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VIDEO SUPPLEMENT

Unified Huntington's Disease Rating Scale: Reliability and Consistency.

Huntington Study Group

Summary: The Unified Huntington's Disease Rating Scale (UHDRS) was developed as a clinical rating scale to assess four domains of clinical performance and capacity in HD: motor function, cognitive function, behavioral abnormalities, and functional capacity. We assessed the internal consistency and the intercorrelations for the four domains and examined changes in ratings over time. We also performed an interrater reliability study of the motor assessment. We found there was a high degree of internal consistency within each of the domains of the UHDRS and that there were significant intercorrelations between the domains of the UHDRS, with the exception of the total behavioral score. There was an excellent degree of interrater reliability for the motor scores. Our limited longitudinal database indicates that the UHDRS may be useful for tracking changes in the clinical features of HD over time. The UHDRS assesses relevant clinical features of HD and appears to be appropriate for repeated administration during clinical studies. **Key Words:** Huntington's disease—Cognitive function—Behavioral abnormalities—Functional capacity—Clinical research.

Huntington's disease (HD) is an autosomal dominant neurodegenerative illness characterized by disorders of movement, cognition, behavior, and functional capacity. The investigators of the Huntington Study Group (HSG) collaborate to develop and examine experimental therapies to treat this progressive disorder. We are particularly interested in assessing interventions that may forestall neuronal degeneration and clinical decline. To this end, the HSG has developed the Unified Huntington's Disease Rating Scale (UHDRS), a comprehensive and reliable instrument to assess the clinical features of HD.

Several instruments and rating scales are currently used to assess features of HD including the quantitated neurological exam (QNE) (1), the HD functional capacity scale (HDFCS) (2), the HD motor rating scale (HDMRS) (3), the Physical Disability and Independence scales (4), Marsden and Quinn's chorea severity scale (5), the HD Activities of Daily Living scale (6), and other relevant measures of HD including the duration of illness, defined as the time from the onset of choreic movements to the time of death (7). The HSG investigators agreed to prospectively evaluate all patients with HD and individuals at risk for HD using a single instrument which combined many of the important elements of these scales.

After several months of pilot experience, the investigators formulated a new hybrid scale to assess four main domains of clinical performance and capacity: motor function, cognitive function, behavioral abnormalities, and functional capacity. Emphasis was placed on clinical features that were likely to show rapid progression and on assessments that could be made during a relatively brief, ~30 min, examination. Neurologists, psychiatrists, neuropsychologists, and other professionals participated in the drafting of the final version of the UHDRS. We report our analysis of the internal consistency and interrelationships of the four domains of the UHDRS and provide pilot results on longitudinal changes in UHDRS scores.

METHODS

Subjects

Data from the final version of the UHDRS have been collected prospectively on 489 patients with

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manifest HD from 20 sites in North America and Europe. Of this group, 229 were men (46.8%), 227 were women (46.4%), and the gender of 33 (6.7%) was unknown. Four hundred and twenty-six patients were white (87.1%), 15 black (3.1%), and race was unknown in 48 (9.8%). One hundred and ninety patients (38.9%) inherited HD from their mother, 211 inherited from their father (43.1%), and the affected parent was unknown in 88 (18.0%). Other characteristics of the cohort are listed in Table 1.

UHDRS

The final version of the UHDRS has four components assessing motor function, cognition, behavior and functional abilities. The UHDRS is reprinted in Appendix 2 and copies of the examination guidelines are available by request.

The motor section of the UHDRS assesses motor features of HD with standardized ratings of oculomotor function, dysarthria, chorea, dystonia, gait, and postural stability. A demonstration of the techniques of the motor exam and examples of each grade of abnormality are provided on the accompanying videotape. The total motor impairment scores is the sum of all the individual motor ratings, with higher scores indicating more severe motor impairment than lower scores.

Cognitive operations are assessed by a phonetic verbal fluency test (8), Symbol Digit Modalities Test (9), and the Stroop Interference Test (10). The Stroop Test results are reported as the raw number of correct answers given in a 45-s period. Results for the other tests are reported as the raw number of correct responses. Higher scores indicate better cognitive performance.

