Emotion recognition and experience in Huntington’s disease: Is there a differential impairment?

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Abstract

Findings on affective processing deficits in Huntington’s disease (HD) have been inconsistent. It is still not clear whether HD patients are afflicted by specific deficits in emotion recognition and experience. We tested 28 symptomatic HD patients and presented them with pictures depicting facial expressions of emotions (Karolinska-Set) and with affective scenes (International Affective Picture System; IAPS). The faces were judged according to the displayed intensity of six basic emotions, whereas the scenes received intensity ratings for the elicited emotions in the viewer. Patients’ responses were compared with those of 28 healthy controls. HD patients gave lower intensity ratings for facial expressions of anger, disgust and surprise than controls. Patients’ recognition deficits were associated with reduced functional capacity, such as problems with social interactions. Moreover, their classification accuracy was reduced for angry, disgusted, sad and surprised faces. When judging affective scenes for the elicitation of happiness, disgust and fear, HD patients had a tendency to estimate them as more intense than controls. This finding points to a differential impairment in emotion recognition and emotion experience in HD. We found no significant correlations between emotion experience/recognition ratings and CAG repeats, symptom duration and UHDRS Motor Assessment in the patient group.

1. Introduction

Huntington’s disease (HD) is an autosomal dominant, neurodegenerative disorder. The disease is caused by an unstable expansion of the trinucleotide repeat cytosine–adenine–guanine (CAG) of the gene IT-15 on the short arm of chromosome 4 that codes for the protein huntingtin (Htt). CAG expansion beyond 35 repeats is associated with the expression of HD (Langbehn et al., 2004). Intraneuronal inclusions of the aggregated mutant Htt lead to progressive cerebral degeneration starting in the caudate nucleus and in the putamen (Douaud et al., 2006; Bohanna et al., 2008). HD is characterised by increasing deterioration in motor function, psychiatric disturbance and cognitive impairment (Bohanna et al., 2008; Henley et al., 2009). There is a close relationship between the number of CAG repeats and age of disease onset as well as its progress (Langbehn et al., 2004). HD patients usually develop changes in personality and emotional functioning during the progress of disease, e.g., increase of hostility, impulsivity, depression and anxiety (Thompson et al., 2002; van Duijn et al., 2007).

One particular deficit in affective processing that has been identified in several studies relates to a selective disgust recognition deficit in preclinical gene-carriers (Gray et al., 1997; Hennenlotter et al., 2004; Sprengelmeyer et al., 2006). Such a deficit can be explained by neurobiological emotion models suggesting that basic feelings (e.g., disgust and fear) are mediated by specific central affect programs initiated in localized brain regions (Calder et al., 2001). Several studies led to the assumption that recognition of disgust stimuli relies on the basal ganglia (particularly the putamen) and the anterior insula (Phillips et al., 1997; Sprengelmeyer et al., 1998; Hennenlotter et al., 2004). Results by Kipps et al. (2007) showed a strong linear correlation between disgust recognition and insula volume in HD.

However, findings on symptomatic HD do not support the idea of a specific disgust processing deficit. Three studies on clinically manifest HD found impairments for various negative emotions but all investigated very small samples (Sprengelmeyer et al., 1996; n = 13; Wang et al., 2003; n = 6; Montagne et al., 2006; n = 8). Furthermore, Milders et al. (2003) revealed for 20 symptomatic HD patients that fear recognition was most severely impaired, but there were also problems in recognising facial expressions of anger, disgust and sadness. Snowden et al. (2008) found poorer performance in 10 symptomatic HD patients for the recognition of anger and to a lesser extent disgust. The manifest HD group in the study of Henley et al. (2008) was significantly worse than controls at recognising facial expressions of surprise, disgust, anger...
and fear. Impaired recognition of each emotion was associated with striatal volume loss. The results of Johnson et al. (2007), studying a large sample of 475 pre-symptomatic individuals, revealed that already gene-carriers performed worse than controls concerning all negative emotions.

