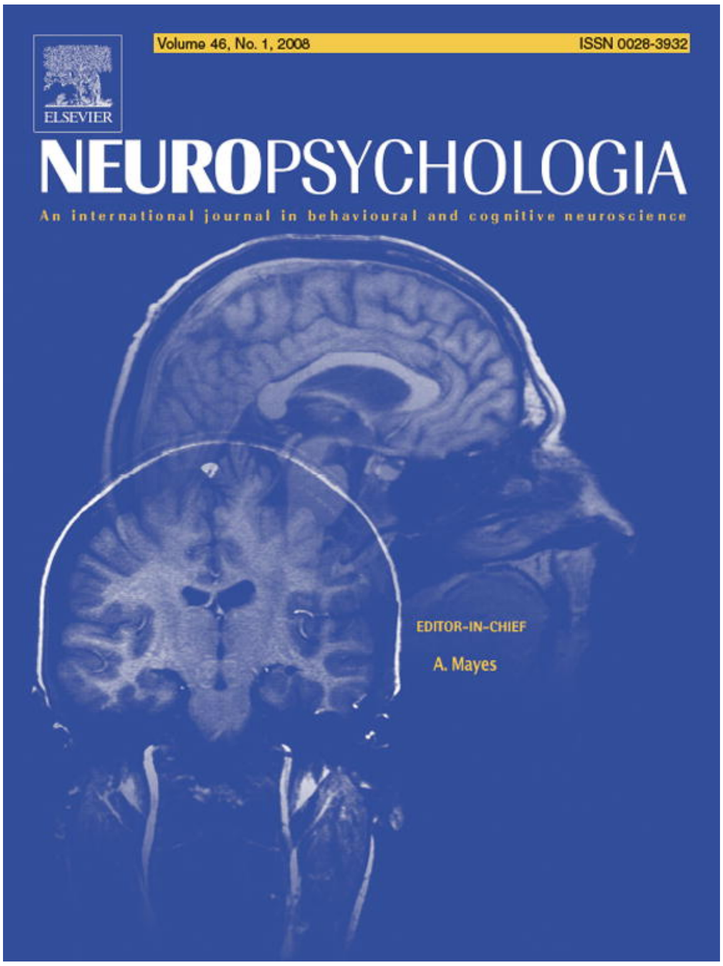
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Note

Huntington's disease impairs recognition of angry and instrumental body language

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Abstract

Patients with Huntington's disease (HD) exhibit motor impairments as well as cognitive and emotional deficits. So far impairments in the ability to recognize emotional stimuli have mostly been investigated by using facial expressions and emotional voices. Other important emotional signals are provided by the whole body. To investigate the impact of motor deficits on body recognition and the relation between motor disorders and emotion perception deficits, we tested recognition of emotional body language (instrumental, angry, fearful and sad) in 19 HD patients and their matched controls with a nonverbal whole body expression matching task. Results indicate that HD patients are impaired in recognizing both instrumental and angry whole body postures. Furthermore, the body language perception deficits are correlated with measures of motor deficit. Taken together the results suggest a close relationship between emotion recognition (specifically anger) and motor abilities.

Keywords: Emotion; Striatum; Action representation

1. Introduction

Huntington's disease (HD) attacks primarily basal ganglia structures (mostly caudate nucleus and putamen) leading to severe motor deficits (Vonsattel et al., 1985). At the same time HD is accompanied by deficits in recognizing emotional expressions, particularly facial expressions of disgust (Sprengelmeyer et al., 1996; Wang, Hoosain, Yang, Meng, & Wang, 2003).

