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Cognitive deficits in Huntington's disease on the Repeatable Battery for the Assessment of Neuropsychological Status

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Abstract

Huntington's disease (HD) is associated with a variety of cognitive deficits, as well as motor and psychiatric disturbances. As clinical trials for HD evolve, briefer screening instruments will be needed to determine cognitive effects of interventions. The Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) may fill this gap. Seventy-five participants diagnosed with HD were evaluated with the RBANS, as well as several other scales typically used in HD. RBANS performances for these participants fell significantly below expectations for the Total Scale score, all five Indexes, and 11 of the 12 individual subtests. Cognitive scores on the RBANS were also significantly related to other markers of HD, including motor abnormalities, functional abilities, and other cognitive scores. Although additional research is needed, the current study supports the clinical applicability of the RBANS in patients with HD.

Introduction

Huntington's disease (HD) is an autosomal dominant, progressive neurodegenerative disorder due to an expanded CAG (cytosine-adenine-guanine) repeat on chromosome 4, which leads to neuronal death in the striatum (Aylward, et al., 2000; Aylward, et al., 1997; Aylward, et al., 2003) and parts of the cortex (Nopoulos, et al., 2007; Paulsen, Magnotta, et al., 2006; Rosas, et al., 2002). Although HD is typically diagnosed by motor abnormalities (e.g., chorea, oculomotor dysfunction, decreased motor speed), cognitive and psychiatric disturbances can be identified early in the disease (Paulsen & Conybeare, 2005; Paulsen, Ready, Hamilton, Mega, & Cummings, 2001) and lead to greater functional disability (Hamilton, et al., 2003; Marder, et al., 2000; Rothlind, Bylsma, Peyser, Folstein, & Brandt, 1993). The psychiatric symptoms associated with HD include depression, obsessive-compulsiveness, and apathy (Beglinger, et al., 2007; Paulsen, et al., 2005; Paulsen, et al., 2001).

Cognitive impairments in HD are observed on tests of attention, verbal fluency, psychomotor speed, executive functioning, learning and memory, emotional processing, and visuospatial functioning (Beglinger, et al., 2005; Brandt, et al., 1996; Brandt, Leroi, O'Hearn, Rosenblatt, & Margolis, 2004; Butters, Wolfe, Granholm, & Martone, 1986; Filoteo, et al., 1995; Henry, Crawford, & Phillips, 2005; Ho, et al., 2003; Massman, Delis, Butters, Levin, & Salmon, 1990; Montoya, et al., 2006; Paulsen & Conybeare, 2005; Roman, et al., 1998; A. E. Rosser & J. R. Hodges, 1994; Snowden, Craufurd, Griffiths, Thompson, & Neary, 2001; Stout & Johnson, 2005). These cognitive changes typically develop gradually; however, at later stages of the disease, the cognitive decline can progress more rapidly (Montoya, et al., 2006). As the disease progresses, it may become difficult to comprehensively evaluate the cognitive

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functioning of patients with HD with lengthy batteries and briefer screening measures of cognitive abilities are needed.

The Repeatable Battery for the Assessment of Neurological Status (RBANS) (Randolph, 1998) is a brief cognitive screening battery that has been used to evaluate neuropsychological functioning in a wide range of neuropsychiatric conditions, but, to our knowledge, only one study (Randolph, Tierney, Mohr, & Chase, 1998) has used this battery to evaluate patients with HD. In this study, Randolph et al. (1998) compared a small group of patients with HD to patients with Alzheimer's disease (AD) and healthy controls on the RBANS. The HD patients performed worse on the Attention and Visuospatial/Constructional Indexes compared to AD patients and worse on all five Indexes compared to controls. Although this study provided preliminary support for the RBANS in characterizing the cognitive deficits associated with AD and HD, it had a number of limitations. First, only 20 patients with HD were examined, which questions the generalizability of the data to other patients with HD. Second, besides being diagnosed with HD, very little information was presented that characterized the HD patients (e.g., severity of motor dysfunction, psychiatric symptoms, performance on other cognitive tests). Lastly, in their original study, Randolph et al. only presented Index scores and no data has been presented on the twelve individual subtests of the RBANS. Therefore, the purpose of the present study was to further validate the RBANS as a tool to assess cognitive functioning in a larger, better-characterized sample of HD patients. In addition, relationships between RBANS scores and other markers of HD will be examined. Finally, comparisons will be made to other relevant clinical studies in the literature that used the RBANS. As clinical trials in HD expand, a well-validated cognitive measure could be useful in determining the effectiveness of treatments.

