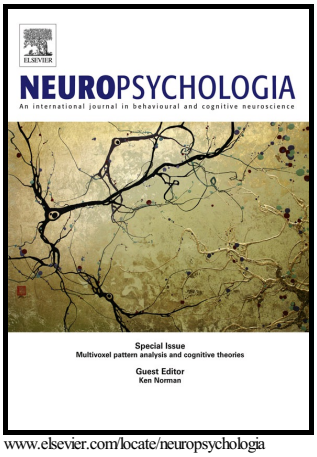

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Exploring emotion regulation and emotion recognition in people with presymptomatic Huntington's disease: the role of emotional awareness

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Abstract

Interest in the role of both emotion regulation and recognition in our understanding of mental health has been steadily increasing, especially in people with chronic illness who also have psychological difficulties. One illness which belongs to this category is Huntington's disease. Huntington's disease (HD) is a chronic neurodegenerative disorder that can cause a number of cognitive and psychological difficulties, including emotion recognition deficits, even before the onset of the symptoms required to make a formal diagnosis. Despite the lack of definite evidence, recent studies have suggested that deficits of emotion regulation and recognition may be expected to play a pivotal role in the early cognitive manifestations of HD.

In this study, we hypothesised that the ability to regulate emotions can be impaired in people with presymptomatic HD, and that such impairment may be associated with a deficit of emotion recognition. To test this, an online survey was carried out with 117 English and Italian-speaking people with presymptomatic HD, compared to 217 controls matched for age and education.

The results suggest that, in presymptomatic participants, emotion regulation and emotion recognition are generally not significantly impaired, and no significant relationships between performances on the two constructs were observed. However, a specific impairment in emotional awareness (a subscale on the Difficulties in Emotion Regulation Scale, DERS) was observed, which appears to be enhanced by the co-occurrence of depressive symptoms, even

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at a subclinical level. Consequently, it is suggested that difficulties in emotional awareness may represent a precursor of more general emotion recognition impairments, which only become apparent as the disease reaches a more symptomatic level. Clinical implications of the findings are discussed and directions for future research are proposed.

1 Introduction

1.1 Emotion regulation

Emotion regulation is defined as the process of managing one's emotions, as well as when and how one experiences or expresses them (Gross, 1998; Mayer, 2001). It involves both negative and positive emotions and its successful operation for good mental health has been widely recognised, with a substantial increase in the number of empirical investigations addressing this broad research area in the last two decades (e.g., Eftekhari, Zoellner, & Vigil, 2009; Gross, 2013; Gross & Muñoz, 1995). The most popular framework that has been developed to explain its functioning is the Process Model of Emotion Regulation (Gross, 1998), which identifies five fundamental families of regulatory emotional processes: a) *Situation selection*, i.e. taking necessary actions to approach or avoid situations potentially involving emotional responses; b) *situation modification*, i.e. modifying a situation in order to affect its emotional impact; c) *attentional deployment*, i.e. deploying or distracting attention in a situation to alter the emotional response; d) *cognitive change*, i.e. changing point of view or perspective on a situation in order to change the emotional response; e) *response modulation*, i.e. taking direct action on managing the behavioural and physical components of the emotional response. The latter two processes are responsible for the two main strategic outcomes of emotion regulation: *reappraisal*, which originates from cognitive change and involves actively rethinking a situation to alter the emotional response, and *emotion suppression*, which belongs to the response modulation family and promotes the decrease of emotion expression (Gross, 2013). While reappraisal is a cognitively-oriented strategy that has proven to be particularly beneficial for the regulation of negative emotions and the promotion of positive experiences, the evidence on emotion suppression (which, by contrast, is behaviourally-oriented) shows limited benefits and suggests the potential for mainly detrimental effects such as increased negative experiences, and memory difficulties (for a meta-analysis, see Webb et al., 2012).

Regardless of their specific efficacy, however, the successful implementation of emotion regulation strategies is based on the accurate functioning of a number of fundamental physiological, behavioural, and cognitive mechanisms that are known to be involved in emotional processing and response (Mauss et al., 2005). These include the ability to recognise emotions in other people, which represents a key cognitive and social skill and whose impairment is likely to cause emotional dysregulations. This is especially true in

clinical conditions where emotion recognition is frequently impaired, such as in neurodegenerative disorders (Löffler et al., 2015).

1.2 Emotional difficulties in neurodegenerative disorders: the case of Huntington's disease

Huntington's disease (HD) is a hereditary progressive neurodegenerative disorder which affects 12.3 people per 100,000 in the UK (Evans et al., 2013). Typical symptoms include involuntary movements (chorea), involuntary abnormal postures (dystonia), cognitive deterioration (dementia) and significant psychological problems (Novak and Tabrizi, 2005). Since the transmission mechanism is autosomal-dominant, every affected individual has a 50% probability of transmitting it to their children. The mean age of onset is 40-50 years, and the mean life expectancy after the diagnosis is typically 20 years (Folstein, 1989). Genetic testing is available for individuals at risk, allowing them to know if they carry the mutated gene. Individuals with this gene mutation will develop the disease at a certain time in their life, and are defined as 'presymptomatic'. The full diagnosis of Huntington's disease is based on the development of motor symptoms along with a familiar history (i.e., proof of an affected parent). Cognitive and psychological changes, including emotional problems, do not have to be present for a diagnosis to be made, even though they can arise much earlier than motor impairments, thus still affecting 'presymptomatic' individuals.