In contrast to emotion identification the elicitation of feelings by the presentation of emotion-relevant scenes has hardly been studied in HD. The only study by Hayes et al. (2007) with 15 HD patients found that fewer scenes were classified as disgusting by symptomatic HD patients than by controls. Similar to disgust recognition it has been suggested that the experience of disgust feelings recruits the insula. In an fMRI investigation by Stark et al. (2007) participants’ reported disgust feelings were correlated with activation of the insular cortex. However, other studies did not observe a specific involvement of a basal gangia-insular circuit in the central representation of disgust experience (e.g. Schienle et al., 2005).

In summary, findings on affective processing dysfunctions in HD are inconsistent. One reason for this concerns small sample sizes in most studies, which limits the power to detect more subtle group differences and enhances the possibility of sample-dependent results. Furthermore, severity of symptoms, influenced by CAG length and disease duration, differed between studies. Unspecific symptoms can precede motor symptoms, which are commonly used as markers of clinical manifestation (Langbehn et al., 2004). Approximately 60% of patients start with mental and emotional dysfunctions (Di Maio et al., 1993), which are often not immediately attributed to HD. Specificity of emotion recognition deficits might vary with disease progression, starting with impaired disgust perception in asymptomatic gene-carriers, as revealed in two studies (Gray et al., 1997; Sprengelmeyer et al., 2006).

Adequate emotional experience and understanding other persons’ emotional behaviour are important for social functioning and quality of life. HD results in a complex symptomatology especially during progressed stages, including impairment of voluntary motor functions, cognition, language and mental functions. HD is related to multiple psychosocial problems in the afflicted persons and their families. Abnormal emotional processing has negative implications for patients’ daily lives, especially for social interactions. Findings of previous studies already demonstrated that the psychosocial well-being of patients with HD is strongly affected (e.g., Helder et al., 2001).

The primary objective of this study was to determine whether the impairment of facial disgust recognition in symptomatic HD extends to the experience of visually induced disgust. We asked the participants to rate facial expressions according to the displayed emotion intensities for all basic emotions (graded choice). This allowed the analysis of emotion recognition deficits that might vary with disease progression, starting with impaired disgust perception in asymptomatic gene-carriers, as revealed in two studies (Gray et al., 1997; Sprengelmeyer et al., 2006).

The Trait scale of the Questionnaire for the Assessment of Disgust Proneness (QADS; Schienle et al., 2002a) measures disgust propensity and describes 37 situations, which have to be judged on a 5-point scale with regard to the experienced disgust (0 = ‘not disgusting’, 4 = ‘very disgusting’); e.g., ‘You are just about to drink a glass of milk as you notice that it is spoiled’. The Cronbach’s alpha of the total scale is 0.90. The Trait scale of the State–Trait Anxiety Inventory (STAI; Laux et al., 1981) measures the frequency of anxious feelings on a 4-point scale. The Cronbach’s alpha of the scale is 0.88.

Habitual emotional reactivity was assessed by self-report inventories: The Beck Depression Inventory (BDI; German version: Hautzinger et al., 1994) assesses depressive symptomatology. Cronbach’s alpha is 0.88. The Questionnaire for the Assessment of Disgust Proneness (QADS; Schienle et al., 2002a) measures disgust propensity and describes 37 situations, which have to be judged on a 5-point scale with regard to the experienced disgust (0 = ‘not disgusting’, 4 = ‘very disgusting’); e.g., ‘You are just about to drink a glass of milk as you notice that it is spoiled’. The Cronbach’s alpha of the total scale is 0.90. The Trait scale of the State–Trait Anxiety Inventory (STAI; Laux et al., 1981) measures the frequency of anxious feelings on a 4-point scale. The Cronbach’s alpha of the scale is 0.88.

The Trait scale of the State–Trait Anxiety Inventory (STAI; Schwenkmezger et al., 1992) assesses trait anger as well as anger expression. All items are rated on 4-point scales. Internal consistency of the STAI is 0.90.