Methods

Participants and Procedures

Seventy-five participants were recruited from the HD clinic at the University of Iowa's Huntington's Disease Society of America Center of Excellence. All participants included in the current analyses were diagnosed with HD by a movement disorder specialist and were rated as 90% or greater confidence that the motor abnormalities were due to HD. All participants in the current study provided informed consent prior to data collection, giving permission for their clinical data to be used for research purposes. All participants were individually evaluated by a clinical research team with the following measures.

Measures

The Unified Huntington's Disease Rating Scale (UHDRS) (Huntington Study Group, 1996) is a clinical research tool for examining motor, cognitive, psychiatric, and daily functioning in patients with HD. Several summary measures from the UHDRS were used as outcome variables: Total Motor score, Total Functional Capacity, and Total Psychiatric score. Briefly, the Total Motor score is the sum of the ratings across 31 different motor items (e.g., ocular pursuit, finger taps, chorea), and ranges from 0 to 124, with higher scores indicating more impaired motor functioning. The Total Functional Capacity score (Shoulson, Kurlan, & Rubin, 1989) assesses a patient's ability to perform both basic and instrumental activities of daily living, which is derived from reports of the patient and his/her companion, and ranges from 0 to 13, with higher scores indicating more intact functioning. The Total Psychiatric score (Beglinger, et al., 2007) is the sum of the product of frequency and severity for 11 psychiatric symptoms (e.g., anxiety, hallucinations, depression), and ranges from 0-176, with higher scores indicating increased psychiatric symptoms. In addition to the other measures, three cognitive tests are part of the UHDRS: phonemic fluency, Symbol Digit Modalities, and Stroop Color Word Test. Phonemic fluency reflects the number of correct words produced across three 1-

minute trials. Symbol Digit Modalities reflects the number of correct items produced in 90 seconds. Lastly, the number of correct items on the interference trial of the Stroop produced in 45 seconds is used. For all three cognitive tests, higher scores reflect better cognitive abilities.

The RBANS is a brief, individually administered test measuring attention, language, visuospatial/constructional abilities, and immediate and delayed memory. It consists of twelve subtests, which yield five Index scores and a Total Scale score. Normative information from the manual, which is used to calculate the Index and Total scores, is based on 540 healthy adults who ranged in age from 20-89 years old. The Index and Total scores are age-corrected standard scores (M = 100, SD = 15). All subtests were administered and scored as defined in the manual, with the exception of the Figure Copy and Figure Recall, which are further described elsewhere (Duff, et al., 2007). Briefly, the original scoring criteria were modified to be less stringent, which leads to more “normal” scores in healthy samples.

In addition to the RBANS, a number of other neuropsychological measures were collected on these patients, including the Wide Range Achievement Test-3 (WRAT-3) (Wilkinson, 1993) Reading Subtest to estimate premorbid intellect (standard score), Trail Making Test (TMT) (Reitan, 1955) Parts A and B to assess visual scanning and set shifting (seconds to complete), and the Beck Depression Inventory – II (BDI-II) (Beck, Steer, & Brown, 1996) to assess depression (raw score). However, not all participants completed all tests due to time restraints, participant fatigue, or severity of cognitive impairments.

Data Analyses

Primary analyses—To validate the utility of the RBANS in patients with HD, one sample t-tests were used to compare Index scores of the HD patients to an expected value of 100 (i.e., age-corrected score at the 50^{th} percentile). Since the twelve subtests of the RBANS are not routinely corrected for age, z-scores were generated for these scores for all participants by comparing them to their appropriate normative data (e.g., all participants who were under 40 were compared to the RBANS 20 – 39 year old normative group, all participants who were 40 – 49 were compared to the RBANS 40 – 49 year old normative group, and so on). One sample t-tests were also used to compare subtest age-corrected z-scores to expected value of 0 (i.e., age-corrected z-score at the 50^{th} percentile). Cohen's d, a measure of effect size, was calculated from the respective means and standard deviations. Finally, Pearson correlations were calculated between RBANS scores and other markers of HD (e.g., Total Motor score, Total Functional Capacity, Total Psychiatric score, and three cognitive scores of the UHDRS) and the other neuropsychological measures. Due to the number of comparisons, an alpha of 0.01 was used throughout.

