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Emotion perception and electrophysiological correlates in Huntington's disease

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HIGHLIGHTS

- This study confirms an emotional face processing deficit in Huntington's disease (HD).
- This study demonstrates the electrophysiological underpinnings of the emotional processing deficit in HD.
- This study suggests that the emotional processing deficit in HD is caused by a generic visual processing deficit.

ABSTRACT

Objective: This study aimed to characterise, emotion perception deficits in symptomatic Huntington's disease (HD) via the use of event-related potentials (ERPs).
Methods: ERP data were recorded during a computerised facial expression task in 11 HD participants and 11 matched controls. Expression (scrambled, neutral, happy, angry, disgust) classification accuracy and intensity were assessed. Relationships between ERP indices and clinical disease characteristics were also examined.
Results: Accuracy was significantly lower for HD relative to controls, due to reduced performance for neutral, angry and disgust (but not happy) faces. Intensity ratings did not differ between groups. HD participants displayed significantly reduced visual processing amplitudes extending across pre-face (P100) and face-specific (N170) processing periods, whereas subsequent emotion processing amplitudes (N250) were similar across groups. Face-specific and emotion-specific derivations of the N170 and N250 ('neutral minus scrambled' and 'each emotion minus neutral', respectively) did not differ between groups.
Conclusions: Our data suggest that the facial emotion recognition performance deficits in HD are primarily related to neural degeneration underlying 'generalised' visual processing, rather than face or emotional specific processing.
Significance: ERPs are a useful tool to separate functionally discreet impairments in HD, and provide an important avenue for biomarker application that could more-selectively track disease progression.

1. Introduction

With the development of up and coming drug trials in Huntington's disease (HD), there is an urgent need to identify potential biomarkers that can sensitively track disease progression and importantly that are functionally relevant (Georgiou-Karistianis et al., 2013a,b,c). Previous studies have identified emotion perception deficits in premanifest HD (pre-HD) individuals 15 years prior to estimated onset (and in the absence of cognitive change), suggesting that emotion alterations may be one of the earliest quantifiable behavioural changes observed preclinically (Gray et al., 1997; Stout et al., 2011). To this end further investigation of emotion perception in HD may offer new insights regarding early functional changes, which could provide an important avenue for biomarker development.

Specific emotion perception deficits in HD were initially documented by Sprengelmeyer et al. (1996), who used six basic

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emotions (happiness, sadness, surprise, anger, disgust, fear) and showed that of these, perception of disgust was the most severely impaired. Other published studies have reported similar findings in HD (Hennenlotter et al., 2004; Montagne et al., 2006). However, studies with differing findings questioned the degree of impairment for disgust and implicated a universal deficit across basic negative emotions, primarily anger and fear (Calder et al., 2010; Henley et al., 2008; Milders et al., 2003; Snowden et al., 2008). Negative behavioural symptoms include apathy, irritability and an increased incidence of depression, whilst there is an additional decrease in self-care and personal hygiene (Rosenblatt, 2007; Snowden et al., 2008). These types of behavioural changes are likely the result of basal ganglia dysfunction together with alterations in prefrontal function (Georgiou-Karistianis et al., 2013a,c; Gray et al., 2013). Conversely their origins may lie in the neural structures underlying perceptual and emotional processing of facial expression stimuli (Gray et al., 2007; Henson et al., 2003). Self-report questionnaires provide insight into self-assessed emotional responsiveness, which in some HD studies have shown trends and significance for lower self-assessed emotion (Sprengelmeyer et al., 1996). Ratings of emotion intensities and perceived emotionality have not been previously investigated in HD and may further substantiate a broader emotional effect that is not specifically a perceptual problem.

Further understanding of the neural substrates and circuitry involved in emotion perception may provide insight into the behavioural and psychiatric symptoms of HD. A sensitive technique that can examine emotion perception in HD is event related potential (ERP) methodology, a derivation of the electroencephalography (EEG). To our knowledge there are currently no published ERP emotion perception studies in HD. The use of ERPs has however been implemented in other areas of HD research (for review see Nguyen et al., 2010), where attenuated amplitudes (reduced coherent neural firing rates) across a range of ERP indices have been reported (e.g., Antal et al., 2003; Beste et al., 2008; Munte et al., 1997). Emotional faces can similarly be studied using ERPs. Of particular relevance is that following more generic visual processing (0–100 ms), specific structural encoding of the face occurs at circa 170 ms (termed the N170), followed by valance-dependent processing of the face at circa 250 ms (termed the N250). The N170 is primarily contributed to by fusiform area (FFA) and superior temporal gyrus (STG), whereas the N250 has less discreet sources (Bentin et al., 1996; Campanella et al., 2002; Henson et al., 2003). Assessment of the ERPs underlying emotional face processing can thus help delineate the disparate processes involved in this ecologically relevant function, and in particular can separate them from the motor-related functions required in performance measures such as reaction time and accuracy.