2.3 Stimuli for the picture perception tasks

All participants viewed emotional scenes and facial expressions on a computer screen (notebook, 15 in.). The participants sat at about 50 cm from the screen. Before starting the experiment participants were asked for their understanding of basic emotions by a short verbal description.

a) Forty-two pictures with emotional facial expressions depicting happiness (6), fear (6), sadness (6), anger (6), disgust (6), surprise (6), and a neutral affective state (6) from the Karolinska-Set (Lundquist et al., 1998) were presented. Half of the poses were female, half were male.

b) Twenty-four emotion-relevant scenes for the induction of happiness (6), fear (6), disgust (6), and an affectively neutral state (6) were presented. Most scenes were taken from the International Affective Picture System (IAPS; Lang et al., 2001). Inducing-disguising pictures were created by Schienle et al. (2002b) and included scenes with animals (magnets, bluebottles, and sloths), a dirty toilet, cactus and an ecremumphate face. The fear-inducing pictures showed threatening situations either through attacks of animals (‘dog with its teeth bared’, IAPS 1300; ‘white shark’; developed by the authors) or human attacks (‘man threatening a woman with a knife’, IAPS 6330; ‘men with pistol’, IAPS 6230; ‘war scene’, IAPS 6940; ‘masked robber’, IAPS 6370). Happy pictures included animals (‘baby seal’, IAPS 1440, M = 9.75, S.D. = 3.20). Handedness was taken as the hand used to write with. All patients with exception of one woman were right-handed. Fourteen of the patients (50%) received no pharmacologic treatment. Antidepressants or/and antipsychotics were applied to fourteen patients (antidepressants: 7 patients, antipsychotics: 2 patients; both: 5 patients). A conducted analysis of variance with the factors medication (medicated vs. non-medicated) and emotion (fear, anger, sadness, disgust, happiness, and surprise) revealed a non-significant mediation effect for the ratings of affective faces and scenes and a non-significant mediation effect interaction (all Fs < 1.36, all Ps > 0.260).

We further tested 28 mentally healthy subjects, 17 men and 11 women, matched for sex, age and socio-economic status. They had been recruited by advertisements in a local newspaper. The control group underwent the Mini-DIPS to exclude the presence of mental disorders. Age of controls ranged between 31 and 63 years, with a mean age of 47.2 years (S.D. = 7.5). Sixteen subjects of the control group (57.1%) were employed. Mean education level was 10.7 years (S.D. = 1.6). Controls were right-handed with the exception of one man and one woman. Groups did not differ in age (t(54) = 0.52, n. s.) and years of education (t(54) = 1.97, n. s.).

Exclusion criteria for patients and for controls were dementia (TDD-Score < 35; Ihl et al., 2000), substance abuse, and current use of sleep-inducing drugs or sedatives. The presence of neurological disorders for controls and of other neurological disorders besides HD for patients was also an exclusion criterion.

The project had been approved by the ethics committee of the University of Graz (vote number 18–138 ex 06/07). All participants gave informed written consent to the study.
controls: M = 3.89; t(no group effects for trait anxiety (STAI; patients: M =4.29, S.D. = 2.34; t(54) = −8.22, P = 0.001), with patients showing lower scores. Groups did not differ in depressive symptoms (BDI; patients: M = 4.89, S.D. = 4.50; controls: M = 3.61, S.D. = 3.33; t(54) = 1.22, n.s.) and disgust propensity (QADS; patients: M = 2.13, S.D. = 0.93; controls: M = 2.01, S.D. = 0.70; t(54) = 0.56, n.s.). We also found no group effects for trait anxiety (STAI; patients: M = 34.66, S.D. = 8.16; controls: M = 32.50, S.D. = 7.43; t(54) = 0.94; n.s.) and trait anger (STAXI; patients: M = 15.19, S.D. = 4.08; controls: M = 16.62, S.D. = 3.89; t(54) = −1.33; n.s.).

The stimulus material had been matched for item difficulty, complexity, brightness and colour. Since the IAPS does not include pictures which reliably induce anger, sadness and surprise these categories were omitted. It is known that IAPS scenes which should induce sadness or anger usually produce mixed emotions (e.g. 50% anger and 50% sadness). Affective neutral stimuli were included as a control (reference) condition.

