Research report

Embodied emotion impairment in Huntington's Disease

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ABSTRACT

Theories of embodied cognition suggest that perceiving an emotion involves somato-visceral and motoric re-experiencing. Here we suggest taking such an embodied stance when looking at emotion processing deficits in patients with Huntington's Disease (HD), a neurodegenerative motor disorder. The literature on these patients' emotion recognition deficit has recently been enriched by some reports of impaired emotion expression. The goal of the study was to find out if expression deficits might be linked to a more motoric level of impairment. We used electromyography (EMG) to compare voluntary emotion expression from words to emotion imitation from static face images, and spontaneous emotion mimicry in 28 HD patients and 24 matched controls. For the latter two imitation conditions, an underlying emotion understanding is not imperative (even though performance might be helped by it). EMG measures were compared to emotion recognition and to the capacity to identify and describe emotions using alexithymia questionnaires. Alexithymia questionnaires tap into the more somato-visceral or interoceptive aspects of emotion perception. Furthermore, we correlated patients' expression and recognition scores to cerebral grey matter volume using voxel-based morphometry (VBM). EMG results replicated impaired voluntary emotion expression in HD. Critically, voluntary imitation and spontaneous mimicry were equally impaired and correlated with impaired recognition. By contrast, alexithymia scores were normal, suggesting that emotion

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representations on the level of internal experience might be spared. Recognition correlated with brain volume in the caudate as well as in areas previously associated with shared action representations, namely somatosensory, posterior parietal, posterior superior temporal sulcus (pSTS) and subcentral sulcus. Together, these findings indicate that in these patients emotion deficits might be tied to the "motoric level" of emotion expression. Such a double-sided recognition and expression impairment may have important consequences, interrupting empathy in nonverbal communication both ways (understanding and being understood), independently of intact internal experience of emotion.

1. Introduction

Patients with Huntington's Disease (HD), a rare, inherited neurological disorder, causing motor, cognitive and emotional dysfunctions, are impaired at recognizing emotional facial expressions. In recent years, consensus has emerged that most emotions are concerned (Henley et al., 2008, 2012; Johnson et al., 2007; Milders, Crawford, Lamb, & Simpson, 2003; Novak et al., 2012; Snowden et al., 2008). Recognizing joy/happiness was long thought to be relatively spared, however, a recent study demonstrated that the impairment extends to positive emotions when the number of positive and negative stimuli is balanced (Robotham, Sauter, Bachoud-Levi, & Trinkler, 2011). However, an integrative explanatory model of the emotion recognition deficit is still outstanding, possibly because emotion processing has mostly been tested from the recognition side only. Here, we will adopt a perspective of embodiment (see Decety & Jackson, 2004; Gallese, 2007; Keysers & Gazzola, 2007; Niedenthal, 2007), arguing that our perception of actions and emotions in others builds upon our own action and emotion representations. Our question then is whether HD patients show impaired representations for both own and others' emotions. There are two different aspects to sharing emotions: a) on the motor level of emotion expression, and b) on the level of internal experience. Evidence for both levels has been gathered by different experimental studies: a) Carr, Iacoboni, Dubeau, Mazziotta, and Lenzi (2003, see also Blair, 2005) have suggested that an emotion recognition deficit in autism might stem from an impaired action-based network (see also Dapretto et al., 2006). Overlapping fMRI activation for observing and imitating emotional facial expressions has been found in a network comprising posterior superior temporal sulcus (pSTS), posterior parietal, anterior insula (AI), amygdala and premotor cortices (Carr et al., 2003; Hennenlotter et al., 2005). Briefly, the respective contributions of these areas might be as follows. pSTS relays higher order visual information, such as information coding gaze, expression, and lip movement (Atkinson & Adolphs, 2011; Halgren, Raij, Marinkovic, Jousmaki, & Hari, 2000; Haxby, Hoffman, & Gobbini, 2000; Hein & Knight, 2008; Kesler-West et al., 2001; Pizzagalli et al., 2002; Said, Haxby, & Todorov, 2011), and biological motion in general (Giese & Poggio, 2003). Information is forwarded to posterior parietal neurons, which code kinesthetic aspects, and further to inferior frontal (BA 44/45) neurons, coding action goals. Somatosensory cortex also plays an important part in the network (Adolphs, Damasio, Tranel, Cooper, & Damasio, 2000; De Gelder, 2006, 2016), perhaps by representing aspects of the body and body surface (Keysers, Kaas, & Gazzola, 2010; Keysers et al., 2004). In sum, this network seems to support shared visuo-motor action representations (Blakemore & Decety, 2001; Grezes, Armony, Rowe, & Passingham, 2003; Grezes & Decety, 2001). b) On the other hand, evidence for a shared network on an internal experiential level has been demonstrated in the AI and anterior cingulate cortex (ACC) for observing and experiencing pain (Jackson, Meltzoff, & Decety, 2005; Singer et al., 2004), but further for a wide range of shared emotions, such as pleasant affect, social exclusion, disgust and anger (summarized in Bernhardt & Singer, 2012). The insular cortex is known to integrate diverse forms of "interoceptive" information (Craig, 2002; Critchley, Wiens, Rotshtein, Ohman, & Dolan, 2004; Kurth, Zilles, Fox, Laird, & Eickhoff, 2010), i.e., the ensemble of information about the visceral and physiological states of the body including itch, coldness, hunger, sensation of fatigue after physical exercise, pain, etc. and also visceral affective states (Craig, 2002). Interoceptive information, mapped to more posterior insular segments, is subsequently re-represented in the AI, where it may become more consciously accessible. This enables various subjective affective experiences and global (homeostatic) feeling states (Craig, 2002, 2009). The cingulate cortex may represent the motivational-premotor counterpart for the sensory-perceptual affective feelings integrated by the insula, conjointly implementing general monitoring and control processes across multiple domains (Paus, 2001). Note that AI-A/MCC stand as neuroanatomical markers of more extensive and complex networks that include brainstem, and midbrain (see Damasio, Damasio, & Tranel, 2013). Further, beyond the interoceptive brain representations that might be shared between emotions, additional differential neural representations presumably exist, tied to each emotion's intrinsic psychobiological functions. Since with regards to HD, consensus has emerged that their emotion processing deficit is not specific to one emotion (Henley et al., 2008, 2012; Milders et al., 2003; Robotham et al., 2011; Snowden et al., 2008), we do not detail these here. Importantly, AI activity during affect sharing (Bernhardt & Singer, 2012; Bird et al., 2010) and reflecting on feelings across the emotion spectrum (Silani et al., 2008), is correlated with empathy self-report scores

and trait alexithymia, the latter referring to difficulties describing and identifying emotions (Sifneos, 1996).