This study aimed to investigate for the first time the relationship between emotion perception deficits and underlying neurophysiological indices in symptomatic HD participants. We adopted a similar behavioural paradigm to that used previously (see Johnson et al., 2007), with negative (disgust and anger), neutral and positive (happy) emotion types to further substantiate whether deficits in emotion perception are disgust-specific or generalise to another negative emotion (anger). Moreover, by measuring ERPs, this study also characterised the nature of the reported neurophysiological modulation in HD in response to emotional faces. Consistent with previous research it was hypothesised that HD participants would display decreased *accuracy* of facial expression identification, particularly for negative emotions (anger and disgust), compared with controls. Further, and based on the assumption that the poorer accuracy would correspond to 'less' emotional processing, it was hypothesised that the (subjective) emotional *intensities* of these expressions would be reduced in HD compared to controls, particularly for negative expressions. Finally, given that the literature suggests that the emotional face processing deficit in HD is specific to negative expressions, it was hypothesised that HD participants would display attenuated *ERP amplitudes* relative to controls for the emotional decoding index (N250) but not the structural encoding index (N170), particularly for negative expressions.

2. Method

2.1. Participants

Twenty-two right handed [Edinburgh Handedness Inventory, EHI (Oldfield, 1970)], individuals aged 40–70 participated in the study. There were 11 HD participants (eight male, three female), all clinically diagnosed by a qualified neurologist (A.C). Disease progression was assessed via the Unified Huntington's Disease Rating Scale (UHDRS) motor examination (Huntington Study Group, 1996). Symptomatic HD participants[1] had a UHDRS motor score of >5. HD participants had previously undertaken genetic testing and CAG repeat length ranged from 40–47[2]. The HD sample was age, gender and IQ [National Adult Reading Test 2nd edition, NART-2 (Nelson et al., 1992)] matched to control participants.

In order to characterise the groups, participants completed a battery of neurocognitive scales and questionnaires, which were compared using independent samples Mann–Whitney U tests [Beck Depression Inventory – 2nd Edition, BDI-II (Beck et al., 1961), with HD participants scoring higher than control ($p < 0.001$); Hospital Anxiety and Depression Scale, HADS (Zigmond and Snaith, 1983), with HD participants scoring higher than control on depression ($p = 0.036$) but not the anxiety ($p = 0.17$) subscale; Positive and Negative Affect Schedule, PANAS (Watson et al., 1988), with reduced Positive (PA; $p = 0.03$) and a trend to increased Negative Affect (NA; $p = 0.07$ in the HD group); Olatunji et al. (2007) modification of the Disgust Scale-Revised (DS-R), with HD scoring higher on 'animal' ($p = 0.03$) and a trend towards higher scores on 'contamination' ($p = 0.060$) subsets, but no difference on 'core' ($p = 0.188$) compared to controls; Emotion Regulation Questionnaire, ERQ (Gross and John, 2003), where no group differences were found; and Orientations to Happiness Measure, OTH (Peterson et al., 2005), where no group differences were found]. See Table 1 for demographic, clinical data, and scores on all scales.

The current study was approved by the Monash University Human Ethics Board and each participant gave informed, written consent.

2.2. Procedure

Participants were prepped for the EEG and then moved into the EEG recording facility, where they rested for 5 min and then completed the facial expression task followed by the questionnaires and scales. For the facial expression task participants were provided with a thorough explanation, including familiarisation with each of the questions and response choices available, followed by a 5-min practice period during which care was taken to ensure understanding, and then the main task was given.

2.3. Materials and apparatus

2.3.1. Facial expression task

The emotional face perception task utilised three basic emotions, *happiness, anger* and *disgust,* and also included *neutral* as an

[1] UHDRS motor scores for two HD participants were not available.
[2] CAG repeat length for one HD participant could not be confirmed.