An impairment in the ability to recognise emotions has been widely noted in empirical studies in people with symptomatic HD over the last few decades, with both cross-sectional and longitudinal investigations consistently reporting evidence of early deterioration in the facial recognition of negative emotions, more specifically anger, fear, and disgust (e.g., Bates et al., 2014; Dumas et al., 2013; Henley et al., 2012; Johnson et al., 2007; Robotham et al., 2011). These findings were further underlined by a recent meta-analysis on disorders of social cognition in HD (Bora et al., 2015). Evidence in people with presymptomatic HD for the same set of deficits is more sparse and contrasting; some evidence from small samples (i.e., below 50) reports no significant impairment of emotion recognition (Kipps et al., 2007; Milders et al., 2003), while other investigations on larger samples observed a significant impairment of negative emotion (e.g., the results from the TRACK-HD study; Labuschagne et al., 2013 – for a systematic review on emotion recognition specifically, see Henley et al., 2012). On the other hand, very little is currently known about emotion regulation in people with neurodegenerative disorders, and HD in particular. For example, a recent review (Löffler et al., 2015) was able to retrieve only one study, which found no differences between people with

symptomatic HD and healthy controls in the self-reported usage of emotion suppression and reappraisal (Croft et al., 2014). Moreover, no studies were retrieved on emotion regulation in people with presymptomatic HD, and to our knowledge none has ever been carried out.

However, as previously anticipated, emotion recognition plays a pivotal role in the successful implementation of emotion regulation, and impairments in emotion recognition are likely to contribute to the development of emotion regulation difficulties, especially in clinical populations (Cecchetto et al., 2014; Gray and Tickle-Degnen, 2010; Löffler et al., 2015). In particular, according to emotional intelligence theory (Mayer, 2001; Salovey and Mayer, 1990), emotion regulation can only occur after emotions have been recognised (Izard et al., 2001; Yoo et al., 2006). With specific reference to the abovementioned Process Model, the inability to recognise emotions effectively seems likely to impair the regulatory processes that are based on the accurate emotional assessment of people and situations. Such processes could include situation selection (e.g., not being able to recognise those that are potentially negative), attentional deployment (e.g., not recognising relevant emotional cues), and cognitive change (e.g., basing change on misrecognised emotions). This is further corroborated by current evidence of a significant correlation between deficits of emotion recognition and emotion regulation difficulties in other clinical populations, such as people with anorexia nervosa (Harrison, Sullivan, Tchanturia, & Treasure, 2009), bulimia (Harrison et al., 2010), and borderline personality disorder (Domes et al., 2009). Moreover, recent evidence on anorexia nervosa has also shown associations between specific components of emotion regulation and psychological difficulties such as depression and anxiety (Racine and Wildes, 2013). Thus, in the particular case of HD, it would be expected that emotion regulation and emotion recognition could play a major role in the early cognitive and psychological difficulties that occur prior to formal diagnosis. This in turn represents the main argument of a currently ongoing debate on whether more comprehensive diagnostic criteria should be considered for the disease (Loy and McCusker, 2013; Paulsen, 2011; Reilmann et al., 2014).

Consequently, considering the evidence of potential early emotion recognition deficits in presymptomatic individuals (Henley et al., 2012) as well as the link between emotion recognition deficits and regulation impairments observed in other clinical populations (Harrison et al., 2010; Harrison et al., 2009; Racine and Wildes, 2013), the overarching aim of this study was to investigate the hypothesis that emotion regulation abilities can be impaired in presymptomatic HD gene carriers, and that such an impairment may be associated with a deficit of emotion recognition. More specifically, the following hypotheses were formulated

for this study: a) People with presymptomatic HD were predicted to report significantly more emotion regulation difficulties when compared to healthy controls; b) performance on emotion recognition was predicted to be worse in people with presymptomatic HD when compared to healthy controls; c) a significant positive relationship was predicted between difficulties in emotion recognition and emotion regulation, with more difficulties in emotional recognition correlating with more difficulties in emotional regulation. Moreover, in order to control for the possible interactions between emotion regulation and recognition with psychological difficulties, depression and anxiety were investigated as potential covariates.

2 Materials and Methods

2.1 Design

This study adopted an online survey to explore emotion regulation and recognition in people with presymptomatic Huntington's disease with age-matched (non-affected) controls. The survey was developed with the Qualtrics® software, and included measures for emotion regulation, emotion recognition, as well as anxiety and depression. Both English and Italian versions of the survey were developed. The Italian version was developed via an ongoing collaboration between the Division of Health Research (DHR) at Lancaster University and the Italian League for Research on Huntington and Related Disease (LIRH Foundation) at the Mendel Institute of Human Genetics in Rome. The aim was to expand the sample size and was possible due to the availability of standardised translations of all the included measures that report the same validity as the English version. Separate links were generated to facilitate the dissemination among the target populations.