With this in mind, one might therefore ask if emotion recognition deficits in HD are accompanied by deficits on an expression level and/or on a level of internal experience. Two former studies have investigated emotion *expression* in HD. Hayes and colleagues (Hayes, Stevenson, & Coltheart, 2009), focusing on disgust, reported impaired spontaneous expression of disgust (other emotions were not evaluated), as well as impaired instructed expressions of all emotions. Trinkler, Cleret de Langavant, & Bachoud-Levi (2013) compared recognition and voluntary expression of the six basic emotions (Ekman, 1999) and found both equally impaired. However, these results could have come about either by a motor expression impairment, or by impairments on the level of internal experience. Here, we reasoned that if HD patients show impaired imitation of emotion expressions, this would be corroborating evidence for a motor expression impairment, since imitation does not necessarily require accessing the internal representation. Further, if emotion identification on a level of internal experience is intact, then, emotion expression on command is not impaired due to a lack of understanding what the emotion means. Moreover, the anatomical underpinnings of emotion recognition deficits should be consistent with the selected hypothesis, i.e. involving the abovementioned network of pSTS, posterior parietal, somatosensory and premotor cortices, rather than AI and ACC.

Here, we used electromyography (EMG) to assess spontaneous and voluntary imitation of emotional facial expressions, and voluntary expression from words. These EMG measures were complemented by a recognition test of the six basic emotions (anger, disgust, fear, joy, sadness, surprise) as well as alexithymia ratings. Additionally, for a subset of HD patients, structural brain images were available and cerebral grey matter was regressed against EMG and recognition scores using voxel-based morphometry (VBM) (Ashburner & Friston, 2000).

Electromyograms capture electric signals of underlying emotional motor reactions on the skin surface. Even though emotions are naturally composed of a large array of facial muscle activations (Ekman & Friesen, 1978), key placements permit to capture typical muscle activations accompanying certain emotions, see below. One can readily measure volitional imitation of facial expressions using EMG, even though this technique has mainly been used to study spontaneous imitation ("mimicry") of another person's non-verbal displays (famously: Dimberg, 1982). Here we measured EMG activity 1) over the eyebrows to capture an ensemble of action units (Ekman & Friesen, 1978) including the *corrugator supercilii*, typically involved in frowning in an angry face, and 2) over the cheek to capture *zygomatic major* muscle activity underlying smiling. These two facial muscle-emotion pairs are classically used in mimicry experiments (e.g. Achaibou, Pourtois, Schwartz, & Vuilleumier, 2008; Dimberg, 1982). Additionally, we explored expression of disgust since disgust has been the most widely studied facial emotion in HD. For this we applied additional electrodes 3) to the sides at the back of the nose, to capture *levator labii superioris alaeque nasi* activity underlying nose wrinkle (Ekman & Friesen, 1978; Waller, Cray, & Burrows, 2008).

Complementarily, in line with the abovementioned authors (Silani et al., 2008), we assessed participants' difficulty to identify and describe emotions using an alexithymia questionnaire.

2. Materials and methods

2.1. Participants

Twenty-eight genetically confirmed HD patients [12 female, 16 male; mean age (±SD) = 50 ± 8 years; 12 ± 3 years of formal education] and 24 control participants (12f, 12 m; mean age: 49 ± 10 years; 13 ± 3 years of formal education) were recruited within the biomarker program approved by the ethical committee of Henri Mondor Hospital. Both groups were left ignorant of the purpose and hypotheses of the study. HD patients and controls were matched for age (t = .33, p = .74), years of formal education (t = −.72, p = .47), and handedness (HD: 2 left; controls: 2 left; Chi^2 = .026, p = .87). None of the participants had any previous or current neurological or psychiatric history except HD. Six HD patients had no medical treatment at time of test, four were on tricyclic, four on tetracyclic antidepressants, five on benzodiazepine anxiolytics, five on vesicular monoamine transport (VMAT) inhibitors against chorea, five on antipsychotics (risperidone or olanzapine) for treatment of chorea and behavioral problems, two on lithium carbonate antidepressants, 13 on SSRI antidepressant/anxiolytics and one on beta-blockers.

2.2. Background neurological and neuropsychological assessment

HD patients were evaluated on the Unified Huntington's Disease Rating Scale (UHDRS, Huntington Study Group, 1996). Its motor part includes 15 items about voluntary and involuntary movements, various gestures and walking capacity, the best possible score is 0. The behavioral part provides a composite score for counting anomalies in frequency and severity (best = 0). The cognitive part of the UHDRS includes literal fluency, the Stroop task and the Symbol Digit Code. Norms are mentioned in Table 1. HD patients were further tested on the Montgomery and Asberg Depression Rating Scale (MADRS, Montgomery & Asberg, 1979), the French adaptation of the Hopkins Verbal Learning Test (FHVLT, Rieu, Bachoud-Levi, Laurent, Jurion, & Dalla Barba, 2006), the Mattis Dementia Rating Scale (MDRS, Mattis, Bellak, & Karasu, 1976, pp. 77−121) and the Trail Making Test, part B (TMT B, Reitan, 1955; Tombaugh, 2004).

2.3. Stimuli

For recognition, eight faces for each of the six basic emotions (anger, disgust, fear, joy, sadness, surprise) were used from the Ekman series (Ekman & Friesen, 1976) and the Karolinska Inventory (Lundqvist, Flykt, & Öhman, 1998), as before (Trinkler et al., 2013). For observation and imitation eight faces of each of joy, anger and disgust were used. During expression, the emotion words COLÈRE (anger), JOIE (joy), and DÉGOÛT (disgust) appeared centrally on the screen (font size = 48).

Table 1 – Background neurological and neuropsychological assessment of patients (mean ± SD).

			HD patients (N = 28)	Published norms (If applicable)
Years since disease onset			6.7 ± 4.1	−
Unstable triplet repeat (CAG) sequences			44.1 ± 2.5	<28
Total functional capacity			11.3 ± 1.4	13
UHDRS	Motor subscale	Total motor score (range = 0–124)	24.4 ± 14.7	≤5
		Dysarthria (range = 0–4)	.4 ± .5	Symptom-free = 0
		Tongue protrusion (range = 0–4)	.4 ± .7	Symptom-free = 0
		Pronosupination (range = 0–8)	1.8 ± 1.1	Symptom-free = 0
		Luria test (also "Fist-edge-palm") (range = 0–4)	.6 ± .9	Symptom-free = 0
		Bucco-oro-lingual chorea (range = 0 to 4)	.8 ± .7	Symptom-free = 0
		Facial chorea (range = 0–4)	.9 ± .7	Symptom-free = 0
	Psychiatric subscale	Apathy (range = 0–16)	.3 ± .6	Symptom-free = 0
			(Only 5 of 28 patients had a score > 0)	
	Cognitive subscale	Stroop (number of correct color words in 45 sec)	27.8 ± 13.4	>35
		Fluency (number of words for 'P', 'R', and 'V' in 2 min)	45.3 ± 24.3	54
		Symbol digit (correct symbol-digit associations in 90 sec)	32.0 ± 14.0	>37
FHVLT	Delayed recognition (max = 12)	Corr. recognition minus false positives	8.6 ± 3.2	>9.7
MDRS	Total (of 144)		129.2 ± 11.0	≥136
	Attention	(Max = 37)	35.5 ± 1.5	>31
	Initiation	(Max = 37)	30.5 ± 6.5	>28
TMT B	Executive function	(Cutoff = 240 sec)	160.8 ± 70.7	≤135
MADRS		(Range = 0–60)	10.7 ± 6.8	<7

