New perspectives on emotional processing in people with symptomatic Huntington's disease: impaired emotion regulation and recognition of emotional body language

Nicolò Zarotti[a1], Ian Fletcher[b], Jane Simpson[c]

[a, b, c] Division of Health Research, Faculty of Health and Medicine, Lancaster University, Lancaster, UK

[1] Corresponding author. Tel.: +44 01524 592858. E-mail: n.zarotti@lancaster.ac.uk. Address: Division of Health Research, Faculty of Health and Medicine, Lancaster University, LA1 4YG, Lancaster, UK.

Abstract

Objective: Emotion regulation and emotional body language (EBL) recognition represent two fundamental components of emotional processing that have recently seen a considerable surge in research interest, in part due to the role they play in optimising mental health. This appears to be particularly true for clinical conditions that can profoundly affect emotional functioning. Among these is Huntington's disease (HD), a neurodegenerative disorder that is associated with several psychological difficulties and cognitive impairments, including well-established deficits in facial emotion recognition. However, although the theoretical case for impairments is strong, the current evidence in HD on other components such as emotion regulation and EBL recognition is sparse.

Method: In this study, it was hypothesised that emotion regulation and recognition of EBL are impaired in people with symptomatic HD, and that these impairments significantly and positively correlate with each other. A between-subjects design was adopted to compare 13 people with symptomatic HD with 12 non-affected controls matched for age and education.

Results: The results showed that emotion regulation and EBL recognition were significantly impaired in individuals with HD. Moreover, a significant positive correlation was observed between facial and EBL recognition impairments, while EBL performance was negatively related to the disease stage. However, emotion regulation and recognition performances were not significantly correlated.

Conclusions: This investigation represents the first evidence of a deficit of emotion regulation and EBL recognition in individuals with HD. The clinical implications of these findings are explored, and indications for future research are proposed.

Keywords: Huntington’s disease, Emotional processing, Emotion regulation, Emotion recognition, body language.

Introduction

In the past few decades psychological research into human emotions has seen a surge of interest, especially due to the comprehensive conceptualisation of constructs such as emotional intelligence. Emotional intelligence is defined as the set of cognitive processes that allows the accurate expression and appraisal of emotions in others and the self (Goleman, 1995; Salovey & Mayer, 1990). In particular, the identification, understanding, facilitation, and management of emotions have been recognised as the four fundamental areas required for the successful processing of emotions. Within this framework, a pivotal role in social and affective functioning is played by emotion recognition and emotion regulation (Ochsner, 2009).

Emotion recognition can be defined as the process of correctly perceiving and identifying emotions in other people, as well as in artificial representations such as drawings or music (Mayer, Caruso, & Salovey, 1999). Historically, the most researched medium of emotion recognition is whole facial expression, such as pictures of faces of actors expressing basic emotions such as anger or fear (Henley et al., 2012). However, emotion recognition is a process mediated by a number of different features other than facial clues, and recognition via eyes, voices, and body language have also been investigated (Baron-Cohen, Wheelwright, Hill, Raste, & Plumb, 2001; Beatrice de Gelder & Van den Stock, 2011; Lima, Castro, & Scott, 2013). The latter medium in particular is gaining increased attention (de Gelder, 2006; Van den Stock, Righart, & de Gelder, 2007), since emotional body language (EBL) recognition has so far been neglected, despite being deeply involved in fundamental social cognitive skills such as empathy and decision-making (de Gelder & Hortensius, 2014).

Emotion regulation is defined as the "processes by which individuals influence which emotions they have, when they have them, and how they experience and express these emotions" (Gross, 1998; p. 275). More specifically, it involves the processes of selecting and modifying potential emotional situations, deploying attention, changing one's perspectives on emotions, and modulating emotional responses (Gross, 1998; 2015). In the last 20 years this area has seen a considerable increase in interest due to the recognition of its importance for psychological resilience and mental health (Aldao, Nolen-Hoeksema, & Schweizer, 2010; Ghorbani, Khosravani, Sharifi Bastan, & Jamaati Ardakani, 2017; Gross & Muñoz, 1995; van der Meer, van Duijn, Giltay, & Tibben, 2015).

Based on the theory of emotional intelligence, emotion recognition and emotion regulation are deeply interconnected processes, since emotions need to be correctly recognised before they can be regulated (Izard et al., 2001; Mayer, 2001; Salovey & Mayer, 1989; Yoo, Matsumoto, & LeRoux, 2006). This is also supported by evidence on the neural bases of both processes, which involve similar subcortical structures such as the limbic system and the basal ganglia (Gross, 2013). Not surprisingly, deficits of emotional processing are observed in many neurodegenerative conditions that involve damage to those structures, such as Parkinson's disease, Alzheimer's disease, multiple sclerosis, and – with a particularly well-established impact on emotion recognition skills – Huntington's disease (Löffler, Radke, Morawetz, & Derntl, 2015).

Huntington's disease (HD) is a genetic neurodegenerative disorder whose typical symptoms include involuntary movements (chorea), cognitive deterioration, and considerable psychological problems (Novak & Tabrizi, 2005). Its prevalence across North America, Europe, and Australia is 5.7 people per 100,000 (Pringsheim et al., 2012). The transmission mechanism is autosomal-dominant, meaning that affected individuals' children have a 50% probability of inheriting the gene, and genetic testing is available to ascertain gene status (in which case the term 'presymptomatic' is used). The onset is usually around age 40, and disease progression can be divided into five stages, starting with mild motor symptoms, cognitive impairment and relative independent functioning (Stage I), and ending with a need for full-time care due to severe motor impairment and dementia (Stage V; Shoulson & Fahn, 1979).

One of HD's most frequently observed cognitive impairments is a deficit of emotion recognition, particularly negative emotions such as anger, fear, and disgust (Bates, Tabrizi, & Jones, 2014). However, while the evidence on this set of impairments is well established, it has been traditionally investigated only through tests based on facial expressions, with very few studies based on different stimuli such as emotional body language (see Henley et al., 2012 for a review). In fact, to our knowledge only two studies have investigated EBL in HD, showing preliminary evidence that a deficit of EBL recognition can also be part of the manifestations of the disease (de Gelder et al., 2008), although it may not be observed in presymptomatic individuals (Aviezer et al., 2009).

In addition to the sparseness of evidence on EBL, very little is also known about emotion regulation in HD. Indeed, a recent review (Löffler et al., 2015) identified only one study where this is specifically addressed and which concluded no differences between people with symptomatic HD and healthy controls (Croft, McKernan, Gray, Churchyard, & Georgiou-Karistianis, 2014). However, the measure adopted by this study – the Emotion Regulation Questionnaire (ERQ; Gross et al., 2003) – only assesses the use of two regulatory strategies (suppression and reappraisal), and does not allow for the exploration of any other specific components of emotion regulation, such as impulse control or emotional awareness.

As mentioned previously, emotion recognition (both facial and EBL) and emotion regulation are likely to influence each other (Ochsner, 2009) and play an essential role in the successful operation of social skills as well as psychological resilience (de Gelder & Hortensius, 2014; Ghorbani et al., 2017; Gross & Muñoz, 1995). Given the theoretical arguments for evidence of such deficits in individuals with HD, a deeper understanding of the extent to which the disease affects these cognitive components would allow for a refinement of current cognitive and behavioural approaches to care and treatment. Moreover, this carries the potential for shedding new light on the neural bases that characterise them and the relationship between cognition and neurobiology, in particular in relation to EBL recognition (de Gelder, 2006). Both these implications have, in turn, the potential to contribute to an ongoing debate which focuses on whether the current diagnostic criteria for HD, which are based on motor manifestations only, should also include early signs of cognitive impairment. (Loy & McCusker, 2013; Paulsen, 2011; Reilmann, Leavitt, & Ross, 2014)

Consequently, the aim of this study was to investigate the hypothesis that both facial and EBL recognition and emotion regulation are impaired in individuals affected by symptomatic Huntington's disease, and that such impairments show a significant relationship with one another. The study design included a comparison with non-affected matched controls and the use of more comprehensive tests of emotion recognition and regulation. More specifically, the following hypotheses were formulated: a) People with HD were predicted to report significantly more emotion regulation difficulties than the control group when assessed on a number of different emotion regulation components; b) emotion recognition was predicted to be significantly impaired in people with HD compared to the controls on both facial and EBL tasks; c) a significant relationship was expected to be observed between emotion regulation difficulties and emotion recognition impairment. In addition, due to the evidence of relationships between psychological difficulties and emotional processing (Cisler, Olatunji, Feldner, & Forsyth, 2010; Joorman & Gotlib, 2010; Martin & Dahlen, 2005), depression and anxiety measures were also included.

Methods

Design and participants

This study adopted a 2-group between-subjects design with matched controls. In total, 25 participants took part, split across two groups (HD and Ctrl) consisting of 13 symptomatic individuals (four male, nine female) and 12 matched non-affected controls (five male, seven female). The sample size of the HD group was comparable to the majority of studies investigating emotion recognition in HD that have been identified by a recent systematic review (i.e., six to 40; Henley et al., 2012). For the HD group, participation was limited to individuals in early to moderate stages of the disease (i.e., I-III). This was decided due to the difficulties in undertaking cognitive tasks that are likely to arise in the later stages of the condition. HD stage was screened through the Total Functional Capacity scale (TFC; Shoulson & Fahn, 1979). More specifically, one participant (7.7%) belonged to stage I, seven (53.8%) to stage II, and five (38.5%) to stage III.