2.2 Participants

In total, 334 participants took part in the present study. The power calculation showed that, assuming a small effect size (d = .2), a minimum sample of 188 participants (99 for each group) was required to achieve a minimum statistical power of .8 using a probability value of 0.05. The first group (Pre-HD) consisted of 117 people with presymptomatic HD, of which 83 were English-speakers and 34 Italian-speakers. The second group (Ctrl) consisted of 217 age-matched controls, of which were 69 English-speakers and 148 Italian-speakers. An initial self-report question was included for the Pre-HD versions of the survey in order to measure the participants' presymptomatic status (i.e., "Have you had a positive genetic test for

Huntington's disease, but you're not experiencing relevant movement problems?"). The presymptomatic participants and the controls did not present any significant differences in terms of age [$t(305.767) = 1.789$, p = ns], years in full-time education [$t(189.749) = -1.864$, p = ns], and gender [$X^2(2, N = 334) = 1.606$, p = ns]. See Table 1 for the full demographic details of the participants.

Table 1

Demographics of the participants.

	Pre-HD		Ctrl	
	N	Mean (SD) Min-Max	N	Mean (SD) Min-Max
Language (EN/IT)	83/34		69/148	
Gender (M/F)	35/82		59/158	
Age (yrs)	117	37.38 (11.06) 19-70	217	40 (15.39) 18-74
Education (yrs)	117	14.49 (2.77) 11-21	217	13.94 (2.11) 11-17
Test-time (yrs)	117	5.09 (5.34) 0-30		

Note. Ctrl = control group; F = female; M = male; Min-Max = minimum-maximum value; N = count; Pre-HD = presymtomatic group; SD = standard deviation; yrs = years.

The participants were enrolled across the UK and other English-speaking countries (e.g., USA, Canada, Australia and New Zealand), as well as Italy and San Marino via social media and with the help of local and international Huntington's disease associations. All the participants reported to be native speakers of the respective languages.

2.3 Measures

2.3.1 Emotion recognition measures

2.3.1.1 Reading the Mind in the Eyes test

The Reading the Mind in the Eyes test (RME; Baron-Cohen et al., 2001) is a test consisting of 36 still pictures of the facial eye regions expressing different emotional states. The participant is asked to match a list of provided emotional tags to the emotions displayed in the pictures. The test yields a total score out of 36, and higher scores indicate higher recognition performance. The RME is used worldwide and has been adopted with many clinical conditions, including schizophrenia (Kettle et al., 2008), autism (Baron-Cohen et al., 2001) and anorexia nervosa (Harrison et al., 2009), as well as Huntington's disease (Allain et al., 2011). It has previously shown acceptable construct validity when compared to other emotion recognition tasks (Alaerts et al., 2011), as well as acceptable internal consistency (Cronbach's α = .63 for men, .60 for women; Voracek and Dressler, 2006). The Italian

translation by Vellante and colleagues (2013) was used for the Italian version of the survey, which has shown good construct (discriminant and convergent) validity and acceptable internal consistency (Cronbach's α = .60).

2.3.2 Emotion regulation measures

2.3.2.1 Difficulties in Emotion Regulation Scale

The Difficulties in Emotion Regulation Scale (DERS; Gratz & Roemer, 2004) is a validated self-report questionnaire consisting of 36 items rating emotion regulation on a 5-point Likert scale. It includes 6 subscales: non-acceptance of emotional responses (NONACCEPT), difficulties engaging in goal directed behaviour (GOALS), impulse control difficulties (IMPULSE), lack of emotional awareness (AWARE), limited access to emotion regulation strategies (STRATEGIES), lack of emotional clarity (CLARITY). It yields a subscore for each subscale, as well as a total score (SUM) out of 180. As the focus of the test is on difficulties, higher scores equal poorer emotion regulation. To our knowledge, the DERS has never been adopted with HD; however, it has been used with several other clinical populations, including participants with both psychological (Fowler et al., 2014) and physical conditions (Kökönyei, Urbán, Reinhardt, Józan, & Demetrovics, 2014). The DERS has previously shown good construct validity (Kökönyei et al., 2014), even when adopted across different cultural and ethnical groups (Ritschel et al., 2015). It also showed very good internal consistency, with a Cronbach's α of .93 for the total score (SUM), and figures ranging between .80 and .89 for the subscales (Gratz and Roemer, 2004). The Italian validation by Sighinolfi and colleagues (2010) was adopted for the Italian version of the survey, which has showed psychometric properties comparable to the English version.