Apathy = multiplication of UHDRS-PSY scores for apathy frequency and severity; FHVLT = French Hopkins verbal learning test; MDRS = Mattis Dementia Rating Scale; TMT B = Trail Making Test B, MADRS = Montgomery and Asberger Depression Rating Scale.

2.4. EMG recording

EMG was recorded using a BioSemi Active-Two amplifier system, using six active electrodes corresponding to three distinct bipolar montages. Three sets of facial electrodes were placed in accordance with EMG guidelines and/or previous literature (Achaibou et al., 2008; Dimberg, 1982; Fridlund & Cacioppo, 1986): Two were placed over the eyebrows capturing *corrugator supercilii* involved in frowning, two over the cheeks capturing *zygomatic major* involved in smiling, and two to the nose back, capturing *levator labii superoris alaeque nasi* and *nasalis* involved in nose wrinkle as part of the disgust expression (Waller et al., 2008).

Two EEG reference and ground electrodes were also placed using an EEG cap (http://www.biosemi/faq/cms&drl.htm). The EMG was continuously recorded at 512 Hz, with a .1–417 Hz band-pass filter.

2.5. Experimental procedure (EMG and recognition experiment)

After EMG electrodes were placed, participants were seated in front of a computer screen (32 × 27 cm) at a distance of 70 cm, with a digital camera placed behind the screen that recorded the whole session. Subjects performed three EMG tasks: observation/spontaneous micro-mimicry (SPONT), voluntary imitation of emotional facial expressions (IMIT), and voluntary production of emotional facial expressions (PROD). For SPONT and IMIT they were presented with 96 stimuli (4 repetitions × 8 face identities × 3 emotions) and for PROD with 12 stimuli (4 repetitions × 3 emotion words). Stimuli were presented at full screen height for 4000 msec each, in random order, separated

by a varying intertrial interval (1500–3000 msec). Tasks followed the same sequence for all participants and were separated by instructions and 3 practice trials (one for each emotion, except for SPONT). During SPONT participants were instructed to simply study the facial expressions that were displayed to them and told that the task served stimulus familiarization. During IMIT participants were instructed to try and imitate voluntarily the exact same facial expression as they saw on the screen, and during PROD they mimed the facial expression according to the emotion word that appeared on the screen. After EMG recording, electrodes were removed and participants performed the recognition experiment (again on the computer screen), in which they were presented randomly with the 48 facial emotion stimuli described above. To the side of the face stimulus the six emotion words appeared followed by a number, and participants responded using the number keys, which emotion they thought corresponded best to the face seen. The experimental conditions are summarized in Fig. 1.

2.6. EMG data preprocessing and analysis

Raw data were segmented offline into 4500 msec epochs, including a 500 msec pre-stimulus baseline, and digitally filtered with a 20–256 Hz band-pass in Brain Vision Analyzer (Brain Products GmbH). These parameters for facial EMG acquisition and analysis were concordant with published guidelines (van Boxtel, 2001; de Luca, 1997) and previous EMG studies (Achaibou et al., 2008). Bipolar montages were then calculated from electrode pairs for each muscle in Matlab 7.1. and the magnitude of the EMG signal was determined by calculating the root-mean-square (RMS) of each bipolar montage over 125 msec interval bins after the onset of each
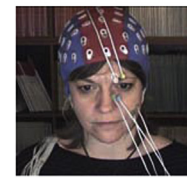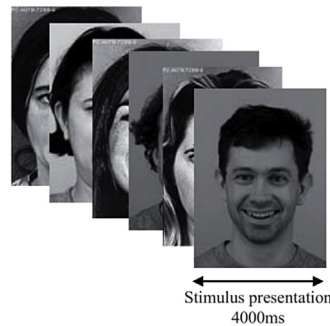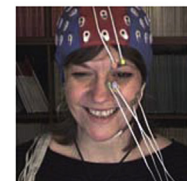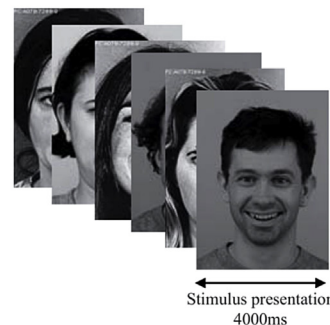
EMG Task/Block

1) SPONT
Measurement of spontaneous micro-mimicry during observation of emotional facial expressions (3 emotions x 8 face identities x 4 repetitions)
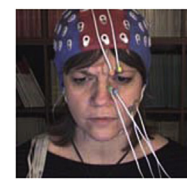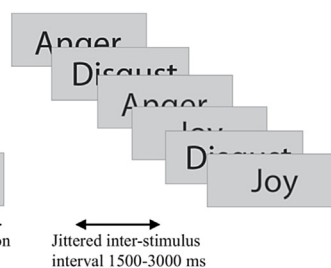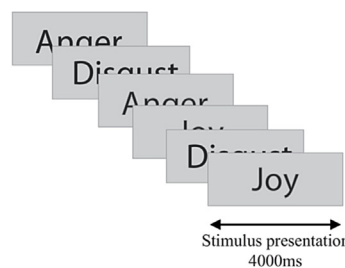
2) IMIT
Voluntary imitation of emotional facial expressions (3 emotions x 8 face identities x 4 repetitions)

3) PROD
Voluntary production of emotional facial expressions from words (3 emotions x 4 repetitions)

Stimuli presented to subjects

Stimulus presentation 4000ms

Jittered inter-stimulus interval 1500-3000 ms

Stimulus presentation 4000ms

Jittered inter-stimulus interval 1500-3000 ms

Anger
Disgust
Anger
Joy
Disgust
Joy

Anger
Disgust
Anger
Joy
Disgust
Joy

Stimulus presentation 4000ms

Jittered inter-stimulus interval 1500-3000 ms

EMG reaction

unvoluntary micro-activity

voluntary overt activity

voluntary overt activity

Fig. 1 – Electromyography experimental design.