The two groups did not present any significant differences in terms of age [$t(23) = .490$, p = ns], years of education [$t(23) = -1.023$, p = ns], or gender [$X^2(1, N = 25) = .322$, p = ns]. See Table 1 for the full demographic details.

Table 1 around here please

The participants of the HD group were recruited with the help of the Regional Care Advisory Service of the Huntington's Disease Association (HDA) in the UK. The participants of the Ctrl group were recruited from partners and caregivers of the participants of the HD group. While this may have not represented the most optimal control group due to potential interactions between the participants, it was considered appropriate as it allowed the recruitment of controls with similar demographic and social characteristics. To limit the risk of confounds, the partners/caregivers were not in the room when the patients were performing the required tasks.

Measures

HD severity measures

Total Functional Capacity Scale (TFC; Shoulson & Fahn, 1979)*:*

The TFC is a standardised tool that assesses everyday functional capacities such as working, handling money, taking care of domestic chores, performing self-care tasks, and living independently. It is part of the larger Unified Huntington's Disease Rating Scale (UHDRS; Huntington Study Group, 1996). The total score ranges from 13 (normal capacity) to 0 (severe disability) and its intervals can be used to determine the stage of the disease: 13-11 = Stage I, 10-7 = Stage II, 6-3 = Stage III, 2-1 = Stage IV, 0 = Stage V. The TFC is characterised by excellent internal consistency (Cronbach's $\alpha = .95$) as well as high interrater reliability (Huntington Study Group, 1996).

Emotion recognition measures

Bochum Emotional Stimulus Set (BESST; Thoma, Soria Bauser, & Suchan, 2013):

The BESST is a validated set of 4490 emotional stimuli consisting of pictures of both male and female facial expressions and emotional body language (EBL). It investigates the recognition of six emotions (fear, disgust, happiness, sadness, surprise and anger) plus neutral expressions. The facial expressions are computer-generated, while the EBL stimuli are based on photographs of actors and actresses. All the stimuli feature multiple ethnic groups. For this study, 10 frontal stimuli from the BESST were randomly selected for each emotion and each expression modality (facial or EBL), half male and half female, to a total of 140 stimuli for two blocks (70 + 70). Thus, the test in this study yielded a total score out of 70 for each modality, as well as a sub score out of 10 for each emotion. The BESST reports excellent norms (Abramson, Marom, Petranker, & Aviezer, 2017), with overall high recognition rates for the whole corpus (83.3% for faces, 85.5% for bodies; Thoma et al., 2013). Other measures of EBL recognition are available in the literature, such as the Bodily Expressive Action Stimulus Test (BEAST; de Gelder & Van den Stock, 2011). However, the latter only consists of the body language component and does not include stimuli for disgust. Therefore, as the recognition of negative emotions plays a particularly important role in HD (Bates, Tabrizi, & Jones, 2014), the BESST was preferred in this study due to its inclusion of disgust, as well as for being currently the only test to include both facial and EBL stimuli within a single set.

Emotion regulation measures

Difficulties in Emotion Regulation Scale (DERS; Gratz & Roemer, 2004)

The DERS is a self-report questionnaire based on 36 items rated on a 5-point Likert scale. It explores emotion regulation on the basis of 6 subscales: non-acceptance of emotional responses, difficulties engaging in goal directed behaviour, impulse control difficulties, lack of emotional awareness, limited access to emotion regulation strategies, lack of emotional clarity. A subscore is yielded for each subscale, which can then be summed to create a total score out of 180 for the whole questionnaire. Higher scores correspond to more difficulties in emotion regulation. To our knowledge, the DERS has never been adopted with people affected by symptomatic HD, but it has been utilised with a number other clinical conditions (Fowler et al., 2014; Kökönyei, Urbán, Reinhardt, Józan, & Demetrovics, 2014), showing very good construct validity (Ritschel, Tone, Schoemann, & Lim, 2015) and internal consistency (Cronbach's α = .93/.89 for total score/subscales Gratz & Roemer, 2004).

Mood and anxiety issues measures

Hospital Anxiety and Depression Scale (HADS; Zigmond & Snaith, 1983)

The HADS is currently one of the most adopted measures of mood and anxiety symptoms in clinical populations and consists of a self-report questionnaire based on 14 items rated on a 4-point scale. The outcome consists of individual scores out of 21 for both anxiety and depression. The HADS has been previously validated with people affected by HD (De Souza, Jones, & Rickards, 2010), and features good construct validity and internal consistency, with a Cronbach's α = .83/.82 for anxiety/depression respectively (Bjelland, Dahl, Haug, & Neckelmann, 2002). The suggested cut-off point for clinical depression and anxiety is 8/21, which guarantees good sensitivity (anxiety/depression = .90/.83) and specificity (anxiety/depression = .78/.79).

Procedure

In general, all the questionnaires were completed by hand by the participants directly. However, in case of difficulties due to motor impairments, the questions were read out to the participants and their responses were recorded by the first author on their behalf. While this approach posed a certain risk of response bias due to misunderstandings, it was adopted to allow the participants with more severe motor impairments to feel as comfortable as possible during the data collection. In order to minimise the risk of bias, during recording the responses, the first author asked for clarifications regarding the participants' answers whenever necessary.

The two blocks of the BESST were administered by the first author on a 15-inch laptop. Each stimulus was presented singularly on a black background in an 834x834 pixel format along with seven emotional labels on the right corresponding to the emotions investigated by the test. The participants were asked to name the label corresponding to the presented stimulus. This seven-alternative method differed from the way the test was administered in the validation study (Thoma et al., 2013), which consisted of a two-alternative forced choice task. The seven-alternative method was considered the most appropriate as it has been previously adopted with the BESST (Abramson et al., 2017; Soria Bauser, Thoma, & Suchan, 2012). It is also included in tests which are considered the 'gold standard' of facial and EBL recognition tests, , such as the Reading the Mind in the Eye test (RME, Baron-Cohen et al., 2001), the Bodily Expressive Action Stimulus Test (BEAST, de Gelder & Van den Stock, 2011), the CANTAB Emotion Recognition Task (Sahakian & Owen, 1992), the Emotion Hexagon Task (Calder et al., 1996), and the original Ekman 60 Faces Test (Ekman, Friesen, 1976). As the answers were provided verbally by the participants, no direct interaction was required between them and the laptop. A practice session consisting of seven stimuli (one for each emotion) was administered prior to the beginning of each block, to allow for familiarisation with the task. The order was kept constant among the participants, with the face block being administered prior to the body language block. No time limit was set for responses. However, the participants were asked to perform the tasks as quickly as possible. Figure 1 illustrates examples of neutral, positive, and negative stimuli administered via the BESST.

Statistical analysis

All the statistical analyses were performed with IBM SPSS Statistics® programme v23 (Armonk, NY: IBM Corp). On account of the relatively small sample size, non-parametric statistics were adopted. This is a common choice when working with small sample of symptomatic HD participants, and especially when investigating emotional processing (e.g., Croft et al., 2014; Snowden et al., 2008; Trinkler, de Langavant, & Bachoud-Levi, 2013). Mann-Whitney tests were performed to make comparisons between the two participant groups, while two-tailed Spearman's correlations were utilised to investigate the relationship between the two main outcome variables. Effects sizes were calculated with Cohen's *d.* In order to avoid loss of power due to the adoption of conservative corrections with a small sample size, significance levels were conventionally set at $p = .05$ with no correction for multiple comparisons. This was in line with several previous studies on emotion recognition in small samples of people with HD (Croft et al., 2014; Ille, Holl, et al., 2011; Ille, Schafer, et al., 2011; Novak et al., 2012; Robotham et al., 2011; Snowden et al., 2008; Sprengelmeyer, Schroeder, Young, & Epplen, 2006; van Asselen et al., 2012), as well as in other rare clinical populations (e.g., frontotemporal dementia; Keane, Calder, Hodges, & Young, 2002).

Ethics approval

This study was reviewed and approved by the Faculty of Health and Medicine Research Ethics Committee at Lancaster University (ref: FHMREC15043).

Results

Measure reliability

All the adopted measures generally showed good levels of reliability comparable to the levels reported in the literature (Bjelland et al., 2002; Gratz & Roemer, 2004). More specifically, high internal consistency was shown by HADS for both anxiety (Cronbach's $\alpha = .808$) and depression (Cronbach's $\alpha = .805$).. With regards to the BESST, the total scores showed high internal consistency in both the facial (Cronbach's $\alpha = .758$) and emotional body language (Cronbach's $\alpha = .863$) modalities. The single emotions scores generally showed acceptable figures, with Cronbach's α ranging between .616 and .896. However, low levels were found for the fear facial score (Cronbach's $\alpha = .567$), and for both the facial and emotional body language scores for surprise (Cronbach's $\alpha = .531$ and .352). As for the DERS, high internal consistency was found throughout the whole measure, with a Cronbach's α of .941 for the SUM score, and figures ranging from .763 to .855 for the subscales.

Participants' scores

Table 2 shows the mean scores of the participants of both the HD and Ctrl groups for the outcome variables, and Figure 2 provides a graphical illustration of the scores. According to the recommended clinical cut-off for the HADS (8/21; Bjelland et al., 2002), six of the participants of HD group (i.e., 46.15%) showed clinical levels of anxiety, while seven (i.e., 53.8%) showed clinical levels of depression. On the other hand, only three of the participants of the Ctrl (i.e., 25%) group showed clinical levels of anxiety, and only one (i.e., 8.3%) reported clinical levels of depression.

