limited by the motor or visual aspects of form-filling, carers could administer the questionnaires orally to enable patients to provide their own opinions on the issues raised in the questionnaires. It was emphasized that patients and controls were to express their *own* opinions in their *respective* questionnaires, and that carers were not to fill in their assessments of what the patients were experiencing.

Participants were sent the two HRQoL questionnaires (SF-36, SIP) at two time points, with the Beck Depression Inventory (BDI) accompanying the initial send-out. The average time interval between test and retest was 6 weeks (± 0.84). Because HD patients were unlikely to change over such a short period of time, a 6-week period was appropriate to estimate test–retest reliability to minimize carryover and recall effects. At an average of 2 weeks (± 0.53) after receipt of the Time 1 questionnaires, a follow-up telephone call was made and the Telephone Interview of Cognitive Status (TICS)[4] was administered. Data from a total of 65 patients and 56 carers were analyzed for Time 1, and subsequently 44 patients and 38 carers participated in the retest study at Time 2.

Demographic and Clinical Characteristics

All participants reported their age; patients also self-reported the duration of disease (from first manifestation of symptoms) and self-rated UHDRS Independence score. The maximum score of 100 indicates total independence and 0 total dependency, being tube fed with total bed care. Patients typically are assessed every 6 months in the clinic and at all appointments the UHDRS is administered. Therefore, the most recent UHDRS total motor score (maximum score of 124, higher scores indicate poor motor functioning) was obtained as well as their clinician-rated UHDRS Independence score. Participants' general cognitive performance was assessed using the TICS where higher scores (maximum of 41) indicated better performance. Participants' mood state was also evaluated using the BDI, where higher scores

TABLE 1. *Demographics and disease characteristics*

	HD patients	Carers
Male (%)	48	41
Married (%)	83	83
Full-time employed (%)	10.8	21.4
Part-time employed (%)	15.4	30.4
Voluntary work (%)	1.5	3.6
Retired (%)	60	39.3
Employment state affected by HD (%)	38.5	NA
Unemployed (other reasons) (%)	12.3	10.7
Average age left education (yr)	20 ± 9.20	19.72 ± 8.85
Contact with carer		
Daily (%)	89	NA
2 or 3 times per week (%)	4	NA
Weekly (%)	7	NA
Age (yr)	55.24 ± 11.92	54.65 ± 11.40
BDI	10.65 ± 7.95	7.65 ± 8.50
TICS[a]	27.94 ± 6.16	34.78 ± 2.51
Duration of HD	7.92 ± 5.69	NA
UHDRS motor score	40.68 ± 20.68	NA
Self-rated UHDRS Independence score	81.34 ± 19.90	NA
Clinician-rated UHDRS Independence score	77.28 ± 19.91	NA

Values are mean ± SD, unless otherwise indicated.
[a]Significant at $P < 0.05$ (two-tailed).
BDI, Beck Depression Inventory; TICS, Telephone Interview of Cognitive Status; UHDRS, Unified Huntington's Disease Rating Scale; HD, Huntington's disease.

indicated more depressed mood (maximum score of 63). These data are presented in Table 1.

HRQoL Measures

The SF-36[5] comprises 36 items with 8 functional dimensions (physical functioning, physical role limitations, mental health, emotional role limitations, social functioning, energy/vitality, pain, and general health perceptions), which can be summarized into two aggregate scores (physical and mental). All scales range from 0 (poorest health state) to 100 (best health state). More recently, a single index of health has been derived from the SF-36.[6] This index is a preference-based measure and provides an indication of the relative value of health, ranging from 0.3 (poorest health state) to 1.0 (best health state).