However, in natural circumstances facial expressions are rarely seen in isolation, but occur in the context of expressions by the whole body. Our ability to perceive these emotional body expressions and their representation in the brain is now becoming an important research topic (de Gelder, 2006), and exploring how HD patients recognize emotional body language may significantly contribute to novel insights. In previous studies with neurologically intact observers, we used fMRI to clarify how the brain recognizes happiness or fear expressed by a whole body (de Gelder, Snyder, Greve, Gerard, & Hadjikhani, 2004; Grezes, Pichon, & de Gelder, 2007). Our results indicate that observing fearful body expressions produces increased activity in brain areas associated with perception of emotional faces, but also in areas involved in representation of action and movement, including caudate nucleus and putamen. Caudate nucleus and putamen are known for their involvement in motor tasks but have also been associated with motivational and emotional task components (Bhatia & Marsden, 1994; Grillner, Hellgren, Menard, Saitoh, & Wikstrom, 2005; Kampe, Frith, Dolan, & Frith, 2001). Therefore, we conjectured that HD is also associated with a deficit in recognizing emotions expressed by the

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whole body. However, since our pilot data indicated that static bodily expressions of disgust are difficult to distinguish from fear (both postures consisting in moving backwards and putting hand palms forward), we did not include disgust.

2. Methods

2.1. Participants

Nineteen HD patients (10 early HD at stage I and nine at stage II, using the classification based on the total functional capacity scale (Shoulson, 1981)) and 19 control subjects participated. HD patients were recruited from an out-clinic follow-up program within the framework of interventional therapy approved by the ethical committee of the Henri Mondor Hospital. They had no previous neurological or psychiatric history and their HD diagnosis was genetically confirmed. All subjects gave informed consent. Patients were evaluated using the Mattis dementia rating scale (MDRS) (Mattis, 1976), and the United Huntington's disease rating scale of which the cognitive part contains the Stroop test, the verbal fluency task and symbol digit test. All patients were administered by the same rater. Atrophy of the caudate was assessed in 11 patients with MRI by calculating an adjusted bicaudate ratio, which took cortical atrophy into account (the minimal distance between the caudate indentations of the frontal horns divided by the width of the brain along the same line multiplied by 100). We opted for this adjustment since there are now several studies showing rather widespread cortical pathology in HD gene carriers (Kassubek et al., 2004a; Kassubek, Gaus, & Landwehrmeyer, 2004b; Thieben et al., 2002).

Control subjects were healthy volunteers with normal or corrected to normal vision and no previous neurological history. They were matched on age, $t(36) = 1.37$, $p = 0.180$, sex, $\chi^2 = 1.18$, $p = 0.669$, dexterity, $\chi^2 = 0.01$, $p = 0.958$ and years of education, $t(36) = 0.29$, $p = 0.775$ (see Table 1 for demographic and general assessment data).

2.2. Stimuli and procedure

Video recordings of eight semi-professional actors (half of them women, age 22–35 years) were used for stimulus construction. Actors performed fearful, angry, and sad expressive gestures with their whole body. The actors also performed instrumental but emotionally neutral actions (pouring water into a glass, combing one's hair, putting trousers on, opening a door, talking on the telephone, and drinking a glass of water). These instrumental displays elicit action representation (Johnson-Frey et al., 2003) and are thus appropriate to use as controls for investigating emotional body expressions, also eliciting action representation and implicit movement perception.

Table 1
Demographic data of HD patients and control subjects

Demographic data	HD	Controls
N	19	19
Sex	9F/10M	8F/11M
Age in years (S.D.)	52.0 (9.1)	48.2 (8.0)
Educational level in years (S.D.)	13.9 (4.6)	14.4 (5.1)
Evolution duration in years (S.D.)	5.9 (4.2)	n.a.
CAG-repeats (S.D.)	42.7 (2.6)	n.a.
Laterality	18R/1L	18R/1L
General assessment data		Normal published range
Total functional capacity	10.0 (2.1)	13
UHDRS motor score	32.4 (19.0)	0
MDRS	129.8 (7.7)	≥136
Stroop color/word	20.3 (10.0)	$\geq 35^{\%}$
Fluency "P" in 2 min	16.8 (7.1)	$18^{\S}$
Symbol digit code	21.9 (8.6)	$\geq 37^{\mu}$
Bicaudate ratio's*	20.7 (4.3)	$<10^{\#}$

n.a. = not applicable *N = 11. The norms are issued from % (Golden, 1978); § (Cardebat, Doyon, Puel, Goulet, & Joanette, 1990); μ (Wechsler, 1981); # (Starkstein et al., 1989).