TABLE 1. *UHDRS baseline characteristics (n = 489)*

	Mean	SD	Range
Age	49.6	13.1	11–91
Age onset HD	41.0	12.7	6–74
Duration HD	8.9	5.2	1–35
Total motor score	47.2	22.0	5–106
Verbal fluency	16.3	10.2	0–62
Symbol digit	21.0	10.8	0–50
Stroop test			
Color	52.7	21.3	0–100
Word	38.8	16.5	0–92
Color word	21.3	10.9	0–58
Total behavior score	11.9	10.3	0–51
Subscales			
Mood	6.1	6.3	0–29
Behavior	4.0	4.2	0–14
Psychosis	0.4	1.6	0–10
Obsessive	1.5	3.1	0–16
Behavioral milestones			
Confused (%)	17.0		
Demented (%)	43.8		
Depressed (%)	30.4		
Requiring treatment for depression (%)	39.8		
Functional checklist score	15.9	7.2	0–25
Independence scale	72.3	19.9	10–100
TFC	6.6	3.8	0–13

The behavioral assessment measures the frequency and severity of symptoms related to affect, thought content and coping styles. The total behavior score is the sum of all responses; however, this score may have less usefulness than the individual subscale scores for mood, behavior, psychosis and obsessiveness which are created by summing the responses to the corresponding questions. The evaluator is also requested to provide a clinical impression as to whether the patient, at the time of the evaluation, has clinical evidence of confusion, dementia, or depression and whether the patient requires antidepressant therapy, according to preset definitions in the examination guidelines. Higher scores on the behavior assessments indicate more severe disturbance than lower scores.

The functional assessments include the HDFCS, the Independence scale and a checklist of common daily tasks. For the latter items, the investigator indicates if the patient could perform the task. The checklist is summed by giving a score of 1 to all "yes" replies. The HDFCS is reported as the total functional capacity (TFC) score. This scale has established psychometric properties including interrater reliability and validity, based on radiographic measures of disease progression (11,12). The independence scale is rated from 0 to 100. Higher scores on the function scales indicate better functioning than lower scores.

Internal Consistency

We performed Cronbach's alpha analyses to examine the internal consistency of the motor, cognitive, behavioral and functional checklist components of the UHDRS. We performed correlational analyses comparing the four components of the UHDRS. Specifically, Spearman rank order correlation coefficients were calculated comparing the total motor score, each cognitive test, the behavior score and each subscale, and the three functional scores.

Interrater Reliability

The reliability of the motor component of the UHDRS was examined among three clinicians who were experienced with the evaluation of patients with HD. Twenty-four patients were each rated by two of the three clinicians, with the clinicians eval-

uating the patients independently. Each clinician rated 16 patients. Raters did not discuss their ratings with each other after individual assessments and remained unaware of each other's scores throughout the study. The interrater reliability of the total motor scores, and of the chorea and dystonia scores, were assessed by intraclass correlation.

Longitudinal Data

Longitudinal rates of change for the motor, HD-FCS, and cognitive components were available since these items had not changed substantially from prior versions of the UHDRS. Rates of change were obtained by fitting a least squared line to the data from patients who had been reevaluated with examinations at least 4 months apart and are expressed as the rate of change per 6 months. For the motor and functional sections, only exams performed by the same clinician were included in the analyses.

RESULTS

Internal Consistency

There was a high degree of internal consistency in each of the four components of the UHDRS. Cronbach's alpha values were 0.95 for the motor scale, 0.90 for the cognitive tests, 0.83 for the behavioral scale, and 0.95 for the functional checklist. Correlational analyses (Table 2) showed that four components of the UHDRS were highly intercorrelated, with the exception of the total behavioral score which did not correlate with any of the other assessments. However, higher mood subscale scores correlated with better motor performance, and higher psychosis and obsessive subscale scores correlated with lower functional scores.

Interrater Reliability

The 24 patients in the reliability study included 14 men and 10 women, with an average age of 48.0 ± 16.4 (mean ± SD) and duration of HD for 9.0 ± 5.5 years. TFC scores were 8.0 ± 3.4 with a range of 3–13. The intraclass correlation coefficient was 0.94 for the total motor score, 0.82 for the chorea score, and 0.62 for the dystonia score.