Each picture was presented for a maximum of 15 s. Pre-tests had displayed that this time was reported as sufficient for identifying the pictures also by HD patients. The presentation could be terminated early by pressing a button on a three-button device, which should induce sadness or anger usually produce mixed emotions (e.g. 50% anger and sadness and surprise these categories were omitted. It is known that IAPS scenes which should induce sadness or anger usually produce mixed emotions (e.g. 50% anger and 50% sadness). Affective neutral stimuli were included as a control (reference) condition.

All statistical analyses were carried out using PASW Statistics 18.0 for Windows. We computed mean emotion intensity ratings for affective scenes and facial expressions for each emotional condition and all six basic emotions. Repeated measures analyses of variance (2 × 6 ANOVAs) were carried out with the independent factors group (HD and Control) and sex (male and female) and the repeated measure factor emotion (fear, disgust, sadness, anger, happiness, and surprise). Alpha level significance was set at 0.05 for all statistical tests. To account for multiple comparisons we used the method by Shaffer (1986) with a family type I error of α = 0.05 for all exploratory analyses. Effect size were calculated by Cohen’s d.

3. Results

3.1. Group comparisons for the questionnaires

Cognitive performance (TFDD) between HD patients and controls differed significantly (patients: M = 39.18, S.D. = 3.92; controls: M = 46.29, S.D. = 2.34; t(54) = −8.22, P = 0.001), with patients showing lower scores. Groups did not differ in depressive symptoms (BDI; patients: M = 4.89, S.D. = 4.50; controls: M = 3.61, S.D. = 3.33; t(54) = 1.22, n.s.) and disgust propensity (QADS; patients: M = 2.13, S.D. = 0.93; controls: M = 2.01, S.D. = 0.70; t(54) = 0.56, n.s.). We also found no group effects for trait anxiety (STAI; patients: M = 34.66, S.D. = 8.16; controls: M = 32.50, S.D. = 7.43; t(54) = 0.94; n.s.) and trait anger (STAXI; patients: M = 15.19, S.D. = 4.08; controls: M = 16.62, S.D. = 3.89; t(54) = −1.33; n.s.).

3.2. Group comparisons for the picture perception tasks (Table 1, Figs. 1 and 2)

3.2.1. Facial expressions

a) Intensities of target emotions: analyzing the intensity ratings of the six target emotions (anger, disgust, sadness, fear, happiness, and surprise; e.g., disgust intensity in faces depicting disgust) we revealed a significant main effect for emotion (F(5, 260) = 20.69, P = 0.001), for group (F(1, 54) = 8.55, P = 0.005), and a significant emotion × group interaction (F(5, 260) = 2.30, P = 0.046). Patients estimated target emotions in affective faces less intense than controls (mean difference (md) = 0.85), concerning anger (md = 1.40, P = 0.001; effect size: Cohen’s d = 0.98), disgust (md = 1.27, P = 0.023; d = 0.63) and surprise (md = 0.88, P = 0.024; d = 0.63; see Fig. 1, left panel). Also, patients perceived more happiness than controls in angry, fearful and sad facial expressions (all mds > 0.73, all Ps < 0.037). We found no significant sex × group interaction for intensity estimations of affective faces.

b) Classification accuracy of target emotions: for assessing participants’ classification accuracy of facial expressions we calculated the difference between the rated target emotion intensity and the mean of all non-target intensity ratings (e.g., classification accuracy for a disgust expression = disgust intensity minus mean intensity (anger, fear, sadness, happiness, and surprise.

Analyses of classification accuracy of target emotions relative to non-target emotions revealed a significant main effect for emotion (F(5, 260) = 57.13, P = 0.001) and for group (F(1, 53) = 11.90, P = 0.001), and a significant emotion × group interaction (F(5, 260) = 2.89, P = 0.015). Patients showed lower classification accuracy concerning anger (md = 1.91, P = 0.001; d = 1.12), disgust (md = 1.72, P = 0.003, d = 0.85), surprise (md = 1.15, P = 0.028, d = 0.60), and sadness (md = 1.09, P = 0.050, d = 0.54; see Fig. 2) than controls.

c) Perception of neutral faces: analyses of neutral faces’ ratings revealed a main effect for the six basic emotions (F(5, 260) = 139.7, P = 0.001), a marginally significant group effect (F(1, 52) = 2.85, P = 0.098) and a significant emotion × group interaction (F(5, 260) = 4.21, P = 0.006). Patients reported marginally higher intensities for neutral faces than controls. Pairwise comparisons showed that patients perceived higher intensities of happiness (md = 1.17, P = 0.012; d = 0.70) and surprise (md = 1.54, P = 0.004; d = 0.81) in neutral faces than controls (see Table 1). Controls assessed sadness highest in neutral faces, differing significantly from all other emotions (all mds > 1.35, P < 0.010).