Secondary analyses—Since no control data is presented, comparisons were made between the RBANS performances of the current HD sample and other relevant clinical studies in the existing literature. Specifically, the means and standard deviations for patients with Alzheimer's disease (AD) (Duff, et al., 2008), Parkinson's disease (PD) (Beatty, et al., 2003), and Multiple Sclerosis (MS) (Beatty, 2004) were compared with our HD patients, which yielded estimated t-tests and effect sizes. Additional details about these other clinical samples can be obtained from the original articles.

Results

Primary analyses

The HD participants in the current sample were nearly evenly distributed for gender (36 male and 39 female), with a mean age of 46.8 (SD = 12.9) years and a mean education of 13.3 (SD = 2.3) years. All participants with genetic testing had CAG repeat lengths in the expanded

range (i.e., >38, mean CAG repeat = 46.3 [SD = 5.8]). Although only 30 of our 75 participants had genetic testing to confirm the expanded CAG length, we have little doubt about those without genetic testing. The neurologists in our clinic typically order the genetic test only if the clinical exam is unclear. So those participants without genetic testing were likely clearer cases of HD on examination. Additionally, participants with and without genetic testing were comparable on all major indices from the UHDRS (e.g., Total Motor score, Total Functional Capacity, Total Psychiatric score, three cognitive tests), RBANS Total score, and BDI-II (all p's>0.05). Motor and psychiatric functioning was variable for the group, but clearly suggested impairments consistent with HD (UHDRS Total Motor mean = 30.5 [SD = 16.0], Total Behavioral mean = 19.2 [SD = 17.9]). Functional disability was also present in this sample (UHDRS Total Functional Capacity mean = 9.3 [SD = 3.4]). Premorbid intellect was estimated to be in the average range (WRAT-3 Reading mean = 91.7 [SD = 14.6]). Compared to this premorbid estimate and normative data, UHDRS cognitive measures were below expectations (phonemic fluency mean = 18.8 [SD = 12.8], Symbol Digit Modalities mean = 27.3 [SD = 12.4], Stroop interference mean = 26.9 [SD = 12.6]).

One sample t-tests for the RBANS Indexes indicated that all fell significantly below 100 (see Table 1). Additionally, 11 of the 12 individual subtests also fell significantly below the expected value of 0 (see Table 1). With the exception of two subtests, effect sizes (i.e., Cohen's d) for all other RBANS Indexes and subtests were large.

Relationships between the RBANS Indexes and other markers of HD are presented in Table 2. Briefly, several important clinical characteristics of HD were significantly correlated with the Total Scale score of the RBANS, including Total Motor (r = -0.47, p=.001), Total Functional Capacity (r = 0.52, p<.001), and all three UHDRS cognitive tests (phonemic fluency r = 0.74, p<.001; Symbol Digit Modalities r = 0.77, p<.001; Stroop interference r = 0.59, p<.001). Similar relationships existed between the RBANS Indexes and subtests and these markers of HD.

Secondary analyses

The means and standard deviations of the RBANS Indexes for the HD sample are reiterated in Table 3, as are the RBANS scores for the samples of AD, PD, and MS. Individual subtest scores can be obtained from the first author or from the original articles. Compared to patients with AD, the participants with HD perform significantly poorer on the Attention Index of the RBANS (p<0.01, d=0.56), whereas patients with AD performed significantly worse on Immediate Memory (p=0.02, d=0.39), Delayed Memory (p<0.01, d=1.02), and Total Scale (p=0.04, d=0.34) scores. AD and HD differences occurred on 9 of the 12 RBANS subtests, with HD subjects performing significantly better than the AD subjects on all 9 subtests (List Learning [p<0.01, d=0.96], Story Memory [p<0.01, d=0.59], Picture Naming [p<0.01, d=0.56], Semantic Fluency [p<0.01, d=0.55], Coding [p=0.05, d=0.33], List Recall [p<0.01, d=1.56], List Recognition [p<0.01, d=0.94], Story Recall [p<0.01, d=1.74], Figure Recall [p<0.01, d=2.07]).