stimulus. Trials in which the difference of activity between a given time-bin and the following one was superior to three standard deviations of the mean value for the whole trial were rejected. Finally, the RMS of the baseline was subtracted from each time interval. For the overt expression conditions IMIT and PROD, EMG magnitude per emotion - muscle-region pair was defined as the trapezoid interval $\left(\frac{1}{n}\int_0^n (c(t) - p(t))dt\right)$ over the 4000 msec stimulus display period in order to capture most closely the time-intensity area observed. Integrals are compared in emotion-muscle-pair × group repeated measures analysis of variance (ANOVAS) and subsequent t-tests. Where variances were not homogeneous, Greenhouse-Geisser correction was used. For VBM analysis (see below) patients' integrals were z-transformed on controls' mean and standard deviation. For the covert spontaneous mimicry condition SPONT, in accord with pre-existing literature (Dimberg, 1982), trials were averaged over the 4000 msec stimulus display period, separately for each pair of emotion-muscle-region. We first conducted the classical angry–happy comparison

(Achaibou et al., 2008; Dimberg, 1982) with a 2 (cheek–frown) × 2 (happy–angry) × 2 (HD patients-controls) repeated measures ANOVA. In a more exploratory vein, we then oppose disgusted-happy in the same way. EMG-data for one control subject was excluded because of spasmophylia. Data of another control data-set was rejected for the SPONT condition because her mean activation was more than three standard deviations above the control mean throughout. One patient data-set had to be excluded for the nose-electrodes during the IMIT condition because of a faulty signal and another patient data-set for the PROD condition due to technical failure.

2.7. Alexithymia questionnaire

As in our previous study (Trinkler et al., 2013), we used the French version (Marchand & Loas, 1995) of the twenty item Toronto Alexithymia Scale (TAS20, Taylor, Bagby, & Parker, 1997), and added 4 × 6 additional items asking specifically about the six basic emotions (anger, disgust, fear, joy, sadness, surprise). Half of these items were probed in an inverted statement, e.g., "en general je ne sais pas ce qui provoque ma

frayeur" ("in general, I don't know what provokes my fear") and "Je ressens immédiatement quand je suis effrayé(e)" ("I am immediately aware when I am frightened") in order to control for response bias. In the original TAS20, 5 of 20 items are inverted in the same way. Participants respond on a five-point scale, ranging from "désaccord complet" ("completely disagree") to "accord complet" ("completely agree"). Eighteen of the controls and 24 of the HD patients taking part in this study filled in this questionnaire after they completed the EMG experiment.

2.8. Structural MRI (sMRI) acquisition and VBM analysis

Whole-brain 3D T1-weighted image volumes were acquired on a 1.5 T (Siemens) scanner using a MPRAGE sequence with TR = 2400 msec, TE = 3.72 msec, FA = 8°, slice thickness = 1.5 mm. Structural data were available in 17 HD patients. VBM was performed using Statistical Parametric Mapping version 8 (www.fil.ion.ucl.ac.uk/spm). Initial tissue classification was performed using the unified segmentation process with rigid alignment. DARTEL (Ashburner, 2007) was used to create a study-specific template. The initial grey matter segmentation was spatially normalized using high dimensional warping onto the template then modulated to account for volume change during the normalization process and smoothed with a 4 mm full width at half maximum kernel.

We ran linear regression models using z-scores from EMG results, emotion recognition as well as TAS20 questionnaire scores. Age and unstable triplet repeat (CAG) sequence length were included as covariates in the model. An explicit binary mask was used for the analysis, generated using the optimal thresholding technique (Ridgway et al., 2009). We report significant grey matter voxels at $p < .001$, with a minimum cluster size of 10 voxels, without correcting for multiple comparisons. Therefore, we restrict our interpretations to areas for which we have a priori hypotheses based on pre-existing literature. In addition we report significant grey matter voxels correlating with IMIT, PROD and SPONT and TAS20 scores within 16 mm-spheres of significant voxels correlating with recognition (small volume correction, $p < .001$). Anatomical localizations are based on Duvernoy's brain atlas (Duvernoy, 1999).

3. Results

3.1. Background neurological and neuropsychological data

Average patient data on neurological and neuropsychological tests are summarized in Table 1. All UHDRS motor scores listed in Table 1 differed from zero (all ts > 2.27). HD patients had relatively mild motor impairment. Stroop, MDRS total and MDRS attention scores were lower than the norm (one-sample t-tests, $t_{Stroop} = -2.76$, $p = .011$; $t_{MDRS\_tot} = -3.15$, $p = .004$; $t_{MDRS\_attention} = 15.40$, $p < .001$), all other cognitive scores were within the normal range ($t < \pm 1.96$). Only five of 28 patients had apathy ratings higher than zero, so no statistical test was performed (but we ran an additional EMG analysis without

these five individuals, see below). Patients had an average depression score higher than normal (>6, $t_{MADRS} = 3.53$, $p = .002$), but within the range considered mild.

3.2. Facial emotion recognition test

Average recognition data are shown in Fig. 2. Compared to controls, HD patients were impaired overall (6 emotions × 2 groups repeated measures ANOVA, main effect of group: $F = 43.0$, $p < .001$; main effect of emotion: $F = 23.5$, $p < .001$; interaction: $F = 4.71$, $p = .001$). Recognition was lower in HD patients than in controls for each single emotion: anger: $t = -6.15$, $p < .001$; disgust: $t = -5.57$, $p < .001$; fear: $t = -6.17$, $p < .001$; joy: $t = -2.92$, $p = .01$; sadness: $t = -5.53$, $p < .001$; surprise: $t = -4.24$, $p < .001$.

3.3. Alexithymia

A visual scan of the questionnaire data yielded that two patients persevered and responded mostly "completely agree" throughout, thus showed a strong response bias. These data were removed from the analysis. For the remaining data, each item was assigned a score from 1 ("completely disagree") to 5 ("completely agree"), and 5 to 1 for inverted items. Summed scores are shown in Table 2. HD patients and controls performed similarly on the TAS20 items ($t = .403$, $p = .69$) and also on the six basic emotions (overall: $t = .35$, $p = .73$; anger: $t = .66$, $p = .51$; disgust: $t = 1.25$, $p = .22$; fear: $t = 1.89$, $p = .06$; joy: $t = -1.41$, $p = .16$; sadness: $t = -.73$, $p = .47$; surprise: $t = -.23$, $p = .82$), showing absence of alexithymia in both groups.

3.4. EMG

For the voluntary production (PROD) and imitation (IMIT) conditions, trapezoid integral are reported for the five areas which yielded overt activation in controls (see Figs. 3 and 4): CHEEK muscle region for happy faces, FROWN and NOSE muscle regions for angry and disgust.