3.2.2. Affective scenes

a) Intensity of target emotions: intensity analyses for the three target emotions (happiness, fear, and disgust) showed significant main
effects for emotion ($F(2, 104) = 18.35, P<0.001$) as well as a marginally significant main effect for group ($F(1, 52) = 3.26, P=0.077$). Patients experienced emotions marginally more intense than controls. We found no significant sex×group interaction for intensity estimations of affective scenes (see Fig. 1, right panel).

b) Experience of neutral scenes: ratings of the neutral scenes displayed a main effect for the six basic emotions estimated ($F(5, 260) = 54.64, P<0.001$) and a significant emotion × group interaction ($F(5, 260) = 6.60, P=0.001$). The main effect for group reached marginally significance ($F(1, 52) = 3.82, P=0.056$). Patients tended to estimate neutral scenes as more intense than controls. Patients reported a higher intensity of happiness (md = 1.65, $d = 0.96$) and marginally also for surprise (md = 0.89, $P = 0.098; d = 0.45$) than controls.

### 3.2.3. Correlative analyses

a) Controlling for confounding variables: For both groups cognitive performance (TFDD) was not associated with emotion intensity ratings neither for recognition nor experience (all $r<0.32; n.s.$). There were no significant correlations between recognition and experience of target emotions in patients and controls (all $r<0.21; n.s.$).

b) Habitual emotion reactivity: patients’ fear experience and anxiety sensitivity were correlated ($r=0.46, P=0.014$). Their disgust experience (intensity) was correlated with anxiety sensitivity ($r=0.53, P=0.004$), trait anxiety ($r=0.42, P=0.027$) and trait anger ($r=0.48, P=0.014$). For controls correlations reached significance between fear experience and anxiety sensitivity ($r=0.60, P=0.001$).

### Table 1

|                  | HD        | Controls   |
|------------------|-----------|------------|
|                  | $N = 28$  | $N = 28$   |
| **Neutral faces**|           |            |
| Disgust          | 2.38 (1.72)| 2.13 (1.26)|
| Anger            | 3.24 (1.88)| 3.21 (1.60)|
| Fear             | 2.85 (1.57)| 2.94 (1.78)|
| Sadness          | 4.05 (1.94)| 4.17 (1.89)|
| Happiness        | 3.78 (1.84)| 2.61 (1.42)|
| Surprise         | 4.35 (2.17)| 2.81 (1.59)|
| **Neutral scenes**|          |            |
| Disgust          | 1.60 (0.80)| 1.54 (0.94)|
| Anger            | 1.80 (0.92)| 1.74 (1.01)|
| Fear             | 1.65 (0.82)| 1.70 (0.96)|
| Sadness          | 1.70 (1.00)| 1.64 (1.08)|
| Happiness        | 4.74 (1.79)| 3.09 (1.66)|
| Surprise         | 3.83 (2.15)| 2.95 (1.77)|

4. Discussion

In this study we investigated emotion recognition and emotion experience in symptomatic HD. The participants had been asked to judge the intensity of affective facial expressions as well as the intensity of emotional experiences elicited by affective scenes using graded choice over six basic emotions.

The analysis of the recognition task revealed lower intensity ratings of target emotions for angry, disgusted and surprised faces in HD patients compared to controls. This points to a quantitative recognition deficit in HD patients. Effect sizes were large for anger and medium for disgust and surprise. Besides this quantitative deficit, patients showed lower classification accuracy concerning facial expressions of anger and disgust (large effect sizes), and for sadness and surprise (medium effect sizes). The described deficits of our patients are in line with findings of Snowden et al. (2008) and Henley et al. (2008) who also identified the strongest impairment in HD for anger recognition. Fear recognition was comparably poor in patients as well as in healthy controls. This is in accordance with previous studies on healthy subjects where facial expressions of fear were typically recognised more poorly than other expressions (e.g., Ekman and Friesen, 1976).