The general results for emotion recognition showed a slightly better performance on the body language modality compared to the facial one in both groups. However, in this study the BESST constituted quite an arduous task for all the participants, as rather low overall recognition rates were observed for both the HD group (32.6% for faces, 40.2% for bodies) and the controls (45.6% for faces, 56.5% for bodies). These represented lower rates compared to the ones reported by the validation study (i.e., 83.3/87%; Thoma et al., 2013), but were in line with those reported in studies that adopted the BESST with a multiple forced-choice paradigm (e.g., 50%; Abramson et al., 2017). In terms of specific emotions, in the face task the least recognised emotion in both groups was sadness (HD: 10.8%, Ctrl: 19.2%), while the most easily identified was happiness (HD: 70%, Ctrl: 92.5%). The results on these two emotions were in line with the findings of the validation study. On the other hand, in the body language modality the lowest scores were observed on disgust for both groups (HD: 10%, Ctrl: 17.5%), while the highest were again on happiness (HD: 75.4%, Ctrl: 78.3%), along with neutral stimuli (HD: 70%, Ctrl: 92.5%). Contrary to the facial modality, this result was opposite to the validation data, which found happiness body stimuli to be least recognised.

With regard to emotion regulation difficulties, the total score (SUM) of the HD group was significantly higher than the available data with general adult populations (e.g., 77/180; Ritschel et al., 2015), meaning that considerably more emotion regulation difficulties were reported by the HD participants. Instead, on average the Ctrl group scored rather lower (67/180) compared with the normative data.

Group comparison analysis

The group comparison analysis showed that the participants affected by HD had a significantly greater level of depression when compared to the controls (U = 27.0, z = -2.787, p = .005); however, no significant difference was found for anxiety levels (U = 72.0, z = -.328, p = ns).

With regard to emotion recognition, the overall performance of the HD group on the BESST was significantly poorer for both the facial (U = 16.5, z = -3.352, p < .001 and body language (U = 32.0, z = -2.510, p = .012) modalities. In terms of specific emotions, the facial modality revealed specifically greater impairments in the HD group for disgust (U = 34.5, z = -2.402, p = .016) and anger (U = 42.0, z = -2.082, p = .012), while the body language modality yielded poorer performances for fear (U = 25.0, z = -2.914, p = .004), sadness (U = 33.0, z = -2.481, p = .013), and neutral stimuli (U = 32.0, z = -2.144, p = .012).

In terms of emotion regulation, a significantly greater level of total difficulties (DERS SUM) was reported by the HD group (U = 29.5, z = -2.639, p = .008). When comparing the specific components of the DERS, significant differences were observed on impulse control difficulties (IMPULSE; U = 28.5, z = -2.730, p = .007), and lack of emotional clarity (CLARITY; U = 30.5, z = -2.595, p = .008).

The effect size analysis showed a very large group effect on the overall scores for facial emotion recognition (d = -1.54), emotional body language recognition (d = -1.378), and emotion regulation (d = 1.011). Moreover, in spite of a lack of statistical significance, several medium to large effect sizes were observed for specific components of the outcome variables, thus showing group effects at a trend level. These included limited access to regulation strategies (STRATEGIES; d = 1.016), difficulties in engaging in goal directed behaviour (DERS GOALS; d = .679), lack of emotional awareness (DERS AWARE; d = .656), facial recognition of happiness (d = -.491), sadness (d = -447), and surprise (d = -.769), as well as body language recognition of disgust (d = -.452), surprise (d = -.599), and anger (d = -.466).

Correlation analysis

In light of the significant differences observed in symptomatic individuals on the group comparison, a correlation analysis was carried out to explore whether the impairments on emotion regulation and recognition in the HD group were correlated with the demographic characteristics and the measures of psychological difficulties. Table 4 illustrates Spearman's coefficients for the correlation analysis of the HD group among the all the variables. The results showed that, with regard to emotion recognition, the overall performance for the facial modality of the BESST (BESST_F_SUM) was strongly correlated with the overall performance for the body language modality (BESST_B_SUM; r_s = .739, p < .001), confirming the relationship between the two emotion recognition components. This was also confirmed by the observation of significant relationships across the modalities between the single scores for neutral stimuli (r_s = .606, p = .028), disgust (r_s = .582, p = .037), and anger (r_s = .589, p = .034), as well as linear trends close to significance for fear (r_s = .526 p = .065) and surprise (r_s = .499, p = .082). In addition, the total score for the body language modality (BESST_B_SUM) shared a significant strong negative relationship with HD stage (r_s = -.675, p = .011), meaning that the recognition of emotional body language of the participants affected by HD deteriorated in line with disease progression. The total score for the facial modality (BESST_F_SUM), showed a similar trend towards HD stage (r_s = -.533, p = .060).

In terms of emotion regulation, the overall level of emotion regulation difficulties (DERS_SUM) shared a very strong positive correlation with levels of anxiety (HADS_A; r_s = .905, p < .001), as well as a strong correlation with levels of depression (HADS_D; r_s = .629, p = .021). In particular, the two components that were specifically impaired in the HD group, IMPULSE and CLARITY, were respectively related to anxiety (r_s = .675, p = .011) and depression (r_s = .717, p = .006).

Discussion

Overview of main findings

This aim of this study was to investigate whether facial and EBL recognition and emotion regulation were impaired in people with symptomatic HD when compared to matched controls, and whether such impairments were significantly correlated. In addition, to our knowledge this was the first study with this specific population both to explore emotion regulation at the same time as emotion recognition and to include facial and EBL recognition modalities together. The results showed significant impairments in the HD group in emotion regulation, as well as emotion recognition in both the facial and EBL modality. This was in line with our initial predictions and confirmed our first two hypotheses.

In terms of specific components of emotion regulation, significant differences were found for impulse control difficulties (DERS IMPULSE) and lack of emotional clarity (DERS CLARITY) in the HD group when compared to healthy controls. This appears to be consistent with several previous observations of impulse control and executive functioning deficits in people with HD (Duff et al., 2010; Galvez et al., 2017; Gray et al., 2013; Mörkl et al., 2016), that are often due to the impact of the disease on prefrontal brain areas (Dogan et al., 2014; Gray et al., 2013) and are likely to play a pivotal role in the clarity and control of emotional experiences. No significant differences were observed for the remaining components of emotion regulation, including DERS STRATEGIES. This particular finding was in line with the only other study on emotion regulation in people with HD, which only explored the use of regulatory strategies and found no significant differences with matched controls (Croft et al., 2014). In addition, since no authors have previously carried out a comprehensive investigation of emotion regulation in HD which includes all its components, the significant difference on the DERS SUM observed in this study represents the first preliminary evidence of a general impairment of emotion regulation in this specific population.

The observed impairment for facial emotion recognition adds further confirmation to the already well-known deficit reported in the literature (for a review, see Bates, Tabrizi, & Jones, 2014). Moreover, the results on the single emotion scores also confirmed the known specific deficit of negative emotions such as disgust and anger, even though no significant difference was found for the facial recognition of fear and sadness. The impairment on the EBL modality was partially in line with the only other study that investigated this construct in people with symptomatic HD and which found a significant impairment in the recognition of anger and emotionally neutral instrumental stimuli, but no deficit for fear and sadness (de Gelder et al., 2008). Indeed, a specific impairment for neutral (yet not instrumental) stimuli was found in the our study too, although the comparisons on the single emotion scores in this study yielded almost opposite results, with a significant impairment for fear and sadness, but no significant deficit for anger. As de Gelder and colleagues (2008) did not include stimuli for happiness, surprise, and disgust, it is not possible to know whether other emotions were impaired, and to what extent our results differ. As a consequence, the finding of our study also represents the first preliminary evidence of an impairment of emotional body language (EBL) recognition in people with symptomatic HD through a comprehensive assessment that includes both positive and negative emotional stimuli, as well as the first study to assess the impact of the disease on the recognition of disgust via body language.

With regards to the relationship between emotion regulation and emotion recognition, the correlation analysis showed that the observed impairments did not significantly correlate. This finding was contrary to the study's third hypothesis. Moreover, it was also inconsistent with what has been previously reported in other clinical populations, such as in people with anorexia nervosa (Harrison et al., 2009). On the other hand, the overall level of emotion regulation difficulties shared a significant correlation with anxiety and depression. In addition, impulse control difficulties and lack of emotional clarity – the two emotion regulation components that were specifically impaired in the HD group – shared significant relationships with anxiety and depression respectively.

These findings suggest that, in the HD group, anxiety and depression might have played a pivotal role in the operationalisation of emotion regulation. While the small participant number makes this impossible to test statistically through more sophisticated analyses, the results are consistent with previous reports of associations between mood and anxiety problems and deficits of emotion regulation (e.g., Ehring et al., 2008; Loas et al., 1997; for a review on anxiety, see Cisler, Olatunji, Feldner, & Forsyth, 2010), and in particular between impulse control and anxiety (e.g., with Parkinson's disease; Voon et al., 2011), and emotional clarity and depression (Dixon-Gordon et al., 2014; Thompson, Boden,

& Gotlib, 2017). Thus, the significant difference on the DERS in the HD group when compared to the Ctrl group may represent a reflection of the significantly higher level of depression reported by the symptomatic participants.

