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Self-awareness of Motor Dysfunction in Patients with Huntington's Disease in Comparison to Parkinson's Disease and Cervical Dystonia

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Abstract

Individuals suffering from Huntington's disease (HD) have been shown to present with poor self-awareness of a variety of symptoms. The aim of this study was to better assess the self-awareness of motor symptoms and activities of daily living (ADL) impairment in HD, in comparison to Parkinson's disease (PD) and cervical dystonia (CD). In particular, the anosognosia/anosodiaphoria of involuntary movements has been investigated. Self-awareness was tested in 23 patients with HD by comparing patient and caregiver ratings in reference to clinical control groups (25 PD with dyskinesias, PDdys; 21 PD without dyskinesias, PDndys; and 20 with CD). Patients were assessed neurologically by relevant rating scales. Self-awareness was tested using a scale based on 15 films demonstrating 3 types of motor symptoms (chorea/dyskinesias, parkinsonism, torticollis) as well as the Self-Assessment Parkinson's Disease Disability Scale. General cognitive status, verbal learning, cognitive control, and mood were also analyzed. Our results indicate that self-awareness of choreic movements was affected more severely in HD than in PDdys, despite comparable cognitive status. Patient–proxy agreement on ADL impairment was roughly similar in all clinical groups. The results are discussed in the context of orbitofrontal–limbic pathology as a potential trigger of anosognosia/anosodiaphoria in individuals with HD. (*JINS*, 2011, *17*, 788–795)

Keywords: Movement disorders, Dementia, Chorea, Dyskinesias, Cervical dystonia, Awareness

INTRODUCTION

Huntington's disease (HD) is an autosomal dominant neurological disease, characterized by motor, cognitive and psychiatric features (Craufurd & Snowden, 2002; Kremer, 2002; White, Vasterling, Koroshetz, & Myers, 1992). Chorea in HD significantly impairs patients' activities of daily living (ADL). However, poor quality of life in HD is directly associated with functional decline and not the severity of motor symptoms alone (Ho, Gilbert, Mason, Goodman, & Barker, 2009). Importantly, alike patients with HD, also individuals with Parkinson's disease (PD) and cervical dystonia (CD) develop involuntary movements that have been linked to the dysfunction of basal ganglia circuits (see: Draganski, Thun-Hohenstein, Bogdahn, Winkler, & May, 2003; Jankovic, 2005). Moreover, choreic movements, typical and quite stable during the day in HD, appear also in advanced PD as a disease-specific drug-induced dyskinesias (Schrag, Jahanshahi, & Quinn, 2000), associated with the "on" state and with better motor function (Jankovic, 2005). Thus, since patients with HD, PD, and CD present with motor symptoms discernible to a neurologically naive observer, they may all suffer because of social stigmatization.

Of interest, it has been recently shown that, despite marked and progressive disability, patients with HD frequently exhibit limited self-awareness of motor, cognitive, and psychiatric symptoms (Deckel & Morrison, 1996; Ho, Robbins, & Barker, 2006; Hoth et al., 2007; Snowden, Craufurd,

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Griffiths, & Neary, 1998; Vitale et al., 2001), a phenomenon known as anosognosia or, in less severe cases, as anosodiaphoria (see: Heilman & Harciarek, 2010). Moreover, there is evidence to suggest that poor insight may even precede clinical diagnosis of HD (Duff et al., 2010). By comparison, in PD, patient–proxy agreement is typically satisfactory in non-demented patients with regard to motor disability, executive function, and quality of life (QoL) (Fleming, Cook, Nelson, & Lai, 2005; Leritz, Loftis, Crucian, Friedman, & Bowers, 2004; Martínez-Martín et al., 2003; Mathias, 2003; McRae, Diem, Vo, O'Brien, & Seeberger, 2002). Of note, self-awareness of motor symptoms in CD has never been investigated.