Prior to the recordings the actors were briefed with a set of standardized instructions. For the instrumental body actions instructions specified the action to be performed. For emotional body actions instructions specified a familiar scenario (for example, opening a door and finding an armed robber in front of you). Static images were obtained from the videos by selecting the most informative frame from the video file and converting it to grayscale pictures. To exclude that face recognition would play a role in recognition of the whole body stimuli, the faces were blocked with a grey mask. Stimulus selection for the present experiment was based on the results of a pilot study in which the images were presented one by one on a PC screen and shown for 4000 ms with a 4000 ms interval. For the emotional bodies, a total of 120 trials were used (3 expressions × 8 identities × 5 repetitions). Participants were instructed to categorize each stimulus in a forced choice procedure as quickly and as accurately as possible by pressing one of the three response buttons corresponding to the three emotions. Overall correct recognition rate was between 100 and 65% (average 93%). For each stimulus category the six highest ranked exemplars were chosen (all recognized at 100% accuracy) for use in the present study. The same procedure was used for the instrumental gestures. Overall correct recognition rate in the pilot study was between 100 and 90% (average 99%), and for each gesture the six highest ranked were chosen (all recognized at 100% accuracy) for the present study.

The experiment consisted of randomized simultaneous presentation of three images on each trial, one at the top (target) and two probes right en left underneath. Target and probe pictures were always of three persons of the same sex, but one probe displayed the same expression as the target and the other one a different expression. Participants were required in a two alternative forced choice task to select the probe with the same expression as the target. They responded by pressing the corresponding button (see Fig. 1A and B for stimulus examples). Stimuli were presented until response. During the intertrial interval (3000 ms) a blank screen was shown. The experiment consisted of a block with emotional stimuli and a block with instrumental stimuli.

The session started with four familiarization trials, followed by 75 experimental trials for the emotional block (3 emotions × 5 stimuli × 5 repetitions) and 48 trials for the instrumental block (6 actions × 4 stimuli × 2 repetitions). Similar to a previous study (Van den Stock, Righart, & de Gelder, 2007), we used a matching task instead of a naming or categorization task because we were primarily interested in tapping into the motor perception processes. A naming or categorization task appeals more to verbal labeling of stimuli and also necessitates a significant language component, abilities which are impaired in HD. The 2AFC task used here requires that matching be based on the basis of movement similarities between stimuli in the same emotion category.

3. Results

Mean accuracy scores for the two groups, separated by expression, are displayed in Fig. 1C. We carried out a repeated measures ANOVA with expression (four levels: instrumental, anger, fear, and sadness) as within-subjects variable and group (two levels: HD and controls) as between-subjects variable. This revealed significant effects of type of expression, $F(3, 108) = 23.54$, $p < 0.001$; group, $F(1, 36) = 22.25$, $p < 0.001$; and a significant interaction, $F(3, 108) = 3.77$, $p = 0.013$. To follow up on the interaction effect, we performed for every expression Bonferroni corrected t tests between both groups. This showed significant differences between the groups on the instrumental and angry expressions, $t(36) > 4.73$, $p < 0.001$. In order to explore differences between stage 1 and stage 2 HD, we performed a post hoc repeated measures ANOVA on the data of the patient group with expression (four levels) as within-subjects variable and stage (two levels) as between-subjects variable. This showed no main effect of group or interaction with expression.

To investigate a possible relationship between whole body recognition abilities on the one hand and behavioral and neu-