Implications for clinical practice

These findings have important implications for clinical practice, as emotion regulation deficits have the potential to disrupt people's daily life in a large number of ways(Gross & Muñoz, 1995). In particular, current evidence suggests that they may cause issues with affective experience (e.g., a decrease in experience of positive emotions), cognitive functioning (e.g., lower memory performance), as well as social skills, such as theory of mind and communication (Gross & Award, 2002). All these issues appear to be even more relevant for people with symptomatic HD, as they are likely to add to (and potentially worsen) the affective, cognitive, and communicative impairments already caused by other symptoms of the condition (Eddy & Rickards, 2015; Hartelius, Jonsson, Rickeberg, & Laakso, 2010; Hubers et al., 2012; Paulsen, 2011; Zarotti, 2016; Zarotti, Simpson, & Fletcher, 2017).

As a consequence, a more in-depth understanding of emotional processing in HD currently plays a pivotal role in clinical practice, since it has the potential to help revise current therapeutic and communicative protocols, as well as informing new ones. Indeed, the possibility of enhancing patients' cognitive reserves through cognitive training interventions has proven to be a very promising approach to delay or control the onset of cognitive symptoms in neurodegenerative diseases (Papoutsi, Labuschagne, Tabrizi, & Stout, 2014). With regard to this, recent preliminary evidence suggests that addressing emotion recognition impairments at both presymptomatic and early stage HD via selfguided computerised cognitive training can lead to significant improvement in recognition accuracy (Kempnich, Wong, Georgiou-Karistianis, & Stout, 2017). In particular, the case of HD represents an ideal model for the exploration of this type of cognitive training, due to its genetic nature, the availability of predictive testing, and the consequent well-established underlying pathological mechanisms (Papoutsi et al., 2014).

Perhaps even more significantly, the finding that emotion regulation deficits are strongly related to levels of depression and disease stage may have a number of important clinical and therapeutic consequences. Indeed, emotion regulation strategies and depressive symptoms have been reported to share stable significant relationships across many different populations, including adolescents, adults, elderly and people with psychological difficulties (Berking, Wirtz, Svaldi, & Hofmann, 2014; Garnefski & Kraaij, 2006). Moreover, evidence has identified depression as a critical factor in triggering a general decrease in coping skills and resilience (Penland, Masten, Zelhart, Fournet, & Callahan, 2000), especially in people affected by neurodegenerative disorders (Baquero, 2015). Considering the pivotal role played by emotion regulation strategies in the successful implementation of coping skills and resilience (Ghorbani et al., 2017; Gross & Muñoz, 1995; Hasking et al., 2010; van der Meer et al., 2015), it could be hypothesised that the combinations of these mechanisms may contribute to the development of a unhelpful circle of regulation difficulties and psychological difficulties. More specifically, suboptimal coping and resilience due to depression would lead to depleted emotion regulation skills, which would in turn contribute to higher levels of depression, in a mechanism that increases in severity along with disease progression.

This hypothesis appears consistent with results from our previous investigation with people with premanifest HD where, despite the absence of general clinically significant levels of depression or emotion regulation deficits, subclinical depressive symptoms were found to significantly predict early difficulties on specific components of emotion regulation (Zarotti, Simpson, Fletcher, Squitieri, & Migliore, 2018). As a consequence, addressing emotion regulation deficits along with depression in people with HD may open up new avenues for alternative forms of psychological intervention. Indeed, a growing body of evidence suggests that emotion regulation may represent a transdiagnostic construct in psychological difficulties (Sloan et al., 2017).

For example, Berking and colleagues (2008) suggested that replacing parts of standard cognitive behavioural therapy (CBT) treatment with training in emotion regulation may enhance the effectiveness of CBT. In particular, the authors applied the Integrative Training of Emotional Competencies(ITEC; Berking 2007)28an intervention based on the intensive practice of a number of emotion regulation skills including progressive muscle and breathing relaxation, non-judgemental awareness of emotions, acceptance and tolerance of negative emotions, effective self-support, analysis of emotional cues, and quantitative and qualitative modification of emotional reactions, which all yielded a significant enhancement of the effects of CBT, These findings appear even more important for HD, as CBT currently represents one of the most adopted approaches to psychotherapy in this condition (Anderson et al., 2011; Ghosh & Tabrizi, 2013; Novak & Tabrizi, 2011). Thus, the inclusion of emotion regulation as a treatment target in psychotherapy may yield greater beneficial effects for both people with presymptomatic and symptomatic HD.

Limitations and future directions

A number of limitations should be considered along with the results of this study. First, the data collection sessions, which occurred within a single day at the participants' home, proved to be challenging for some of the participants. For this reason, no cognitive screening was performed prior to the administration of the research materials. While it is recognised that this prevented a more precise understanding of the participants' level of overall cognitive functioning, which would have allowed for a better clinical depiction of the stage of disease and cognitive performance more generally, it also allowed the cognitive load to remain manageable throughout the data collection. Thus, avoiding the risk of increased fatigue affecting the performance of the participants on the experimental measures was prioritised over the potential benefits of adding of a cognitive screening as a covariate.

Secondly, the overarching aim of this study was to investigate whether emotion regulation and both facial and EBL recognition were impaired in people affected by symptomatic HD. As a consequence, no clinical control group was included in the design and this could have isolated factors specific to people with HD as opposed to other neurodegenerative diseases more generally. However, while this decision fitted the purpose of this study, it is recognised as a limitation.

Thirdly, the generally low recognition rates on the BESST showed that the emotion recognition tasks were somewhat difficult for both the HD and Ctrl group as compared to the available normative data. This is likely due to the differences in the way the tasks were administered compared to the validation study (Thoma et al., 2013), which was based on a two-alternative forced choice task with a 3000ms limit, while the present study featured a seven-alternative forced choice task with no time limit. Indeed, lower recognition rates have also been reported when adopting the BESST with the same method as the present study, i.e. with tasks based on four or more alternatives (Abramson et al., 2017). In addition, the general better performance observed in both groups on the EBL recognition component as opposed to the facial one may be due to an effect of familiarisation due the order of presentation of the tasks (facial first, EBL second), which was kept constant among the participants.

Finally, an important caveat to be noted is the potential effect of the relatively small sample size, despite it being in line with most of the current studies on emotion recognition in HD (Bates, Tabrizi, & Jones, 2014). Indeed, the effect size analysis showed that most of the observed inconsistencies with the results in the previous literature in fact represented differences at a trend level characterised by medium to large effect sizes (d = -.452 – 1.016). This could be also applied to some of the results of the correlation analysis that were approaching significance, such as the correlation between facial emotion recognition and HD stage. With regard to this, it is worth noting that the sample size of the present study may have partially affected effect sizes, potentially making them deviate from the real population effect sizes farther than a larger sample. However, considering the current direction of the evidence available from other investigations, it seems reasonable to hypothesise that the effect sizes observed in this study were in fact not significantly affected by the sample size. Consequently, the adoption of a larger sample would yield significant differences on fear, sadness, and anger in line with the findings in the previous literature, as well as significant correlations in line with the ones that were found in this study. Thus, the conclusion that HD directly impacts emotion regulation should be considered preliminary and taken cautiously until additional evidence is obtained with larger samples.

Future research should aim at further exploring emotion regulation and emotion recognition in larger samples of people with different stages of symptomatic HD, in order to obtain a better understanding of the potential relationship between these two constructs. In particular, more investigations are needed on the inclusion of emotion regulation as treatment target in psychotherapy for people with this condition. As for emotional body language, other measures of EBL recognition should also be adopted with HD populations, in order to control for the convergent validity of the BESST and to build a comprehensive corpus of data similar to the one currently available for facial stimuli. In particular, more data are warranted on the optimal use of the BESST stimuli when based on a multiple-choice forced task, in order to avoid potential floor effects in participant performance. From this perspective, the adoption of EBL measures would benefit from the inclusion in large multi-centre studies, which would also allow the integration of comprehensive cognitive screenings.Finally, clinical control groups of people affected by diseases that share common symptoms with HD (e.g., people with Parkinson's disease) should be included, in order to clarify the role of the different factors that may contribute to the development of emotion regulation and EBL recognition impairments.

Conclusion

This study has shed new light on emotional processing in people with symptomatic Huntington's disease by providing different sources of evidence that emotion regulation and emotional body language (EBL) recognition are significantly impaired in this population, and that the latter is negatively related to the stage of disease. It also provided the first preliminary evidence of a significant direct correlation between deficits of facial and body language emotion recognition in HD, although emotion regulation and emotion recognition were not related.

Altogether, these findings support the suggestion that better insight into emotion recognition and regulation issues in HD, along with their connections to mood and anxiety disorders, would allow the development of psychological and pharmacological interventions that are tailored around the emotional needs of each patient.

