

The impairment of emotion recognition in Huntington's disease extends to positive emotions.

Laura Robotham, Disa Sauter, Anne-Catherine Bachoud-Lévi, Iris Trinkler

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1 The impairment of emotion recognition in Huntington's disease extends to positive
2 emotions
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4 Running page heading: recognition of positive emotions in HD
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8 Laura Robotham*^{1,2,3}, Disa A. Sauter*⁴ Anne-Catherine Bachoud-Lévi ^μ ^{1,2,3,5} and Iris
9 Trinkler ^μ ^{1,2,3}
10
11

12 * and ^μ These two authors contributed equally to this work.
13
14
15
16

17 ¹ INSERM U955, E1, Equipe de Neuropsychologie Interventionnelle, Institut Mondor de Recherche
18 Biomédicale, Créteil, France
19

20 ² Département d'Etudes Cognitives, Ecole Normale Supérieure, Paris, France
21
22

23 ³ Université Paris Est, Faculté de Médecine, Créteil, France
24

25 ⁴ The Max Planck Institute for Psycholinguistics, Nijmegen, The Netherlands.
26

27 ⁵ AP-HP, Centre de Référence Maladie de Huntington, Service de Neurologie, CHU Henri Mondor,
28 Créteil, France
29
30

31 Corresponding author :
32
33

34 Anne-Catherine Bachoud-Lévi
35

36 Service de neurologie
37

38 Hôpital Henri Mondor
39

40 51 av du Mal de Lattre de Tassigny
41

42 94010 Créteil cedex, France
43

44 Tel +33 1 49 81 37 93
45

46 Fax + 33 1 49 81 23 26
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48 bachoud@gmail.com
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4 Abstract
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6 Patients with Huntington's Disease (HD) are impaired in the recognition of emotional
7 signals. However, the nature and extent of the impairment is controversial: It has
8 variously been argued to disproportionately affect disgust (e.g., Sprengelmeyer et al.,
9 1996), to be general for negative emotions (Snowden et al., 2008), or to be a
10 consequence of item difficulty (Milders et al., 2003). Yet no study to date has
11 included more than one positive stimulus category in emotion recognition tasks, and
12 most studies have focused on the recognition of emotions from facial stimuli. In this
13 study, we test the hypothesis that patients with HD may be impaired in their
14 recognition of positive as well as negative emotional signals, by examining the
15 recognition of a range of positive emotions from vocal cues. We present a study of 14
16 Huntington's patients and 15 controls performing a forced-choice task with a
17 previously validated set of negative and positive non-verbal emotional vocalizations
18 (Sauter and Scott 2007). Although HD patients performed above chance for each
19 emotion, they were found to be impaired in both positive and negative emotions,
20 including pleasure, fear and anger. These findings complement previous work by
21 demonstrating that impairments in emotion recognition in HD extend to positive and
22 negative emotions, which may imply a general deficit.
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50 Keywords: Huntington's disease; Emotion; Vocalizations; Affect
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Introduction

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Huntington's disease (HD) is a rare, inherited, neurodegenerative disease characterized by motor impairments. It is associated with numerous cognitive and attentional deficits, as well as difficulty in processing and interpreting emotions. But although it is well established that patients with HD are impaired in the recognition of emotional signals, there is little agreement about the nature and extent of this impairment. Earlier studies found an impairment that disproportionately affected the recognition of disgust expressions in clinical (e.g. Sprengelmeyer et al., 1996) and a circumscribed deficit for disgust in pre-clinical individuals with HD (e.g., Gray et al. 1997). However, several studies have documented a more general impairment encompassing several negative emotions in clinical patients (Calder et al., 2010) or virtually all negative emotions across both pre-clinical and clinical individuals (e.g., De Gelder et al., 2008; Johnson et al., 2007; Snowden, et al., 2008). Some authors have argued that the selective impairment for disgust stimuli found in some studies may have been a consequence of item difficulty (Milders et al., 2003). Recently, Snowden et al. (2008) employed ten different tasks assessing recognition of emotion from facial and vocal cues in patients with manifest HD. They found that patients were impaired in the recognition of several negative emotions across their extensive battery of tasks and concluded that HD may cause a general impairment in the identification of negative emotions.

