

# Huntington's Disease Patients Have Selective Problems With Insight

Aileen K. Ho, PhD,<sup>1,2\*</sup> Anna O.G. Robbins, BSc,<sup>2</sup> and Roger A. Barker, MRCP, PhD<sup>2,3</sup>

<sup>1</sup>*School of Psychology, University of Reading, Reading, United Kingdom*

<sup>2</sup>*Cambridge Centre for Brain Repair, University of Cambridge, Cambridge, United Kingdom*

<sup>3</sup>*Department of Neurology, Addenbrooke's Hospital, Cambridge, United Kingdom*

**Abstract:** The objective of this study was to determine insight in patients with Huntington's disease (HD) by contrasting patients' ability to rate their own behavior with their ability to rate a person other than themselves. HD patients and carers completed the Dysexecutive Questionnaire (DEX), rating themselves and each other at two time points. The temporal stability of these ratings was initially examined using these two time points since there is no published test–retest reliability of the DEX with this population to date. This was followed by a comparison of patients' self-ratings and carer's independent ratings of patients by performing correlations with patients' disease variables, and an exploratory factor analysis was conducted on both sets of ratings. The DEX showed good test–

retest reliability, with patients consistently and persistently underestimating the degree of their dysexecutive behavior, but not that of their carers. Patients' self-ratings and carers' ratings of patients both showed that dysexecutive behavior in HD can be fractionated into three underlying components (Cognition, Self-regulation, Insight), and the relative ranking of these factors was similar for both data sets. HD patients consistently underestimated the extent of only their own dysexecutive behaviors relative to carers' ratings by 26%, but were similar in ascribing ranks to the components of dysexecutive behavior. © 2005 Movement Disorder Society

**Key words:** Huntington's disease; dysexecutive syndrome; carers; cognition; self-regulation; insight.

Behavioral and cognitive changes related to impaired frontal lobe functioning are often referred to as a dysexecutive syndrome. These changes include impulsiveness, apathy, lack of insight, and disorganization<sup>1</sup> and have been linked to compromised frontostriatal integrity<sup>2</sup> as occurs in Huntington's disease (HD).<sup>3</sup> In HD, it is well known that cognitive and behavioral dysexecutive changes together with motor impairment constitute the cardinal triad of symptoms in this condition.

The dysexecutive symptoms of patients with HD are typically provided by informants or by clinicians in a clinical interview and assessment. Anecdotally, the impression is that HD patients may be unaware of certain aspects of their behavior or may have a different or

distorted self-perception of this. A common way of determining awareness of impairment is to contrast a patient's self-rating with a more objective rater, such as a spouse or carer who has intimate knowledge of the patient's day-to-day behavior.<sup>4</sup> To our knowledge, there has not been a systematic investigation between such informant observations and the observation of HD patients themselves. This comparison of the perspectives in a patient–carer dyad is of interest since the aspects of behavior most evident to each party may not be the same, and it provides not only useful information on the patients' level of awareness but also valuable insight into the management approach most likely to succeed. Carers are in a unique and important position of being able to observe the patient continually, in a variety of naturalistic situations. Although carers' ratings of dysexecutive symptoms are perhaps inevitably tinged by their inherent emotional involvement, there is evidence for carers' ratings to be more objective than patients' self-ratings since the former have been found to show a stronger correlation with neuropsychological tests of executive func-

\*Correspondence to: Dr. Aileen K. Ho, School of Psychology, University of Reading, Earley Gate, Reading RG6 6AL, United Kingdom. E-mail: aileenkho@netscape.net

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tion.<sup>5,6</sup> Apart from providing a relatively more objective benchmark for evaluating patients' awareness, carers' ratings also reflect their personal perspectives of the patients' dysexecutive behavior. Thus, an analysis of patient-carer perspectives is also likely to prove useful in a practical sense, since any differences may have implications in the dynamics of the dyad and therefore implications for management and quality-of-life issues.

In this study, we used the Dysexecutive Questionnaire (DEX) from the Behavioral Assessment of the Dysexecutive Syndrome (BADs) battery,<sup>5</sup> which has been validated in a general neurological population,<sup>7</sup> to examine the ability of HD patients to perform self-rater assessments of their own behavior to demonstrate their degree of insight, and to perform independent assessments of the behavior of another person (their carer). The temporal stability of these ratings was examined using assessment at two time points since there are no published test-retest reliability data on the DEX with this population to date. A comparison of patients' self-ratings and carer's independent ratings was then examined by performing correlations with patients' disease variables and finally exploratory factor analysis on both sets of ratings was conducted to determine the underlying components of dysexecutive behavior from both perspectives.