References

Abramson, L., Marom, I., Petranker, R., & Aviezer, H. (2017). Is fear in your head? A comparison of instructed and real-life expressions of emotion in the face and body. *Emotion*, *17*(3), 557–565. https://doi.org/10.1037/emo0000252

Aldao, A., Nolen-Hoeksema, S., & Schweizer, S. (2010). Emotion-regulation strategies across psychopathology: A meta-analytic review. *Clinical Psychology Review*. https://doi.org/10.1016/j.cpr.2009.11.004

Anderson, K., Craufurd, D., Edmondson, M. C., Goodman, N., Groves, M., van Duijn, E., … Goodman, L. (2011). An international survey-based algorithm for the pharmacologic treatment of obsessive-compulsive behaviors in Huntington's disease. *PLoS Currents*, *3*, RRN1261. https://doi.org/10.1371/currents.RRN1261

Aviezer, H., Bentin, S., Hassin, R. R., Meschino, W. S., Kennedy, J., Grewal, S., … Moscovitch, M. (2009). Not on the face alone: perception of contextualized face expressions in Huntington's disease. *Brain : A Journal of Neurology*, *132*(Pt 6), 1633–1644. https://doi.org/10.1093/brain/awp067

Baquero, M. (2015). Depressive symptoms in neurodegenerative diseases. *World Journal of Clinical Cases*, *3*(8), 682. https://doi.org/10.12998/wjcc.v3.i8.682

Baron-Cohen, S., Wheelwright, S., Hill, J., Raste, Y., & Plumb, I. (2001). The "Reading the Mind in the Eyes" Test revised version: a study with normal adults, and adults with Asperger syndrome or high-functioning autism. *Journal of Child Psychology and Psychiatry, and Allied Disciplines*, *42*(2), 241–251. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/11280420

Bates, G., Tabrizi, S., & Jones, L. (2014). *Huntington's Disease*. (G. P. Bates, S. J. Tabrizi, & L. Jones, Eds.). New York: Oxford University Press.

Berking, M. (2007). *Training Emotionaler Kompetenzen [Affect regulation training]*. Heidelberg: Springer.

Berking, M., Wirtz, C. M., Svaldi, J., & Hofmann, S. G. (2014). Emotion regulation predicts symptoms of depression over five years. *Behaviour Research and Therapy*, *57*(1), 13–20. https://doi.org/10.1016/j.brat.2014.03.003

Berking, M., Wupperman, P., Reichardt, A., Pejic, T., Dippel, A., & Znoj, H. (2008). Emotion-regulation skills as a treatment target in psychotherapy. *Behaviour Research and Therapy*, *46*(11), 1230–1237. https://doi.org/10.1016/j.brat.2008.08.005

Bjelland, I., Dahl, A. A., Haug, T. T., & Neckelmann, D. (2002). The validity of the Hospital Anxiety and Depression Scale: An updated literature review. *Journal of Psychosomatic Research*, *52*(2), 69–77. https://doi.org/10.1016/S0022-3999(01)00296-3

Calder, A. J., Young, A. W., Rowland, D., Perrett, D. I., Hodges, J. R., & Etcoff, N. L. (1996). Facial emotion recognition after bilateral amygdala damage: Differentially severe impairment of fear. *Cognitive Neuropsychology*, *13*(5), 699–745. https://doi.org/10.1080/026432996381890

Cisler, J., Olatunji, B., Feldner, M., & Forsyth, J. (2010). Emotion regulation and the anxiety disorders: an integrative review. *Journal of Psychopathology and Behavioral Assessment*, *32*(1), 68–82. https://doi.org/10.1007/s10862-009-9161-1.Emotion

Croft, R. J., McKernan, F., Gray, M., Churchyard, a, & Georgiou-Karistianis, N. (2014).

Emotion perception and electrophysiological correlates in Huntington's disease. *Clinical Neurophysiology : Official Journal of the International Federation of Clinical Neurophysiology*, *125*(8), 1618–1625. https://doi.org/10.1016/j.clinph.2013.12.111

de Gelder, B. (2006). Towards the neurobiology of emotional body language. *Nature Reviews Neuroscience*, *7*(3), 242–249. https://doi.org/10.1038/nrn1872

de Gelder, B., & Hortensius, R. (2014). The many faces of the emotional body. In *New Frontiers in Social Neuroscience* (Vol. 21, pp. 153–164). https://doi.org/10.1007/978-3-319-02904-7

de Gelder, B., & Van den Stock, J. (2011). The Bodily Expressive Action Stimulus Test (BEAST). Construction and validation of a stimulus basis for measuring perception of whole body expression of emotions. *Frontiers in Psychology*, *2*(August), 181. https://doi.org/10.3389/fpsyg.2011.00181

de Gelder, B., Van den Stock, J., Balaguer Rde, D., Bachoud-Levi, A. C., Balaguer, R. D. D., & Bachoud-Lévi, A.-C. (2008). Huntington's disease impairs recognition of angry and instrumental body language. *Neuropsychologia*, *46*(1), 369–373. https://doi.org/10.1016/j.neuropsychologia.2007.10.015

De Souza, J., Jones, L. A., & Rickards, H. (2010). Validation of self-report depression rating scales in Huntington's disease. *Movement Disorders : Official Journal of the Movement Disorder Society*, *25*(1), 91–96. https://doi.org/10.1002/mds.22837

Dixon-Gordon, K., Dovidio, J., Gilbert, K., Hilt, L., Marroquin, B., Mclaughlin, K., … Aldao, A. (2014). Impaired emotional clarity and psychopathology: a transdiagnostic deficit with symptom-specific pathways through emotion regulation. *Journal of Social and Clinical Psychology*, *33*(4), 319–342. https://doi.org/http://dx.doi.org/101521jscp2014334319

Dogan, I., Saß, C., Mirzazade, S., Kleiman, A., Werner, C. J., Pohl, A., … Reetz, K. (2014).

Neural correlates of impaired emotion processing in manifest Huntington's disease. *Social Cognitive and Affective Neuroscience*, *9*(5), 671–680. https://doi.org/10.1093/scan/nst029

Duff, K., Paulsen, J. S., Beglinger, L. J., Langbehn, D. R., Wang, C., Stout, J. C., … Predict, H. D. I. of the H. S. G. (2010). "Frontal" behaviors before the diagnosis of Huntington's disease and their relationship to markers of disease progression: evidence of early lack of awareness. *Journal of Neuropsychiatry and Clinical Neuroscience*, *22*(2), 196–207. https://doi.org/10.1176/appi.neuropsych.22.2.196

Eddy, C. M., & Rickards, H. E. (2015). Interaction without intent : The shape of the social world in Huntington ' s disease. *Social Cognitive and Affective Neuroscience*, 1–36. https://doi.org/10.1093/scan/nsv012

Ehring, T., Fischer, S., Schnülle, J., Bösterling, A., & Tuschen-Caffier, B. (2008). Characteristics of emotion regulation in recovered depressed versus never depressed individuals. *Personality and Individual Differences*, *44*(7), 1574–1584. https://doi.org/10.1016/j.paid.2008.01.013

Ekman, P., Friesen, W. V. (1976). *Pictures of Facial Affect*. Palo Alto: Consulting Psychologists Press.

Fowler, J. C., Charak, R., Elhai, J. D., Allen, J. G., Frueh, B. C., & Oldham, J. M. (2014). Construct validity and factor structure of the Difficulties in Emotion Regulation Scale among adults with severe mental illness. *Journal of Psychiatric Research*, *58*, 175–180. https://doi.org/10.1016/j.jpsychires.2014.07.029

Galvez, V., Fernandez-Ruiz, J., Bayliss, L., Ochoa-Morales, A., Hernandez-Castillo, C. R., DIáz, R., & Campos-Romo, A. (2017). Early Huntington's disease: Impulse control deficits but correct judgment regarding risky situations. *Journal of Huntington's Disease*, *6*(1), 73–

78. https://doi.org/10.3233/JHD-160223

Garnefski, N., & Kraaij, V. (2006). Relationships between cognitive emotion regulation strategies and depressive symptoms: A comparative study of five specific samples. *Personality and Individual Differences*, *40*(8), 1659–1669. https://doi.org/10.1016/j.paid.2005.12.009

Ghorbani, F., Khosravani, V., Sharifi Bastan, F., & Jamaati Ardakani, R. (2017). The alexithymia, emotion regulation, emotion regulation difficulties, positive and negative affects, and suicidal risk in alcohol-dependent outpatients. *Psychiatry Research*, *252*(August 2016), 223–230. https://doi.org/10.1016/j.psychres.2017.03.005

Ghosh, R., & Tabrizi, S. J. (2013). Clinical aspects of Huntington's disease (pp. 3–31). Springer, Berlin, Heidelberg. https://doi.org/10.1007/7854_2013_238

Goleman, D. (1995). *Emotional intelligence*. New York: Bantam Books.

Gratz, K. L., & Roemer, L. (2004). Multidimensional assessment of emotion regulation and dysregulation: development, factor structure, and initial validation of the Difficulties in Emotion Regulation Scale. *Journal of Psychopathology and Behavioral Assessment*, *26*(1), 41–54. https://doi.org/10.1023/B:JOBA.0000007455.08539.94

Gray, M. A., Egan, G. F., Ando, A., Churchyard, A., Chua, P., Stout, J. C., & Georgiou-Karistianis, N. (2013). Prefrontal activity in Huntington's disease reflects cognitive and neuropsychiatric disturbances: The IMAGE-HD study. *Experimental Neurology*, *239*(1), 218–228. https://doi.org/10.1016/j.expneurol.2012.10.020

Gross, J. (2013). *Handbook of emotion regulation, Second Edition*. Guilford Publications.