The SIP[7] consists of 136 items that describe psychosocial and physical health behaviors in 12 categories of function (sleep and rest, eating, work, home management, recreation and pastimes, ambulation, mobility, body care and movement, social interaction, alertness behavior, emotional behavior, and communication), which can be summarized into two aggregate scores (a physical dimension and psychosocial dimension) and then further into a single total score. The physical dimension aggregate was derived from the sum of the "body care and movement," mobility," and "ambulation" dimensions, whereas the psychosocial dimension aggregate was obtained from the sum of the "emotional behavior," "social interaction," alertness behavior," and "communication" dimensions. The overall SIP total score was the sum across all dimensions, and scores indicate percentage of dysfunction ranging from 0 (best health state) to 100 (poorest health state).

RESULTS

The demographic details of respondents are described in Table 1. A one-way analysis of variance showed that there was no significant difference between HD patients and controls in terms of age ($F(1,120) = 0.078$; $P = 0.780$) or mood state as assessed by the BDI ($F(1,101) = 3.405$; $P = 0.068$). However, HD patients in line with group definitions, predictably demonstrated lower general cognitive status (TICS score) than their healthy carers ($F(1,106) = 60.718$; $P<0.0001$).

Figure 1a summarizes quality of life scores on the SF-36 questionnaire for the patients and carers, with reference to normal values (on eight dimensions) derived from an age-equivalent healthy control sample[8] as a comparison. The figure shows that carers' ratings are very similar to population norms, but patients show a significantly lower score by approximately 20 points (indicating poorer quality of life) on the dimensions of "physical functioning" ($F(1,102) = 17.84$; $P<0.0001$), "social functioning" ($F(1,106) = 7.67$; $P = 0.007$), "physical role limitations" ($F(1,103) = 9.93$; $P = 0.002$), and "general health perceptions" ($F(1,102) = 4.05$; $P = 0.047$), as well as aggregate scores ("physical summary component" ($F(1,97) = 8.65$; $P = 0.004$) with only a trend for poorer scores on the "mental summary component" ($F(1,97) = 3.71$; $P = 0.057$).

Patients' and carers' quality of life scores on the SIP are summarized in Figure 1b, with reference to normal values from an age equivalent healthy control sample[1] as a comparison. Carers' ratings are again very similar to population norms, whereas patients show a higher sickness impact score (indicating poorer quality of life) on all dimensions and aggregate scores ($P < 0.05$). "Work" and "alertness behavior" were the most affected for patients, although the dimensions of "home management," "recreation and past times," and "communication" were all severely impaired as well (mean score > 20). Only on the "eating" dimension were patients not impaired (mean score < 6), although they were significantly more impaired than carers. On the aggregate scales, the "psychosocial dimension" was at the thresh-