For PROD, as controls, HD patients show emotion-specific activation above zero (all integrals > 0, all ts ≥ 3.02, all ps ≤ .005), but EMG-activity is significantly smaller (5 emotion-muscle pairs × 2 groups repeated measures ANOVA, significant effect of group: $F = 9.13$, $p = .004$; emotion-muscle pair: $F = 2.88$, $p < .048$; no interaction: $F = .74$, $p = .051$). Patients differed from controls for CHEEK-happy (t-3.73, $p = .001$), FROWN-angry ($t = -3.28$, $p = .003$), FROWN-disgusted ($t = -7.03$, $p < .001$) and NOSE-angry ($t = -4.99$, $p < .001$) muscle-emotion pairs, with a trend also for NOSE-disgusted ($t = 2.043$, $p = .051$). See Fig. 3.

For IMIT, EMG-activity was largely identical to PROD and also smaller in HD patients compared to controls (5 emotion-muscle pairs × 2 groups repeated measures ANOVA, significant effect of group: $F = 11.8$, $p = .001$; emotion-muscle pair: $F = 13.4$, $p < .001$; no interaction: $F = 2.41$, $p = .08$). Patients differ from controls for CHEEK-happy ($t = -2.29$, $p = .03$), FROWN-angry ($t = -5.40$, $p < .001$), NOSE-angry ($t = -4.54$, $p < .001$), FROWN-disgusted ($t = -4.41$, $p < .001$) and NOSE-disgusted ($t = -3.53$, $p = 002$) muscle-emotion pairs. See Fig. 4.

In the spontaneous mimicry (SPONT) condition, firstly, the happy-angry comparison yields the following results: the 2 (CHEEK−FROWN) × 2 (happy−angry) × 2 (HD patients-controls)

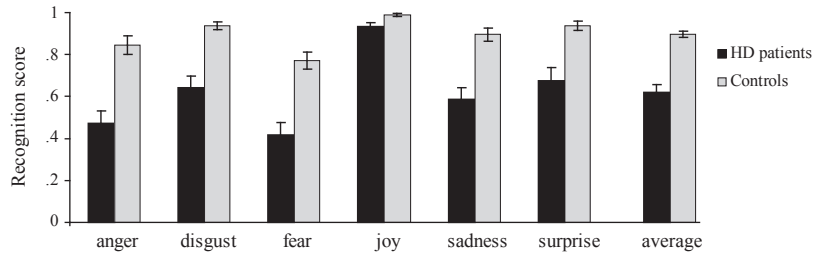**Fig. 2 − Results of facial emotion recognition test.**

Table 2 − Alexithymia scores.

	TAS20	Additional items pertaining to 6 basic emotions					
		Sum across emotions	Anger	Disgust	Fear	Joy	Sadness	Surprise
HD patients	49.6 (15.1)	54.73 (12.04)	9.59 (3.3)	10.86 (3.45)	9.82 (2.77)	6.32 (2.5)	9.36 (3.17)	8.77 (2.65)
Controls	46.4 (9.1)	53.56 (9.4)	9 (2.33)	9.61 (2.75)	8.44 (1.79)	7.5 (2.79)	10.06 (2.71)	8.94 (1.98)

Means across summed scores, standard deviations in brackets. Max = 100 for TAS20, 0−51: "non-alexithymia"; 52−61"possible alexithymia"; >61 "alexithymia". Max = 20 for single emotion items.

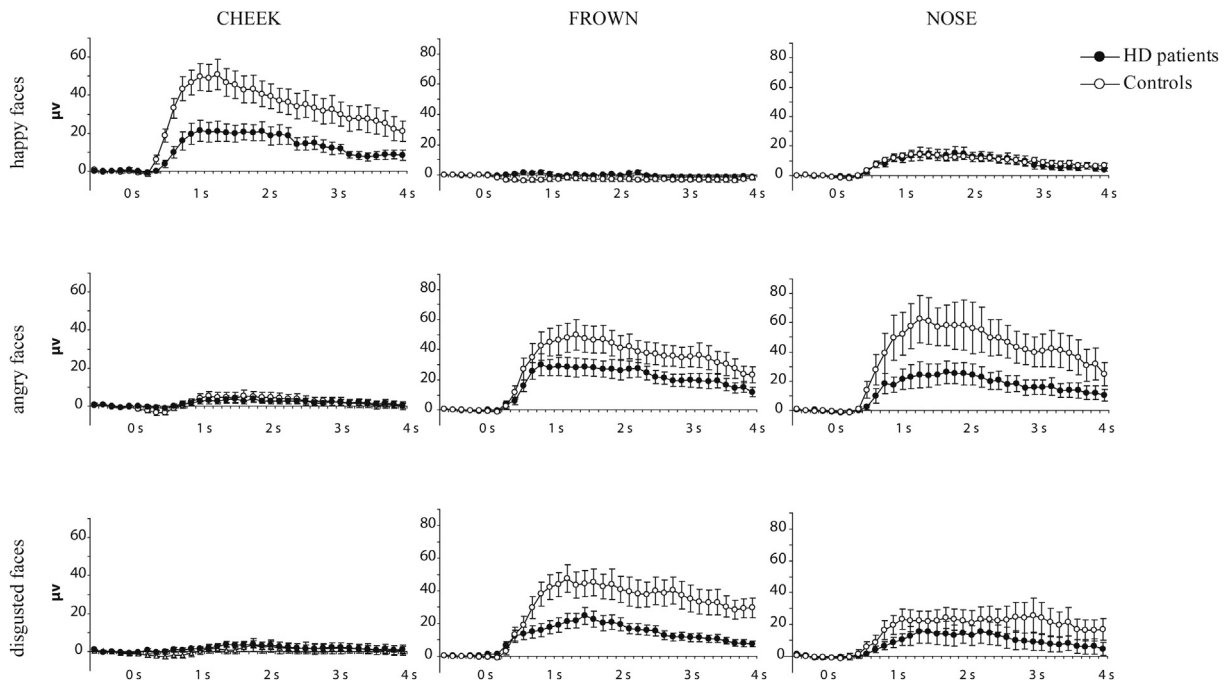## Voluntary production of emotional facial expressions in EMG

Fig. 3 − Voluntary production of emotional facial expressions, average EMG activation per time bin. HD patients and controls show specific muscle activation over corresponding facial areas, CHEEK for happy faces, FROWN and NOSE wrinkle for angry and disgusted faces, but HD patients show significantly less activation.