2.3.3 Mood and anxiety issues measures

2.3.3.1 Hospital Anxiety and Depression Scale

The Hospital Anxiety and Depression Scale (HADS; Zigmond & Snaith, 1983) is a validated self-report questionnaire consisting of 14 items rating anxiety and depression symptoms on a 3-point scale. No unified score is provided at the end of the test. Instead, individual scores on a scale out of 21 are provided for anxiety and depression. The HADS represents one of the most adopted measures in clinical populations and it has been specifically validated with people affected by HD (De Souza et al., 2010). A review of its psychometric properties (Bjelland et al., 2002) reported good construct validity when compared to other common clinical measures, as well as good internal consistency (mean Cronbach's α = .83 for anxiety, .82 for depression). The same study identified a recommended

cut-off point of 8/21 to yield both good sensitivity (anxiety = .90; depression = .83) and specificity (anxiety = .78; depression = .79). The HADS was incorporated in this study to control for the potential confounding effect of depression and anxiety levels on the ability to regulate and recognise emotions. For the Italian version of the survey, the validation by Costantini and colleagues was adopted (1999), which showed comparable high construct validity and internal consistency (Cronbach's α = .89 and .88 for anxiety and depression respectively).

2.4 Statistical analysis

Data were analysed with IBM SPSS Statistics® programme v23 (Armonk, NY: IBM Corp). Independent-sample t-tests and one-way ANOVAs were performed to compare means across the participants groups. Considering the number of repeated comparisons, in order to control for family-wise error-rate (FWER), the Bonferroni correction was applied and the significance level was adjusted from .05 to .005. The relationship between the participants' demographic and clinical characteristics and the outcome variables (i.e., emotion regulation, emotion recognition, depression and anxiety) was investigated through Pearson's correlations (two-tailed).

2.5 Ethics approval

This study was reviewed and approved by the Faculty of Health and Medicine Research Ethics Committee at Lancaster University (ref: FHM REC16015).

3 Results

The mean scores of the participants of the Pre-HD and Ctrl groups on the outcome variables are shown in Table 2. Based on the recommended cut-off values of skewness and kurtosis (West et al., 1995), the scores were normally distributed. Of the participants in the Pre-HD group, 48.7% and 23.1% showed clinical levels of anxiety and depression respectively, by scoring above the recommended clinical cut-off for the HADS (i.e., 8/21; Bjelland et al., 2002). The participants of the Ctrl group showed similar figures, with 46.5% and 23.5% above the cut-off point. In terms of levels of emotion regulation difficulties, the Ctrl group showed total scores (SUM) comparable to the reported data with general adult populations (e.g., 77/180; Ritschel et al., 2015), while the Pre-HD group scored marginally higher, meaning that more emotion regulation difficulties were reported. The mean emotion recognition

performance of both groups was very similar and slightly below the reported normative data for general adult populations (e.g., 26/36; Baron-Cohen et al., 2001). In terms of measure reliability, high levels were shown by the HADS (Cronbach's α = .84/.78 for anxiety/depression) and the DERS (Cronbach's α = .94 for the SUM score, .83 to .89 for the subscales). The RME showed a level of reliability (Cronbach's α = .55) comparable to previously accepted figures in the literature (Voracek and Dressler, 2006).

The group comparison showed that the Pre-HD and Ctrl groups did not differ significantly in terms of total score of emotion regulation difficulties (DERS SUM; [$F(1, 332) = 1.939$, p = ns]), emotion recognition (RME; [$F(1, 332) = 1.291$, p = ns]), as well as anxiety (HADS-A; [$F(1, 332) = 1.472$, p = ns]) and depression (HADS-D; [$F(1, 332) = .393$, p = ns]). When comparing the subscales of the DERS, significant differences were observed only on the AWARE score [$F(1, 332) = 9.359$, $p = .002$]. Effect size analysis indicated no relevant effects of any of the other variables, except for a small effect of the IMPULSE score ($d = .220$). The full details of the comparison results are shown in Table 2.

In addition, no significant correlation was found between the total level of emotion regulation difficulties (DERS-SUM) and emotion recognition performance (RME) in either the presymptomatic participants ($r = -.030$, $N = 117$, p = ns) or the controls ($r = -.050$, $N = 217$, p = ns). Table 3 shows the full details of Pearson's correlation coefficients for the Pre-HD participants' scores across all the variables.

Table 2

Participants' scores across the outcome variables.

		Pre-HD	Ctrl	Between-group comparison			Reliability
		Mean (SD)	Mean (SD)	*F*	*p*	*d*	α
HADS	HADS-A	7.85 (4.78)	7.24 (4.16)	1.472	ns	.136	.84
	HADS-D	4.77 (3.911)	5.04 (3.61)	.393	ns	-.071	.78
DERS	SUM	80.91 (27.59)	76.97 (22.93)	1.939	ns	.155	.94
	NONACCEPT	12.98 (5.59)	13.26 (5.83)	.174	ns	-.049	.87
	GOALS	13.12 (5.30)	12.95 (5.05)	.079	ns	.032	.87
	IMPULSE	11.93 (5.33)	10.86 (4.35)	.946	ns	.220	.85
	AWARE	15.77 (5.91)	13.83 (5.31)	9.359	.002	.345	.83
	STRATEGIES	16.90 (7.54)	16.42 (6.79)	.348	ns	.066	.89
	CLARITY	10.21 (4.28)	9.65 (4.14)	.1335	ns	.132	.84

RME	Total Score	24.07 (3.863)	24.58 (3.97)	1.291	ns	-.130	.55

Note. Clinical cut-off for the HADS: 8/21. Significance level = .005; α = Cronbach's alpha; AWARE = lack of emotional awareness; CLARITY = lack of emotional clarity; Ctrl = control group; DERS = Difficulties in Emotion Regulation Scale; F = ANOVA F value; GOALS = difficulties engaging in goal directed behaviour; HADS-A = HADS anxiety score; HADS-D = HADS depression score; IMPULSE = impulse control difficulties; NONACCEPT = non-acceptance of emotional responses; Pre-HD = presymptomatic group; RME = Reading the Mind in the Eyes test; SD = standard deviation; Sig. = significance; Size = effect size; SUM = DERS total score.