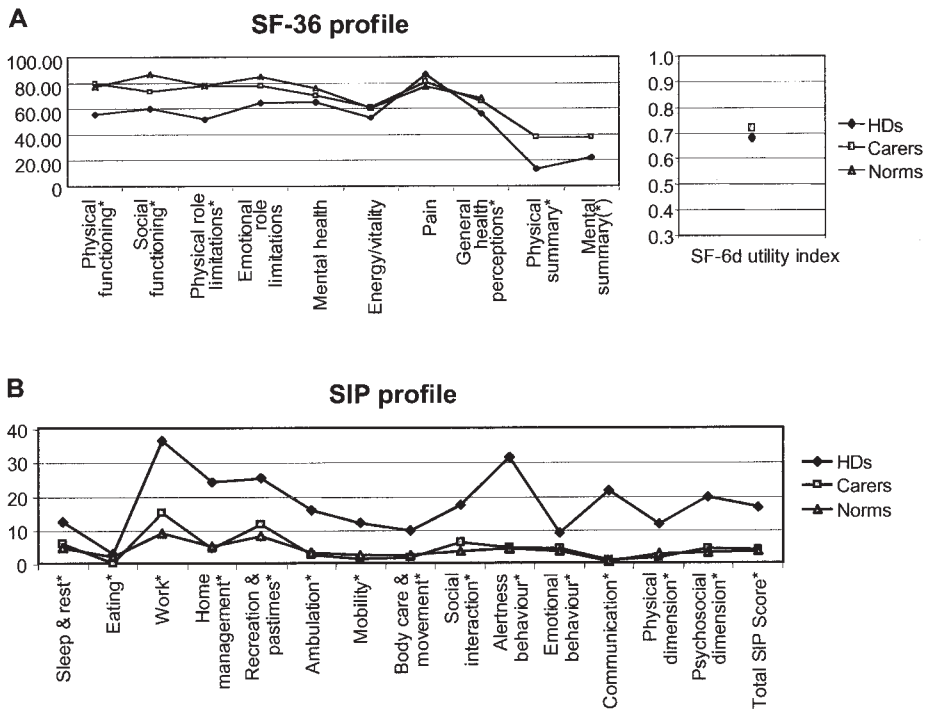

FIG. 1. a: Short Form 36 health survey questionnaire (SF-36) scores (norms from Jenkinson and colleagues[8]) and (**b**) Sickness Impact Profile (SIP) scores for patients and carers (norms from Helder and coworkers[1]). The asterisk indicates significant group difference at $P < 0.05$).

old of severe impairment and rated as more impaired than the "physical dimension."

Reliability: Internal Consistency and Test–Retest Stability

To test the internal consistency of these instruments, Cronbach's α coefficients were used. Both the SF-36 and SIP from HD patients and carers were all above criterion[9] ($\geq$0.8) at Time 1 and Time 2, demonstrating a high degree of internal consistency for both questionnaires.

Test–retest reliability for the SF-36 dimensions and aggregate scales appeared to be highly stable over time for both HD patients and carers for virtually all subscales, as shown by intraclass correlation coefficients (ICCs) that were greater than 0.7.[10] The only exception was for the "emotional role limitation" subscale for patients (0.6383), which just fell short of the criterion. The SIP questionnaire appeared to be fairly reliable for most subscales over the retest period for patients for which all dimensions and aggregate scales had ICCs above 0.7, with two subscales, i.e., "emotional behavior" (0.4819) and "work" (0.6858), falling below criterion. For carers, the SIP again showed reasonable overall test–retest reliability, with three different subscales falling short of criterion, i.e. "mobility" (0.5176), "alertness behavior" (0.3791), and "social interaction" (0.6243).

Construct Validity

An inspection of the correlation matrix between patients' clinical variables and the health dimensions of both questionnaires generally showed that there appeared to be reasonably good convergent validity. Spearman's correlations occurred in a coherent manner and in the direction expected, for example, increasing age in both patients (Table 2) and carers (Table 3) corresponded to poorer ratings primarily on physical dimensions, while more depressed mood was associated with poorer ratings on dimensions that tapped both physical and mental aspects of health. The TICS did not appear to correlate significantly with any of the scales for carers, perhaps due to a ceiling effect for carers who had significantly higher scores than patients. In patients, it appears that poorer TICS scores were associated with poorer attention as there was a significant correlation with SIP "alertness behavior" and with measures of disease severity (e.g., "physical functioning" in the SF-36; e.g., "ambulation" in the SIP). The correlation matrix also shows that the pattern of correlations between quality of life dimensions and patient-rated Independence scores were virtually identical with that shown by clinician-rated scores, and indeed patients and clinicians ratings were very highly correlated ($r^2 = 0.93$; $P < 0.0001$).

TABLE 2. *HD patients' Spearman's correlations between functional measures and subscales of the SF-36 and SIP*