Nonetheless, to date, there is only one comparative study addressing the question of unawareness of dyskinesias in HD and PD (Vitale et al., 2001). The results of this study indicate that in PD the unawareness seems to be inversely related with severity of dyskinesias, while in HD it is directly related to disease duration and its severity. The interpretation of these findings is, however, limited by small sample sizes of both subject with HD and PD. Thus, it remains unclear whether cognitively comparable patients with HD and PD are similarly (un)aware of choreic movements, as underestimation of such movements was reported in HD (Snowden et al., 1998; Vitale et al., 2001), PD (Vitale et al., 2001), drug-induced dyskinesias in schizophrenia (Caracci, Mukherjee, Roth, & Decina, 1990), and in choreas other than HD (Shenker, Wylie, Fuchs, Manning, & Heilman, 2004).

Several factors are believed to contribute to poor self-awareness of symptoms in HD: (1) cognitive: lower cognitive status, memory impairment, cognitive control deficits; (2) emotional: avoidance coping strategies, mood; (3) disease related: disease severity and sensory deficit (the latter in case of awareness of motor symptoms) (see: Hoth et al., 2007; Snowden et al., 1998).

This study aimed at assessing self-awareness of motor symptoms and ADL impairment in HD in comparison to PD (both with dyskinesias, PDdys; and without them, PDndys) and CD. A direct comparison of patients with HD and PDdys (with equal cognitive status) enabled assessing perception of choreic movements as an early, persistent, and core symptom (in HD), and as a late and variable symptom (PDdys). Also, by comparing individuals with PD who did with those who did not develop dyskinesias, we investigated the perception of parkinsonism at different disease stages. PDndys and CD served as reference groups. Our study used a larger N than previous studies for HD/PD comparisons, a video basis for judgment of motor symptoms instead of questionnaires/interview that were previously applied (Snowden et al., 1998; Vitale et al., 2001) and control populations to verify the following hypotheses.

Based on the clinico-pathological features of each of these conditions, we expected that, if anosognosia is a product of prefrontal pathology deficits in self-awareness of the disease-specific symptoms should be primarily identified in HD and PDdys groups. If so, anosognosia/anosodiaphoria of motor symptoms would be expected to correlate with the performance on cognitive measures of dorsolateral prefrontal function. Alternatively, self-awareness of symptoms may be more deficient in HD than in PD, as it seems to be one of the early symptoms of HD (Duff et al., 2010), probably resulting from a more widespread neurodegeneration process encompassing frontostriatal and orbitofrontal–limbic pathways (Douaud et al., 2009). Such lesions may, in turn, lead to a failure to attach significant negative value to particular impairments in behavior.

Summing up the study aimed at assessing self-awareness of symptoms in HD with reference to PD and CD with the emphasis on the self-awareness of choreic movements in HD and elucidating the underlying causes of poor self-awareness of symptoms in HD.

METHODS

Procedure

The patients were recruited from a specialty outpatient Movement Disorders Clinic and Dystonia Center in St. Adalbert Hospital in Gdansk, Poland. All consecutive patients with the diagnosis of HD, PD, or CD were asked to participate in the study by the examiner trained in movement disorders (by J.S., W.S., or M.S.). The duration of patients' recruitment was 8 months, and patients were enrolled if they agreed to participate and fulfilled the inclusion criteria during the neurological visit. All HD patients recruited for the study participated in the REGISTRY study by European Huntington's Disease Centre in Gdansk. All the participants volunteered for the study.