## PATIENTS AND METHODS

This study was part of a larger study of quality of life in patients with a genetic diagnosis of HD and their respective carers (for further details, including satisfactory response rates, see Ho et al.<sup>8</sup>). Briefly, the sample comprised respondents of a mail-out to patients in the local HD clinic in Cambridge. Participants received the DEX at two time points, with the Beck Depression Inventory (BDI)<sup>9</sup> accompanying the initial send-out. All participants reported their age; patients also self-reported the duration of disease (from first manifestation of symptoms) and self-rated Unified Huntington's Disease Rating Scale (UHDRS) Independence Score (maximum score of 100 indicates total independence). Patients are typically reviewed every 6 months in the clinic and the motor UHDRS assessment is administered along with questions on Total Functional Capacity and an Independence Scale Score ascribed; no detailed psychiatric assessment was conducted. Therefore, the most recent UHDRS total motor score (maximum of 124, indicating poorest motor functioning) was obtained as well as their clinician-rated UHDRS Independence Score.

The average time interval between test and retest of the DEX was  $6 \pm 0.84$  weeks. At an average of  $2 \pm 0.53$  weeks after receipt of the time 1 questionnaires, a follow-up telephone call was made and the Telephone In-

**TABLE 1.** Demographics and disease characteristics

	HD patients	Carers
Demographics		
Male:female	37:38	31:36
Age	$53.91 \pm 12.63$	$54.65 \pm 11.40$
General cognition (TICS) <sup>a</sup>	$28.61 \pm 5.98$	$34.79 \pm 2.51$
Mood (BDI)	$9.68 \pm 7.68$	$7.65 \pm 8.50$
Disease characteristics		
Duration of disease	$7.02 \pm 5.77$	NA
UHDRS motor score	$34.47 \pm 23.62$	NA
UHDRS total functional capacity	$11.89 \pm 0.76$	NA
UHDRS Independence Scale, patient-rated	$82.62 \pm 19.99$	NA
UHDRS Independence Scale, clinician-rated	$79.73 \pm 20.30$	NA

<sup>a</sup>Significant at  $P < 0.05$ .

terview of Cognitive Status (TICS)<sup>10</sup> was administered to assess participants' general cognitive status.

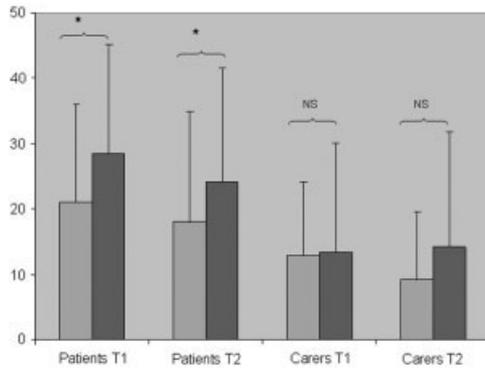
The DEX<sup>5</sup> is a 20-item questionnaire constructed to reflect the range of problems encountered in the dysexecutive syndrome, covering four areas: emotional/personality changes, motivational changes, behavioral changes, and cognitive changes. Items are scored using a five-point Likert scale ranging from 0 (never) to 4 (very often). Higher scores reflect greater dysexecutive characteristics. There are two identical versions of the DEX, one to be completed by the patient (self-rated version) and the other by a close relative or carer (independent rater version) who is well acquainted with the patient. In this study, both patients and carers completed self-rated versions (rating themselves) as well as independent rater versions (rating the other party).

The participants comprised 75 HD patients at all stages of disease and 67 carers. Group demographics are presented in Table 1. Patients and carers were not significantly different in terms of age [ $t(133) = 0.36$ ;  $P > 0.05$ ] and mood on BDI [ $t(112) = 1.33$ ;  $P > 0.05$ ]. Consistent with their disease status, patients' general cognitive status was lower than that of carers [ $t(112) = 7.19$ ;  $P < 0.05$ ], although importantly not reaching scores typically associated with dementia.

## RESULTS

Summary data of all DEX measures are presented in Figure 1. Spearman's correlations show that test-retest reliability was significantly and similarly high for all measures ( $r^2 > 0.7$ ;  $P < 0.001$ ) conducted by patients and carers.

It was also established that patients did indeed demonstrate significantly more dysexecutive symptoms than carers as evidenced by self-ratings (time 1:  $t(143) = 3.59$ ,  $P < 0.05$ ; time 2:  $t(76) = 2.72$ ,  $P < 0.05$ ) as well



**FIG. 1.** Patients' and carers' self- and independent DEX ratings (mean and standard deviation) at time 1 (T1) and at time 2 (T2). Light gray bars, self rating; dark gray bars, others' rating. Asterisk, significant at  $P < 0.05$ ; NS, not significant.

as independent carers' ratings of them (time 1:  $t(119) = 4.85$ ,  $P < 0.05$ ; time 2:  $t(72) = 2.27$ ,  $P < 0.05$ ).