	Age	UHDRS	Disease duration	PatIS	ClinIS	TICS	BDI
SF-36							
Physical functioning	−0.07	−0.54[a]	−0.38[a]	0.64[a]	0.70[a]	0.51[a]	−0.04
Physical role limitations	0.00	−0.25	−0.15	0.42[a]	0.45[a]	0.27	−0.35[b]
Mental health	0.16	0.27[b]	−0.05	0.02	0.06	−0.11	−0.62[a]
Emotional role limitations	−0.11	0.13	0.05	0.09	0.04	0.01	−0.41[a]
Energy/vitality	0.16	0.04	−0.12	0.25	0.35[b]	0.13	−0.52[a]
Pain	−0.05	0.03	−0.16	0.31	0.30	−0.06	−0.41[a]
General health perceptions	0.13	−0.03	−0.25	0.44[a]	0.46[a]	0.09	−0.49[a]
Physical summary	−0.03	−0.26	−0.25	0.57[a]	0.59[a]	0.36[b]	−0.49[a]
Mental summary	0.02	0.07	−0.05	0.28	0.29	0.09	−0.66[a]
SF-36 Utility Index	0.06	−0.23	−0.35[b]	0.51[a]	0.54[a]	0.32[b]	−0.36[b]
SIP							
Sleep and rest	0.05	0.06	0.14	−0.36[a]	−0.38[a]	−0.22	0.26
Eating	0.32[b]	0.15	0.02	−0.30[b]	−0.34[b]	−0.33[b]	0.15
Work	−0.36[a]	0.14	0.28[b]	−0.24	−0.29	0.01	0.36[a]
Home management	0.20	0.24	0.14	−0.45[a]	−0.46[a]	−0.21	0.15
Recreation and pastimes	0.12	0.19	0.03	−0.66[a]	−0.52[a]	−0.20	0.52[a]
Ambulation	0.17	0.46[a]	0.41[a]	−0.69[a]	−0.71[a]	−0.35[b]	0.15
Mobility	0.06	0.26	0.28[b]	−0.58[a]	−0.61[a]	−0.26	0.30[b]
Body care and movement	0.01	0.30[b]	0.21	−0.57[a]	−0.57[a]	−0.12	0.22
Social interaction	−0.01	0.02	0.07	−0.36[a]	−0.18	−0.23	0.59[a]
Alertness behaviour	0.07	0.13	0.18	−0.39[a]	−0.35[a]	−0.28[b]	0.56[a]
Emotional behaviour	0.07	−0.09	0.06	−0.20	−0.10	0.10	0.29[b]
Communication	0.13	0.42[a]	0.22	−0.59[a]	−0.59[a]	−0.34[b]	0.31[b]
Physical dimension	0.09	0.42[a]	0.35[a]	−0.72[a]	−0.72[a]	−0.29[b]	0.29[b]
Psychosocial dimension	0.08	0.17	0.16	−0.51[a]	−0.40[a]	−0.27	0.60[a]
Total SIP score	0.10	0.32[b]	0.24	−0.69[a]	−0.64[a]	−0.33[b]	0.47[a]

Note: UHDRS is the Unified Huntington's Disease Rating Scale motor scale (higher scores = poorer performance). PatIS is the patient's self-rated score on their level of functioning/independence level (higher scores = better performance). ClinIS is the clinician's rating of patients' level of functioning/independence level (higher scores = better performance). The TICS (Telephone Interview of Cognitive Status) is a measure of general cognition/mental status (higher scores = better performance), The BDI (Beck Depression Inventory) is a self-rated measure of depressive mood (higher scores = poorer performance).

[a]Significant at $P < 0.01$ (two-tailed).

[b]Significant at $P < 0.05$ (two-tailed).

HD, Huntington's disease; SF-36, Short Form 36; SIP, Sickness Impact Profile.

To more formally summarize these observations, a reliability analysis was performed on patients' correlations between the SF-36 scales and clinical variables (age, UHDRS, disease duration, clinician-rated Independence Scale, patient-rated Independence Scale, TICS, BDI) and separately for the SIP to provide a measure of agreement for each set of correlations. The SF-36 demonstrated a higher reliability between correlations (ICC = 0.74) than the SIP scales (ICC = 0.64). A similar analysis for carers using fewer clinical variables (age, TICS, BDI) showed a similar pattern favoring the SF-36 (ICC = 0.80) over the SIP (ICC = 0.66). This finding suggests that the SF-36 appears to show relatively higher construct validity as a measure of health-related quality of life.