Table 3

Correlation coefficients for pre-HD participants' scores.

	1	2	3	4	5	6	7	8	9	10	11	12	13	14
1. Gender														
2. Age	-.157													
3. Education	-.080	-.001												
4. Test	-.036	.286**	.057											
5. HADS-A	.157	-.098	.030	-.130										
6. HADS-D	-.002	-.006	-.093	-.146	.669**									
7. DERS SUM	.104	-.065	.055	-.133	.758**	.760**								
8. DERS NONACCEPT	.141	-.006	.067	-.070	.572**	.501**	.796**							
9. DERS GOALS	.147	-.040	.117	-.094	.667**	.628**	.826**	.570**						
10. DERS IMPULSE	.110	.019	.072	-.131	.618**	.580**	.803**	.561**	.619**					
11. DERS AWARE	-.055	-.072	-.085	-.148	.400**	.513**	.662**	.423**	.384**	.391**				
12. DERS STRATEGIES	.115	-.074	.093	-.057	.764**	.745**	.925**	.709**	.786**	.762**	.434**			
13. DERS CLARITY	.040	-.153	-.016	-.181	.643**	.727**	.841**	.587**	.655**	.547**	.606**	.751**		
14. RME	.153	-.026	.194*	.012	-.058	-.184*	-.030	.061	.058	-.058	-.155	-.018	-.030	

Note. * = p<.05; ** = p<.01; AWARE = lack of emotional awareness; CLARITY = lack of emotional clarity; DERS = Difficulties in Emotion Regulation Scale; GOALS = difficulties engaging in goal directed behaviour; HADS-A = HADS anxiety score; HADS-D = HADS depression score; IMPULSE = impulse control difficulties; NONACCEPT = non-acceptance of emotional responses; RME = Reading the Mind in the Eyes test; SUM = DERS total score; Test = time since predictive test.

Table 4

Standardised coefficients for the multiple regression models of the AWARE subscore.

	Pre-HD		Ctrl	
	β	*p*	β	*p*
HADS-A	.128	ns	.129	ns
HADS-D	.669	.000	.221	ns

Note. AWARE = lack of emotional awareness; β = standardised coefficient; Ctrl = control group; HADS-A = HADS anxiety score; HADS-D = HADS depression score; *p* = significance; Pre-HD = presymptomatic group.

Despite not being initially hypothesised but in order to explore this result further, multiple regressions were conducted to assess the contribution of anxiety and depression to the AWARE subscale, as it was the only one to show a significant difference across the groups. The

standardised regression coefficients are summarised in Table 4. Using the enter method, it was found that the model (which consisted of two variables) explained a significant amount of the variance of the AWARE score in both the Pre-HD ($F(2, 114) = 20.947$, $p < .001$, $R^2 = .269$, $R^2_{Adjusted} = .256$) and Ctrl group ($F(2, 214) = 5.623$, $p = .004$, $R^2 = .050$, $R^2_{Adjusted} = .041$). The analysis in the Pre-HD group showed that the AWARE score was not significantly predicted by anxiety ($\beta = .128$, p = ns), but it was significantly predicted by depression ($\beta = .669$, $p < .001$). On the other hand, in the Ctrl group neither anxiety ($\beta = .129$, p = ns) nor depression ($\beta = .221$, p = .ns) significantly predicted the AWARE score.

4 Discussion

To our knowledge, the present study has been the first to investigate emotion regulation abilities in a population of participants with presymptomatic Huntington's disease (HD) by comparing these with those of matched healthy controls. In addition, it has also been the first to explore the relationship between emotion regulation and emotion recognition, as well the potential interaction with depression and anxiety, in this type of population. The results showed no significant differences between the two groups in terms of general difficulties in regulating emotions and performance on an emotion recognition task. Moreover, no significant correlation was found between emotion regulation and emotion recognition scores in both groups. As a consequence, none of our initial hypotheses was confirmed by the results, suggesting that the ability both to regulate and recognise emotions does not deteriorate early in non-symptomatic carriers of the gene Huntington's disease, and these two abilities are not significantly correlated. The result on emotion recognition appears to be inconsistent with what found by the TRACK-HD study, which reported significant impairments for the recognition of disgust, fear, and surprise (Labuschagne et al., 2013). Also, in this current study, there was no correlation with emotion regulation – which contradicts previous findings in other clinical populations (Harrison et al., 2010; Harrison et al., 2009; Racine and Wildes, 2013). However, part of these inconsistences may be explained by a number of methodological differences and, in particular, the type of measures adopted in this and the other studies. In particular, the measure adopted by the TRACK-HD study (the Ekman and Friesen face stimulus set; Ekman and Friesen, 1976) featured a subscore for each of the seven investigated emotions, while the RME only features an overall total score. Thus, the lack of impairment in emotion recognition found in this study may be at least partially due to the lack of specific subscores for selectively impaired emotions. Also, the measure of emotion regulation we adopted (DERS) focused specifically on difficulties rather than other

components (e.g., the use regulatory strategies). Therefore, the lack of correlation in HD may be at least partially due to the specific adoption of the DERS, and its interaction with the RME.

