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Perception of Emotional Facial Expressions at Different Intensities in Early-Symptomatic Huntington's Disease

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Key Words
Neuropsychology · Huntington's disease · Basal ganglia disorders · Affective processing

Abstract
Background: While there is abundant evidence that patients with Huntington's disease (HD) have an impairment in the recognition of the emotional facial expression of disgust, previous studies have only examined emotion perception using full-blown facial expressions. ***Objective:*** The current study examines the perception of facial emotional expressions in HD at different levels of intensity to investigate whether more subtle deficits can be detected, possible also in other emotions. ***Method:*** We compared early symptomatic HD patients with healthy matched controls on emotion perception, presenting short video clips of a neutral face changing into one of the six basic emotions (happiness, anger, fear, surprise, disgust and sadness) with increasing intensity. Overall face perception ability as well as depressive symptoms were taken into account. ***Results:*** A specific impairment in recognizing the emotions disgust and anger was found, which was present even at low emotion intensities. ***Conclusion:*** These results extend previous findings and support the use of more sensitive emotion perception paradigms, which enable the detection of subtle neurobehavioral deficits even in the pre- and early symptomatic stages of the disease.

Introduction

Huntington's disease (HD) is characterised by motor impairments, psychiatric disturbances and dementia [1], but the onset and severity of these symptoms differ. Behavioural changes [2] and impairments in affective processing [3, 4] are often reported in the early stages. With respect to the perception of facial emotional expressions, a specific deficit in the perception of the emotion disgust has consistently been reported [3, 5–9], which may be related to dysfunction of the basal ganglia and insular cortex [9]. Deficits in the perception of other emotions, i.e. fear, anger, surprise and sadness, have been reported as well [3, 7, 9], but appear to be less prominent. These findings are clinically relevant, since the perception of facial expressions is important for efficient social interaction in everyday life.

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Previous studies in pre-symptomatic HD have examined the labelling of full-blown facial expressions [7]. Although HD patients are impaired in recognizing full-blown expressions of disgust, ceiling effects have been reported even in patients with emotion-perception deficits [10]. Hence, static pictures of computer-generated morphs displaying mixtures of two emotions (e.g., a morphed facial expression containing 10% sadness and 90% disgust) have been used to make the task more sensitive, the so-called Emotion Hexagon Task [3, 11, 12], again indicating that HD patients are poor at identifying expressions containing the emotion disgust. However, although the facial expressions we encounter in daily life are generally dynamic (i.e., we see a neutral face change into an angry one), all previous studies on emotion perception in HD have used static stimuli. Furthermore, previous studies have almost exclusively used facial stimuli from the Ekman and Friesen [13] set containing black and white pictures that appear somewhat out of date. The current study investigates in more detail the perception of affective facial expressions at different levels of intensity in early symptomatic HD, using a newly developed task in which short video clips showing coloured pictures of actors mimicking all six basic emotions are presented morphing from a neutral face into different levels of emotional intensity.

Subjects and Methods

Eight HD patients (5 men; mean age 46.4 years, SD = 11.2), were recruited from the Department of Neurology at the Leiden University Medical Centre (LUMC) in the Netherlands. All patients were clinically diagnosed, supported by genetic tests, and in the early symptomatic stage as confirmed by the Unified Huntington Disease Rating Scale motor scale (mean total motor score 17.1, SD = 6.2) [10]. The Mini Mental State Examination (MMSE) [10] was performed in order to exclude patients with possible dementia; 1 female patient obtained an MMSE score of 21 and was excluded from all further analyses. The mean MMSE for the remaining patients was 29.1 (SD = 1.22, range 27–30). While it should be noted that the MMSE does not screen all neurocognitive domains, all patients were independently functioning in activities of daily living and hence did not fulfil the DSM-IV criteria of dementia syndrome [14]. Thirty age- and education-matched healthy control subjects participated (14 men, mean age 39.0 years, SD = 11.1), without a history of psychiatric or neurological disease and without depressive symptoms as measured with the Beck Depression Scale and the Hospital Anxiety and Depression Scale [15]. No significant differences were found between the HD patients and the controls for age or education level (all t values <2.1). The study was approved by the Medical Ethical Committee of the LUMC, and informed consents were obtained from all participants.