Longitudinal Data

Longitudinal data were available for motor scores on 180 patients, for the TFC on 202 patients, and for cognitive tests in ~130 patients. Patients were followed for 8.0 ± 2.4 months (range, 4.1–19.6 months). Table 3 lists the average change in each score, over a 6-month period.

CONCLUSION

We found a high degree of internal consistency among the motor, behavioral, cognitive, and functional components of the UHDRS. We also found there was a high degree of reliability among three different raters performing the motor assessment. The scores on the motor, cognitive and functional components, including the previously validated TFC scale, were highly intercorrelated, although the total behavioral score did not correlate well with the other sections. However, the mood subscale was associated with better motor performance, a finding consistent with the predominance of mood disorders in early HD. Additionally, the psychotic and obsessive disturbance subscales were associated with functional impairment, consistent with

TABLE 2. *Intercorrelations of UHDRS assessments*

	Motor	VF	Sym D	S-W	S-C	S-CW	Behav T	S-M	S-B	S-P	S-0	FC	IND
Motor	1												
VF	*−0.60	1											
Symbol digit	*−0.65	*0.64	1										
Stroop word	*−0.63	*0.61	*0.77	1									
Stroop color	*−0.64	*0.64	*0.73	*0.85	1								
Stroop color word	*−0.57	*0.58	*0.69	*0.72	*0.66	1							
Behavioral total	−0.10	0.07	0.05	0.08	0.04	−0.03	1						
Mood subscale	*−0.19	0.15	0.14	0.12	0.09	0.06	*0.81	1					
Behavior subscale	0.01	0.03	0.06	0.10	0.05	−0.02	*0.71	*0.31	1				
Psychosis subscale	0.04	0.00	−0.05	−0.01	0.02	−0.01	*0.28	0.10	*0.19	1			
Obsessive subscale	0.04	0.02	−0.04	−0.02	−0.02	−0.10	*0.51	*0.21	*0.29	*0.22	1		
Functional checklist	*−0.75	*0.59	*0.65	*0.60	*0.61	*0.58	−0.07	0.06	−0.13	*−0.14	*−0.14	1	
Independence	*−0.75	*0.58	*0.63	*0.62	*0.63	*0.54	−0.05	0.09	−0.13	*−0.14	−0.10	*0.90	1
TFC	*−0.72	*0.58	*0.62	*0.58	*0.61	*0.52	−0.07	0.06	*−0.14	*−0.13	−0.12	*0.94	*0.92

* $p < 0.005$.

S-C, Stroop color; S-W, Stroop word; S-CW, Stroop color word; Behav T, behavioral total; S-M, subscale mood; S-B, subscale behavior; S-P, subscale psychosis; S-O, subscale obsessive; FC, functional checklist; IND, independence; VF, verbal fluency; TFC, total functional capacity

TABLE 3. *Longitudinal changes in UHDRS (expressed as units of change per 6 months)*

	Mean	SD	Range
Total motor (n = 180)	3.2	8.4	−21.6 to 33.5
VF (n = 142)	0.1	5.3	17.7 to 21.7
Symbol digit (n = 131)	−0.1	4.2	11.4 to 21.7
Stroop color word (n = 139)	−1.3	13.1	−103.0 to 44.7
Functional checklist (n = 171)	−0.9	3.0	−9.1 to 12.8
Independence (n = 180)	−3.8	8.6	−41.3 to 29.0
TFC (n = 202)	−0.3	2.1	−7.2 to 19.6
TFC where initial score ≥ 3 (n = 176)	−0.5	1.6	−7.2 to 6.6

the predominance of these disorders in the later stages of HD. These findings reflect the fact that behavioral abnormalities, unlike the slow and fairly steady deterioration in the other domains, are heterogeneous, episodic and without clear additive temporal progression. Also behavioral disturbances are the aspects of HD that are most amenable to symptomatic intervention, and therefore are likely to be less consistently observed over time.

The UHDRS assesses relevant clinical domains of HD and was designed for repeated administrations during clinical research studies. We continue to prospectively collect UHDRS data and now have accrued >1,500 individuals with HD and individuals at risk for HD. The preliminary longitudinal data suggest that the UHDRS may be useful for tracking clinical changes longitudinally in HD patients, although further data are needed to establish its utility. The UHDRS may be particularly suitable for tracking clinical changes in the setting of controlled trials of experimental interventions. The UHDRS yields several scores assessing the primary features of HD (motor, cognitive, behavioral) as well as the overall functional impact of these features; therefore, the primary hypothesis of the clinical trial should determine the selection of the primary response variable, while the other scales may serve as secondary response measures. Studies of the usefulness of the UHDRS in clinical trials are in progress.