ANOVA over average of timebins 0−4000 msec yields a main effect of muscle ($F = 7.08$, $p = .011$), a significant interaction of emotion × muscle ($F = 8.14$, $p = .006$), and of emotion × muscle × group ($F = 6.32$, $p = .015$). Controls do show a significant muscle regions × emotion interaction (2 × 2 ANOVA, $F = 8.34$, $p = .009$), but patients do not ($F = .11$, $p = .74$). Thus, controls show spontaneous EMG modulation in the direction of more activity over CHEEK for happy faces, and more

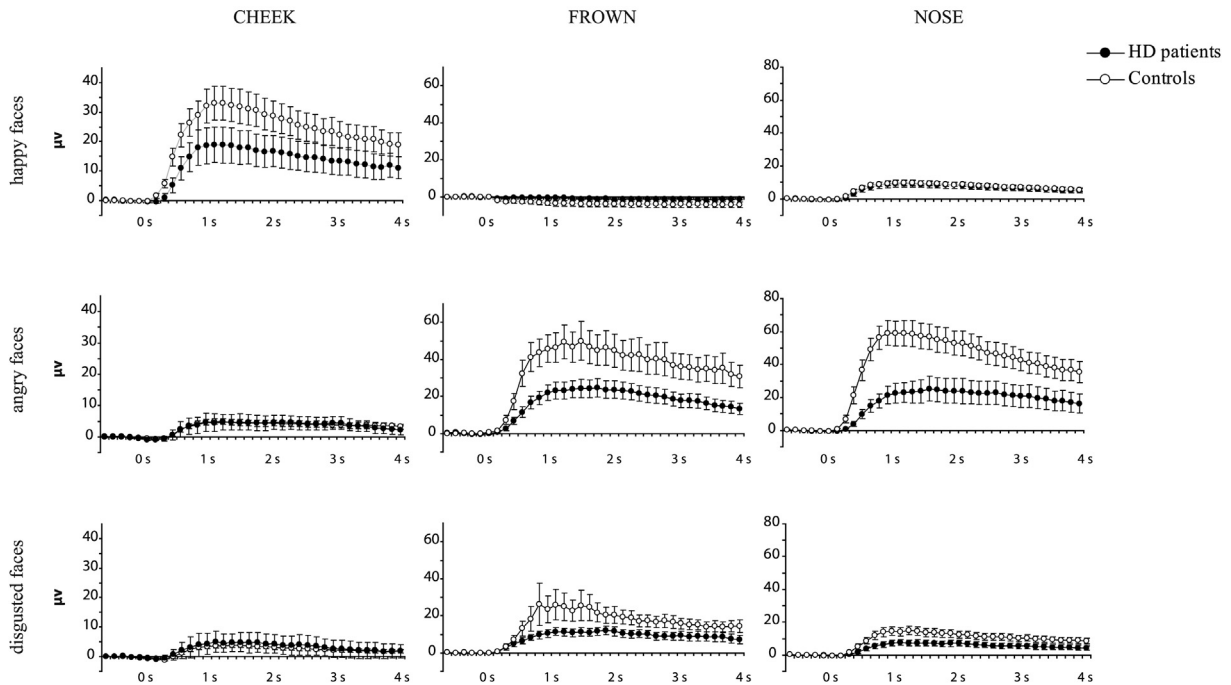## Voluntary imitation of emotional facial expressions in EMG

Fig. 4 – Voluntary imitation of emotional facial expressions, average EMG activation per timebin. Similarly as for production, HD patients and control subjects show specific muscle activation over corresponding facial areas, but HD patients show significantly less activation.

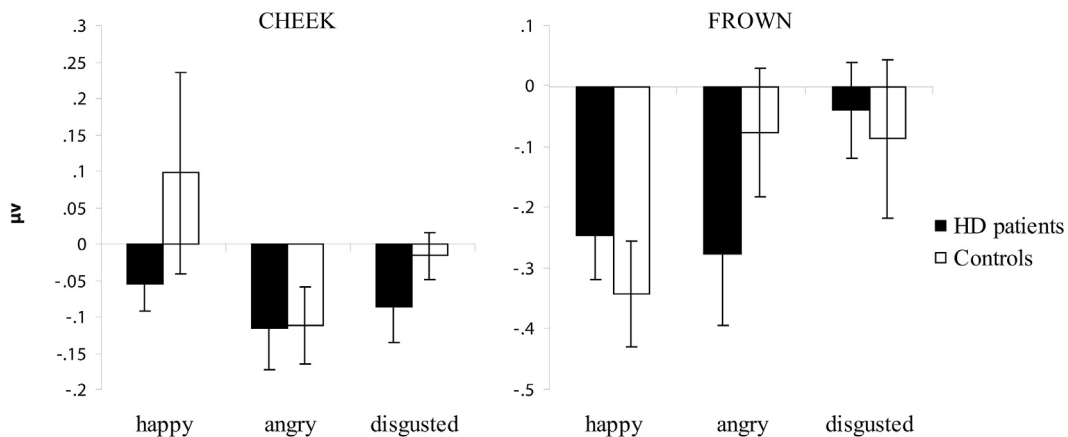## Spontaneous mimicry while viewing faces

Fig. 5 – Average mean EMG activation for spontaneous mimicry in reaction to passively viewing facial expressions. Control subjects show a significant muscle regions by emotion interaction. This modulation is lost in HD.

FROWN activation for angry faces compared to happy faces. This modulation is lost in HD. See Fig. 5. Secondly, for the happy-disgusted comparison, the 2 × 2 × 2 ANOVA was run over CHEEK–FROWN on the basis that our PROD results have shown most important natural activation for overt disgusted expressions on FROWN (and not NOSE) in controls. It yielded a main effect of muscle ($F = 8.31$, $p = .006$), and a significant emotion × muscle interaction ($F = 7.66$, $p = .008$), but no emotion × muscle × group interaction and no effect of group (both $F < 1.11$).

3.5. Medication effects

Most medication groups were too small (n ≤ 5) to conduct meaningful analyses on their effect on performances. Nevertheless, we tested for differences between SSRI takers (n = 13), and non-takers (n = 15) in PROD, IMIT, SPONT, recognition and TAS20 but found no significant differences (all t ≤ 1.19, p ≥ .25). Finally, we re-ran the main comparisons for PROD, IMIT, SPONT, recognition, excluding the five patient subjects with antypsychotics. All t-tests remained significant (all t ≥ 2.99, p ≤ .005).

3.6. Apathy

In order to exclude the possibility that EMG differences for HD were due to apathy, we repeated the main analyses for RECOGNITION, PROD and IMIT excluding the five subjects with apathy scores >0. All t-tests remained significant ($t_{REC} = -5.4$, $p < .001$; $t_{PROD} = -4.17$, $p = .001$; $t_{IMIT} = -3.59$, $p = .002$; 2.36, $p ≤ .028$).