Clinical diagnosis, supported by neuroimaging (Computed Tomography or Magnetic Resonance Imaging) and laboratory tests, was established by a movement disorders specialist according to the broadly accepted clinical criteria for PD (Litvan et al., 2003), HD (Kremer, 2002), and CD (Albanese et al., 2006). In HD, the diagnosis was in all cases confirmed by genetic testing. Individuals were included if their Mini-Mental State Examination score was $\geq$20 points. Patients with concurrent neurological dysfunction or alcohol abuse were excluded from the study. Further exclusion criteria were the following: inability to complete the study protocol due to severe oculomotor/motor impairment (inability to remain in a sitting position during few hours and inability to read) and lack of proxy to provide ratings. Proxies were required to have spent most of their time with the patients and knew them well premorbidly. From the total of 97 patients who initially agreed to participate, 14 subjects were excluded from the study: 3 with HD (1 with severe thyroid dysfunction, 1 after rupture of intracranial aneurysm, 1 with history of alcohol abuse), 3 with PDdys (1 with severe dementia with MMSE score below 20, 1 without proxy, 1 after pallidotomy), 1 with PDndys (lack of proxy), and 7 with CD (1 after mild head trauma, 6 without proxy). The study was approved by Bioethic committee of the Medical University of Gdansk and conducted in accordance with the Helsinki Declaration.

The testing was performed during the day, in case of PD patients always in the "on" phase. Eighty-three patients were tested at the clinic (73 on the outpatient basis, 10 on the inpatient basis), while 6 individuals were tested at home. In all cases, the examination was performed in a quiet room with good lightning. The questionnaires were administered to the

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patients and their proxies independently. The patients filled in the questionnaires assessing motor, memory, and executive function before neuropsychological assessment to assess the general self-awareness of symptoms, and to avoid the confounding effect of the testing procedure on the patient's ratings.

Patients

Eighty-nine patients participated in the study (23 with HD, 25 with PDdys, 21 with PDndys patients, and 20 with CD). Group demographics and disease characteristics are presented in Table 1. The groups were matched in terms of sex and years of education. Due to the heterogeneity of treatment regimens only levodopa dosage in PD patients was calculated and is presented in Table 1.

Due to a difference in the average age of symptom onset between HD, PD, and CD, and owing to the fact that choreic movements are early symptoms in HD, whereas in PD, they occur after several years of levodopa treatment, our groups could not have been matched in terms of age and

Table 1. Demographics and disease characteristics of HD, PD, and CD patients

	HD $N = 23$ [a][1]	PDdys $N = 25$ [b]	PDndys $N = 21$ [c]	CD $N = 20$ [d]	F/H/t/U/χ^2 tests[2]
Demographics					
Age	49.83±11.12[3] [b,c]	65.68±10.03 [a,d]	64.67±7.59 [a,d]	51.75±12.98 [b,c]	$F(3;85) = 14.11$; $p < .001$
Education (years)	12[4]	12	13	12	H(3, $N = 89$) = 1.99; $p = 57$; s.i.
Male : female	14: 9	12 : 13	15 : 6	8 : 12	$\chi^2 = 4.92$; $p = .18$; s.i.
Disease characteristics					
Duration of disease	5 [b]	12 [a,c]	4 [b]	8 [—]	H (3, $N = 89$) = 28,76 $p < .0001$
UPDRS III	NA	22.04 (9.14)	18.29 (10.38)	NA	$t(44) = 1.31$ $p = 0.20$, s.i.
Daily levodopa dose	NA	1000	500	NA	$U = 90.00$; z = 3.04 $p = .002$
UHDRS motor	38.09 (±14.33)	NA	NA	NA	NA
TWSTRS severity	NA	NA	NA	15.55 (±6.41)	NA
MADRS	10.00 [—]	15.00 [c]	7.00 [b]	10.50 [—]	H(3, $N = 89$) = 11.47 $p = .0094$
Medication					
– Neuroleptics	$n = 23$	—	—	—	
– Antidepressants	$n = 19$	$n = 6$	$n = 4$	—	
– Myorelaxants	$n = 1$	—	—	$n = 1$	
– Levodopa	—	$n = 25$	$n = 20$	—	
– Dopamine agonists	—	$n = 18$	$n = 14$	—	
– Amantadine	—	$n = 11$	$n = 7$	—	
– Benzodiazepine derivatives	$n = 2$	$n = 3$	—	—	
– Botulinum toxin injections	—	—	—	$n = 20$	
– Acetylocholinesterase inhibitors	—	$n = 3$	—	—	
– Selegiline	—	—	$n = 3$	—	
Proxy					
– Partners	56%	72%	76%	60%	
– Children	9%	28%	10%	10%	
– Parents	17%	0%	0%	5%	
– Siblings	9%	0%	0%	20%	
– Friends	9%	0%	14%	5%	