Patients' self-ratings and carers' independent ratings of patients were significantly different at time 1 [ $t(63) = 3.83$ ;  $P < 0.05$ ], and this persisted at time 2 [ $t(34) = 2.25$ ;  $P < 0.05$ ]. Figure 1 shows that on both occasions, patients' self-ratings were underestimated relative to the carers' ratings. However, patients' independent rating of carers' behavior was not significantly different compared to carers' self-rating of their own behavior at time 1 [ $t(52) = 0.24$ ;  $P > 0.05$ ] and at time 2 [ $t(34) = 1.68$ ;  $P > 0.05$ ], so showing that it is a relatively selective lack of insight than a general inability to perform such assessments.

In the correlational analysis, patients' self-ratings correlated significantly with quality-of-life measures, mood, and patient-rated Independence Scale Score (Table 2), such that greater DEX self-rating scores were associated with lower quality of life, level of independence, and mood. As with patients' self-ratings, carers' ratings of patients correlated significantly with quality-of-life measures, but importantly they also correlated with UHDRS measures of disease severity (motor score, functional capacity, clinician-rated independence score) and general cognition as well as patient-rated scores (independence level, mood). Therefore, carers' rating of higher dysexecutive behavior in patients was more objective since their judgement was significantly associated with greater clinical disease severity. It was also noted that there was a moderate and significant correlation between patients' self-rating and carers' independent rating for time 1 ( $r^2 = 0.461$ ;  $P < 0.05$ ;  $n = 64$ ) and for time 2 ( $r^2 = 0.539$ ;  $P < 0.05$ ;  $n = 64$ ).

Finally, the exploratory factor analysis (varimax rotation, eigenvalues  $>1$  reported) of patients' self-rated

DEX scores revealed a three-factor model (Self-regulation, Cognition, and Insight) that accounted for 64.47% of the variance. When carers' ratings were subject to factor analysis, a four-factor model (Cognition, Self-regulation, Psychosocial self-regulation, and Insight), which accounted for 71.70% of the variance, emerged (Table 3).

Table 3 shows that the patients' self-rated three-factor model is extremely similar to the carers' independent-rated four-factor model, in that patients' Self-regulation factor is essentially an amalgamation of carers' self-regulation and Psychosocial self-regulation factors. Therefore, these two closely related factors will be combined into one general Self-regulation factor in the next analysis, where the DEX item scores (which loaded onto each factor) were averaged to provide a composite DEX score for each factor. (Figure 2 shows that patients' self-reported Cognition factor was significantly higher than Insight [ $t(73) = 5.03$ ;  $P < 0.05$ ], which was in turn significantly higher than Self-regulation [ $t(73) = 2.14$ ;  $P < 0.05$ ]. For carers' responses, the Cognition factor was also rated the most affected, but this was not significantly different from the Insight factor [ $t(60) = 1.61$ ;  $P > 0.05$ ], with both being equally affected and significantly higher than the Self-regulation factor [ $t(60) = 5.47$ ,  $P < 0.05$ ;  $t(60) = 2.98$ ,  $P < 0.05$ , respectively].

## DISCUSSION

This study aims to redress the dearth of studies examining patients' and carers' viewpoints on disease-related

**TABLE 2.** Spearman's correlations for HD patients' dysexecutive behavior

	Self-rating	Others' rating
Patient details and disease variables		
Age	-0.207	0.061
Duration of disease	0.182	0.164
UHDRS motor score	-0.024	0.055 <sup>a</sup>
UHDRS total functional capacity	0.020	0.326 <sup>b</sup>
UHDRS Independence Scale, patient-rated	-0.278 <sup>a</sup>	-0.285 <sup>a</sup>
UHDRS Independence Scale, clinician-rated	-0.229	-0.285 <sup>a</sup>
General cognition (TICS)	-0.069	-0.399 <sup>a</sup>
Mood (BDI)	0.784 <sup>b</sup>	0.376 <sup>b</sup>
Quality-of-life measures		
SF-36 physical component	-0.457 <sup>b</sup>	-0.436 <sup>b</sup>
SF-36 mental component	-0.558 <sup>b</sup>	-0.460 <sup>b</sup>
SIP physical dimension	0.306 <sup>b</sup>	0.363 <sup>b</sup>
SIP psychosocial dimension	0.642 <sup>b</sup>	0.539 <sup>b</sup>
SIP total score	0.474 <sup>b</sup>	0.480 <sup>b</sup>

<sup>a</sup>Significant at  $P < 0.05$ .

<sup>b</sup>Significant at  $P < 0.001$ .

SF-36, Short Form 36 health survey; SIP, Sickness Impact Profile.