3.7. Correlations between recognition, imitation, production and alexithymia

We correlated average REC with TAS20 as well as average EMG IMIT and PROD scores. Average IMIT and PROD scores, in order to equally weight all emotions, were calculated as follows: [CHEEK−happy + (FROWN−angry + NOSE-angry)/2 + (FROWN-disgusted + NOSE-disgusted)/2]/3. Average REC was correlated with TAS20 ($\rho = -.41$, $p = .006$), IMIT ($\rho = .55$, $p < .001$) and PROD ($\rho = .54$, $p < .001$) over all subjects and effects remain for patients only ($\rho_{TAS20} = -.50$, $p = .01$; $\rho_{IMIT} = .49$, $p < .009$; $\rho_{PROD} = .46$, $p < .02$). In a second step, we re-ran correlations between REC

and IMIT/PROD, partialling out TAS20. Both correlations persisted, over all subjects ($r_{IMIT} = .51$, $p = .001$; $r_{PROD} = .54$, $p < .001$) and for patients only ($r_{IMIT} = .43$, $p = .050$; $r_{PROD} = .43$, $p < .050$).

3.8. VBM results

Grey matter voxels in left posterior fusiform gyrus, left somatosensory cortex and posterior parietal cortex, bilateral pSTS, posterior bank of the lateral fissure, bilateral caudate nucleus, left subcentral gyrus, and right middle temporal gyrus (BA21/22) were correlated with average recognition (z-) scores (REC). See Fig. 6. Two posterior clusters were found to correlate with IMIT, in the occipital lobe and posterior cingulate. Within the 16 mm-sphere search volume of clusters found for REC, we additionally found several clusters in the middle temporal gyrus/right anterior STS (BA21/22), see Fig. 6b. No correlations were observed for grey matter and PROD or SPONT. All significant clusters are summarized in Supplementary Table 1.

4. Discussion

Patients with HD are globally impaired at recognizing emotion expressions. Here we followed up on the recently reported emotion expression deficits in these patients (Hayes et al., 2009; Trinkler et al., 2013). Following theories of embodied cognition (Decety & Jackson, 2004; Gallese, 2007; Keysers & Gazzola, 2007; Niedenthal, 2007), which state that perceiving emotions in others ties to our own emotional representations, we asked if HD patients show impaired representations for both own and others' emotions. Our goal was to find out if the
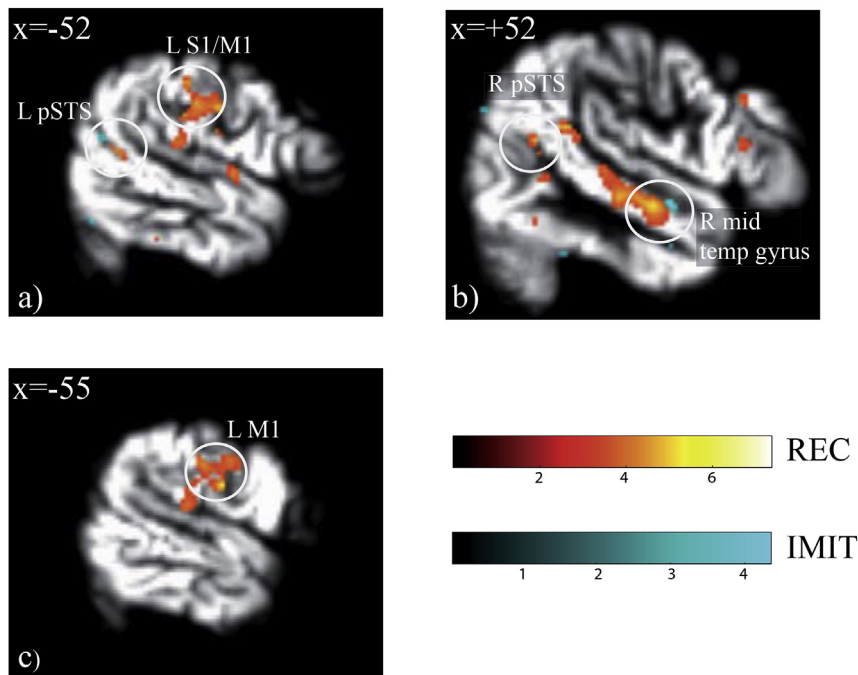

Fig. 6 — Brain areas in which grey matter volume correlated with emotion recognition (REC, orange) and EMG-imitation scores (IMIT, cyan). a) left (L) somatosensory (S1), motor (M1) and posterior superior temporal sulcus (pSTS), b) right (R) pSTS and anterior middle temporal gyrus, c) L motor cortex (M1). Displayed on one patient's segmented, DARTEL-transformed and smoothed 3D T1 image at p = .005 uncorr. for better visibility.

joint expression — recognition deficit might be tied to a more motoric level of impairment. For this, we used EMG to measure not only expression on command, but also spontaneous and voluntary imitation which does not necessarily draw on intact internal experience. Complementarily, we tested emotion identification on the level of internal experience using alexithymia questionnaires.

We found impaired EMG signals of imitation, paralleling impaired EMG signals of voluntary production of emotional facial expressions. These impairments were accompanied and correlated with impaired emotion recognition, but contrasted with intact emotion identification on an internal level of emotional experience, thus permitting to tie the impairment to the motoric level of emotion expression in HD.

4.1. Impaired voluntary imitation of emotional facial expressions

Former studies have shown impaired production of instructed expressions across all emotions in HD (Hayes et al., 2009; Trinkler et al., 2013), as well as impaired spontaneous disgust expressions (Hayes et al., 2009). Here we replicated these findings using EMG. We found significantly reduced EMG activity in HD compared to controls for the voluntary production of happy, angry and disgusted expressions. Note that we were somewhat unsuccessful to capture disgust expressions in both patients and controls in our third electrode location nose wrinkler (NOSE). This might be related to the fact that the disgust expression is more variable and has no single muscle activation exclusively and universally attributed to it (Ekman, Friesen, & Hager, 2002; Waller et al., 2008), whereas for happy and angry expressions there exist necessary muscle activations, including *zygomaticus major* and *corrugator supercilii* respectively. In our study, both disgust and angry expressions were mostly captured by a combination of FROWN and NOSE in normal controls, congruent with reports that FROWN also captures some *levator labii superioris alacque nasi* activation (Tassinari, Cacioppo, & Green, 1989), attributed to disgust. Notably, our EMG results demonstrate that HD patients show qualitatively similar EMG activity as normal controls, but to a significantly lesser extend. Since the activation curves are clearly distinct per emotion-muscle, we are confident that we measured emotion-specific facial muscle activation.