Note. NA=not assessed; s.i. = statistically insignificant; MADRS = Montgomery-Asberg Depression Rating Scale; UHDRS = Unified Huntington's Disease Rating Scale; UPDRS = Unified Parkinson's Disease Rating Scale; TWSTRS = Toronto Western Spasmodic Torticollis Rating Scale; HD = Huntington's disease; PD = Parkinson's disease; CD = cervical dystonia; PDdys = PD with dyskinesias; PDndys = PD without dyskinesias.

[1]Letters a–d denote significant intergroup differences as indicated in the first row of the table.

[2]The differences between the two groups were analyzed either with *t*-unpaired test, U-Mann-Whitney test, or chi-square test. The differences among the four groups were tested either with one-way analysis of variance test with Scheffe *post hoc* comparisons or with H Kruskal-Wallis test with *post hoc* comparisons. Significant inter-group differences are indicated by a–d as indicated in the first row.

[3]Mean ± standard deviation is reported in case of normal data distribution.

[4]Median is reported in case of non-normal data distribution.

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Table 2. Neuropsychological assessment data of HD, PD, and CD patients

	HD $N = 23$ [a][1]	PDdys $N = 25$ [b]	PDndys $N = 21$ [c]	CD $N = 20$ [d]	F/H/t/U tests[2]
MMSE	26[3]	27	28	28.50	H(3, $N = 89$) = 14.48; $p = .02$; s.i
AVLT I-V	29.5±8.33[4] [c,d]	38.2±11.78 [—]	41.33±12.65 [a]	44.60±11.02 [a]	$F(3,85) = 7.54$ $p = .002$
AVLT delayed recall	5.52±2.69 [c,d]	7.68±3.24 [—]	8.67±2.99 [a]	9.90±3.68 [a]	$F(3,85) = 7.47$ $p = .002$
AVLT- % after delay	71.33±21.70 [—]	79.39±18.38 [—]	86.39±17.39 [—]	87.21±19.83 [—]	$F(3,85) = 3.18$ $p = .03$; s.i.
Stroop CWIT	0.10 [c,d]	0.11 [c,d]	0.02 [a,b]	0.02 [a,b]	H(3, $N = 87$) = 28.77 $p < .0001$

Note. AVLT = Auditory Verbal Learning Test; CWIT = Colour-Word Interference Test; MMSE = Mini-Mental State Examination; s.i. = statistically insignificant; HD = Huntington's disease; PD = Parkinson's disease; CD = cervical dystonia; PDdys = PD with dyskinesias; PDndys = PD without dyskinesias.
[1]Letters a–d denote significant intergroup differences as indicated in the first row of the table.
[2]The differences between two groups were analyzed either with *t*-unpaired test or Mann Whitney *U* test. The differences among the 4 groups were tested either with one-way analysis of variance test with Scheffe *post hoc* comparisons or with H Kruskal-Wallis test with *post hoc* comparisons. Significant inter-group differences are indicated by a–d as indicated in the first row.
[3]Median is reported in case of non-normal data distribution.
[4]Mean ± standard deviation is reported in case of normal data distribution.

disease duration. PDdys and PDndys groups differed in the presence of dyskinesia and severity of other PD symptoms. Montgomery Asberg Depression Rating Scale (MADRS) assessment identified more depressive symptoms in PDdys than in other groups. HD and PDdys groups were matched for cognitive status (see Tables 1 and 2).