Using the sample of patients with PD with dementia from Beatty et al. (2003), HD patients performed comparably on all RBANS Indexes. PD patients without dementia, however, performed significantly better than the HD patients on nearly all RBANS Indexes (Immediate Memory [p=0.02, d=0.54], Visuospatial Constructional [p<0.01, d=0.83], Language [p<0.01, d=0.76], Attention [p<0.01, d=1.35], Total Scale [p<0.01, d=0.89]; Delayed Memory was not significantly different; individual subtest scores were not reported in this article).

Beatty et al. (2004) similarly split MS patients into groups of those with and without cognitive impairments. Not surprisingly, MS patients without cognitive impairments outperformed our HD patients on all RBANS Indexes (Immediate Memory [p=<0.01, d=1.39], Visuospatial

Constructional [p<0.01, d=1.46], Language [p<0.01, d=0.95], Attention [p<0.01, d=1.42], Delayed Memory [p<0.01, d=1.07], Total Scale [p<0.01, d=1.57]). MS patients with cognitive impairments still outperformed HD patients on 3 of the RBANS Index scores (Visuospatial Constructional [p<0.01, d=1.29], Attention [p<0.01, d=1.13], Total Scale [p=0.04, d=0.77]). Individual subtest scores were also not reported in Beatty et al. (2004).

Discussion

The cognitive deficits in HD have been widely documented (Beglinger, et al., 2005; Brandt, et al., 2004; Butters, et al., 1986; Filoteo, et al., 1995; Henry, et al., 2005; Montoya, et al., 2006; Paulsen & Conybeare, 2005; A. Rosser & J. R. Hodges, 1994; Rothlind, et al., 1993; Snowden, et al., 2001; Stout & Johnson, 2005), and HD remains an important disorder for neuropsychology, as it represents a protypical "subcortical" dementia. However, as clinical trials continue to advance for this disease, brief yet sensitive batteries will be needed to measure cognitive functioning in these patients. The RBANS might fill this important gap.

Consistent with the preliminary report of Randolph et al. (1998), patients with HD from the current study performed poorly on the RBANS. The cognitive deficits were severe (e.g., Total Scale score averaging at the 2nd percentile, large effect sizes for the majority of RBANS measures), and pervasive (e.g., scoring significantly below expectations on the Total Scale score, all five Indexes, and 11 of the 12 subtests). The HD patients in the current sample, however, appeared more impaired than those reported by Randolph et al. (e.g., Total Scale mean = 69.7 vs. 84.9, respectively), which might have lead to differences in the findings of the two studies.

Despite the significant deficits on the RBANS by patients with HD, the current data cannot determine if this reflects a true "cognitive footprint" of HD or if this reflects a more general neuropsychological profile of patients with a variety of neurodegenerative disorders. The secondary analyses in the current study can only begin to address this question. First, the RBANS appears able to adequately separate classical "cortical" and "subcortical" cognitive profiles. The HD patients in the current study performed very differently than the AD patients of Duff et al. (2008), but very similarly to the PD with dementia patients of Beatty et al. (2003). Although this distinction of cortical/subcortical profiles is not definitive within neuropsychology (Arango-Lasprilla, et al., 2006; Salmon & Filoteo, 2007), two of the subcortical conditions (i.e., HD and PD) were separated from the cortical condition (i.e., AD) on the RBANS. The patients with MS, which also reflect a more prominent subcortical condition, were considerably higher functioning than the HD patients, although the two groups did perform comparably on tasks of immediate and delayed memory and language. The inclusion of additional cognitive testing (e.g., executive functioning) and psychiatric measures (e.g., depression) may have further separated these groups. Second, higher functioning patients with neurodegenerative disorders (e.g., PD without dementia, MS without cognitive impairment) did perform significantly better than the patients with HD in the current sample on nearly all Indexes of the RBANS. This suggests that the RBANS may be an appropriate tool to track cognitive decline in these disorders, although additional data is needed in milder cases of HD and AD. Third, there does not appear to be sufficient evidence to clearly identify a "cognitive footprint" of HD on the RBANS. As noted earlier, the RBANS seems to adequately separate patients with HD from patients with AD, but it does not appear to be able to separate patients with HD from patients with PD with dementia. The one caveat is that only Index scores were available to compare the HD patients with the PD and MS patients. It remains possible that individual subtest differences on the RBANS do exist to separate these groups of subcortical patients.