Materials

To measure the perception of facial expressions, a task was used to assess the accuracy in labelling different levels of intensity of emotional facial expressions, which included the emotions anger, disgust, fear, happiness, sadness and surprise [16]. Stimuli for the facial expressions were coloured pictures taken from actors (2 males and 2 females) who were asked to express the six different emotions and a neutral face. A computer-generated program, developed from algorithms designed by Benson and Perrett [17], constructed morphing stages between two endpoint facial expressions (one of which was always neutral) of the same actor. This resulted in morphed images between neutral (0% emotion) and full-blown expression (100% emotion) that were used to construct video clips that incrementally increase the degree of expression by 10% steps, starting at a 20% intensity level (see figure 1 for a schematic overview). The video clips were presented beginning with the lowest intensities (i.e., first all video clips showed expressions changing from neutral into 20% emotion, subsequently all clips changing from neutral into 30% emotions, and so on until the full-blown 100% expressions). The different types of emotion of the same intensity were presented in pseudo-random order to control for possible order effects of previously encountered emotion types that may influence the response [18]. In each trial, the subject was required to make a forced choice between one of six emotional expression labels displayed on the screen after the presentation of each clip, without time restrictions. Additionally, general facial perception was measured with the short version of the Benton Test of Facial Recognition [15].

Results

The accuracy for the various emotional intensities was calculated by means of the number of correctly recognized emotions at each intensity level. For each emotion, a 9 × 2 repeated measures ANOVA (GLM) was conducted with the group (HD versus control) as the between-subject variable, and intensity (20, 30, 40, 50, 60, 70, 80, 90, and 100%) as the within-subject variable. Group differences were found for the emotions disgust ($F_{1, 35} = 10.28$, $p < 0.005$) and anger ($F_{1, 35} = 4.50$, $p < 0.05$; fig. 1), but not for the emotions fear, happiness, surprise and sadness (all Fs <2.8). No interactions were found between group and intensity for any of the emotions (all Fs <1.51). In order to compare our results with previous studies using full-blown expressions, independent-sample t tests were performed for the emotions disgust and anger at a 100% intensity level. This revealed a significant difference between the patient group and the controls for the emotion disgust ($t_{35} = 2.97$, $p = 0.005$), but not for anger ($t_{35} = 0.40$, $p = 0.69$). These impairments were not due to an overall deficit in recognizing faces, as the performances on the Benton Test of Facial Recognition were all in the normal range (mean scores: patients = 21.1, controls = 22.9).