The patients rated angry, fearful and sad faces as happier compared to controls. Also, they perceived neutral faces as happier and as more surprised than the healthy subjects. In contrast, the controls rated neutral faces as slightly sad, which is in line with earlier findings (e.g., Katsikitis, 1997; Lee et al., 2008). We hypothesize that this ‘positivity bias’ might be a consequence of patients’ focusing on easy to decode emotions, such as happiness. Negative emotions are more difficult to decipher and the patients clearly displayed recognition deficits for affects with negative valence. Due to the reduced recognition of negative emotions, the sensitivity for signs of positive emotions might be enhanced.

Altogether, HD patients showed quantitative and qualitative deviations from healthy subjects as to emotion recognition. There were differences regarding the perceived intensity as well as the distinction of emotions with negative and inconclusive valence. We could also display that these deviations were not specifically linked with patients’ poorer cognitive performance.
Our second main objective was to determine if the impairment in emotion recognition extends to experience. Interestingly, patients had a tendency to give higher intensity ratings for all experienced emotions (happiness, fear, and disgust) than controls. The group difference in experienced emotion intensity was only marginally significant and therefore should be interpreted with caution. However, our data clearly show that HD patients responded differently to affective scenes (increased intensity ratings) compared to affective faces (decreased intensity ratings). Perhaps HD patients start to focus more on their own emotions because the decoding of emotions in others becomes more and more difficult as the disease progresses. The diminished perception of emotions in others might lead to a (compensatory) enhancement of one’s own feelings. Another explanation is also possible. Since HD patients experience strong emotions and are overwhelmed by them, this might interfere with the correct perception of emotional cues in others.

Moreover, patients and controls rated neutral scenes differently. They displayed a positivity bias (similar to the positivity bias for neutral faces) and experienced more happiness and surprise when viewing neutral scenes relative to the healthy participants. Future studies are needed in order to replicate this observation.

Hence our data show an HD-related differential impairment in emotion recognition and experience. Such a deficit seems possible as the two processes recognition and experience were independent from each other. The intensity ratings for emotional faces and scenes (for a given target emotion) were not correlated with each other, neither in patients nor in controls.

We also investigated whether heterogeneous findings on disgust recognition and experience in HD may be due to an influence of personality and gender-related factors. But we found no difference between patients and controls in habitual emotional reactivity concerning disgust, anxiety, anger and depression. This is in line with results of previous questionnaire assessments (Sprengelmeyer et al., 2006; Hayes et al., 2007; Johnson et al., 2007). We also found no gender-specific effects in HD. Associations between experience of visual emotional stimuli and habitual emotional reactivity concerned anxiety sensitivity, trait anxiety, disgust sensitivity, and trait anger being related to experienced disgust and fear. This corresponds with previous studies (e.g., Schienle et al., 2005).

Although no findings exist from previous studies on symptomatic HD we expected symptom severity to affect emotion processing. We found associations between functioning in everyday life and emotion recognition deficits. Huntington disease has a significant impact on the social life. Deficient comprehension of other persons’ affect can lead to a breakdown in interpersonal relationships, especially when linguistic functionality becomes worse with progression of disease. Especially impaired perception of others’ anger and fear may result in negative consequences for communication. Our results point to a relationship between these deficiencies and everyday functioning. Therefore, therapeutic approaches should address this aspect and supply emotional recognition trainings for symptomatic patients.

As a limitation of the present study it needs to be mentioned that the two applied emotional tasks differed in their difficulty. The emotion recognition task included six emotions, whereas the experience task only contained three conditions. This might have influenced the ratings. Furthermore, we plan to study a larger sample of HD patients in order to replicate the findings on enhanced emotion experience.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at doi:10.1016/j.psychres.2011.04.007.

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