As noted earlier, HD has typically been viewed as a "subcortical" dementia (Bonelli & Cummings, 2008), which is contrasted with "cortical" dementias as having slower psychomotor speed, relatively better recognition memory compared to recall, and more prominent executive dysfunction. Some of these subcortical characteristics can be examined with the RBANS. For example, speeded tasks on the RBANS include Semantic Fluency and Coding. Compared to normative data, these two subtests had the largest effect sizes (1.6 and 2.2, respectively), which suggests a disproportionate deficit on these tasks. A comparison of List Recall and List Recognition provides some conflicting information about this indicator of subcortical dysfunction. List Recall of 10 words after a brief delay averaged 3.6 words for the HD patients, whereas List Recognition averaged 17.6 correct (out of a possible 20 [10 hits, 10 correct rejections]). Further examination of the List Recognition scores indicated that participants averaged 8.4 (SD = 1.9) "hits" and 0.8 (SD = 1.4) "false positive errors." Looking solely at these raw scores, it appears that recognition performance is relatively better than recall (i.e., only 3 words recalled, but 8 correctly identified on recognition with few false positives). Looking solely at the effect sizes of difference from theoretical 50^{th} percentile (Table 1), these two scores appear comparable (List Recall d = 1.2; List Recognition d = 1.0). Looking solely at age-corrected z-scores based on normative data, List Recall appears less impaired than List Recognition (z = -1.5 and -2.8, respectively). However, we remind the readers that the normative values of these two different subtests can skew the resulting z-scores and effect sizes. For example, the normative data on 40 – 49 year olds (i.e., the modal age in the current sample) for List Recall is M = 6.3, SD = 1.9 and for List Recognition is M = 19.7, SD = 0.6. Given these normative values, the worst possible performance on the List Recall subtest (i.e., 0 correct) for 40 – 49 year olds is z = -3.3, whereas very poor performances on the List Recognition subtest yield much worse standardized scores (e.g., 0 correct [out of 20] = z of -32.8; 10 correct = z of -16.2; 15 correct = z of -7.8). The RBANS List Recognition subtest is not unique in this regard, as many recognition trials on memory tests (e.g., Brief Visuospatial Memory Test – Revised, Hopkins Verbal Learning Test – Revised, Wechsler Memory Scale - III) do not provide standardized scores for total correct but instead give some general indication of intact/impaired (e.g., > or < 16^{th} percentile). Given this potential skewness of z-scores and effect sizes, we cautiously interpret the raw scores to suggest that List Recognition is relatively better than List Recall, which is consistent with other memory studies in HD (Butters, et al., 1986; Massman, et al., 1990; Montoya, et al., 2006). Lastly, executive dysfunction cannot be directly assessed with the RBANS, as it contains no subtests that purport to measure this deficit. Nonetheless, as can be seen in Table 2, most RBANS Indexes and subtests strongly correlated with Trail Making Test Part B, a commonly identified measure of executive functioning. Overall, the RBANS appears able to evaluate many of these indicators of a "subcortical profile," even if they are not specific to HD.

As with other neuropsychiatric disorders, the presentation of HD varies considerably between individuals (Rosas, et al., 2008). Some individuals present with more noticeable motor dysfunction, whereas others present primarily with cognitive or psychiatric disturbances. It is informative to see how these symptoms of HD co-occur. As observed in the correlational analyses, the cognitive difficulties observed on the RBANS appear to vary with the motor abnormalities and functional declines in patients with HD. It is also interesting to note that RBANS performances did not vary with psychiatric symptoms, either the UHDRS Total Psychiatric score or the BDI-II. A lack of association with psychiatric symptoms might actually make the RBANS a good choice for clinical trials in HD, where depression and obsessive-compulsiveness are common (Beglinger, et al., 2007; Paulsen, et al., 2005). If the RBANS is to be considered for clinical trials in HD, then it possesses other key features of adequate neurocognitive endpoints, such as the availability of alternate forms (e.g., four forms available through test publisher), brevity of administration, and availability of translations for multinational studies (e.g., translated into over 20 languages).