A multivariate regression model was used to assess the association between disease severity (as indexed by duration of disease) and the aggregate scales of both questionnaires. The SF-36 (with three response variables, i.e., physical component summary, mental component summary, and utility index) showed a significant effect of duration of disease multivariately on all three outcomes (Wilk's Lambda = 0.822; $P = 0.037$). Further examination of univariate test of effect showed that the outcome variables driving the significance of the multivariate test was the utility index ($P = 0.02$) and to a lesser extent the physical component summary ($P = 0.08$). A separate multivariate regression model with three response variables for the SIP (i.e., physical dimension, psychosocial dimension, and total score) showed a significant effect of duration of disease multivariately on all three outcomes as well (Wilk's lambda = 0.761; $P = 0.004$). The outcome variables driving the significance of this multivariate test was the physical aggregate score ($P = 0.002$) and the total score ($P = 0.05$).

Finally, we examined the extent to which illness-related clinical variables were able to explain the variance on the SIP and SF-36 aggregate scores. Separate multiple regression analyses with SF-36 (Table 4, top) and SIP (Table 4, bottom) aggregate scores were used as

TABLE 3. *Carers' Spearman's correlations between functional measures and subscales of the SF-36 and SIP*

	Age	TICS	BDI
SF-36			
Physical functioning	−0.43[a]	0.10	−0.42[a]
Physical role limitations	−0.28[b]	−0.08	−0.59[a]
Mental health	−0.14	0.09	−0.77[a]
Emotional role limitations	−0.30[b]	−0.09	−0.50[a]
Energy/vitality	−0.21	−0.17	−0.66[a]
Pain	−0.28[b]	−0.02	−0.48[a]
General health perceptions	−0.03	−0.04	−0.62[a]
Physical component summary	−0.30[b]	−0.03	−0.71[a]
Mental component summary	−0.25	−0.02	−0.77[a]
SF-36 utility index	−0.19	0.20	−0.57[a]
SIP			
Sleep & rest	0.13	−0.18	0.63[a]
Eating	0.29[b]	−0.19	0.24
Work	0.06	−0.03	0.37[a]
Home management	0.13	0.06	0.53[a]
Recreation & pastimes	0.24	0.08	0.62[a]
Ambulation	0.12	−0.05	0.27
Mobility	0.19	−0.06	0.34[b]
Body care & movement	0.09	−0.07	0.36[a]
Social interaction	0.15	−0.07	0.61[a]
Alertness behaviour	−0.02	−0.15	0.55[a]
Emotional behaviour	0.06	−0.15	0.51[a]
Communication	0.31	−0.18	0.14
Physical dimension	0.20	−0.07	0.41[a]
Psychosocial dimension	0.14	−0.10	0.66[a]
Total SIP score	0.24	−0.04	0.74[a]

[a]Significant at $P < 0.01$ (two-tailed).
[b]Significant at $P < 0.05$ (two-tailed).
SF-36, Short Form 3b; SIP, Sickness Impact Profile; TICS, Telephone Interview of Cognitive Status; BDI, Beck Depression Inventory.

criteria, and then age, UHDRS, disease duration, clinician-rated Independence Scale, patient-rated Independence Scale, TICS, and BDI were chosen as predictors. It was shown that comparatively high R^2 values (>0.40) were obtained on all aggregates scales for both questionnaires. From the table, the BDI was clearly the most important predictor for aggregate scores of both instruments.

DISCUSSION

This study examined the health-related quality of life in a group of mild–moderately severe HD patients and carers using both the SIP and SF-36. For the first time, the suitability of these two instruments was evaluated psychometrically in these patients and their carers. HD patients were found to experience poorer quality of life than their carers (who had very similar ratings with normative data from the general population) on several dimensions, including "general health perceptions," "physical functioning," "physical role limitations," and "social functioning" of the SF-36. The former two dimensions were similarly affected in a recent study,[3] although they also found decreased "pain" for patients, which we did not. The impaired dimensions were reflected in a reduced physical summary score, and a trend for a reduced mental summary score. Patient's overall utility index was similar to that of controls, indicating that patients preference-based quality of life rating was still similar to controls, despite differences on the other descriptive aggregate scales.