In addition, the adoption of the RME offers the opportunity to formulate an alternative interpretation of the findings on emotion recognition. Indeed, the RME has been initially constructed as a task of affective theory, and in particular a test of theory of mind (ToM, i.e. the ability to predict other people's intentions, reactions, beliefs, and mental states; (Green et al., 2008; Mathersul et al., 2013; Mitchell and Phillips, 2015) through the recognition of complex emotions (Baron-Cohen et al., 2001). However, it is also often considered and adopted as an emotion recognition measure (Adolphs et al., 2002; Guastella et al., 2010; Harrison et al., 2010; Harrison et al., 2009; Quintana et al., 2012; Vellante et al., 2013), and, as pointed out by a recent review (Mitchell and Phillips, 2015), the RME represents "a likely point of interface between the two concepts" (p. 4). Thus, the dual nature of the measure allows for the opportunity to conceptualise in more detail the non-impaired results on the RME and the lack of correlation with the DERS from the ToM perspective as well. In particular, the recent meta-analysis by Bora and colleagues on social cognitive deficits in HD (2016) has highlighted that presymptomatic individuals in general show no significant impairment of ToM when compared to healthy controls. This was in contrast to the significant ToM impairment which was generally observed in fully symptomatic individuals. Thus, within this perspective, the results on the RME of the present study are coherent with current research on impairments of ToM in people with presymptomatic HD. With specific regard to a possible correlation between ToM and emotion regulation, the already noted lack of investigations of the construct in people with HD (Löffler et al., 2015) does not allow comparisons with any previous results. However, when investigated in other clinical conditions, such as borderline personality disorder (BPD), no significant correlation was found between emotion regulation and ToM in either adolescents (Sharp et al., 2011) or adults (Ghiasi et al., 2016). Therefore, the lack of correlation between the results on the RME and DERS in the present study may also be interpreted as a lack of correlation between ToM and emotion regulation consistent with previous findings.

When analysing more specific components of emotion regulation, a significant difference was found between the two groups on one subscale, with the presymptomatic participants reporting significantly greater lack of emotional awareness (DERS-AWARE), i.e. difficulties in the ability to attend to and acknowledge emotions. A possible explanation of this could lie in the development of some of HD's early cognitive symptoms. Indeed, general

difficulties of awareness and emotional processing have been linked to poorer functioning of prefrontal brain regions such as the anterior insular (AIC) and anterior cingulate (ACC) cortices (Craig, 2009; Lane et al., 1998; Phillips, Drevets, Rauch, & Lane, 2003), which are known to be affected by HD (Dogan et al., 2014; Gray et al., 2013) and are often responsible for a wide range of emotional difficulties in both symptomatic (Craufurd et al., 2001; Ho et al., 2006; Hoth et al., 2007; Mörkl et al., 2016) and presymptomatic patients (Klöppel et al., 2010). Therefore, the difference observed for the presymptomatic participants on the AWARENESS subscale of the DERS may represent an expression of the many subtle biological and cognitive changes that presymptomatic individuals can experience before a formal diagnosis of HD is made (e.g., Tabrizi et al., 2011).

In particular, it could be interpreted in light of the early development of the well-established impairments in emotion recognition in people with symptomatic HD. This appears to be supported by multiple evidence of a significant predictive role played by alexithymia (a condition characterised by pervasive deficits of emotional awareness; Lane et al., 2000) in emotion recognition impairments both when it manifests alone (Lane et al., 2000, 1996), as well as when it is part of other clinical conditions, such as autism (Cook et al., 2013) and eating disorders (Brewer et al., 2015). Moreover, alexithymia measures often show high correlation levels with many of the DERS subscales, including the AWARE one (Ghorbani et al., 2017; Stasiewicz et al., 2012). In contrast, current evidence on alexithymia in HD is extremely limited, as a recent systematic review identified only one study that reported no significant impairment in symptomatic individuals (Ricciardi et al., 2015). Therefore, the significantly greater level of difficulties observed with the presymptomatic participants might represent a precursory gateway to the development of the emotion recognition impairment found in fully symptomatic individuals, which in turn is likely to affect the other components of emotion regulation. This hypothesis requires further exploration, as it is currently uncertain whether emotional awareness difficulties may lead to a fully alexithymic condition in people with symptomatic HD. In addition, in this study the particular structure and sensitivity of the RME might have prevented the observation of any significant correlations between the specific AWARE subscale and emotion recognition performance (see Limitations section).