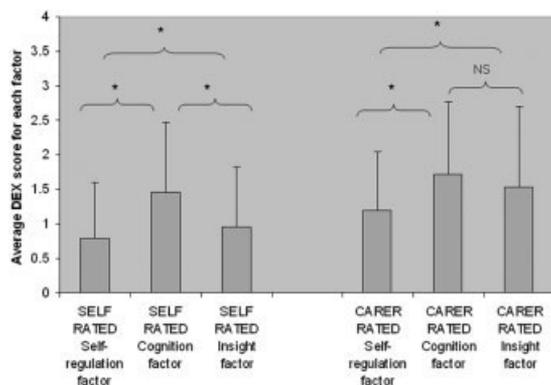
**TABLE 3.** Factor loadings of DEX items

DEX question summary	Patients' self-rating			Carers' independent rating			
	Factor 1: Self-regulation	Factor 2: Cognition	Factor 3: Insight	Factor 1: Cognition	Factor 2: Self-regulation	Factor 3: Psychosocial self-regulation	Factor 4: Insight
1. Comprehension difficulties		0.68		0.67			
2. Acting without thinking	0.59				0.69		
3. Confabulation	0.60					0.75	
4. Planning problems		0.85		0.89			
5. Euphoria	0.69			0.49			
6. Poor temporal sequencing		0.65		0.63			
7. Poor insight			0.53	0.76			
8. Lethargy and apathy		0.66		0.74			
9. Socially embarrassing conduct	0.81				0.81		
10. Variable motivation	0.73				0.82		
11. Difficulty showing emotion			0.74				0.64
12. Quick-tempered	0.66					0.66	
13. Unconcerned about own behavior			0.65			0.62	
14. Perseverative behavior	0.66				0.79		
15. Restlessness		0.47				0.61	
16. Poor self-control	0.82				0.67		
17. Word-action inconsistency	0.77				0.79		
18. Easily distracted		0.72		0.78			
19. Poor decision-making		0.83		0.77			
20. Unconcerned about others' feelings			0.63				0.44

behavioral changes in neurodegenerative disease. The results establish that there is good test-retest reliability for use of the DEX for HD patients and their carers. Patients with HD are indeed more dysexecutive relative to their neurologically intact carers, both when each party rated themselves and when they rated each other. The dysexecutive behavior of patients (from patients' and carers' perspectives) was linked to patients' quality-

of-life indexes. Patients' self-ratings also correlated with self-ratings of mood and independence level. However, carers' ratings of the patients further correlated with clinical measures suggesting that their ratings were indeed more accurate and objective, and a suitable comparison against which patients' level of insight can be determined.

Although patients were able to provide accurate ratings of their carers and so were able to perform this assessment accurately with a third party, they persistently and selectively underestimated the degree of only their own dysexecutive behavior by 26%. This figure is lower than that of a mixed sample of traumatic brain injury patients,<sup>4</sup> but it is approximately twice that found in very elderly rehabilitation patients without neurodegenerative disease.<sup>11</sup> Normal adults typically show no difference between DEX self-rating and informant rating of their behavior<sup>12</sup> as did patients with a related basal ganglia disorder of mild Parkinson's disease.<sup>13</sup> Thus, HD patients have a significant inability to perceive correctly the degree to which their behavior is dysexecutive and show a lack of insight in terms of judging *absolute extent*. As such, they may not recognize that certain



**FIG. 2.** Self-rated and carer-rated average DEX scores for factors. Asterisk, significant at  $P < 0.05$ ; NS, not significant.

behaviors are beyond the socially accepted norm or certain tasks are beyond the normal realm of difficulty for them.

The similarity of the structure of patients' behavior (for self-rated and carer-rated data) from the exploratory factor analysis provides support that HD patients' dys-executive symptoms can indeed be fractionated into three underlying components comprising Self-regulation, Cognition, and Insight. Another important parameter of insight is that of *relative extent*, i.e., the ability to rank order behaviors relative to each other. The pattern of patients' self-ratings show that cognitive problems are most keenly felt, followed by difficulty with insight and then difficulty with self-regulation. However, carers perceived both cognitive problems and lack of insight to be the major problems to an equal extent, with self-regulation difficulties relatively less evident. The similar relative ranking of these three summary DEX factors shows that HD patients generally concur with carers regarding the area they are most (cognition) and least (self-regulation) affected by, and judging *relative extent* appears to be more intact than *absolute extent*. It also provides support that patients' were in fact discriminately self-rating the items relative to each other, but were simply too lenient across the board.

Patients' lack of insight regarding the extent of their dysexecutive behavior has broad implications for patient management both in the clinic and home and may influence practical decisions such as vocational choices, treatment, and/or rehabilitative compliance. This study shows that while HD patients may fail to appreciate fully the degree to which their behavior is compromised, their relative ranking of problem areas are nonetheless likely

to reflect closely the more objective and comprehensive observations of an external party.

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