On the SIP, patients showed significantly lower quality of life ratings for all dimensions of well-being and all aggregate scores were lower than carers (who had very similar ratings with normative data from the general population), particularly on the psychosocial dimension. This profile was similar to a previous study[1] and provides further support for a HD-specific profile as identified by the SIP. However, the bias toward greater impairment on the psychosocial dimension may be overstated and somewhat misleading because the "communication" dimension, which contributes to this aggregate is largely based on the physical restriction of increased speech motor impairment rather than the desire for verbal communication. Furthermore, "alertness behavior" also contributes to the aggregate psychosocial score, although some items actually contain a clearly motor or physical component (i.e., statements including "I have more minor accidents: for example, I drop things, I trip and fall, or I bump into things"), which may result in overestimation of the aggregate score. As such, while fewer dimensions of the SF-36 appear to be sensitive to disease status, the SF-36 aggregate scores appear to project a more accurate HD profile of impaired physical and mental functioning, with a greater emphasis on the former.

Internal consistency was uniformly high for both instruments in both groups and at both time points. For patients and carers, test–retest reliability of both questionnaires was generally high, with only one ("physical role limitations" for patients) and five ("work" and "emotional behavior" for patients; "mobility," "social interaction," and "alertness behavior" for carers) dimensions of the SF-36 and SIP, respectively, showing reliability indices that fell short of criterion. On this basis, the SF-36 appeared to be more stable over time, for patients and carers alike.

For both instruments, there appeared to be good convergent reliability between the many dimensions that tapped physical function with clinical indices of disease extent, symptom severity and functional ability. However, only one dimension (SIP "alertness be-

TABLE 4. *Results of multiple regression analyses with clinical variables as predictors and SF-36 and SIP aggregate scores as criteria*

	Age	UHDRS	Disease duration	PatIS	ClinIS	TICS	BDI	R^2
SF-36								
Pcs								0.51
β	−0.18	0.33	−0.06	0.06	0.50	0.18	−0.35	
p	0.24	0.18	0.69	0.86	0.21	0.36	0.03	
Mcs								0.48
β	−0.20	0.45	−0.14	−0.03	0.31	0.13	−0.56	
p	0.20	0.08	0.41	0.94	0.45	0.52	0.00	
Utility index								0.41
β	0.01	0.41	−0.31	−0.14	0.64	0.62	−0.20	
p	0.95	0.15	0.13	0.72	0.15	0.54	0.27	
SIP								
Phy								0.66
β	0.12	−0.19	0.24	−0.22	−0.64	−0.00	−0.12	
p	0.33	0.34	0.07	0.45	0.05	0.98	0.37	
Psy								0.46
β	0.20	0.11	0.11	−0.54	0.34	−0.13	0.45	
p	0.20	0.65	0.51	0.14	0.40	0.53	0.01	
SIP Total								0.59
β	0.19	−0.05	0.09	−0.31	−0.32	−0.05	0.22	
p	0.17	0.83	0.53	0.32	0.35	0.78	0.12	

Note: For the SF-36, Pcs = Physical component summary, Mcs = Mental component summary. For the SIP, Phy = Physical dimension, Psy = Psychosocial dimension.

SF-36, Short Form 36; SIP, Sickness Impact Profile; UHDRS, Unified Huntington's Disease Rating Scale; PatIS, patient's self-rated score; ClinIS, clinician's rating of patient functioning; TICS, Telephone Interview of Cognitive Status; BDI, Beck Depression Inventory.

havior") was associated with cognitive status as measured using the TICS. This finding is likely to have arisen because there were fewer dimensions that directly tapped mental functioning, and it is more difficult to gauge this more-abstract dimension of well-being. SIP dimensions such as "alertness behavior" and "communication," which contribute to the aggregate score for the psychosocial dimension are in fact influenced by movement-related items. Other SIP dimensions such as "emotional behavior" and "social interaction" are difficult to represent and, therefore, also tend to be less stable over time.