Moreover, a considerable contribution to poorer emotional awareness may come from social and environmental factors, and more specifically from the type of family context, as many gene carriers often grow up in contact with a parent affected by symptomatic HD. Indeed, current evidence suggests that the family environment and the emotional climate in

which a child is raised are deeply related to the successful development of emotional processing skills, and especially emotion regulation (for a review, see Morris et al., 2007). In particular, parents' emotional responses to their children's emotions have been linked with emotional awareness (Eisenberg et al., 1998; Schultz et al., 2001; Sim et al., 2009). Thus, the fact of living in a family context characterised by challenging emotional responses due to close contact with a symptomatic parent may have hampered the successful learning of the ability to acknowledge emotions in the presymptomatic participants of this study, ultimately contributing to the development of a deficit of emotional awareness in particular. This hypothesis needs further exploration with individuals for whom more details regarding personal and clinical history are available – in particular with the aim of discerning whether this influence contributes to predispose the development of emotional impairments that emerge after onset (as suggested in this study), or whether it constitutes a potential independent process that may trigger deficits separately at the presymptomatic stage.

Further insight was also provided by the results of the regression models, which showed that depression explained a considerable portion of variance on the AWARE subscale in the Pre-HD group only. This differential effect of depression on presymptomatic participants could again represent an expression of the early cognitive manifestations of the disease. In particular, this may limit patients' coping abilities and overall resilience in the face of depression, even when they have not yet reached clinical levels, in a way that is common to many neurodegenerative conditions (for a review, see Baquero, 2015). The predictive role of depression is also corroborated by findings from studies with clinically depressed individuals; these often show problems with many components of emotion regulation (Ehring et al., 2008; Loas et al., 1997), and in particular emotional awareness (Boden and Thompson, 2015).

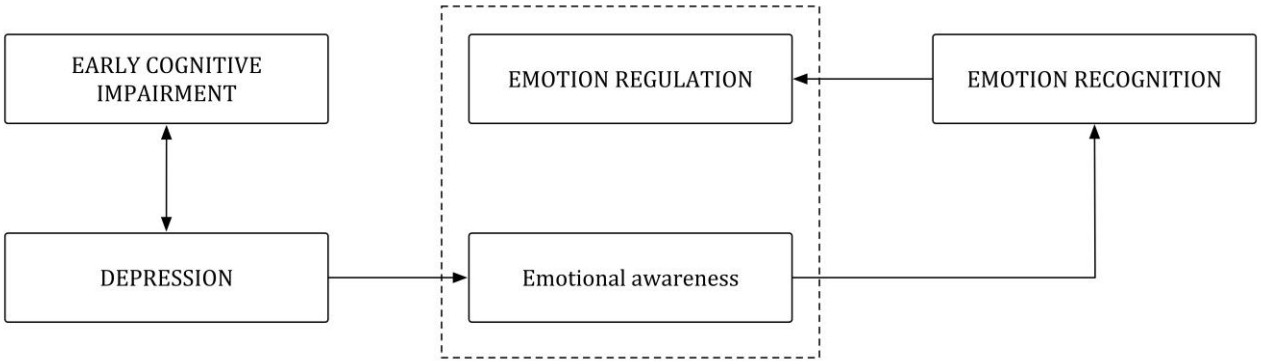

Figure 1: hypothesised interaction among the discussed constructs. Arrowed lines identify direct influence. Upper case identifies main constructs, lower case subcomponents. Dotted lines identify inclusion within the same construct.

Thus, due to the concurrent development of psychological difficulties within the context of suboptimal psychological resilience due to HD's early cognitive symptoms, depression might show a disproportionate effect on presymptomatic people even at subclinical levels. This may contribute to a greater lack of emotional awareness, and eventually lead to the development of emotion recognition impairments which, as previously mentioned, may in turn affect the other components of emotion regulation, thus establishing a symptomatic vicious circle. Figure 1 shows a schematisation of the hypothesised interaction.

However, despite the evidence reported above, it is worth noting that a possible alternative conceptualisation of emotional awareness (and in particular how it is operationalised in the DERS) might identify the construct as more akin to the cognitive side of emotional processing, thus being more closely related to emotion recognition rather than emotion regulation. Indeed, it is important to consider such view in relation to the potential effect on other measures. More specifically, it could be plausible that the inability to cognitively recognise one's own emotional states may have an impact on the results yielded by self-reported measures such as the DERS and the HADS (e.g., not indicating a difficulty regulating emotions due to failing to recognise emotionally challenging states). While this conceptualisation does not appear to be incompatible with the precursory role hypothesised in this study, more investigations are required to shed light on the cognitive and affective nature of emotional awareness.