Measures

Neurological assessment

Neurological examination comprised of the motor section from Unified Huntington's Disease Rating Scale (UHDRS) (Huntington Study Group, 1996) for HD, the Unified Parkinson's Disease Rating Scale (UPDRS) Part II-IV (Paulson & Stern, 1997) for PD and the Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) (Consky, Basinski, Bele, Ranawaya, & Lang, 1990) for CD. For PD groups, scores only scores from UPDRS III and for CD group only scores from TWSTRS-severity are reported in Table 1.

Assessment of the self-awareness of symptoms

Questionnaires filled in by both the patient and his/her proxy included: Motor Impairment Scale (MIS) based on a series of 15 films demonstrating different motor symptoms [5 from UHDRS (Reilmann et al., 2009), 5 from UPDRS (Goetz et al., 1995), and 5 from TWSTRS (Comella et al., 1997)] and Self-Assessment Parkinson's Disease Disability Scale (SPDDS) (Brown, MacCarthy, Jahanashi, and Marsden, 1989). In both the MIS and the SPDDS, higher scores correspond to greater impairment.

The MIS, assessing the severity of motor symptoms (chorea, core parkinsonian symptoms, torticollis) from a patient/caregiver perspective, was created specifically for the purposes of the current study. This scale was based on training films for neurologists (UHDRS items: chorea in trunk, lower limbs, upper limbs, face, and buccolingual area; UPDRS items: hand tremor, posture, bradykinesia, leg tremor, and gait; TWSTRS items: laterocollis, anterocollis, retrocollis, rotation, and shoulder elevation; see Appendix for item choice, testing procedure, reliability and validity data). The global score on the MIS ranges from 0 to 15, subscores for each of the assessed domains (choreic movements/dyskinesias, parkinsonism, torticollis) range from 0 to 5.

The SPDDS, assessing the impairment in the activities of daily living caused by motor symptoms, has a minimum score of 24 and maximum score of 120 and it comprises 24 items. The patient and the observer can fill the SPDDS in; as such procedure was used in a validation study (Brown et al., 1989).

For patient–proxy discrepancies, average discrepancy score (based on item differences between patient and caregiver) was computed as described by Hoth et al. (2007) for each scale separately. Using average scores instead of sum of discrepancies made the results independent of differences in scale length used for different domains.

Mood assessment

The patients' mood was assessed by means of Montgomery-Asberg Depression Rating Scale (MADRS) (Montgomery & Asberg, 1979). Rating scale filled in by the examiner (not by the patient) was chosen because of the possibly impaired insight in HD.

Neuropsychological assessment

Neuropsychological assessment addressed global cognitive function (MMSE) (Folstein, Folstein, & McHugh, 1975), verbal learning (Auditory Verbal Learning Test AVLT)

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Table 3. Patient–caregiver (P-C) discrepancies in Motor Impairment Scale (MIS) (based on films) and Self-Assessment Parkinson's Disease Disability Scale (SPDDS)

	HD $N = 23$	PDdys $N = 25$	PDndys $N = 21$	CD $N = 20$	H/U tests[1]
MIS – global assessment P-C average difference score	−0.20[2]	0.00	−0.07	0.03	H(3, $N = 89$) = 5.52 $p = 0.14$; s.i.
MIS – choreic movements P-C average difference score	−0.40	0.20	NA	NA	$U = 154.00$; $z = -2.76$ $p = 0.005$
MIS – parkinsonism P-C average difference score	NA	0.00	−0.20	NA	$U = 220.50$; $z = 0.93$ $p = 0.35$; s.i.
SPDDS P-C average difference score	−0.21	0.04	−0.04	0.00	H(3, $N = 89$) = 2.74 $p = 0.43$; s.i.

Note: NA = not assessed; s.i. = statistically insignificant; HD = Huntington's disease; PD = Parkinson's disease; CD = cervical dystonia; PDdys = PD with dyskinesias; PDndys = PD without dyskinesias.
[1]The differences between the two groups were analyzed with Mann Whitney U test. The differences among the 4 groups were tested either with H Kruskal-Wallis test with *post hoc* comparisons.
[2]Median value.