A consistent finding across previous studies investigating emotion recognition in HD patients is that the recognition of positive emotions has been spared in the recognition of facial as well as vocal stimuli (e.g., Calder et al., 2010, Snowden et al., 2008, but see Sprengelmeyer et al., 1996). However, one limitation is that they have all included

1 only a single positive emotion category (joy/happiness). Recent work, however, has
2 shown that a range of positive states can be reliably communicated using non-verbal
3 signals such as posture, voice, and touch (see Sauter, 2010). In particular, non-verbal
4 vocalizations, such as laughs, cheers, and sighs, reliably signal joyful emotions
5 including amusement, achievement, and pleasure (Sauter et al. 2010; Sauter and Scott,
6 2007), as well as negative emotions such as anger, disgust, fear and sadness. Non-
7 verbal vocalizations thus provide a class of emotional signals in which the recognition
8 of a range of both positive and negative emotions can be tested.
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22 The aim of the current study was to examine emotion recognition in HD using several
23 negative and positive emotions. In order to test the specificity of the impairment, we
24 employed an emotion identification task using previously validated non-verbal
25 affective vocalization stimuli of positive and negative emotions (Sauter et al., 2010;
26 Sauter and Scott, 2007). This allowed us to address two hypotheses. One possibility is
27 that the recognition of positive emotions is in fact spared in HD, as previous authors
28 have found. Alternatively, previous studies may have failed to detect the impairment
29 in the recognition of positive emotional stimuli because of the use of only a single
30 class of positive emotional stimuli. This study examined the recognition of more than
31 one type of positive emotional signal in HD.
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48 Methods and Materials

49 Participants

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51 The participants consisted of 14 genetically confirmed patients with HD (6 female,
52 mean age = 51.29 (\pm 7.69) years; mean years of formal education = 12.29 (\pm 3.36)),
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1 of formal education = 12.53 (\pm 2.64)). The patients and controls were matched for age
2 ($t_{27} = -1.25$, $p > 0.2$) and years of education ($t_{27} = .22$, $p > 0.8$). Participants were
3 native French speakers recruited through the biomarker program approved by the
4 ethical committee of Henri Mondor Hospital, Paris and gave informed consent to
5 taking part in the study. The HD patients had no previous neurological or psychiatric
6 history other than HD. None of the participants reported hearing problems in
7 everyday life and their basic auditory perception was assessed using a computer beep;
8 all participants comfortably perceived the test sound at a volume of 3.5 of 10.
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21 Background neurological and neuropsychological assessment of HD patients

22 The patients were at early stages of the disease, according to the Total Functional
23 Capacity Scale (Shoulson, 1981), presenting with apparent but mild movement and
24 cognitive impairments. None of the patients showed signs of depression. Data from
25 the neurological and neuropsychological assessments of the HD patients are
26 summarized in Table 1.
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39 Emotional vocalization stimuli

40 The emotional vocalization stimuli were obtained from a previously validated set of
41 affective non-verbal vocalizations, such as laughs, sighs, and screams, lasting
42 approximately one second per stimulus (Sauter et al., 2010; Sauter and Scott, 2007).
43 The categories included in the current study were from three positive emotions
44 (achievement, amusement, and pleasure), and four negative emotions (anger, disgust,
45 fear, and sadness). Ten tokens were used per emotion category, resulting in a set of 70
46 stimuli, with equal numbers of male and female vocalizations.
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Procedure