Gross, J. J. (1998). The emerging field of emotion regulation: an integrative review. *Review of*

General Psychology, *2*(5), 271–299. https://doi.org/10.1017.S0048577201393198

Gross, J. J. (2015). The extended process model of emotion regulation: elaborations, applications, and future directions. *Psychological Inquiry*, *26*(July), 130–137. https://doi.org/10.1080/1047840X.2015.989751

Gross, J. J., & Award, E. C. (2002). Emotion regulation: Affective, cognitive, and social consequences. *Psychophysiology.*, *39*, 281–291.

Gross, J. J., & John, O. P. (2003). Individual differences in two emotion regulation processes: Implications for affect, relationships, and well-being. *Journal of Personality and Social Psychology*, *85*(2), 348–362. https://doi.org/10.1037/0022-3514.85.2.348

Gross, J. J., & Muñoz, R. F. (1995). Emotion regulation and mental health. *Clinical Psychology: Science and Practice*, *2*(2), 151–164. https://doi.org/10.1111/j.1468-2850.1995.tb00036.x

Harrison, A., Sullivan, S., Tchanturia, K., & Treasure, J. (2009). Emotion recognition and regulation in anorexia nervosa. *Clinical Psychology and Psychotherapy*, *16*(4), 348–356. https://doi.org/10.1002/cpp.628

Hartelius, L., Jonsson, M., Rickeberg, A., & Laakso, K. (2010). Communication and Huntington's disease: qualitative interviews and focus groups with persons with Huntington's disease, family members, and carers. *International Journal of Language & Communication Disorders / Royal College of Speech & Language Therapists*, *45*(3), 381–393. https://doi.org/10.3109/13682820903105145

Hasking, P. A., Coric, S. J., Swannell, S., Martin, G., Thompson, H. K., & Frost, A. D. J. (2010). Brief report: Emotion regulation and coping as moderators in the relationship between personality and self-injury. *Journal of Adolescence, 33*(5), 767–773. https://doi.org/10.1016/j.adolescence.2009.12.006

Henley, S. M. D., Novak, M. J. U., Frost, C., King, J., Tabrizi, S. J., & Warren, J. D. (2012). Emotion recognition in Huntington's disease: a systematic review. *Neuroscience and Biobehavioral Reviews*, *36*(1), 237–253. https://doi.org/10.1016/j.neubiorev.2011.06.002

Hubers, a a M., Reedeker, N., Giltay, E. J., Roos, R. a C., van Duijn, E., & van der Mast, R. C. (2012). Suicidality in Huntington's disease. *Journal of Affective Disorders*, *136*(3), 550–557. https://doi.org/10.1016/j.jad.2011.10.031

Huntington Study Group. (1996). Unified Huntington's Disease Rating Scale: Reliability and-consistency. *Movement Disorders*, *11*(2), 136–142. https://doi.org/10.1002/mds.870110204

Ille, R., Holl, A. K., Kapfhammer, H.-P. P., Reisinger, K., Schäfer, A., Schienle, A., … Schienle, A. (2011). Emotion recognition and experience in Huntington's disease: is there a differential impairment? *Psychiatry Research*, *188*(3), 377–382. https://doi.org/10.1016/j.psychres.2011.04.007

Ille, R., Schafer, A., Scharmuller, W., Enzinger, C., Schoggl, H., Kapfhammer, H.-P. P., … Schienle, A. (2011). Emotion recognition and experience in Huntington disease: a voxel-based morphometry study. *Journal of Psychiatry & Neuroscience*, *36*(6), 383–390. https://doi.org/10.1503/jpn.100143

Izard, C., Fine, S., Schultz, D., Mostow, A., Ackerman, B., & Youngstrom, E. (2001). Emotion knowledge as a predictor of social behavior and academic competence in children at risk. *Psychological Science*, *12*(1), 18–23. https://doi.org/10.1111/1467-9280.00304

Joorman, J., & Gotlib, I. H. (2010). Emotion regulation in depression: Relation to cognitive inhibition. *Cognition & Emotion*, *24*(2), 281–298. https://doi.org/10.1080/02699930903407948.Emotion

Keane, J., Calder, A. J., Hodges, J. R., & Young, A. W. (2002). Face and emotion processing in frontal variant frontotemporal dementia. *Neuropsychologia*, *40*(6), 655–665. https://doi.org/10.1016/S0028-3932(01)00156-7

Kempnich, C. L., Wong, D., Georgiou-Karistianis, N., & Stout, J. C. (2017). Feasibility and efficacy of brief computerized training to improve emotion recognition in premanifest and early-symptomatic Huntington's disease. *Journal of the International Neuropsychological Society : JINS*, *23*(4), 314–321. https://doi.org/10.1017/S1355617717000145

Kökönyei, G., Urbán, R., Reinhardt, M., Józan, A., & Demetrovics, Z. (2014). The Difficulties in Emotion Regulation Scale: Factor structure in chronic pain patients. *Journal of Clinical Psychology*, *70*(6), 589–600. https://doi.org/10.1002/jclp.22036

Lima, C. F., Castro, S. L., & Scott, S. K. (2013). When voices get emotional: a corpus of nonverbal vocalizations for research on emotion processing. *Behavior Research Methods*, *45*(4), 1234–1245. https://doi.org/10.3758/s13428-013-0324-3

Loas, G., Dhee-Perot, P., Chaperot, C., Fremaux, D., Gayant, C., & Boyer, P. (1997). Anhedonia, alexithymia and locus of control in unipolar major depressive disorders. *Psychopathology*, *01*, 206–212.

Löffler, L. a. K., Radke, S., Morawetz, C., & Derntl, B. (2015). Emotional dysfunctions in neurodegenerative diseases. *Journal of Comparative Neurology*, *00*, n/a-n/a. https://doi.org/10.1002/cne.23816

Loy, C. T., & McCusker, E. A. (2013). Is a motor criterion essential for the diagnosis of clinical huntington disease? *PLoS Currents*, *5*. https://doi.org/10.1371/currents.hd.f4c66bd51e8db11f55e1701af937a419

Martin, R. C., & Dahlen, E. R. (2005). Cognitive emotion regulation in the prediction of depression, anxiety, stress, and anger. *Personality and Individual Differences*, *39*(7), 1249–

1260. https://doi.org/10.1016/j.paid.2005.06.004

Mayer, J. D. (2001). *A field guide to emotional intelligence*. (J. D. Mayer, Ed.), *Emotional intelligence in everyday life: A scientific inquiry*. New York, NY: Psychology Press.

Mayer, J. D., Caruso, D. R., & Salovey, P. (1999). Emotional intelligence meets traditional standards for an intelligence. *Intelligence*, *27*(4), 267–298. https://doi.org/10.1016/S0160-2896(99)00016-1

Mörkl, S., Müller, N. J., Blesl, C., Wilkinson, L., Tmava, A., Wurm, W., … Painold, A. (2016). Problem solving, impulse control and planning in patients with early- and late-stage Huntington's disease. *European Archives of Psychiatry and Clinical Neuroscience*, *266*(7), 663–671. https://doi.org/10.1007/s00406-016-0707-4

Novak, M. J. U., & Tabrizi, S. J. (2005). Huntington' s disease. *British Medical Journal*, *341*(july). https://doi.org/10.1136/bmj.c3109

Novak, M. J. U., & Tabrizi, S. J. (2011). *Huntington's disease: Clinical presentation and treatment*. *International Review of Neurobiology* (Vol. 98). Elsevier Inc. https://doi.org/10.1016/B978-0-12-381328-2.00013-4

Novak, M. J. U., Warren, J. D., Henley, S. M. D., Draganski, B., Frackowiak, R. S., & Tabrizi, S. J. (2012). Altered brain mechanisms of emotion processing in pre-manifest Huntington's disease. *Brain*, *135*(Pt 4), 1165–1179. https://doi.org/10.1093/brain/aws024

Ochsner, K. N. (2009). The social-emotional processing stream: Five core constructs and their translational potential for schizophrenia and beyond. *Biological Psychiatry*, *64*(1), 48–61.

Papoutsi, M., Labuschagne, I., Tabrizi, S. J., & Stout, J. C. (2014). The cognitive burden in Huntington's disease: Pathology, phenotype, and mechanisms of compensation. *Movement*

Disorders, *29*(5), 673–683. https://doi.org/10.1002/mds.25864

Paulsen, J. S. (2011). Cognitive impairment in Huntington disease: diagnosis and treatment. *Current Neurology and Neuroscience Reports*, *11*(5), 474–483. https://doi.org/10.1007/s11910-011-0215-x

Penland, E. A., Masten, W. G., Zelhart, P., Fournet, G. P., & Callahan, T. A. (2000). Possible selves, depression and coping skills in university students. *Personality and Individual Differences*, *29*(5), 963–969. https://doi.org/10.1016/S0191-8869(99)00247-0

Pringsheim, T., Wiltshire, K., Day, L., Dykeman, J., Steeves, T., & Jette, N. (2012). The incidence and prevalence of Huntington's disease: a systematic review and meta-analysis. *Movement Disorders : Official Journal of the Movement Disorder Society*, *27*(9), 1083–1091. https://doi.org/10.1002/mds.25075

Reilmann, R., Leavitt, B. R., & Ross, C. A. (2014). Diagnostic criteria for Huntington's disease based on natural history. *Movement Disorders*, *29*(11), 1335–1341. https://doi.org/10.1002/mds.26011

Ritschel, L. A., Tone, E. B., Schoemann, A. M., & Lim, N. E. (2015). Psychometric properties of the Difficulties in Emotion Regulation Scale across demographic groups. *Psychological Assessment*, *27*(3), No Pagination Specified. https://doi.org/10.1037/pas0000099

Robotham, L., Sauter, D. A., Bachoud-Levi, A. C., Trinkler, I., Bachoud-Lévi, A.-C., & Trinkler, I. (2011). The impairment of emotion recognition in Huntington's disease extends to positive emotions. *Cortex*, *47*(7), 880–884. https://doi.org/10.1016/j.cortex.2011.02.014

Sahakian, B. J., & Owen, a M. (1992). Computerized assessment in neuropsychiatry using CANTAB: discussion paper. *The Royal Society of Medicine*, *85*(July), 399–402. https://doi.org/10.1177/014107689208500711

Salovey, P., & Mayer, J. (1990). Emotional intelligence. *Imagination, Cognition and Personality*, *9*, 185–211.