APPENDIX 1

The following persons and participating institutions of the Huntington Study Group were involved in the design and implementation of the UHDRS and authorship of this report.

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APPENDIX 2
HUNTINGTON STUDY GROUP
UNIFIED HUNTINGTON'S DISEASE RATING SCALE

MOTOR ASSESSMENT

OCULAR PURSUIT (horizontal and vertical)
- 0 = complete (normal)
- 1 = jerky movement
- 2 = interrupted pursuits/full range
- 3 = incomplete range
- 4 = cannot pursue

SACCADE INITIATION (horizontal and vertical)
- 0 = normal
- 1 = increased latency only
- 2 = suppressable blinks or head movements to initiate
- 3 = unsuppressable head movements
- 4 = cannot initiate saccades

SACCADE VELOCITY (horizontal and vertical)
- 0 = normal
- 1 = mild slowing
- 2 = moderate slowing
- 3 = severely slow, full range
- 4 = incomplete range

DYSARTHRIA
- 0 = normal
- 1 = unclear, no need to repeat
- 2 = must repeat to be understood
- 3 = mostly incomprehensible
- 4 = mute

TONGUE PROTRUSION
- 0 = can hold tongue fully protruded for 10 seconds
- 1 = cannot keep fully protruded for 10 seconds
- 2 = cannot keep fully protruded for 5 seconds
- 3 = cannot fully protrude tongue
- 4 = cannot protrude tongue beyond lips

MAXIMAL DYSTONIA (trunk and extremities)
- 0 = absent
- 1 = slight/intermittent
- 2 = mild/common or moderate/intermittent
- 3 = moderae/common
- 4 = marked/prolonged

MAXIMAL CHOREA (face, mouth, trunk and extremities)
- 0 = absent
- 1 = slight/intermittent
- 2 = mild/common or moderate/intermittent
- 3 = moderate/common
- 4 = marked/prolonged

RETROPULSION PULL TEST
- 0 = normal
- 1 = recovers spontaneously
- 2 = would fall if not caught
- 3 = tends to fall spontaneously
- 4 = cannot stand

FINGER TAPS (right and left)
- 0 = normal) (≥15/5 sec.)
- 1 = mild slowing and or reduction in amplitude (11–14/5 sec.)
- 2 = Moderately impaired. Definite and early fatiguing. May have occasional arrests in movement (7–10/5 sec.).
- 3 = Severely impaired. Frequent hesitation in initiating movements or arrests in ongoing movements (3–6/5 sec.)
- 4 = Can barely perform the task (0–2/5 sec.)

PRONATE/SUPINATE-HANDS (right and left)
- 0 = normal
- 1 = mild slowing and/or irregular
- 2 = moderate slowing and irregular
- 3 = severe slowing and irregular
- 4 = cannot perform

LURIA (fist-hand-palm test)
- 0 = ≥4 in 10 seconds, no cue
- 1 = <4 in 10 seconds, no cue
- 2 = ≥4 in 10 seconds, with cues
- 3 = <4 in 10 seconds with cues
- 4 = cannot perform

RIGIDITY-ARMS (right and left)
- 0 = absent
- 1 = slight or present only with activation
- 2 = mild to moderate
- 3 = severe, full range of motion
- 4 = severe with limited range

BRADYKINESIA-BODY
- 0 = normal
- 1 = minimally slow (? normal)
- 2 = mildly but clearly slow
- 3 = moderately slow, some hesitation
- 4 = markedly slow, long delays in initiation

GAIT
- 0 = normal gait, narrow base
- 1 = wide base and/or slow
- 2 = wide base and walks with difficulty
- 3 = walks only with assistance
- 4 = cannot attempt

TANDEM WALKING
- 0 = normal for 10 steps
- 1 = 1 to 3 deviations from straight line
- 2 = >3 deviations
- 3 = cannot complete
- 4 = cannot attempt