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2 Participants were played the stimuli in a random order via headphones from a
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4 computer running a purpose-written E-prime script. After each stimulus, a list of the
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6 response alternatives was displayed on the screen (1: "accomplissement"
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8 (achievement), 2: "amusement" (amusement), 3: "colère" (anger), 4: "dégoût"
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10 (disgust), 5: "peur" (fear), 6: "plaisir" (pleasure), and 7: "tristesse" (sadness)), and
11
12 participants were asked to judge the emotion expressed in the vocalization they heard.
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14 Participants responded by typing in a number on the keyboard, and no time limit was
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16 enforced.
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Statistical Analysis

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26 In forced-choice tasks, performance for a particular category can be artificially
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28 inflated by the disproportionate use of that response. 'Hu' scores are unbiased hit rates
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30 that take into account each participant's use of the different response alternatives
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32 (Wagner, 1993). A score of zero denotes chance performance and a score of one
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34 perfect performance. Hu scores were calculated for patients' and controls' recognition
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36 rates for each emotion category, and tested against (Hu-score calculated-) chance
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38 level. Due to group sizes and heterogeneous population variances in the data, non-
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40 parametric analyses were employed for all analyses. Like Snowden et al. (2008) and
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42 several previous authors, we employed Mann-Whitney U tests for pairwise-
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44 comparisons, and Friedman tests for within-group comparisons. Further, like
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46 Snowden et al. (2008), correction procedures were not employed in order to avoid
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48 loss of power in these small samples.
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Results

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1 Both the HD patients and the controls were able to identify the emotions from the
2 non-verbal vocalizations: They performed better than chance for the task overall
3 (controls: $z = -3.41$, $p < 0.001$; HD patients: $z = -3.30$, $p < 0.001$). However, the
4 controls performed better than the HD patients (average performance for controls:
5 $0.52 (\pm 0.12)$, and for patients: $0.28 (\pm 0.16)$, $U = 21$, $z = -3.67$, $p < 0.001$).

6
7 Performance varied across the emotion types in the controls, ($\chi^2_r = 29.5$, $p < 0.001$),
8 and there was a trend for variability across emotion categories in the patient group (χ^2_r
9 $= 11.6$, $p = 0.07$). Considering each emotion separately, both controls and HD patients
10 performed above chance for all emotions (controls : $z = -3.35$ for achievement, $z = -$
11 3.41 for amusement, $z = -3.41$ for pleasure, $z = -3.41$ for anger, $z = -3.42$ for disgust, z
12 $= -3.41$ for fear, and $z = -3.42$ for sadness, all $p < 0.001$) and HD: ($z = -2.67$ for
13 achievement; $z = -3.30$ for amusement; $z = -3.23$ for pleasure; $z = -3.23$ for anger; $z =$
14 -3.11 for disgust; $z = -3.23$ for fear; and $z = -3.23$ for sadness; all $p < 0.001$, for
15 achievement: $p = 0.004$). Performance per emotion and group is shown in Figure 1.

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INSERT FIGURE 1 ABOUT HERE

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Recognition performance in HD patients was lower than in controls for achievement
($U = 56.0$, $z = -2.1$, $p = 0.032$), anger ($U = 34.0$, $z = -3.1$, $p = 0.002$), disgust ($U =$
 48.5 , $z = -2.5$, $p = 0.012$), fear ($U = 14.5$, $z = -4.0$, $p < 0.001$), and pleasure ($U =$
 43.5 , $z = -2.7$, $p = 0.006$), with a trend also for sadness ($U = 60.5$, $z = -2.0$, $p =$
 0.051), and amusement vocalizations ($U = 66.5$, $z = -1.7$, $p = 0.093$). Importantly,
recognition performance in HD patients was lower for the average of positive ($U =$
 45.5 , $z = -2.6$, $p = 0.009$) as well as the average of negative emotions ($U = 21.5$, $z = -$
 3.6 , $p < 0.001$).