Most importantly however, impaired production of emotion expression in HD is paralleled by impaired imitation. Imitation does not necessarily require intact emotion identification on a level of internal experience. Therefore, our EMG results provide support that shared emotion representations might be impaired at a motor level in HD. Concordantly, recognition and imitation were highly correlated, and this correlation was not driven by emotional awareness, since it persisted after we partialled out emotional awareness (as measured by the alexithymia questionnaire).

4.2. Impaired spontaneous mimicry

Controls showed differential micro-modulation of cheek and frown activity when observing emotional expressions, replicating the well known facial mimicry effect for happy versus angry facial expressions (Achaibou et al., 2008; Dimberg, 1982).

Controls showed no mimicry effect for disgust. Other studies have also reported inconsistent NOSE mimicry for disgust (Blairy, Herrera, & Hess, 1999). More importantly, the observed mimicry effect in the form of a happy — angry antagonism was completely absent in HD patients. Even if, as has been discussed widely (Hess & Blairy, 2001), mimicry does not necessarily mean that an emotion has been automatically induced, mimicry might serve an important purpose in communication. This is evidenced by the observation that people imitate somebody else's painful face more when they are aware that their expression is visible to the other (Bavelas, Black, Lemery, & Mullett, 1986). In HD, the absence of mimicry might cause the disruption of emotional resonance processes in social interactions. Perhaps their addressees will feel less empathized with. This might add to the perception of family and caregivers that HD patients are selfish and lack empathic care (Snowden et al., 2003).

4.3. Intact emotional awareness

The HD patients tested here clearly displayed no signs of alexithymia, in accordance with a former study (Trinkler et al., 2013). Since however, we could not possibly attempt to take absence of evidence as evidence of absence, alexithymia results are only complementary and may serve to refine our main, EMG, findings. Note further that some authors have previously reported significantly different and also very high alexithymia scores in manifest HD patients (Eddy & Rickards, 2015). Their patients were of comparable age and had comparable executive dysfunction as the group we previously tested (Trinkler et al., 2013) and the group here. This might be because Eddy et al. used an older version of the test, but more importantly, they report no control for response bias. However, since the TAS do only partially control for response bias (five out of 20 items are inverted), it is possible that HD patients might show perseveration and as a result score very highly on this questionnaire. Further studies might follow up on the observed differences and test emotional awareness with more fine-grained but also lengthier procedures such as the Levels of Emotional Awareness Scale (LEAS, Lane, Quinlan, Schwartz, Walker, & Zeitlin, 1990).

Finally, there is additional evidence that emotions conveyed via a non-motoric route, namely via contextual cues (such as a graveyard implying sadness and dirty underwear being suggestive of disgust), are also processed correctly in HD patients (Aviezer et al., 2009), as is semantic emotion comprehension (Snowden et al., 2008). Furthermore, and even more strikingly, HD patients were shown to be more sensitive to the affective scenes of the International Affective Picture System than normal controls (Ille et al., 2011).

4.4. Anatomical correlates

Our VBM findings should be considered preliminary, since T1 images were only available for a subset of patients, hence we also found no brain volume correlations with PROD due to lack of power. Also, by applying no corrections for multiple comparisons in an attempt to avoid loss of power, we restrict our interpretations to areas for which we have *a priori* hypotheses based on pre-existing literature. These limitations

notwithstanding, anatomical correlates are largely compatible with the idea of a shared emotion representation impairment on a motor level in HD: We found brain volume differences correlated to emotion recognition in pSTS, posterior parietal and somatosensory cortices, areas previously attributed to shared visuo-motor action representations (Blakemore & Decety, 2001; Grezes et al., 2003; Grezes & Decety, 2001). They are congruent with areas activated in the observation and imitation of smiling (Hennenlotter et al., 2005) and emotion expressions over all (Carr et al., 2003). The implication of somatosensory cortices in this shared emotion network has further been demonstrated in a large lesion study (Adolphs et al., 2000; see also Keysers et al., 2010). Like previous authors, we found emotion recognition to correlate with striatal volume change in HD patients (Henley et al., 2008). This might be more particularly related to HD, however, one previous fMRI study has also reported striatal involvement in emotion imitation (Dapretto et al., 2006).

4.5. Implications for HD research

In synthesis, here we are able to show that the long and well known emotion recognition impairment in HD (Henley et al., 2012; Snowden et al., 2008; Sprengelmeyer et al., 1996) is part of a more global impairment encompassing emotion expression and even imitation, compatible with the hypothesis of impaired shared emotion representations on a motoric level. Shared emotion representations cover a large fronto-posterior network anatomically. This might be why they are susceptible to disruptions in HD, which is characterized by diffuse fronto-posterior brain atrophy (Tabrizi et al., 2009). The extensive nature of this network might further lie at the cause of emotion recognition being such a sensitive marker for disease progress in HD, making it a potentially powerful biomarker (Novak et al., 2012; Scahill et al., 2013).

4.6. Consequences for patients and caregivers, over and above HD

Here we establish the theory that in HD, emotion recognition is impaired due to a motor-based shared emotion representation impairment. The consequences of this impairment for affected individuals are that they will encounter difficulties not only in reading other people's emotions, but also expressing their own emotions to their family and caregivers non-verbally. We have further shown that different levels exist for sharing emotions, and that a motor level of emotion expression can be dissociated from a level of internal experience. In HD, seemingly, the latter is spared, permitting emotion understanding via other routes, such as verbal-conceptually (see Snowden et al., 2008) or with situational aids (see Aviezer et al., 2009).

What this research teaches us beyond HD, is that in any patient presenting with emotion recognition impairments it might be useful to adopt an embodied stance and consider firstly that the impairment might be two-sided, affecting both the sender and the receiver end of emotion communication. Secondly, within a shared emotion representation impairment, dissociable sub-levels might be preserved, and, typically, faulty expression of emotion can go along with intact feeling. Patients and their caregivers can be helped by focusing on intact levels for facilitating social communication. This applies typically to cases such as Parkinson's Disease (Adolphs, Schul, & Tranel, 1998; Bowers et al., 2006; Clarke, Neargarder, & Cronin-Golomb, 2008; Jacobs, Shuren, Bowers, & Heilman, 1995; Rinn, 1984; Simons, Ellgring, & Pasqualini, 2003; Sprengelmeyer et al., 2003), but perhaps even to psychiatric disorders such as schizophrenia (e.g. Brüne, 2005; Kahn, 2014; Kohler et al., 2003).

In all of these cases it might appear possible to establish dissociable levels of emotion sharing. Moreover, re-education schemes could make use of the partial dissociability of these levels. They might on the one hand re-train motor expression levels wherever possible, and on the other hand provide patients and their social partners with other (verbal-conceptual or contextual) routes to empathy.

Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.cortex.2017.02.019.

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