5 Limitations and future directions

Despite allowing for a large sample, the online nature of this study carries the intrinsic limitation of a lack of direct contact between the researcher and the participants. This includes the inability to obtain some important clinical details about the participants, such as pharmacological therapies and regimes, as well as the number of CAG repeats, which would allow for the calculation of disease burden scores and thus offer a better understanding of proximity to onset. Moreover, some of the demographic characteristics of the presymptomatic group should be taken into consideration when interpreting the results from the present study. In particular, despite the mean age of the participants (37.38 years) being approximately five years younger than the mean age of onset of HD (Folstein, 1989), the total age range was very wide (19-70). This translated into the inclusion of young participants who

may have been very far from onset, as well as participants who, due to advanced age and despite self-identifying as presymptomatic, may in fact represent people with reduced penetrance who may never develop the disease. However, it should be also noted that the range in our study was similar to other large sample investigations (e.g., 18-65 in TRACK-HD; Labuschagne et al., 2013), and only two participants reported to be older than 60, thus accounting for the 1.7% of the total presymptomatic sample.

In addition, the responses to all questions were made mandatory to proceed throughout the survey. While this eliminated the need to control for missing data, it might have also limited the sample size due to participants dropping out before completing their participation. To control for this issue, the number of measures was limited to keep the overall time of the survey below 30 minutes. From this perspective, the RME was chosen in this study as its brevity fit particularly well with an online design, and due to the availability of both an English and Italian version. However, as noted above, despite being widely regarded and utilised as an emotion recognition test (Adolphs et al., 2002; Guastella et al., 2010; Harrison et al., 2010; Harrison et al., 2009; Quintana et al., 2012; Vellante et al., 2013), the measure was originally created as a test of theory of mind (ToM) to assess the recognition of mental states through eyes expressing complex emotions (Baron-Cohen et al., 1997) – and may thus be conceived as a task investigating both constructs (Mitchell and Phillips, 2015). As a consequence, on one hand it may be possible that the RME was not sensitive enough to detect subtle differences in emotion recognition in the specific population of this study, nor may it be able to show potential correlations with emotional awareness. On the other hand, however, if interpreted as a measure of ToM, the RME may have correctly identified a lack of correlation with emotion regulation that would be consistent with reports from other clinical populations. Finally, high levels of anxiety were reported by many participants in both the Pre-HD and Ctrl groups (48.7% and 46.5% respectively). While clinical anxiety has been found to be part of the range of psychological difficulties experienced by presymptomatic individuals (e.g., results from PREDICT-HD; Duff et al., 2007), the levels reported by the control group were generally higher compared to the data available from studies using the HADS with other general populations, although with a certain variability (e.g., 21%, Hinz and Brähler, 2011: 33%, Crawford et al., 2001). Thus, it should be noted that the control group in this study may not be a perfect representation of the general population in terms of anxiety levels.

Future research should aim at adopting more diversified measures of emotion regulation and recognition in face to face studies. In particular, the adoption of emotion recognition tasks that are clearly distinct from ToM and characterised by more comprehensive stimuli based on both faces and body language – such as the recent Bochum Emotional Stimulus Set (BESST; Thoma, Soria Bauser, & Suchan, 2013) – may yield different results in terms of comparison of recognition performance and correlation between emotion recognition and emotional regulation. Moreover, more in-depth measures for emotional awareness and alexithymia should be used to investigate further the precursory role of emotional awareness difficulties hypothesised in this study, as well as the potential alternative conceptualisations of the construct. Further exploration is also warranted on emotion regulation in people affected by HD with the adoption of measures focused on both difficulties and regulatory strategies. Finally, more comprehensive information should be collected on the family context of the participants, and in particular in relation to challenging emotional responses, in order to investigate this potential effect on emotion recognition and awareness in presymptomatic and symptomatic individuals.

All these suggestions could generally benefit from the inclusion in large-scale longitudinal clinical trials, which would allow an increase in our understanding of emotional processing in Huntington's disease over its full clinical course.

6 Conclusion

This online study offered some preliminary insight into emotion regulation in people with presymptomatic Huntington's, as well as further insight into emotion recognition. The findings suggest that presymptomatic individuals show a wide range of normal abilities, as emotion regulation and emotion recognition were not significantly impaired when compared to healthy controls, nor did they share significant relationships with one another. However, one specific emotion regulation component, emotional awareness, was significantly impaired. This could be due to HD's early cognitive manifestations, and a catalytic role may be played by their co-occurrence with psychological difficulties such as depression, even at a subclinical level. Moreover, the greater level of difficulties in emotional awareness shown by presymptomatic people may represent a precursor of the development of the emotion recognition impairments that are often observed in fully symptomatic individuals and that may in turn have a detrimental effect on the other components of emotion regulation.

These findings can have important implications for clinical practice; better management of depression could lead to increased levels of emotional awareness, better emotion recognition performance, and eventually better emotion regulation, as well as everyday communication and quality of life. In addition, the possible precursor role of emotional awareness shows the potential to consider the current diagnostic criteria by shedding new light on early cognitive difficulties in HD, as well as inform new therapeutic protocols and interventions tailored around the emotional and communicative needs of the people affected by this condition.

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Highlights

- Emotion regulation and recognition may be impaired in presymptomatic HD.
- An online survey was conducted with 117 presymptomatic HD, compared to 217 controls.
- Emotion regulation and emotion recognition were not generally impaired.
- A specific impairment in emotional awareness was observed.
- A precursory role of emotional awareness in recognition impairments is suggested.