1 Confusion matrices are shown in Table 2. Errors were largely consistent across
2 patients and controls, and included some cross-valence confusions. Common errors
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4 included sad sounds mistaken for pleasure, anger sounds classified as disgust, and
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6 achievement sounds perceived as signaling amusement.
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17 Discussion

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19 The results of this study show that patients with HD have reduced sensitivity in the
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21 identification of emotional signals overall, for positive as well as negative emotions.
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23 Importantly however, HD patients (as well as controls) were able to perform the task
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25 at better-than-chance levels, overall and for each category of non-verbal emotional
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27 vocalizations. Additionally, the relative difficulty and confusion errors made by the
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29 patients largely mirrored those of the controls (see Table 2). This pattern suggests that
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31 the patients' impairment was not due to a failure to understand or perform the task,
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33 but rather a consequence of a reduced sensitivity to emotional cues in both positive
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35 and negative affective vocal signals. Although the current study did not include a
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37 control task testing higher order auditory processing for non-emotional stimuli, the
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39 general ability of the patients to perform the task and the similarity of their
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41 performance to that of the control participants, suggests that the emotion recognition
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43 impairment in HD was not due to primary auditory deficits. A previous study has also
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45 found intact performance in HD patients on auditory tasks including phoneme
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47 processing (Teichmann, Darcy, Bachoud-Lévi, & Dupoux, 2009).
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51 The results from the current study are consistent with previous work that has found
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53 impaired recognition of several negative emotions in HD (De Gelder et al., 2008;
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1 Johnson et al., 2007; Milders et al., 2003; Snowden et al., 2008), but our findings go
2 beyond earlier research in demonstrating an impairment that extends to positive
3 affective signals, thus highlighting the importance of examining the identification of
4 positive emotion in more depth.
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11 While further work will be needed to confirm whether HD patients are also impaired
12 in the recognition of positive emotional signals in other modalities, our results suggest
13 that the emotion recognition deficit in HD is not specific to a single emotion or to
14 negative emotions. This finding lends some support to the hypothesis that emotion
15 processing may be impaired at a general level in HD. Given that HD patients are
16 impaired in the recognition of emotional cues from facial, bodily, and vocal signals
17 (De Gelder et al., 2008; Snowden et al., 2008), this impairment likely occurs at post-
18 perceptual processing levels.
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31 Although little is known about HD patients' understanding of their own emotions,
32 recent work has suggested that patients' general semantic representations are not
33 impaired (Hayes et al. 2007; Snowden et al., 2008), highlighting the need to examine
34 other mechanisms that may play a causal role in the emotion recognition impairment
35 in HD. It is worth noting that, although our results suggest an overall impairment in
36 the recognition of both positive and negative emotions, the impairment for certain
37 emotions of both valence types appear to be particularly severe. The recognition of
38 anger and fear was especially impaired in the HD patients compared to control
39 participants in our study, like in Calder et al. (2010), who attributed this pattern to
40 insensitivity to social disapproval in HD. However, insensitivity to social disapproval
41 is unlikely to explain the strong impairment for pleasure signals also found in the
42 current study.
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By contrast, given the motor symptomatology in HD, one candidate mechanism may be affective motor representations, which have been suggested to underlie emotion recognition across modalities (Niedenthal 2007).

Another important factor in understanding the underlying mechanisms in causing this impairment is the neuropathology associated with HD. Comparisons of the emotion recognition problems in HD patients with those of other patient groups with brain pathology affecting the striatum, such as Parkinson's Disease (PD), could inform our understanding of the role of striatal regions in emotion processing. Studies on PD patients' emotion recognition in the visual modality have yielded inconclusive results, with some finding an impairment (e.g., Kan et al., 2002), and others not (e.g., Adolphs et al. 1998). For auditory stimuli impaired recognition has been observed for a number of negative emotions (e.g., Lloyd, 1999; Pell, 1996), as well as for positive prosody (Pell and Leonard, 2003). A recent meta-analysis of emotion recognition in PD concluded that the identification of negative emotions is disproportionately impaired in this group of patients (Gray and Tickle-Degnen, 2010). However, no study to date has used more than one category of positive emotion with PD patients. Future comparisons of emotion recognition of a wide range of emotions in HD and PD patients may elucidate the contribution of the basal ganglia to emotion processing.