Despite the supportive findings for using the RBANS in patients with HD, some limitations should be mentioned. First, we did not utilize a control group in the current study, and future studies should attempt to find appropriate comparison participants. Although not ideal, comparing these patients to expectations based on age-matched normative data is more analogous to clinical practice. The lack of a control group also prevents the calculation of sensitivity and specificity values for the RBANS in our HD sample. Although these test characteristics have been reported for HD patients in an earlier version of the RBANS (Mohr, Walker, Randolph, Sampson, & Mendis, 1996), future studies are encouraged to replicate this work. Second, in recruiting patients in our HD clinic, we excluded individuals that were too impaired to complete the RBANS or not impaired enough to warrant a diagnosis based on motor functioning. In this way, our results likely apply to a "window" of HD, rather than the entire spectrum. Third, there are other biomarkers of HD that were not collected as part of routine practice in our HD clinic. For example, future studies might also examine the relationship between the RBANS and neuroimaging (especially the striatum). As noted earlier, only a subset of our sample were genetically confirmed to have the CAG expansion, and future studies might remove any doubt about diagnosis by having all participants undergo this testing. Finally, the RBANS was developed as a screening battery, and more comprehensive neuropsychological batteries will likely detect even more subtle cognitive difficulties in HD and pre-diagnosed cases (Paulsen, Hayden, et al., 2006; Paulsen, et al., 2007). Regardless of these limitations, the current study provides additional support for using the RBANS to assess cognition in patients with HD, and future studies might extend these findings with longitudinal comparisons of HD patients.

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Table 1

RBANS Indexes and subtest scores in the HD sample

RBANS score	Mean (SD)	Range	t (df) p	d
Immediate Memory	70.7 (20.7)	40 – 109	-12.2 (74) <.001	1.7
List Learning	20.5 (7.4)	2 – 36	-9.3 (74) <.001	1.3
Story Memory	11.3 (5.2)	0 – 22	-10.8 (74) <.001	1.5
Visuospatial/Constructional	78.1 (17.6)	50 – 116	-10.6 (71) <.001	1.4
Figure Copy	14.1 (4.0)	2 – 20	-8.9 (71) <.001	1.4
Line Orientation	14.4 (4.4)	2 – 20	-3.6 (74) .001	0.5
Language	82.3 (13.2)	40 – 122	-11.6 (74) <.001	1.3
Picture Naming	9.5 (0.8)	7 – 10	-0.6 (74) .56	0.1
Semantic Fluency	13.7 (4.8)	6 – 28	-12.8 (74) <.001	1.6
Attention	67.7 (17.7)	40 – 106	-15.6 (72) <.001	2.0
Digit Span	8.4 (2.3)	4 – 15	-9.0 (74) <.001	1.0
Coding	26.8 (12.4)	0 – 53	-15.6 (72) <.001	2.2
Delayed Memory	74.5 (2.3)	40 – 124	-9.7 (71) <.001	1.4
List Recall	3.6 (2.8)	0 – 10	-8.9 (74) <.001	1.2
List Recognition	17.6 (2.5)	9 – 20	-6.2 (74) <.001	1.0
Story Recall	5.6 (3.1)	0 – 12	-10.5 (74) <.001	1.4
Figure Recall	10.0 (4.6)	1 – 20	-7.6 (71) <.001	1.1
Total Scale	69.7 (17.2)	43 – 114	-14.8 (70) <.001	1.9

Note. Index scores are left justified and age-corrected using the standardization sample from the RBANS manual. Subtest scores are indented and are raw scores. Means and standard deviations (in parentheses) are presented in the second column. t = t-value from one sample t-test. df = degrees of freedom. p = p-value. d = Cohen's d.