Of interest, the BDI correlated strongly with almost all dimensions of well-being, and as such, we were able to reasonably predict all three aggregate scores for both instruments to more or less the same extent by using our clinical variables as predictors. This was achieved primarily with the inclusion of the BDI, as a past study[1] has shown comparatively poorer prediction rates without a clinical index of mood. Whereas mood is strongly associated with ratings on both physical and mental dimensions, it was not specifically measured in either instrument. Another observation from the correlation matrix is the reassuring consistency between purely patient-rated and clinician-rated independence scale scores during interview.

An incidental finding of this study was the similarity of carers' responses on QoL dimensions on both the SIP and SF-36. This finding supports that of a previous study[3] and may be due to the positive aspects of caring such as heightened self-esteem, and also a response shift in their perception of their own health in comparison to their spouses. While a limiting factor in this study was the low response rate due to the labour intensive nature of study where two questionnaires were administered at two time points, it was not greatly lower than other studies with this patient population that just involved one time point.[3] Furthermore, the demographics of those responding were not significantly different from the nonresponders, suggesting that the group studied was representative of our larger HD cohort.

In summary, the psychometric properties of the SF-36 and SIP show that both questionnaires are generally comparable in terms of validity of dimensions, with a slight advantage for the former, and care must be exercised in interpreting the SIP subscales that relate to the psychosocial dimension, as this is likely to be influenced by physical impairment. The dimensions of the SF-36 also appear to be more stable for patients and carers over the 6-week retest period and, therefore, would emerge as the recommended instrument of choice. The relative psychometric advantage of the SF-36 is also compatible with its practical benefit of being shorter, quicker to administer, and hence, more user friendly. Further work is required to

examine the sensitivity of the SF-36 to determine its utility in tracing patients' and carers' health-related quality of life over the progression of the disease, especially in more advanced stages of disease, and response to emerging treatment interventions.

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Insular Dopamine D_2 Receptors and Novelty Seeking Personality in Parkinson's Disease

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Abstract: **Novelty seeking is a temperament trait characterized by impulsiveness and exploratory behavior. Dopamine has been suggested to be the primary neurotransmitter modulator of novelty seeking, and in young healthy subjects, a correlation between increased novelty seeking and decreased insular cortical dopamine D_2 receptor availability has been reported. The proposed link between dopamine deficiency and reduction in novelty seeking in Parkinson's disease is controversial. The present study examined whether a link between insular D_2 receptor availability and novelty seeking can be replicated in Parkinson's disease patients. [^{11}C]FLB 457 positron emission tomography imaging was carried out in 28 patients with Parkinson's disease, and the data were analyzed using voxel-based statistical analysis. The results demonstrated a negative correlation between the novelty seeking score and the dopamine D_2 availability bilaterally in the insular cortex (corrected $P = 0.001$; $r = -0.74$ [right hemisphere]; $r = -0.66$ [left hemisphere]). The results provide further support for a relationship between novelty seeking and insular D_2 receptors. They indicate that the association is cross-cultural, independent of age, and unaffected by dopaminergic degeneration.** © 2004 Movement Disorder Society

Key words: PET; personality; insular cortex; dopamine D_2 receptor; Parkinson's disease

Novelty seeking, a tendency toward excitement in response to novel stimuli, represents an independent trait of temperament, which is influenced by genetic factors. Individuals who are higher than average in novelty seeking are characterized as impulsive, exploratory, fickle, excitable, quick-tempered, extravagant, and disorderly. Individuals who are low in novelty seeking, in contrast, have been described as often becoming preoccupied with narrowly focused details and require considerable thought before making decisions.[1] There are several neu-

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