The results from the current study are the first to show an impairment of recognition of positive emotional signals from non-verbal vocal signals in HD. The results are consistent with recent findings that have failed to find a specific impairment in the recognition of disgust in HD, and instead point to a global impairment of emotion recognition in HD, affecting both positive and negative emotional signals.

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2 Table 1. Background neuropsychological data for the HD patients, with published
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4 normal ranges where available. Notes: UHDRS = Unified Huntington's Disease
5 Rating Scale (UHDRS; Huntington Study Group, 1996); Motor and Behavioral sub-
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7 scales and TFC (Total Functional Capacity) scale (Shoulson, 1981), MADRS =
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9 Montgomery and Asberg Depression Rating Scale (Montgomery and Asberg, 1979);
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11 FHVLT = French Hopkins Verbal Learning Test (Rieu, et al., 2006); MDRS = Mattis
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13 Dementia Rating Scale (Mattis et al., 1976; normal range norms from Guelfi, 1997);
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15 TMT B = Trail Making Test part B (Tombaugh, 2004).
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24 Table 2. Confusion matrices for controls and Huntington's patients in percentages.
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26 Achieve = Achievement, Amuse = Amusement, Plea = Pleasure. Stimuli categories in
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28 rows and response categories in columns. All rows add to 100. Correct responses are
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30 marked in bold.
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34 35 36 Figure legend

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39 Figure 1. Performance (Hu scores) in the recognition of non-verbal vocalizations of
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41 emotions by Huntington's patients (dotted line) and controls (black line) in a 7-way
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43 forced-choice task. Standard error bars are shown for each category of vocalization
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46 for each group.
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Table 1.

Test	Sub-scale/domain	HD patients	Published normal cut-off
UHDRS	TFC (max 13)	11.4 (1.2)	-
	Motor (max124)	26.3 (10.9)	-
	Behavioural (max 16)	1.9 (2.8)	-
MADRS	Depression (max 60)	11.0 (6.7)	< 15
FHVLT (C)	Delayed recognition (max 12)	8.1 (3.8)	>9.7 ^a
MDRS	Total (max144)	128.9 (11.7)	>136
	Attention	35.6 (1.5)	>31
	Initiation	29.6 (7.4)	>28
TMT B	Executive function	161.9 (58.7)	> 135

^a Cut-off was calculated as mean minus two standard deviations from the published control data (11.3 ± 0.8).

Table 2.

Stimulus type	Response						
	Controls (n = 15)						
	Ach	Amuse	Plea	Anger	Disgust	Fear	Sadness
Achieve	52.7	32.7	12.0	0.7	0.7	1.3	0.0
Amuse	0.7	85.3	13.3	0.0	0.0	0.0	0.7
Plea	13.3	2.0	69.3	0.7	6.0	1.3	7.3
Anger	12.0	6.0	1.3	58.7	14.7	6.7	0.7
Disgust	5.3	2.7	4.0	2.7	84.0	1.3	0.0
Fear	4.7	4.0	2.7	4.0	2.0	80.0	2.7
Sadness	6.7	2.0	19.3	0.7	8.7	2.7	60.0
HD Patients (n = 14)							
	Achieve	Amuse	Plea	Anger	Disgust	Fear	Sadness
Achieve	36.4	27.9	18.6	5.0	2.9	5.7	3.6
Amuse	10.0	67.9	15.0	0.7	1.4	0.7	4.3
Plea	17.1	7.1	48.6	3.6	12.1	6.4	5.0
Anger	12.1	7.9	2.9	38.6	17.1	19.3	2.1
Disgust	7.9	7.9	5.7	6.4	55.7	7.1	9.3
Fear	8.6	18.6	9.3	7.1	10.7	42.9	2.9
Sadness	10.7	6.4	20.7	4.3	7.9	5.0	45.0

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Figure