(Choynowski & Kostro, 1980), and cognitive control (Stroop Control Word Interference Test, CWIT) (Stroop, 1935).

Stroop CWIT was administered in a format used by the European Huntington's Disease Network (www.euro-hd.net) (European Huntington's Disease Network Cognitive Working Group, 2009) with a modified procedure. In each trial (color naming, color reading, interference) 50 stimuli were presented. The proportion of uncorrected errors to reactions (to account for omission errors) in the interference trial was computed and constituted a cognitive control measure (as uncorrected errors represent the failure to successfully monitor one's performance).

To sum up, MIS and SPDDS were administered to patients and caregivers. Neurologist performed UHDRS, UPDRS or TWSTRS. Neuropsychologist administered MMSE, AVLT, MADRS, and Stroop task.

Data Analysis

The reliability of the MIS was tested by means of internal-consistency Cronbach's alpha coefficient. Normality of distribution was tested with Shapiro-Wilk W test and homogeneity of variance was assessed with Levene's test. The between-group differences for the four groups were tested using one-way analysis of variance with *post hoc* Scheffe test or H Kruskal-Wallis test with *post hoc* comparisons (Siegel & Castellan, 1988). Differences between two groups were tested either with Mann-Whitney U test, t unpaired test or χ^2 tests as appropriate. Correlation analyses were performed using Spearman rank correlation coefficients. A conventional alpha of .01 was used in all the analyses.

RESULTS

Assessment of Memory, Cognitive Control, and Mood

Patients with HD exhibited the worst performance in AVLT, while their performance on the Stroop CWIT was comparable to PDdys group (see Table 2).

Assessment of Patient–Caregiver Discrepancies

Overall, patients with HD underestimated their deficits in the motor domain (see Table 3). Differences for ADL assessment were not statistically significant.

Patient–proxy reports of choreic movements differed significantly when average difference scores were analyzed. As the median of average difference score in the HD group has a negative value and median in the PDdys has a positive value, patients with HD as a group had a tendency to underestimate choreic movements, while patients with PDdys tended to overestimate those movements (see Table 3). Patient–proxy agreement on parkinsonism severity was lower in the PDdys than in the PDndys group.

Self-awareness of Symptoms and Other Clinical Variables

The relationship between self-awareness of symptoms and other clinical variables was assessed only for the HD group. For MIS (motor) and SPDDS (ADL) average difference scores were mildly correlated with Stroop score, AVLT scores, MMSE score, MADRS score, and UHDRS motor score. None of these correlations were statistically significant, however.

DISCUSSION

The present study was designed to comparatively assess the self-awareness of motor symptoms and ADL dysfunction in HD in comparison to PD and CD, with special emphasis on the chorea perception. The results of the analyses show that, in comparison to patients with advanced PD, individuals with HD underestimate the intensity of choreic movements. Thus, our findings are consistent with previous reports suggesting deficient self-awareness of chorea in HD (Hoth et al., 2007; Snowden et al., 1998; Vitale et al., 2001). In particular, our data are in concordance with the results from the questionnaire-based study obtained by Snowden et al. (1998), who showed that in HD self-awareness of ADL dysfunction (consequences of motor symptoms) is better preserved than the self-awareness

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of chorea. Our data, based on the movie presentation of involuntary movements, provided a similar pattern of results: better preserved self-awareness of ADL impairment than self-awareness of choreic movements. This discrepancy suggests that the patients' subjective experience of chorea is impaired, while observing its impact on ADL may be possible at the same time. This explanation suggests neurophysiological rather than neuropsychological background of impaired self-awareness of chorea. Alternatively, greater chorea may be associated by the patients with more severe disease stage and as such may be denied on the basis of psychological defense mechanisms.