COGNITIVE ASSESSMENT

VERBAL FLUENCY TEST (raw score)

SYMBOL DIGIT MODALITIES TEST (raw score)

STROOP INTERFERENCE TEST

Color Naming (number correct)
Word Reading (number correct)
Interference (number correct)

BEHAVIORAL ASSESSMENT

Use the following keys to rate both severity and frequency

Severity	*Frequency*
0 = absent	0 = almost never
1 = slight, questionable	1 = seldom
2 = mild	2 = sometimes
3 = moderate	3 = frequently
4 = severe	4 = almost always

Sad/Mood: feeling sad, sad voice/expression, tearfulness, inability to enjoy anything.
Low Self-Esteem/Guilt: self blame, self deprecation including feelings of being a bad or unworthy person, feelings of failure.
Anxiety: worries, anticipation of the worst, fearful anticipation.
Suicidal Thoughts: feels life not worth living, has suicidal thoughts, active suicidal intent, preparation for the act.
Disruptive or Aggressive Behavior: threatening behavior, physical violence, verbal outbursts, threatening, foul, or abusive language.
Irritable Behavior: impatient, demanding, inflexible, driven and impulsive, uncooperative.
Obsessions: recurrent and persistent ideas, thoughts or images
Compulsions: repetitive, purposeful, and intentional behaviors.
Delusions: Fixed false beliefs, not culturally shared
Hallucinations: a perception without physical stimulus: Auditory, Visual, Tactile, Gustatory and Olfactory

Does the investigator believe the subject is confused? Yes or No
Does the investigator believe the subject is demented? Yes or No
Does the investigator believe the subject is depressed? Yes or No
Does the subject require pharmacotherapy for depression? Yes or No

FUNCTIONAL ASSESSMENT Yes or No

Could subject engage in gainful employment in his/her accustomed work?

Could subject engage in any kind of **gainful** employment?

Could subject engage in any kind of volunteer or non gainful work?

Could subject manage his/her finances (monthly) without any help?

Could subject shop for groceries without help?

Could subject handle money as a purchaser in a simple cash (store) transaction?

Could subject supervise children without help?

Could subject operate an automobile safely and independently?

Could subject do his/her own housework without help?

Could subject do his/her own laundry (wash/dry) without help?

Could subject prepare his/her own meals without help?

Could subject use the telephone without help?

Could subject take his/her own medications without help?

Could subject feed himself/herself without help?

Could subject dress himself/herself without help?

Could subject bathe himself/herself without help?

Could subject use public transportation to get places without help?

Could subject walk to places in his/her neighborhood without help?

Could subject walk without falling?

Could subject walk without help?

Could subject comb hair without help?

Could subject transfer between chairs without help?

Could subject get in and out of bed without help?

Could subject use toilet/commode without help?

Could subject's care still be provided at home?

INDEPENDENCE SCALE

Please indicate the most accurage current level of subject's independence (only _0 or _5 selections are acceptable)

100: No special care needed
090: No physical care needed if difficult tasks are avoided
080: Pre-disease level of employment changes or ends; cannot perform household chores to pre-disease level, may need help with finances
070: Self-care maintained for bathing, limited household duties (cooking and use of knives), driving terminates; unable to manage finances
060: Needs minor assistance in dressing, toileting, bathing; food must be cut for patient
050: 24-hour supervision appropriate; assistance required for bathing; eating, toileting
040: Chronic care facility needed; limited self feeding, liquified diet
030: Patient provides minimal assistance in own feeding, bathing, toileting
020: No speech, must be fed
010: Tube fed, total bed care

FUNCTIONAL CAPACITY

OCCUPATION
0 = unable
1 = marginal work only
2 = reduced capacity for usual job
3 = normal

FINANCES
0 = unable
1 = major assistance
2 = slight assistance
3 = normal

DOMESTIC CHORES
0 = unable
1 = impaired
2 = normal

ADL
0 = total care
1 = gross tasks only
2 = minimal impairment
3 = normal

CARE LEVEL
0 = full time skilled nursing
1 = home or chronic care
2 = home

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LEGEND TO THE VIDEOTAPE

Demonstration of the motor examination of the Unified Huntington's Disease Rating Scale and examples of each grade of abnormality.

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