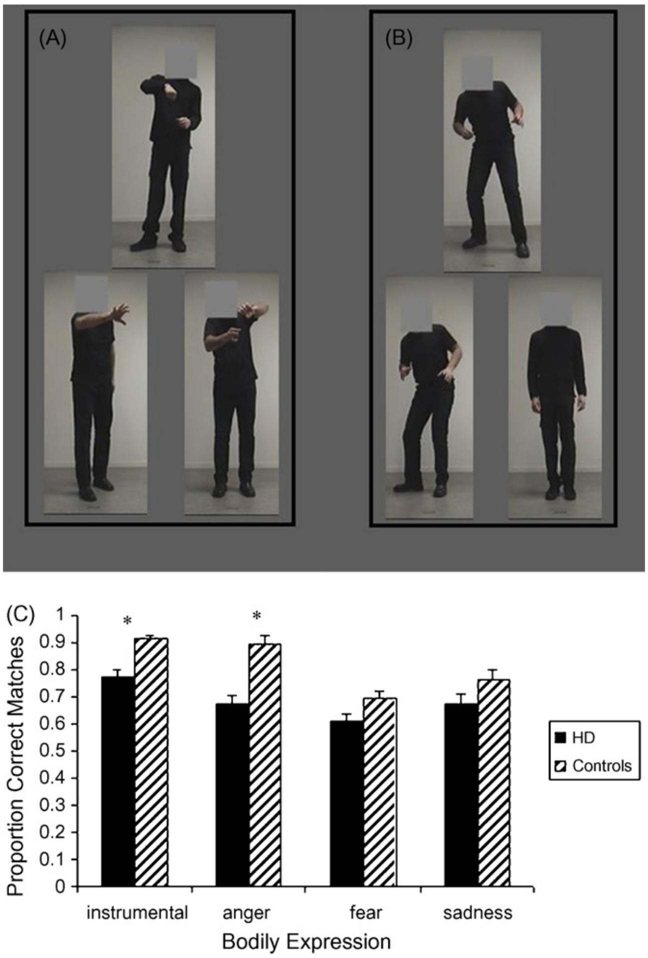

Fig. 1. (A) Stimulus example in the instrumental action block and (B) the emotional block showing a fearful body on top and a fearful (left) and a sad (right) body at the bottom. (C) Proportion correct matches as a function of group and bodily expression. Error bars represent one S.E.M. above the mean. *Significant at the $P < 0.01$ level.

rostructural deficits on the other hand, we performed partial correlation analyses, controlling for scores on the MDRS in order to correct for effects due to general cognitive decline.

Table 2 shows significant correlations between general assessment measures and experimental data. Partial correlations between bicaudate ratio and experimental data were not significant for any bodily expression. Since the UHDRS motor consists of a number of subscales, we performed an exploratory correlation analysis at the level of different components of the UHDRS motor, according to Shannon (Shannon, Raman, & Leurgans, 1999) namely chorea, dystonia, oculo-motor dysfunction, and motor dysfunction. This revealed a significant correlation between the motor dysfunction component and recognition of angry body postures, $r = -0.575$, $p = 0.016$. The correlation between motor dysfunction and recognition of instrumental body postures was marginally significant, $r = -0.476$, $p = 0.053$. When confined to the subjects who underwent both bicaudate and UHDRS measurements ($N = 9$), both correlations remained stable, but at a lower significance level ($r = -0.712$, $p = 0.048$ and $r = -0.643$, $p = 0.085$, respectively). Motor dysfunction takes into account gait disturbance, axial disorders, bradykinesia, rigidity, postural reflexes, and gesture disabilities.

Correlations between bicaudate ratio and MDRS, $r = -0.737$, $p = 0.004$ and between bicaudate ratio and the UHDRS motor dysfunction component, $r = 0.558$, $p = 0.047$ were also significant.

Table 2
Partial correlations (controlling for performance on MDRS) between bicaudate ratio, UHDRS subscales (Shannon et al., 1999) and experimental data

		Bicaudate ratio	Chorea	Dystonia	Motor	Oculo-motor
Body anger						
	r	−0.012	−0.046	−0.445	−0.575	−0.190
	p	0.973	0.861	0.073	0.016*	0.464
	N	11	15	15	15	15
Body fear						
	r	0.111	0.123	0.298	0.006	−0.060
	p	0.761	0.640	0.245	0.983	0.819
	N	11	15	15	15	15
Body sad						
	r	−0.555	0.085	−0.356	−0.182	−0.161
	p	0.096	0.747	0.161	0.484	0.536
	N	11	15	15	15	15
Body instrumental						
	r	−0.231	0.001	−0.291	−0.476	−0.199
	p	0.520	0.996	0.257	0.053 (*)	0.443
	N	11	15	15	15	15

* Significant at the $p < 0.05$ level; (*) marginally significant.