Moreover, the fact that poor self-awareness of symptoms was seen in HD but not in PD supports the hypothesis that anosognosia/anosodiaphoria in HD may be predominantly associated with orbitofrontal–limbic pathology, resulting in failure to attach significant negative value to particular impairments in behavior. What is more, the results of this research are additionally strengthened by the fact that self-awareness of motor symptoms has been evaluated using a movie material, which is likely to have improved comprehension of test items.

Nonetheless, although our study indicates that anosognosia/anosodiaphoria of chorea might be characteristic for HD, the relationship between the severity of motor symptoms and the diminished self-awareness of these symptoms remains unclear. For example, in the study by Vitale et al. (2001), but in contrast to the study by Hoth et al. (2007) greater severity of motor symptoms in HD was associated with poorer self-awareness of these symptoms. This discrepancy is, however, difficult to interpret in the light of our data, since the mean UHDRS motor score in our study was similar to that reported by Hoth et al., whereas the UHDRS mean score was not presented in the study by Vitale et al. Importantly, in our study the magnitude of patient–proxy discrepancy was unrelated to symptom severity. Moreover, the results of our analyses have also suggested that self-awareness of motor symptoms in HD is not related to patients' memory. Thus, this study contrasts some previous reports indicating that in patients with HD poor self-awareness of symptoms is typically associated with memory disturbance (Deckel & Morrison, 1996; Hoth et al., 2007). Along the same line, our findings do not support the somewhat paradoxical observation by Snowden et al. (1998) that better memory (verbal learning either object recall) may be associated with more deficient self-awareness. In our study, no association was noted between mood and the degree of self-awareness. It could be argued that possibly many variables contribute to poor self-awareness and none of them influences it to an extent that could be ascertained in a study with a limited number of subjects.

Previous research have suggested that deficient self-awareness of symptoms in HD is not a result of poor judgment, as it was shown that patients' ability to assess the behavior of other people was preserved (Ho et al., 2006; Hoth et al., 2007). As already mentioned, such a selective deficit might stem from a failure to perceive consequences of one's behavior and to attach negative value to one's actions, reflecting orbitofrontal–limbic pathology. In our study, all participants were asked only to rate their own functioning and not proxies', as rating the intensity of motor dysfunction and its impact on daily function in proxies would be pointless. Comparable Stroop CWIT results in both our HD and PDdys group might also suggest that the cognitive ability to monitor one's performance (detect errors) may not be a crucial factor responsible for deficient self-awareness of symptoms. Thus, further studies are needed to elucidate the underlying causes of deficient self-awareness in HD.

Although deficient self-awareness in HD has been frequently associated with prefrontal pathology (Hoth et al., 2007; Sitek, Slawek, & Wieczorek, 2008), recent studies have shown that orbitofrontal dysfunction, albeit characteristic for HD, may be also seen even in mild PD without dementia (Lyoo, Ryu, & Lee, 2010; Tinaz, Courtney, & Stern, 2011). Moreover, orbitofrontal atrophy, is not selective in HD, and the neurodegeneration in this disorder encompasses also extra-fronto-striatal dysfunction (e.g., corpus callosum as well as posterior cortical areas) (Douaud, 2009; Halliday et al., 1998; Rosas et al., 2010). Thus, since we did not compare the severity of anosognosia/anosodiaphoria of motor symptoms in HD with the extent of brain abnormalities on neuroimaging, the attribution of deficient self-awareness of motor symptoms exclusively to the orbitofrontal–limbic atrophy in HD may be a simplification of this complex phenomenon and requires further empirical evidence.

The present study has several limitations. First, two different scales were used to assess self-awareness, which was controlled for by using average discrepancy scores (Hoth et al., 2007). Second, neuropsychiatric factors, other than depression (such as apathy or anxiety) and neuropsychological factors (others than cognitive control and memory) were not included in the analysis. Third, the effect of pharmacotherapy (e.g., neuroleptic drugs) on our results was not analyzed due to the heterogeneity of treatment regimens. Moreover, the fact that the magnitude of patient–proxy discrepancy was unrelated to symptoms severity might have resulted from the overall underrepresentation of patients with severe symptomatology due to the chosen MMSE cut-off as one of the inclusion criteria.