Shoulson, I., & Fahn, S. (1979). Huntington disease: clinical care and evaluation. *Neurology*, *29*(1), 1–3. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/154626

Sloan, E., Hall, K., Moulding, R., Bryce, S., Mildred, H., & Staiger, P. K. (2017). Emotion regulation as a transdiagnostic treatment construct across anxiety, depression, substance, eating and borderline personality disorders: A systematic review. *Clinical Psychology Review*, *57*(September), 141–163. https://doi.org/10.1016/j.cpr.2017.09.002

Snowden, J. S., Austin, N. a, Sembi, S., Thompson, J. C., Craufurd, D., & Neary, D. (2008). Emotion recognition in Huntington's disease and frontotemporal dementia. *Neuropsychologia*, *46*(11), 2638–2649. https://doi.org/10.1016/j.neuropsychologia.2008.04.018

Soria Bauser, D., Thoma, P., & Suchan, B. (2012). Turn to me: electrophysiological correlates of frontal vs. averted view face and body processing are associated with trait empathy. *Frontiers in Integrative Neuroscience*, *6*(November), 106. https://doi.org/10.3389/fnint.2012.00106

Sprengelmeyer, R., Schroeder, U., Young, A. W., & Epplen, J. T. (2006). Disgust in pre-clinical Huntington's disease: a longitudinal study. *Neuropsychologia*, *44*(4), 518–533. https://doi.org/10.1016/j.neuropsychologia.2005.07.003

Thoma, P., Soria Bauser, D., & Suchan, B. (2013). BESST (Bochum Emotional Stimulus Set)-A pilot validation study of a stimulus set containing emotional bodies and faces from frontal and averted views. *Psychiatry Research*, *209*(1), 98–109. https://doi.org/10.1016/j.psychres.2012.11.012

Thompson, R. J., Boden, M. T., & Gotlib, I. H. (2017). Emotional variability and clarity in depression and social anxiety. *Cognition and Emotion*, *31*(1), 98–108. https://doi.org/10.1080/02699931.2015.1084908

Trinkler, I., de Langavant, L. C., & Bachoud-Levi, A. C. (2013). Joint recognition-expression impairment of facial emotions in Huntington's disease despite intact understanding of feelings. *Cortex*, *49*(2), 549–558. https://doi.org/10.1016/j.cortex.2011.12.003

van Asselen, M., Júlio, F., Januário, C., Campos, E. B., Almeida, I., Cavaco, S., & Castelo-Branco, M. (2012). Scanning patterns of faces do not explain impaired emotion recognition in Huntington disease: Evidence for a high level mechanism. *Frontiers in Psychology*, *3*(February), 31. https://doi.org/10.3389/fpsyg.2012.00031

Van den Stock, J., Righart, R., & de Gelder, B. (2007). Body expressions influence recognition of emotions in the face and voice. *Emotion*, *7*(3), 487–494. https://doi.org/10.1037/1528-3542.7.3.487

van der Meer, L. B., van Duijn, E., Giltay, E. J., & Tibben, A. (2015). Do attachment style and emotion regulation strategies indicate distress in predictive testing? *Journal of Genetic Counseling*, *24*(5), 862–871. https://doi.org/10.1007/s10897-015-9822-z

Voon, V., Sohr, M., Lang, A. E., Potenza, M. N., Siderowf, A. D., Whetteckey, J., … Stacy, M. (2011). Impulse control disorders in parkinson disease: A multicenter case-control study. *Annals of Neurology*, *69*(6), 986–996. https://doi.org/10.1002/ana.22356

Yoo, S. H., Matsumoto, D., & LeRoux, J. A. (2006). The influence of emotion recognition and emotion regulation on intercultural adjustment. *International Journal of Intercultural Relations*, *30*(3), 345–363. https://doi.org/10.1016/j.ijintrel.2005.08.006

Zarotti, N. (2016). Communication in Huntington's disease: What do we know? *FPOP Bulletin*, (136), 35–39.

Zarotti, N., Simpson, J., & Fletcher, I. (2017). ‘I have a feeling I can’t speak to anybody’: A thematic analysis of communication perspectives in people with Huntington’s disease. *Chronic Illness*, 174239531773379. https://doi.org/10.1177/1742395317733793

Zarotti, N., Simpson, J., Fletcher, I., Squitieri, F., & Migliore, S. (2018). Exploring emotion regulation and emotion recognition in people with presymptomatic Huntington’s disease: The role of emotional awareness. *Neuropsychologia*, *112*, 1–9. https://doi.org/10.1016/j.neuropsychologia.2018.02.030

Zigmond, A. S., & Snaith, R. P. (1983). The Hospital Anxiety and Depression Scale. *Acta Psychiatrica Scandinavica*, *67*(6), 361–370. https://doi.org/10.1111/j.1600-0447.1983.tb09716.x

Tables

Table 1

Demographics of the participants.

	HD			Ctrl		
	Mean	SD	Range	Mean	SD	Range
Age (yrs)	53.46	5.109	42-63	52.17	7.907	33-63
Education (yrs)	12.92	2.66	11-18	14	2.594	11-18
Diagnosis time (yrs)	5.54	1.713	3-9			
TFC score	6.92	2.139	13-0			

Note. Ctrl = control group; HD = Huntington's disease group; SD = standard deviation; TFC = Total Functional Capacity; yrs = years.

Table 2

Participants' scores across the outcome variables.

		HD	Ctrl	Between-group comparison			Reliability
		Mean (SD)	Mean (SD)	*U*	*p*	*d*	α
HADS	HADS-A	7.00 (5.80)	5.58 (2.46)	72.00	ns	.318	.808
	HADS-D	8.00 (4.49)	3.00 (2.697)	27.00	.005	1.350	.805
DERS	SUM	90.92 (28.85)	67.00 (16.92)	29.50	.008	1.011	.941
	NONACCEPT	12.69 (5.76)	11.42 (6.08)	63.00	ns	.214	.855
	GOALS	14.00 (5.71)	10.92 (2.91)	60.00	ns	.679	.814
	IMPULSE	14.54 (5.14)	9.33 (3.65)	28.50	.007	1.168	.808
	AWARE	17.54 (5.44)	14.33 (4.27)	44.50	ns	.656	.763
	STRATEGIES	18.38 (6.37)	12.92 (4.14)	43.00	ns	1.016	.819
	CLARITY	13.77 (5.08)	8.08 (2.02)	30.50	.008	1.471	.775
BESST FACES	TOTAL	22.85 (7.06)	31.92 (4.33)	16.50	.000	-1.54	.758
	NEUTRAL	4.69 (2.50)	5.42 (1.68)	69.00	ns	-.342	.634
	FEAR	2.08 (1.38)	2.58 (2.31)	71.00	ns	-.262	.567
	DISGUST	2.38 (1.85)	4.50 (2.27)	34.50	.016	-1.023	.638
	HAPPINESS	7.00 (2.34)	7.92 (1.24)	63.50	ns	-.491	.670
	SADNESS	1.08 (1.44)	1.92 (2.23)	62.00	ns	-.447	.616
	SURPRISE	2.23 (1.73)	3.58 (1.78)	44.50	ns	-.769	.531
	ANGER	3.54 (2.93)	6.00 (2.30)	40.00	.037	-.933	.756

BESST BODIES	TOTAL	28.15 (11.08)	39.58 (3.85)	32.00	.012	-1.378	.863
	NEUTRAL	7.00 (3.27)	9.25 (1.29)	40.50	.032	-.905	.896
	FEAR	2.62 (2.47)	5.67 (1.23)	25.00	.004	-1.563	.680
	DISGUST	1.00 (1.29)	1.75 (1.96)	62.50	ns	-.452	.638
	HAPPINESS	7.54 (1.39)	7.83 (2.12)	58.00	ns	-.156	.627
	SADNESS	4.62 (3.01)	7.58 (1.08)	33.00	.013	-1.309	.815
	SURPRISE	2.62 (1.61)	3.50(1.31)	54.50	ns	-.599	.352
	ANGER	2.77 (2.65)	3.92 (2.27)	55.50	ns	-.466	.740

Note. BESST TOTAL max score: 70. BESST single emotion max score: 10. Clinical cut-off for the HADS: 8/21. AWARE = lack of emotional awareness; CLARITY = lack of emotional clarity; Ctrl = control group; d = Cohen's d effect size; DERS = Difficulties in Emotion Regulation Scale; GOALS = difficulties engaging in goal directed behaviour; HADS-A = HADS anxiety score; HADS-D = HADS depression score; HD = symptomatic HD group; IMPULSE = impulse control difficulties; NONACCEPT = non-acceptance of emotional responses; SD = standard deviation; STRATEGIES = limited access to emotion regulation strategies; SUM = DERS total score; U = Mann-Whitney's U.