4. Discussion

The major finding of this study concerns the deficit of HD patients in recognizing instrumental and angry whole body postures. Recognition of meaningful non-emotional actions was not investigated previously, yet reports in the literature indicate that action related deficits in HD have been observed with different tasks in other settings (Aron, Sahakian, & Robbins, 2003). The present data provide evidence that action related abilities are important for recognition of instrumental actions and bodily expressions of anger. The action component at stake in recognition of whole body expressions of sadness is considerably less important. We conjecture that this is due to the fact that this emotion is typically associated with relaxation and loss of muscle tonus. Similarly, recognition of whole body expressions of fear also implies a reduced action component as fear cannot only lead to flight but is equally associated with freezing of the whole body (LeDoux, 1996). So the observed deficits in recognizing instrumental body actions and bodily expressions of anger are compatible with the idea that the motor deficit of HD patients impairs their ability for action recognition. On the basis of this, a relationship between the UHDRS motor score and experimental data was expected. This is in fact the case. Corrected for general cognitive decline, HD patients were more impaired in recognizing the angry and the instrumental body expressions if their motor symptoms were more severe. The difficulties in perceiving body emotions were not related to abnormal movements like chorea or dystonia but to features that better capture the body posture and the abilities to perform gestures. This suggests a link between perception and production of body gestures that needs to be further investigated. In view of our previous results that caudate nucleus activity was observed in the contrast between "fearful" and "instrumental" expressions of the body (de Gelder et al., 2004) it is surprising that neurodegeneration of the caudate nucleus in these HD patients is not reflected in impaired fear processing. But at present we have no data available directly comparing fear and anger expressions which would allow us to estimate the relative involvement of caudate nucleus in fear vs. anger action perception. This issue clearly needs further investigations.

Research over the last decade has clearly indicated that recognition of instrumental actions involves some of the same brain areas that are involved in performance of that action by the observer himself. The importance of motor areas for action recognition is illustrated by the research on mirror neurons by Rizzolatti and co-workers (di Pellegrino, Fadiga, Fogassi, Gallese, & Rizzolatti, 1992; Grèzes & Decety, 2002; Rizzolatti & Craighero, 2004). Thus the observed impairment in recognition of instrumental actions evokes the concept of motor resonance at the center of motor cognition abilities, which are implemented in premotor cortex, parietal cortex and superior temporal sulcus (STS). Degeneration of the motor areas in HD, predominantly striatum and its connections to parietal and premotor cortex and STS is consistent with the importance of action representation for intact recognition of whole body postures. The areas involved in spontaneous facial expressions (dorsolateral prefrontal cortex, orbitofrontal cortex, anterior cingulate, insula and amygdala) connect with the motor system via de basal ganglia (Alexander & Crutcher, 1990; Damasio, 1999) and this network may constitute an important part of a dedicated mechanism for visuomotor emotion perception.

Correlations between structural and functional cerebral changes and cognitive abilities have been reported in HD, although not consistently (see Montoya, Price, Menear, & Lepage, 2006, for a review). We were unable to find a correlation between our structural anatomical index (adjusted bicaudate ratio) and recognition of body postures. It should be stated that we preferred to include cortical pathology in our anatomical index, in view of recent reports about rather widespread cortical pathology already in preclinical HD (Kassubek et al., 2004a, 2004b; Thieben et al., 2002). However, incorporating cortical atrophy may rule out any kind of specific relationship between decreased volume of the striatum on the one hand and behavioral data on the other hand.

A question for future research concerns the relationship between recognition of emotional faces and bodies. Recently, selective deficits in recognition of angry faces have been reported in patients with damage to the ventral striatum (Calder, Keane, Lawrence, & Manes, 2004). Furthermore, the disgust recognition deficit in HD has also been extended from facial expressions to scenes, odors, vocal expressions and declarative knowledge (Hayes, Stevenson, & Coltheart, 2007). We did not include whole body expressions of disgust, since our pilot data indicated they were very hard to recognize in static stimuli once the facial information is completely blurred.

Finally, in view of the relation we observed between emotion recognition deficits (specifically anger) and motor abilities, an interesting question is whether the same pattern of deficits observed here will also be found when we use dynamic stimuli. We are currently investigating this issue.

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