CONCLUSIONS AND PRACTICAL IMPLICATIONS

The present study highlights the need of interviewing the HD caregivers in clinical practice, as patients with HD tend to underestimate their motor abnormalities. Hence, underreporting of symptoms may have serious consequences in the patient's management, such as inadequate pharmacological treatment failing to address main, albeit unreported problems. Another important implication of the present study is that reducing chorea in HD should not be automatically regarded as a priority in HD pharmacotherapy, mostly because of the limited data on drug effectiveness as well as their possible adverse effects (Mestre, Ferreira, Coelho, Rosa, & Sampaio, 2009), especially in cases of mild chorea. Additionally, psychoeducational intervention should be aimed at HD caregivers', so they may

attempt to accept deficient self-awareness as another symptom of HD and, thus, may learn how to better cope with the devastating impact of HD on the functioning of both the patient's and his/her surroundings.

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APPENDIX

Motor Impairment Scale

I. Item choice

For each symptom, films with patients presenting moderate intensity of a given symptom where chosen.

II. Item order

1 Hand tremor (UPDRS)
2 Shoulder elevation (TWSTRS)
3 Chorea- lower limb (UHDRS)
4 Bradykinesia (UPDRS)
5 Gait (UPDRS)
6 Rotation (TWSTRS)
7 Chorea-buccolingual (UHDRS)
8 Retrocollis (TWSTRS)
9 Leg tremor (UPDRS)
10 Chorea-upper limb (UHDRS)
11 Anterocollis (TWSTRS)
12 Laterocollis (TWSTRS)
13 Chorea-face (UHDRS)
14 Posture (UPDRS)
15 Chorea-trunk (UHDRS)

III. Procedure

Each film was presented till the participant provided the answer (replayed if necessary), but not for a period shorter than 7 seconds (the duration of the shortest film). Each time, all films were shown on the same notebook monitor (HP Pavilion dv5 Notebook PC; dimensions 33×21 cm). When a viewer mentioned that some symptoms were not persistent, but temporary (as in case of dyskinesia in PD), he/she was asked to rate its intensity referring to moments when it was present.

IV. Scoring

Each symptom from each movie was rated by the patient as either absent (0), less pronounced than in the movie (1), of more or less similar intensity as shown (2), or as more pronounced (3). The month preceding the actual testing was suggested as a reference period, with the exception of CD patients and proxies who were asked to rate the symptoms intensity before former botulinum toxin injection (all were treated, but examined after the wash-out period before the next injection).

V. Reliability

Reliability analysis performed for MIS yielded satisfactory results for all subscales (all Cronbach's alpha coefficients were between 0.71 and 0.78). Item–subscale correlation coefficients were moderate (0.40–0.69).

VI. Validity

Validity of the scale was evidenced by inter-group differences for subscores, consistent with diagnosis (HD, PD or CD) and disease severity (in case of PD). Severity of choreic movements/dyskinesias was higher in HD and PDdys than in PDndys and CD, both according to patients' ($H(3, N = 89) = 44.78$; $p < .0001$) and caregivers' ratings ($H(3, N = 89) = 49.01$; $p < .0001$). Parkinsonism severity was higher in PDdys than in HD, PDndys, and CD according to patients ($H(3, N = 89) = 41.87$; $p < .001$) and higher than in HD and CD according to proxies ($H(3, N = 89) = 43.82$; $p < .0001$). Torticollis symptoms severity was rated by the patients as higher in CD than in HD, PDdys, and PDndys ($H(3, N = 89) = 43.97$) and higher in CD than in PDdys and PDndys by the caregivers ($H(3, N = 89) = 26.93$; $p < .001$).

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