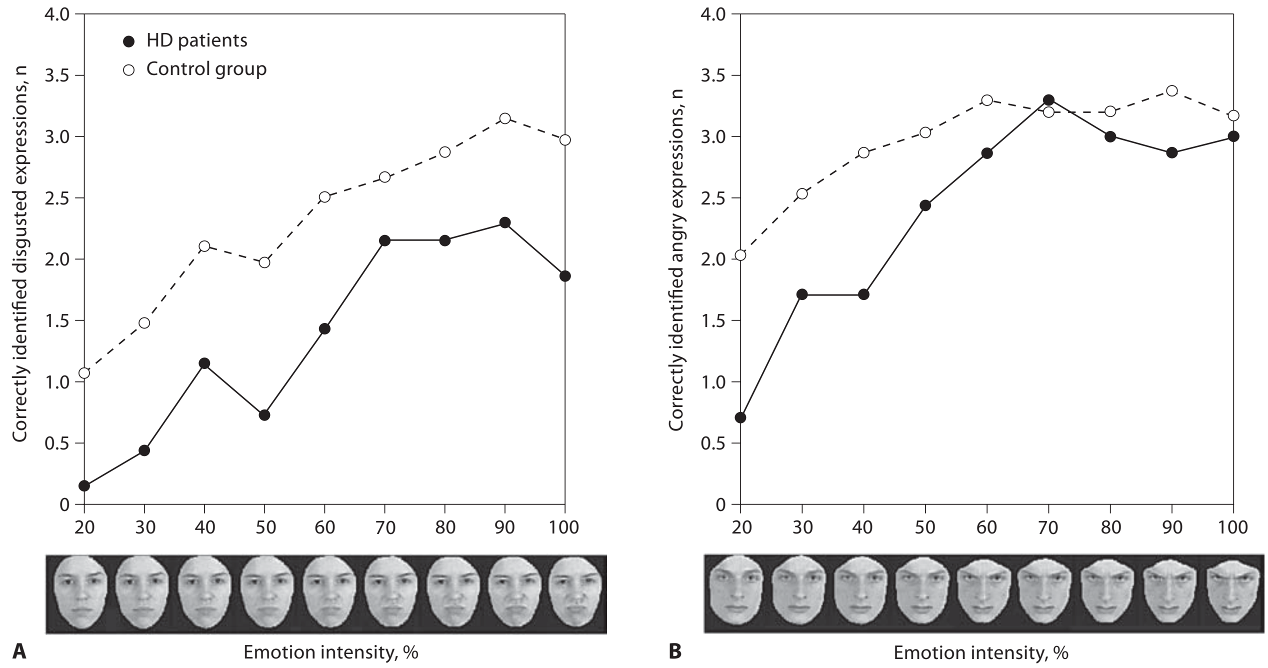

Fig. 1. Mean number of correctly identified emotional expressions (maximum 4) for the patients with Huntington's disease (HD) and healthy controls for the emotions disgust (**A**) and anger (**B**) morphing from a neutral expression to different levels of intensities (0–20, 0–30, etc. until 0–100% emotion).

Discussion

The objective of this study was to examine in detail the perception of emotional facial expressions of increasing intensity in HD patients. The results show specific impairments in recognizing the emotions disgust and anger on all levels of emotional intensities, in line with previous findings showing evidence for impairment in the recognition of facial expressions, specifically disgust, in the early stages of the disease [3, 5–9, 11]. Moreover, HD patients made more errors in recognizing angry faces correctly for all intensity levels taken together compared to the controls, but if only the full-blown expressions were taken into account, no difference was found between the patients and controls. While the recognition of disgust has been related to the basal ganglia and insular cortex [9], the underlying mechanisms of recognition of angry emotional expressions are unclear, but may be related to the ventral striatum [19]. Furthermore, imaging studies have demonstrated atrophy in these brain areas in HD [20]. Thus, the current findings on emotion perception can be related to neural structures that are dysfunctional in HD.

These findings support the use of more sensitive emotion perception tasks, rather than applying only full-blown facial expressions. Compared with the Emotion Hexagon Task, the present paradigm differs in two important aspects. First, the Emotion Hexagon Task always shows expressions mixing two emotion types that differ in intensity. Consequently, it is difficult to establish what the correct answer is in the Emotion Hexagon Task, in that the results always have to be interpreted compared to the control group. For example, healthy participants in most cases see a 30% sad/70% disgusted face as disgusted, whereas generally HD patients view this as sad [3]. While this response may be the result of an inability to perceive disgust, the response as such is not fully incorrect. Also, facial expressions consisting of two different emotions may appear to be somewhat unnatural. These problems are taken into account by the present task which shows expressions of different levels of emotional intensity as opposed to neutral. Since participants always have to make a forced-choice response (in which 'neutral' as a choice option is not possible), this results in responses that are either correct or incorrect. Second, the stimuli of the Emotion

Hexagon Task are static pictures of morphed expressions, while the current task showed dynamic video clips of faces changing from neutral into one of the basic emotions. Possibly, this resembles more closely the emotions we encounter in daily life, which in most cases are not still facial expressions. Methodological differences may also explain why impaired perception of angry faces in HD has not always been reported [3, 7].

The present study shows that deficits in emotion perception in HD are robust and can even be established in small sample sizes. Since establishing the course of HD-specific symptoms is currently already very important for many HD patients and gene carriers, assessment of emotion perception may become a clinically useful addition to the neurocognitive assessment of individual HD patients. Future studies should clarify whether the reduced capacity to recognise facial expressions in others is related to the behavioural changes in HD.

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