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Table 2

Correlations between RBANS scores and other markers of Huntington's disease

RBANS score	Motor	Psych	Function	Phonemic fluency	SDMT	Stroop	WRAT Reading	TMT-A	TMT-B	BDI-II
Immediate Memory	-.50**	.02	.52**	.68**	.70**	.53**	.54*	-.44*	-.50*	-.13
List Learning	-.54**	0.2	.62**	.62**	.68**	.53**	.42	-.50*	-.46*	-.13
Story Memory	-.38*	.02	.46**	.69**	.66**	.56**	.67**	-.44*	-.50*	-.15
Visuospatial/Constructional	-.26	.09	.40*	.48**	.50**	.47**	.40	-.30	-.43*	-.20
Figure Copy	-.42**	.05	.53**	.46**	.57**	.52**	.39	-.52**	-.35	-.20
Line Orientation	-.21	-.06	.34*	.36*	.38*	.37*	.29	-.37*	-.40*	-.30
Language	-.38*	.11	.39*	.54**	.56**	.37*	.32	-.44*	-.61**	-.17
Picture Naming	-.20	.08	.29	.16	.17	.10	.16	-.13	-.45*	.03
Semantic Fluency	-.44**	-.01	.44**	.65**	.63**	.53**	.40	-.53**	-.53*	-.30
Attention	-.43**	-.04	.46**	.75**	.78**	.60**	.47*	-.55**	-.60**	-.21
Digit Span	-.31*	-.07	.35*	.57**	.58**	.44**	.46*	-.47**	-.46*	-.22
Coding	-.66**	.09	.61**	.83**	.92**	.78**	.42	-.72**	-.72**	-.17
Delayed Memory	-.42**	.00	.38*	.62**	.67**	.49**	.43*	-.51**	-.56**	-.16
List Recall	-.41**	-.03	.49**	.64**	.70**	.57**	.42	-.49**	-.55**	-.20
List Recognition	-.32*	-.05	.47**	.49**	.55**	.44**	.47*	-.48**	-.47*	-.19
Story Recall	-.43**	.00	.37*	.64**	.66**	.56**	.58**	-.42*	-.61**	-.12
Figure Recall	-.36*	.05	.50**	.57**	.60**	.56**	.45*	-.47*	-.49*	-.19
Total Scale	-.47**	.03	.52**	.74**	.77**	.59**	.51*	-.53**	-.63*	-.21

Note. Motor = UHDRS Total Motor score, Psych = UHDRS Total Psychiatric score, Function = UHDRS Total Functional Capacity score, SDMT = Symbol Digit Modalities Test, Stroop = Stroop Interference trial, TMT = Trail Making Test, WRAT Read = Wide Range Achievement Test – 3 Reading subtest, BDI-II = Beck Depression Inventory – II,

* = p<.01,

** = p<.001.

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Table 3

RBANS Indexes in the HD and other clinical samples

RBANS score	HD	AD	PDD	PDND	MS impaired	MS intact
Immediate Memory	70.7 (20.7)	63.9 (13.1)*	66.7 (12.0)	81.6 (17.5)*	79.1 (14.3)	96.4 (14.7)**
Visuospatial/Constructional	78.1 (17.6)	82.6 (16.8)	70.9 (18.3)	92.5 (16.9)**	101.3 (21.7)**	102.7 (15.7)**
Language	82.3 (13.2)	81.1 (12.8)	86.4 (10.3)	91.6 (8.1)**	90.6 (11.7)	94.0 (10.9)**
Attention	67.7 (17.7)	77.9 (18.5)**	69.3 (13.6)	91.0 (15.9)**	87.8 (17.9)**	91.7 (15.5)**
Delayed Memory	74.5 (2.3)	54.7 (15.8)**	71.1 (16.3)	83.4 (16.4)	77.6 (22.8)	95.5 (14.9)**
Total Scale	69.7 (17.2)	64.5 (13.1)*	65.9 (11.0)	84.4 (13.6)**	83.1 (19.2)*	94.5 (13.4)**

Note. Means and standard deviations (in parentheses) are presented. HD = Huntington's disease sample from the current study. AD = Alzheimer's disease from Duff et al. (2008). PDD and PDND = Parkinson's disease with dementia and Parkinson's disease no dementia (respectively) from Beatty et al. (2003). MS = Multiple sclerosis from Beatty et al. (2004). All comparisons are with HD group.

* = $p<0.05$,

** = $p<0.01$.

Individual subtest differences can either be obtained from the first author or from the original articles.