Table 3

Spearman's correlation coefficients for the HD group across the variables.

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
1 GENDER																															
2 AGE	.112																														
3 EDUCATION (YRS)	.342	-.127																													
4 DIAGNOSIS (YRS)	-.272	.121	-.073																												
5 TFC SCORE	.271	.326	.092	.125																											
6 HD STAGE	-.501	-.323	-.325	-.025	-.902**																										
7 HADS_A	.022	-.032	-.220	-.204	-.301	.260																									
8 HADS_D	-.201	.170	-.135	-.222	-.593*	.558*	.510																								
9 DERS_SUM	.089	.132	-.316	-.168	-.421	.371	.905**	.629*																							
10 DERS_NONACCEPT	-.067	.029	-.568*	-.017	-.441	.484	.739**	.404	.878**																						
11 DERS_GOALS	.291	-.035	-.218	-.267	-.355	.279	.867**	.436	.919**	.770**																					
12 DERS_IMPULSE	.067	.191	-.396	-.267	-.214	.223	.717**	.457	.866**	.857**	.787**																				
13 DERS_AWARE	.134	-.059	.163	-.548	-.453	.334	.383	.825**	.437	.124	.358	.265																			
14 DERS_STRATEGIES	-.067	.170	-.425	.109	-.550	.558*	.765**	.632*	.916**	.861**	.823**	.724**	.301																		
15 DERS_CLARITY	-.112	.250	.109	-.371	-.459	.491	.351	.687**	.456	.313	.309	.456	.611*	.412																	
16 BESST_F_SUM	.157	-.213	.296	-.345	.393	-.533	.025	.015	-.168	-.404	-.101	-.082	.287	-.434	-.264																
17 BESST_F_NEUTRAL	.383	-.440	.204	-.492	-.080	-.156	-.241	-.211	-.239	-.218	-.156	-.221	.233	-.427	-.299	.377															
18 BESST_F_FEAR	.275	-.264	-.157	-.121	.238	-.248	-.259	-.286	-.342	-.312	-.095	-.305	-.184	-.321	-.595*	.364	.216														

	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
19 BESST_F_DISGUST	.092	-.347	.382	-.216	.408	-.529	.088	-.178	-.179	-.419	-.037	-.159	.105	-.410	-.376	.902**	.293	.273													
20 BESST_F_HAPPNSS	-.519	-.023	-.249	-.142	.263	-.092	-.257	.018	-.274	-.167	-.421	-.070	-.023	-.357	-.182	.357	.106	.126	.219												
21 BESST_F_SADNESS	.310	.131	.248	-.030	-.077	-.040	.079	.590*	.115	-.155	.022	-.049	.678*	.078	.285	.356	.036	.086	.089	-.143											
22 BESST_F_SUPRISE	.271	.197	.375	-.465	-.037	-.207	.254	.338	.206	-.185	.328	.181	.443	.064	.248	.521	.023	.109	.499	-.216	.279										
23 BESST_F_ANGER	-.249	-.086	-.029	.136	.099	-.069	.604*	.376	.458	.282	.382	.386	.196	.339	-.013	.475	-.359	-.017	.496	.290	.220	.217									
24 BESST_B_SUM	.491	-.077	.320	-.238	.521	-.675*	-.207	-.259	-.300	-.511	-.080	-.266	.025	-.458	-.546	.739**	.357	.700**	.691**	.204	.215	.456	.212								
25 BESST_B_NEUTRAL	.409	-.427	.138	-.111	.223	-.322	-.431	-.549	-.429	-.377	-.179	-.441	-.208	-.456	-.731**	.235	.653*	.606*	.311	.119	-.198	-.100	-.275	.654*							
26 BESST_B_FEAR	.551	.261	.309	-.199	.759**	-.878**	-.298	-.396	-.342	-.499	-.179	-.171	-.167	-.524	-.447	.612*	.212	.526	.530	.204	.111	.397	.086	.888**	.473						
27 BESST_B_DISGUST	.195	-.006	.069	-.266	.287	-.420	.410	.018	.151	-.009	.194	.215	.090	-.089	-.036	.656*	.037	.212	.582*	-.109	.245	.494	.418	.314	-.251	.340					
28 BESST_B_HAPPNSS	-.185	-.327	.081	-.291	-.261	.167	.286	.330	.051	-.190	.103	-.276	.424	.009	-.121	.408	.191	.330	.440	.188	.217	.343	.358	.344	.181	-.006	.210				
29 BESST_B_SADNESS	.365	.208	.190	-.083	.496	-.653*	-.137	-.044	-.172	-.438	-.051	-.167	.085	-.272	-.466	.713**	.156	.567*	.628*	.203	.335	.568*	.317	.910**	.424	.835**	.337	.328			
30 BESST_B_SURPRISE	.578*	.036	.747**	-.012	.077	-.372	-.069	.165	-.077	-.383	-.011	-.189	.370	-.175	.047	.462	.185	.140	.347	-.332	.718**	.499	.154	.509	.096	.448	.303	.119	.507		
31 BESST_B_ANGER	.248	-.109	-.055	-.054	.582*	-.470	.158	-.127	.089	-.008	.236	.222	-.038	-.064	-.331	.531	-.114	.455	.530	.313	.094	.119	.589*	.648*	.303	.603*	.255	.041	.574*	.137	

Note. * = p<.05; ** = p<.01; AWARE = lack of emotional awareness; BESST = Bochum Emotional Stimulus Set; BESST_B = BESST Bodies modality; BESST_F = BESST Faces modality; CLARITY = lack of emotional clarity; Ctrl = control group; DERS = Difficulties in Emotion Regulation Scale; GOALS = difficulties engaging in goal directed behaviour; HADS = Hospital Anxiety and Depression Scale; HADS-A = HADS anxiety score; HADS-D = HADS depression score; HD = Huntington's disease; IMPULSE = impulse control difficulties; NONACCEPT = non-acceptance of emotional responses; STRATEGIES = limited access to emotion regulation strategies; SUM = total score; TFC = Total Functional Capacity; yrs = years.

Legend to figures

Figure 1: Example of neutral, positive, and negative emotion stimuli administered via the BESST. On the top are stimuli for the face block, on the bottom are stimuli for the body one. The left column shows neutral stimuli; the central column shows stimuli for happiness; the right column shows stimuli for fear. Each stimulus was presented separately to the participants.

Figure 2: Participants' results on the emotion recognition tasks. Mean (and standard deviation) of correct responses for each of the six-emotion category on the BESST, across both the facial and body language modality (max score = 10).

Figures

Figure 1

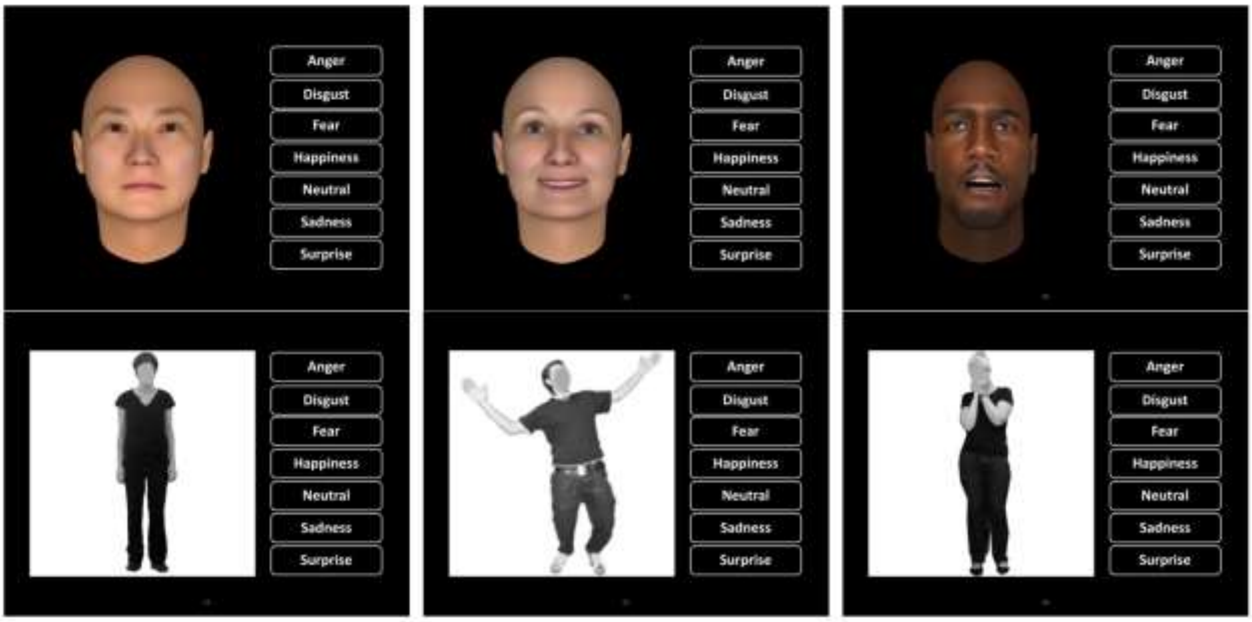

Figure 2